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Ronald J. Scorpio
Editors

Pediatric Surgery

Diagnosis and Treatment

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To our loved ones at home, who wait,
because they understand.

Foreword

In the spirit of disclosure, I wish to state that Drs. Kennedy Jr., Scorpio, and Coppola have been friends of mine for many years. But even more important, they are colleagues who I trust and rely on to provide quality care to all of our children. They have never let me down. With the addition of this new book they have been able to capture their experience and the experiences of the contributors, into words so that we can all learn from them in a timely and succinct manner. This book will provide clinicians with a convenient way of understanding the various conditions that go into pediatric surgery. Even though it is intended to be a handbook, considerable effort has gone into making this book as complete as possible. It should be viewed as a quick and convenient reference and a starting point to the understanding of what it takes to do pediatric surgery.

I sincerely hope that this pocket-sized handbook will become a big part for all physicians who do pediatric surgery. It is to their need for concise and practical information that this book has been written.

Danville, PA, USA
January 2014

Michael Ryan, DO
Janet Weis Children's Hospital

Preface

There are relatively few pediatric surgeons in the world. They tend to be concentrated in tertiary medical centers in metropolitan areas. Many large cities don't have a pediatric surgeon of their own. This is a testament to the kind fact that most children will not need emergency surgery. Pediatric surgery, as a discipline, however is practiced by many more providers, in every hospital that has a child enter their doors. Every time a pediatrician diagnoses a child with abdominal pain, an emergency medicine physician cleans a child's wound, or a nurse chooses a location for an intravenous line in an infant, pediatric surgical care is being delivered. This book is recognition of the fact that every day providers who may not have completed pediatric specialty training need to make rapid decisions in the care of children who could possible need an operation. It is written in a style that allows for rapid assessment and practical selection of diagnostic and therapeutic modalities. It is our hope that it can provide useful guidance across the spectrum from the novice trainee in a clerkship faced with a new consultation in the emergency department to the seasoned family physician making the decision to refer a patient to a pediatric surgeon. We have arranged the material into sections of perioperative resources and specific conditions affecting children across the stages from neonate to adolescent. Every child helped in some small way by this work is a fulfillment of our intentions.

The authors are not responsible for product liability, negligence, injury, or otherwise for any loss, harm, or injury resulting from any material contained in this work. This work contains information relating to general principles of medical care which should not be construed as specific instructions for individual patients. The responsibility for making treatment decisions lies with the providers caring for specific patients. Pharmaceutical product inserts and manufacturers' product information should be reviewed for current information, including contraindications, dosages, and precautions. Users of this text accept these conditions.

Danville, PA, USA

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The editors wish to acknowledge the tireless and compassionate efforts of Ms. Maria Carl in helping us care for children.

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Abbreviations

^{99m}Tc	Technetium-99m
ABC	Airway, breathing, circulation
ABG	Arterial blood gases
ACEI	Angiotensin-converting-enzyme inhibitor
ACIP	Advisory Committee on Immunization Practices
ACS	Acute chest syndrome
ADH	Antidiuretic hormone
AFP	Alpha fetoprotein
AIS	Androgen insensitivity
ALL	Acute lymphoblastic leukemia
ALTEs	Acute life-threatening events
AMD	Actinomycin D
AML	Acute myeloblastic leukemia
Amp	Adenosine monophosphate
ANA	Antinuclear antibodies
Ao	Aorta
AP	Anterior posterior
APTT	Activated partial thromboplastin time
ASA	Aspirin
ATLS	Advanced trauma life support
ATV	All-terrain vehicle
AUA	American Urologic Association
AVN	Avascular necrosis
AXR	Abdominal x-ray
bHCG	Beta human chorionic gonadotropin
BID	Twice daily
BIG	Bone injection gun
BMA	Bone marrow aspiration
BMP	Basic metabolic panel
bp	Blood pressure
BPD	Bronchopulmonary dysplasia

bpm	Beats per minute
BT-UTI	Breakthrough urinary tract infection
BUN	Blood urea nitrogen
BVM	Bag valve mask
C	Celsius
Ca	Calcium
CAH	Congenital adrenal hyperplasia
CAIS	Complete androgen insensitivity
CBC	Complete blood count
CBCD	Complete blood count with differential
CBF	Cerebral blood flow
CCAM	Congenital cystic adenomatoid malformation
CCB	Calcium channel blocker
CDC	Centers for Disease Control
CDH	Congenital diaphragmatic hernia
CFTR	Cystic fibrosis transmembrane conductance regulator
CGRP	Calcitonin gene-related peptide
cGy	Centigray
CHF	Congestive heart failure
CK-MB	Creatinine phosphokinase MB isoenzyme
Cl	Chlorine
CLE	Congenital lobar emphysema
cm	Centimeters
CMV	Cytomegalovirus
CNS	Central nervous system
COG	Children's Oncology Group
COX	Cyclooxygenase
CPAM	Congenital pulmonary airway malformation
CPAP	Continuous positive airway pressure
CPC	Choroid plexus coagulation
CPR	Cardiopulmonary resuscitation
CSF	Cerebrospinal fluid
c-spine	Cervical spine
CT	Computed tomography or chest tube
CXR	Chest x-ray
d	Days
D10W	10 % dextrose in water
D51/2NS	5 % dextrose in 0.45 % normal saline
D5W	5 % dextrose in water
DAI	Diffuse axonal injury
DDAVP	Desmopressin acetate
DHEA	Dehydroepiandrosterone
DIC	Disseminated intravascular coagulopathy
DIOS	Distal intestinal obstruction syndrome
DMSA	Dimercaptosuccinic acid

DNA	Deoxyribonucleic acid
DSD	Disorders of sexual differentiation
DtaP	Diphtheria tetanus pertussis vaccination
DVT	Deep vein thrombosis
E	Esophagus
EBV	Epstein Barr virus
ECF	Extracellular fluid
ECG	Electrocardiogram
ECMO	Extracorporeal membrane oxygenation
ED	Emergency department
EDTA	Ethylenediaminetetraacetic acid
EFS	Event-free survival
EKG	Electrocardiogram
EMS	Emergency medical services
EOM	Extraocular muscles
ERCP	Endoscopic retrograde cholangiopancreatography
ETV	Endoscopic third ventriculostomy
EUS	Endoscopic ultrasound
EVD	External ventricular drainage
F	Fahrenheit
FA	Fluorescein angiography
FAS	Fetal hemoglobin, hemoglobin A and S
FAST	Focused assessment with sonography for trauma
FDA	Food and Drug Administration
FFP	Fresh frozen plasma
FiO2/PaO2	Inspired concentration of oxygen/partial pressure of oxygen
FIP	Focal intestinal perforation
FNA	Fine needle aspiration
FNAB	Fine needle aspiration biopsy
Fr.	French
FS	Full strength
FSH	Follicle stimulating hormone
FSP	Fibrin split products
GBEF	Gallbladder ejection fraction
GCS	Glasgow Coma Scale
GE	Gastroesophageal
GER	Gastroesophageal reflux
GERD	Gastroesophageal reflux disease
GFR	Glomerular filtration rate
GI	Gastrointestinal
gr	Grain
G-Tube	Gastrostomy tube
GU	Genitourinary
H.	Flu Haemophilus influenza
H.	Pylori Helicobacter pylori

H2	Histamine 2
HAART	Highly active antiretroviral therapy
HbCO	Carboxyhemoglobin
HB-MEM	Hepatoblastoma mixed epithelial and mesenchymal phenotype
HCO_3	Bicarbonate
HCP	Hydrocephalus
HGB	Hemoglobin
Hib	Haemophilus influenza type b
HIDA	Hepatobiliary iminodiacetic acid
HIV	Human immunodeficiency virus
HPV	Human papillomavirus
hr	Hours
HS	Hereditary spherocytosis
I&D	Incision and drainage
IA	Innominate artery
ICP	Intracranial pressure
ICU	Intensive Care Unit
IgA	Immunoglobulin A
IgG	Immunoglobulin G
IgM	Immunoglobulin M
IIV	Influenza
IL	Intralesional
IM	Intramuscular
in	Inches
INSS	International Neuroblastoma Staging System
IO	Intraosseous
IPV	Inactivated poliovirus
ITP	Idiopathic thrombocytopenic purpura
IV	Intravenous
IVC	Inferior vena cava
IVF	In-vitro fertilization
IVIG	Intravenous immunoglobulin
JJ	Double J ureteral stent
K	Potassium
kcal	Kilocalorie
KCl	Potassium chloride
KUB	Kidney, ureter, bladder x-ray
LAA	Left aortic arches
lbs	Pounds
LCCA	Left common carotid artery
LDH	Lactate dehydrogenase
LFT	Liver function test
LH	Luteinizing hormone
LP	Lumboperitoneal
LPA	Left pulmonary artery

LSCA	Left subclavian artery
LUQ	Left upper quadrant
LVH	Left ventricular hypertrophy
MAGPI	Meatal advancement and glanuloplasty incorporated
MAP	Mean arterial pressure
MCHC	Mean corpuscular hemoglobin concentration
MCT	Medium-chain triglyceride
MCV	Meningococcal conjugate
MEN	Multiple endocrine neoplasia
MIBG	Metaiobenzylguanidine
MIP	Megameatus intact prepuce
MIS	Minimally-invasive surgery
MMR	Measles/mumps/rubella
MMWR	Morbidity and Mortality Weekly Report
mOsm/L	Miliosmoles/Liter
MPA	Medroxyprogesterone acetate
MRA	Magnetic resonance angiography
MRCP	Magnetic resonance cholangiopancreatography
MRI	Magnetic resonance imaging
MRSA	Methicillin-resistant Staphylococcus aureus
MRU	Magnetic resonance urography
MRV	Magnetic resonance venography
MT	Macrotrabecular
mu	Morphine opioid receptor
MVC	Motor vehicle collision
MyoD	Myogenic differentiation
N	Regional nodes
N_0	Regional nodes not clinically involved
N_1	Regional nodes clinically involved
Na	Sodium
NaCl	Sodium chloride
NAVEL	Nerve artery vein empty lymphatics
NEC	Necrotizing enterocolitis
NFL	Nerve fiber layer
NGT	Nasogastric tube
NICU	Neonatal intensive care unit
NMDA	N-methyl-D-aspartate
NO	Non obstructive
NPO	Nothing by mouth
NS	Nodular sclerosis
NSAIDs	Nonsteroidal anti-inflammatory drug
NSS	Normal saline solution
N_x	Clinical status of regional nodes unknown
NY	New York
O	Obstructive

OF	Osmotic fragility
OGT	Oral gastric tube
OI	Osteogenesis imperfecta
OR	Operating room
PA	Pulmonary artery
PACU	Post-anesthesia care unit
PAIS	Partial androgen insensitivity
PCP	Primary care physician
PCV	Conjugate pneumococcal vaccine
PDA	Patent ductus arteriosus
PET	Positron emission tomography scan
PFA	Protection from abuse
pH	Negative logarithm of hydrogen ion concentration
PICC	Percutaneously inserted central catheter
PO	By mouth
PPI	Proton pump inhibitor
PPN	Peripheral parenteral nutrition
PPV	Patent processus vaginalis
pRBC	Packed red blood cells
PSARP	Posterior sagittal anorectoplasty
PT	Prothrombin time
PT/INR	Prothrombin time/international normalized ratio
PTT	Partial thromboplastin time
q	Every
RAA	Right aortic arches
RBC	Red blood cells
RCCA	Right common carotid artery
RDS	Respiratory distress syndrome
RDW	Red cell distribution width
RES	Reticuloendothelial system
Rh	Rhesus blood factor
RLQ	Right lower quadrant
RMS	Rhabdomyosarcoma
RPA	Right pulmonary artery
RSCA	Right subclavian artery
RTA	Renal tubular acidosis
RUQ	Right upper quadrant
RUTI	Recurrent urinary tract infection
RV	Rotavirus
SCIWORA	Spinal cord injury without radiographic abnormality
SCU	Small cell undifferentiated
Serum	TG Serum triglyceride
SI	Sacroiliac
SOAPME	Suction, oxygen, airway, pharmacy, monitors, extra equipment
SPT	Suprapubic tube

SQ	Subcutaneous
STD	Sexually transmitted disease
SVC	syndrome Superior vena cava syndrome
SWL	Shockwave Lithotripsy
T	Tumor invasiveness
T_1	Confined to anatomic site of origin
T_2	Extension and/or fixation to surrounding tissue
T3	Triiodothyronine
T4	Tetraiodothyronine
TAR	Thrombocytopenia-absent radii
TB	Tuberculosis
TBI	Traumatic brain injury
TBSA	Total body surface area
TCA	Trichloroacetic acid
Tdap	Tetanus-diphtheria and pertussis
TEE	Transesophageal echocardiogram
TEF	Tracheoesophageal fistula
TIP	Tubularized incised plate
TOF	Tetralogy of Fallot
TPN	Total parenteral nutrition
TRALI	Transfusion associated acute lung injury
TSH	Thyroid stimulating hormone
TTP	Thrombotic thrombocytopenic purpura
TX	Texas
UA	Urinalysis
UCF	Urethrocutaneous fistula
UDT	Undescended testis
UG	Urogenital
UGI	Upper gastrointestinal series
UH	Unfavorable histology
UL	Ultralarge
UP	Ureteropelvic
UPJ	Ureteropelvic junction
UPJO	Ureteropelvic junction obstruction
URI	Upper respiratory infection
US	Ultrasound
UTI	Urinary tract infection
UVJ	Ureterovesical junction
VA	Ventriculoatrial
VAC	Vacuum-assisted closure
VAR	Varicella
VATS	Video-assisted thoracoscopy
VCR	Vincristine
VCUG	Voiding cystourethrogram
VF	Ventricular fibrillation

VIP-oma	Vasoactive intestinal peptide tumor
V-IV	Ventriculo-intravenous
VLBW	Very low birth weight
VOC	Vaso-occlusive
VP	Ventriculoperitoneal
VPl	Ventriculopleural
VS	Ventriculosubgaleal
VT	Ventricular tachycardia
VUR	Vesico-ureteral reflux
vWD	von Willebrand disease
vWF	von Willebrand factor
WBC	White blood cell
YAG	Yttrium argon laser

Part I
Perioperative Issues

Fluids and Electrolytes

Alfred P. Kennedy Jr.

Children are not small adults. Babies are not small children. This is particularly true in the management of fluids in electrolytes of the pediatric surgical patient.

1. Newborns:
   (a) Newborns have decreased glomerular filtration rates (GFR) and renal concentrating ability (maximum = 600 mOsm/L).
   (b) The distal convoluting tubule in infants is relatively unresponsive to antidiuretic hormone (ADH) secretion.
   (c) Newborns are unable to excrete excess sodium (Na) and premature infants are considered salt wasters due to the immaturity of the renal tubule. However, both populations are able to excrete relatively large quantities of dilute urine.
   (d) Consequently, there are major shifts in the water content of the intracellular and extracellular compartments and total body water over the first year of life.

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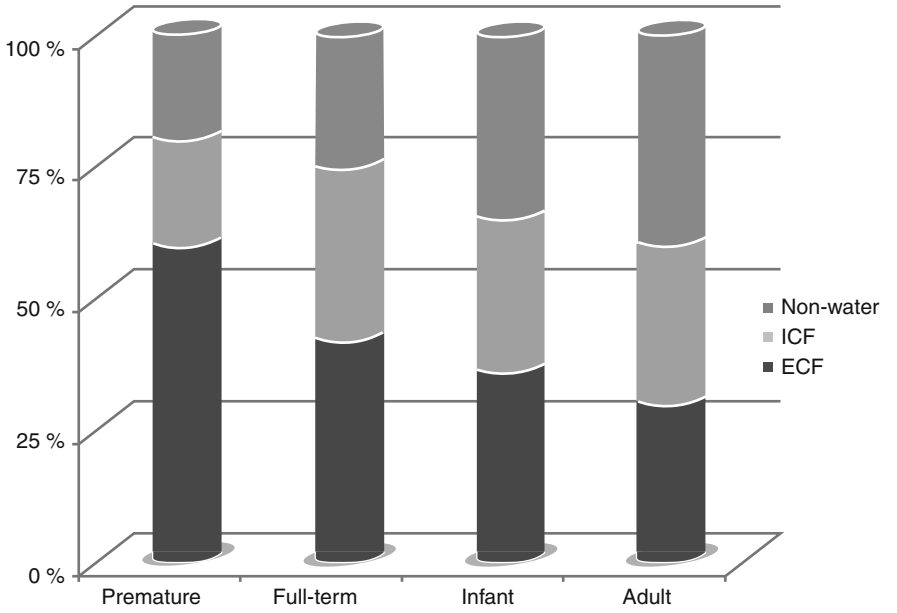

Fig. 1 Relative body fluid compartments by age. As children grow, the relative size of body fluid compartments change. *ECF* extracellular fluid, *ICF* intracellular fluid (*Source*: Christopher Coppola)

2. Urine output:
   (a) Measurement of hourly urine output is essential in critically ill children.
   (b) Catheterization of small infants, especially the male is fraught with complication and should only be undertaken by experienced personnel.
   (c) In order to excrete adequate solute load, the following are approximations of adequate hourly urine output:
      (i) Infant ~ 2 mL/kg.
      (ii) Child ~ 1 mL/kg.
      (iii) Adolescent ~ 0.5 mL/kg.
   (d) These are only approximate volumes and are certainly an underestimate in the critically ill child.
   (e) Other signs of adequate perfusion include capillary refill, skin temperature, heart rate, level of consciousness and fontanel examination in the infant.
3. Fluid requirements: "4 -2 -1 rule".
   (a) 4 mL/kg/h for the first 10 kg of dry body weight.
   (b) 2 mL/kg/h for the next 10 kg of dry body weight.
   (c) 1 mL/kg/h for each additional kg of dry body weight.
   (d) Example: A 20 kg child will need approximately 60 mL/h of maintenance fluid.
   (e) This is only an estimate and may require frequent adjustment depending on urine output, heart rate and other signs of perfusion.

4. Insensible water loss:
   (a) Respiratory: May be mitigated by humidifying inspired gases in those children who require mechanical ventilation.
   (b) Skin.
      (i) Especially significant in premature infants.
      (ii) Mitigated by wrapping extremities, humidified incubators.
5. Common electrolyte anomalies:
6. Sodium (Na): Newborn requires approximately 2 mEq/kg/day.
   (a) Hyponatremia.
      (i) Most common electrolyte abnormality in surgical patients.
      (ii) Usually indicates hypo-osmolality and excess extracellular water.
      (iii) Symptoms are CNS related and include headache, nausea, lethargy, hallucinations and coma.
      (iv) Common Causes in children:
         1. Excess ADH secretion in postoperative period.
         2. Bowel obstruction.
         3. Peritonitis.
         4. Renal impairment.
         5. Hypotonic intravenous fluid administration.
         6. Diuretics.
         7. Mannitol infusion.
      (v) Correction requires treatment of underlying cause and slow adjustment of serum sodium to prevent central pontine myelinolysis.
7. Potassium (K):
   (a) As most potassium is intracellular, serum potassium correlates poorly with the amount of total body potassium.
   (b) Daily requirement is 1–2 mEq/kg.
   (c) Potassium infusion should be undertaken only in the face of normal urine output.
   (d) Hypokalemia:
      (i) Symptoms include muscle weakness, cramps, myalgia, and paralysis.
      (ii) Common causes in children:
         1. Diarrhea.
         2. Diuretics.
         3. Excessive vomiting (e.g. pyloric stenosis).
         4. Metabolic alkalosis.
         5. Prolonged nasogastric tube aspiration.
         6. Inadequate K administration (TPN).

(iii) Treatment requires recognition of underlying cause and most commonly K supplementation either orally or intravenously. When the patient is symptomatic (e.g. arrhythmia) rapid repletion requires placement of a central line. No more than 20 mEq/L should be given via a peripheral IV.

(e) Hyperkalemia:

(i) Symptoms depend on serum levels of potassium and include arrhythmias as well as neuromuscular dysfunction.
(ii) ECG changes include peaked T waves, widened QRS complex and loss of P waves. Wide complex VT, VF or asystole may follow if untreated.
(iii) Common causes in children:

1. Hemolysis (pseudohyperkalemia).
2. Excessive intake.
3. Renal impairment.
4. Succinylcholine.

(iv) Treatment may be emergent in the face of arrhythmias.

1. IV calcium chloride (membrane stabilizer).
2. Sodium polystyrene enema.
3. IV loop diuretics.
4. IV Na HCO_3.
5. Hemodialysis.

8. Calcium (Ca):

(a) Most Ca contained within bone.
(b) Ca levels dependent on serum albumin concentration.
(c) Direct measurement of serum ionized Ca most accurate means of determining ECF Ca.
(d) Hypocalcemia:

(i) Symptoms include perioral paresthesia, carpal pedal spasm, tetany, seizures.
(ii) Chvostek Sign.
(iii) Trousseau Sign.
(iv) ECG changes include prolonged QT interval, peaked t waves and heart block.
(v) Common causes in children include:

1. Hypoalbuminemia.
2. TPN infusion.
3. Sepsis.
4. Pancreatitis.
5. Renal impairment.
6. Neonates are initially hypoparathyroid.

(vi) Treatment requires calcium supplementation.

9. Common Formulas:

   (a) Water deficit = Total body water (see graph) × (1 − serum sodium).
   (b) Plasma osmolality = 2 × Na + glucose (mg %)/18 + BUN (mg %)/2.8.
   (c) FeNa = 100 × (urine Na × plasma Cr) ÷ (urine Cr × plasma Na).

      1. FeNa <1 consistent with pre-renal condition.
      2. FeNa >3 consistent with renal condition.

Table 1 Common IV fluid composition

Fluid	Na (mEq/L)	Cl (mEq/L)	K (mEq/L)	Lactate (mEq/L)	Glucose (g/L)	Osmolarity
Ringer's lactate	130	109	4	28	0	272
Normal saline	154	154	0	0	0	308
½ normal saline	77	77	0	0	0	154
D5W	0	0	0	0	50	252
D10W	0	0	0	0	100	505
3 % saline	513	513	0	0	0	1,026

Nutrition

Alfred P. Kennedy Jr.

It is essential that children receive adequate nutrition for daily metabolic needs, growth, and recovery from illness and operation. The importance of nutrition is compounded by the fact that many children facing surgical illness have a deficit in nutrition caused by illness itself.

1. Caloric requirements: vary by age and must include allowances for stress and growth.
   (a) Total caloric need:
      (i) 0–1-years-old: 90–120 kcal/kg/day.
      (ii) 1–7-years-old: 75–90 kcal/kg/day.
      (iii) 7–12-years-old: 60–75 kcal/kg/day.
      (iv) 12–18-years-old: 30–60 kcal/kg/day.
   (b) Protein calories (15 % of total calories).
      (i) 0–1-years-old: 2–3.5 g/kg/day.
      (ii) 1–7-years-old: 2–2.5 g/kg/day.
      (iii) 7–12-years-old: 2 g/kg/day.
      (iv) 12–18-years-old: 1.5 g/kg/day.
   (c) Lipid Calories: 30–40 % of total calories.
      (i) Begin with 0.5 g/kg/day of lipid and increase by 0.5 g/kg/day to a maximum of 3 g/kg/day.
   (d) Carbohydrate calories: 50 % total calories.
   (e) The Calorie to g Nitrogen ratio should range from >180:1 in newborns to >150:1 in older young adults.

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2. Enteral nutrition:
   (a) Newborns.
      (i) Calorie requirements are 120 kcal/kg/day for newborns and premature children.
      (ii) With adequate nutrition, newborns should gain 20–30 g/day.
      (iii) Breast milk and standard infant formulas have 0.67 kcal/mL or 20 kcal/oz. so they need 180 mL/kg/day to achieve 120 kcal/kg/day.
      (iv) Some newer formulas like Neocate (Nutricia, Gaithersburg, MD) are 1 kcal/cc or 30 kcal/oz.
      (v) Feedings are typically given as boluses but can be given continuously via nasogastric tube or gastrostomy tube. Feedings given via a jejunal tube should always be given continuously.
      (vi) For premature infants and babies with short or damaged gut, such as in necrotizing enterocolitis, a hydrolyzed (partially pre-digested) formula such as Pregestimil (Mead Johnson, Glenview, IL) or Vivonex (Nestle, Vevey, Switzerland) is used. The caloric density of any formula can be increased up to 30 kcal/oz. by adding medium-chain triglyceride (MCT) oil or milk fortifiers if required and is tolerated.
      (vii) Feeding intolerance should be suspected if:
         1. Stool output is >24 mL/kg/day.
         2. Gastric aspirates are >10 mL/kg.
         3. Clinitest (Bayer, Leverkusen, Germany) reveals >2 % reducing substances (sugar) in stool.
         4. Diarrhea.
         5. Distention.
      (viii) Enteral feeding devices: an enterostomy tube that is inadvertently removed should be replaced as soon as possible to avoid spontaneous closure of the site. If the tube was placed more than 2 months prior, the catheter can usually be easily replaced. However, if enteral contents cannot be aspirated from the tube after placement, or if there is any question of improper placement site, tube position should be confirmed by endoscopy or fluoroscopy with contrast instilled into tube. Fill balloon with water. If the tube site has spontaneously closed, an attempt can be made with dilators and Seldinger technique, but an operation to place a new tube may be required.
3. Parenteral nutrition: total parenteral nutrition (TPN) orders are often written with the help of a neonatologist, a pharmacist, or a nutritionist, but the surgeon must have a familiarity with the process. TPN usually starts with a maintenance volume to which nutrients and electrolytes are added to meet the child's nutritional goals.

(a) Glucose:

(i) The initial concentration of glucose is usually 10 %. This is increased daily to a maximum of 25 % for central TPN, a maximum of 12.5 % for peripheral parenteral nutrition (PPN), or until desired calorie supplementation is met.
(ii) The minimum glucose infusion to prevent hypoglycemia in newborns is 4 mg/kg/min. The maximum is 16 mg/kg/min. All children initiated on TPN should have blood glucose measured until stable. Urine is evaluated for glucosuria.

(b) Protein:

(i) Protein requirements vary with age. Newborns need 2–3/5 g/kg/day and have a higher essential amino acid requirement. Protein delivered at >3.5 g/kg/day may predispose to cholestasis. Older children and adolescents require only about 1.5–2 g/kg/day of protein.
(ii) Protein is given as an amino acid solution and is typically begun at a rate of 1 g/kg/day then increased by 1 g/kg/day until the goal is reached.
(iii) Calories derived from protein are usually not counted when calculating total calorie requirements. This is because surgical patients differ slightly from general pediatric patients without wound healing requirements. After an operation, calorie supplementation is provided so that protein stores are not utilized as energy sources but rather as substrates for wound healing.
(iv) Monitor serum BUN, albumin, and total protein.

(c) Fat:

(i) Calories derived from fat should total 25–40 % of non-protein calories and should not exceed 55 % of total calorie intake.
(ii) Fats are given as 10 or 20 % Intralipid (Fresnius Kabi, Bad Homburg, Germany) fat emulsion and started at 1 g/kg/day then advanced to a maximum of 3 g/kg/day.
(iii) 1 mL of 10 % Intralipid = 1.1 kcal.
(iv) 1mL of 20 % Intralipid = 2.0 kcal.

(d) Cholestasis: newborns on TPN often develop cholestasis which can be a major source of morbidity. It is manifest by jaundice and later cirrhosis. Direct hyperbilirubinemia and later transaminasemia are the laboratory indicators of cholestasis. The best treatment is conversion to enteral feeding. When this is impossible, trophic feeds may help. It may help to cycle TPN (infusion for 18–20 h alternating with 4–6 h with infusion off), reduction of protein, elimination of manganese, copper, and vitamins A, D, E, and K. Phenobarbital and ursodiol may help with bile elimination.

Table 1 Parenteral nutrition formulation recommendations

Age	Preterm	Term infants	1–3 years	4–6 years	7–10 years	11–18 years
Energy (kcal/kg/day)	85–105	90–108	75–90	65–80	55–70	30–55
Protein (g/kg/day)	2.5–4	2.5–3.5	1.5–2.5	1.5–2.5	1.5–2.5	0.8–2
Sodium (mEq/kg/day)	2–4	2–4	2–4	2–4	2–4	60–150 mEq/day
Potassium (mEq/kg/day)	2–4	2–4	2–4	2–4	2–4	70–180 mEq/day
Calcium (mg/kg/day)	50–60	20–40	10–20	10–20	10–20	200–800 mg/day
Phosphorus (mg/kg/day)	30–45	30–45	15–40	15–40	15–40	280–900 mg/day
Magnesium (mEq/kg/day)	0.5–1	0.25–1	0.25–0.5	0.25–0.5	0.25–0.5	8–24 mEq/day
Zinc (mcg/kg/day)	325–400	100–250	100	100	50	2–5 mg/day
Copper (mcg/kg/day)	20	20	20	20	5–20	200–300 mcg/day
Manganese (mcg/kg/day)	1	1	1	1	1	40–50 mcg/day
Selenium (mcg/kg/day)	2	2	2	2	1–2	40–60 mcg/day

Pediatric Anesthesiology

Yohannes B. Getachew

There are special anesthetic implications for the newborn and pediatric patient.

1. Anatomy, physiology and pharmacology: "Children are not small adults!" Children's anatomy and physiology evolve with changing age, in terms of organ size and maturity, structure, performance, and proportion of different parts of the body.

   (a) Cardio-pulmonary:

   (i) Intra- to extra-uterine cardiopulmonary transition.
   (ii) Parallel-to-series circuit change in neonates, the three shunts – placenta, ductus arteriosus, and foramen ovale.
   (iii) Transition of the pulmonary system.
   (iv) Changes in pulmonary compliance.
   (v) Airway anatomic differences: Head size and shape, mouth, tongue, epiglottis, location of the larynx, glottis is the narrowest portion of the vairway.
   (vi) Differences in the thoracic cage structure: Mostly cartilaginous at birth but most bones will ossify by middle age.
   (vii) The role of the intercostal and accessory muscles during respiration increases with age.
   (viii) Pulmonary measurements (volumes and capacities change with age).
   (ix) Maturation of components of the cardiovascular system – myocardial compliance improves (constant stroke volume for the first few years of life) and heart rate decreases with age, Components of the blood pressure- constant stroke volume, systemic vascular resistance dependence on autonomic nervous system is less compared to adults.

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(b) CNS:

(i) Brain continues to develop for years after birth, cortical hyperplasia into toddler years.
(ii) Brain tissue's sensitivity to anesthetic drugs, alcohol beverages, etc.
(iii) Effect of anesthesia on the developing brain: A source of controversy due to recent research demonstrating an effect on the brain by multiple anesthetic episodes.
(iv) Fragile intracranial vessels.

(c) Renal system, fluids and electrolytes:

(i) Transition and maturation of the renal system: ability to filter, reabsorb, concentrate urine, and excrete waste changes with growth.
(ii) Body fluid: ratio between extracellular fluid and intracellular fluid, composition of the body fluids and electrolyte treatment.

(d) Pharmacodynamics:

(i) Intravenous induction agents: effects on various organs.
(ii) Narcotics: maturity and concentration of opioid receptors.
(iii) Neuromuscular blocking drugs: effects, sensitivity of monitoring.
(iv) Miscellaneous medications: need and desired or undesired effects.

(e) Pharmacokinetics:

(i) Hepatic enzymes: maturity, glucuronidation.
(ii) Renal excretion of fat-soluble vs. water-soluble drugs.

2. Preoperative evaluation:

(a) "The goals of the preoperative evaluation are to reduce patient risk and morbidity associated with surgery and co-existing diseases, promote efficiency and reduce costs, as well as to prepare the patient medically and psychologically for surgery and anesthesia." (Clinical Anesthesia, Barash).
(b) A thorough history and physical examination to make complete and accurate diagnosis which will dictate whether further labs and intervention are indicated. Early diagnosis and initiation of intervention or preparation always favors better outcome.
(c) Systemic review: pulmonary, cardiovascular system, renal, gastrointestinal-hepatic, endocrine, etc. How much time is there to implement any intervention?
(d) Fluid and electrolyte deficit/management: is there dehydration, fluid overload? Any on-going fluid/electrolyte loss – renal, gastrointestinal, skin, burn? Is the patient receiving the appropriate intravenous fluid in the appropriate amount?
(e) Assess the need for blood products: is an order for products placed? Blood loss from trauma, prior surgery, decreased bone marrow activity (chronic disease, malignancy, etc.)
(f) Optimize blood sugar, blood pressure, urine output, etc.

(g) Full stomach, NPO status (2–3 h for clear liquids, 4 h for breast milk, 6 h for formula and 8 h for solid foods), gastrointestinal obstruction – factors that increase the risk of aspiration.
(h) Antibiotics needed/given?
(i) Communication between the surgeon and anesthesiologist is key to patient safety, continuity of management, anticipation of problems, efficiency and a successful perioperative patient care. The communication should continue throughout the perioperative period.

3. Intra-operative care:

(a) The anesthetic plan is usually made in the preoperative period depending on the type/site of surgery, patient's medical status and maturity, etc.
(b) Some of the factors that affect anesthetic course:

(i) Preexisting medical status.
(ii) Type and site of surgery, use of retractors that can affect organ function (e.g. lung, kidney compression).
(iii) Fluid shift including how much and at what rate blood is being lost, how much is the insensitive water loss from exposed internal organs.
(iv) Intravenous analgesic requirement. The use of local anesthetic agents (especially pre-emptive administration) either by the surgeon or anesthesiologist, as a local infiltration or regional block reduces the intravenous analgesic requirement and reduces the inflammatory response as well as expediting the emergence from anesthesia and the later recovery.
(v) Intra-operative monitoring: blood pressure, heart rate, oxygenation, CO_2 level, temperature, Fluid balance (intravenous fluid administered, urine output, fluid/blood loss), brain function or perfusion monitors, of which the common ones are the bispectral index (BIS) monitor and NIRS (near infrared spectroscopy) which still carry some controversy.

4. Postoperative care:

(a) Even though a last minute change of plan is possible as to where postoperative recovery should take place it is usually determined in the preoperative or early part of the intra-operative periods based on certain criteria.
(b) Most postoperative patients go to either the post anesthesia care unit (PACU) or the intensive care unit (ICU), as determined by several factors. The question that should be addressed is: how intensely should the patient be monitored immediately after surgery? What is anticipated?
(c) Factors that influence port-operative course:

(i) Pre-existing medical condition (cardiac, pulmonary, central nervous system, endocrine, etc.)
(ii) Degree of physiological derangement encountered pre- as well as intra-operatively.

(iii) Type of operation.
(iv) Events that occurred in the operating room, e.g. extensive operation, anesthetic complications (aspiration, malignant hyperthermia, etc.), major fluid shifts including degree of blood loss, unstable vital signs.

(d) When the decision is made to transfer a patient to an ICU the Anesthesia team as well as the surgical team should communicate with the ICU team about the patient's status.
(e) Patients who are going to recover in the PACU will eventually go home, to a floor or the ICU. The decision about where a patient goes after the PACU is usually predetermined again based on the many factors described above. But a patient's status can change.
(f) The criteria used to discharge a patient from a PACU varies little between institutions, but there are three basic factors to be considered:

(i) Wakefulness: how awake is the patient- fully awake, responsive, not responsive, activity level- does the patient move extremities or not?
(ii) Stable vital signs: ability to protect and maintain airway, oxygen saturation level, breaths deep and coughs, verbal response (be it crying or talking), blood pressure and heart rate within age-appropriate range, acceptable pain control, post-operative nausea and vomiting should be under control.
(iii) No evolving surgical complication.

(g) Most patients stay a minimum of 30 min in the PACU and criteria for discharge should be more stringent for those going home. For those being transferred to the ICU, the reason to do so might be lack of improvement or deterioration of one or more of the above criteria.

Procedural Sedation

Richard L. Lambert

1. Procedural sedation:
   (a) The use of procedural sedation outside the operating room to minimize or alleviate pain and/or anxiety has become commonplace in most children's hospitals.
   (b) The approach should be well thought out ahead of time and the person providing sedation should be qualified to handle any situation that may arise during the sedation, including respiratory or cardiac decompensation, anaphylactic reaction and vomiting with aspiration to name a few.
   (c) The process of providing procedural sedation can be divided into the following three components which will be discussed later in detail:
      (i) Pre-sedation.
      (ii) Intra-procedure.
      (iii) Post-sedation.
2. Definitions and levels of sedation:
   (a) The approach to the child and use of medications may vary greatly, i.e. intramuscular midazolam to relieve anxiety for a brief non-painful procedure versus intravenous fentanyl and propofol to achieve deep sedation for a procedure that is both painful and noxious.
   (b) The term "conscious sedation", while still present in the medical vernacular, is a misnomer and should be replaced with "procedural sedation". A child is either conscious and receiving medications to relieve pain and/or anxiety, or they are sedated, with a correspondingly altered mental status (see Tables 1 and 2).

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Table 1 Common definitions used in procedural sedation

Analgesic	Relieves pain by altering perception of nociceptive stimuli
Sedative	Decreases activity, moderates excitement and calms patient
Anxiolytic	Relieves apprehension and fear due to anticipated act or illness
Amnestic	Affects memory; patient is unable to recall events after delivery of drug
Hypnotic	Produces drowsiness and aids in the onset and maintenance of sleep

Table 2 Levels of sedation

	Minimal	Moderate	Deep	General anesthesia
Cognitive function	Normal to partially impaired	Partially to fully impaired	Fully impaired	Fully impaired
Responsiveness	Normal response to verbal commands	Purposeful response to verbal commands and/or light touch	Purposeful response to painful stimulation	No response to stimulation
Airway patency	Normal	May require intervention	Occasionally require intervention	Often require intervention
Spontaneous breathing	Normal	Normal to adequate	May be impaired	Often impaired
Cardiovascular function	Normal	Normal	May be impaired	Occasionally impaired

3. Pre-Sedation Assessment:

   (a) History:

      (i) A general history of each child should be known prior to administering sedative and analgesic medications. Important historical information that may impact the choice of medications administered is summarized in Table 3.

      (ii) NPO guidelines per Table 4.

   (b) Physical Exam:

      (i) A focused physical exam should include assessment of vital signs, cardiopulmonary function and baseline neurological status.

      (ii) Airway.

         1. The pediatric airway is anatomically different than the adult.
         2. A child's tongue is relatively larger compared to the space it occupies in the oropharynx.

            (a) Children with macroglossia, micrognathia or retrognathia may have even more limited space relative to the oropharynx.

Table 3 Pre-sedation assessment

Active medical issues
Current medications
Known allergies
Known adverse reactions to medications
Previous history of sedation or anesthesia and any complications
History of significant snoring or apnea
Known anatomic limitations to opening mouth or moving head and neck
Loose teeth or presence of dental devices
Time since last oral or enteral intake
Recent illnesses (fever, upper respiratory infection, etc.)
Family history of problems with sedation or anesthesia

Table 4 NPO guidelines

Food	Hours of NPO
Clear liquids	2
Breast milk (infants)	4
Formula or light meal	6
Full meal	8

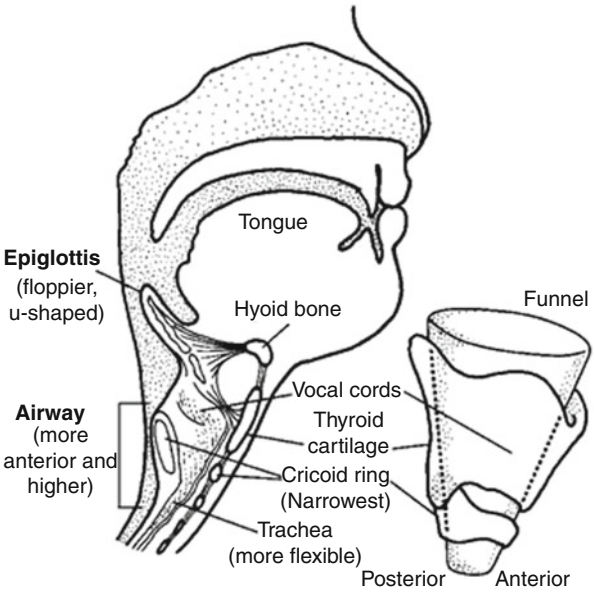

Fig. 1 Pediatric airway, as compared to the adult. The narrowest part of the pediatric airway is the cricoid ring (Included with permission from Susan Gilbert as published at http://www.medartist.com/emergency_pediatrics.html. Downloaded 28 Apr 2013)

Table 5 ASA classification

ASA Classification	
ASA I	Normal healthy patients
ASA II	Patients with mild systemic disease
ASA III	Patients with severe systemic disease that is limiting but not incapacitating
ASA IV	Patients with incapacitating disease which is a constant threat to life
ASA V	Moribund patients not expected to live more than 24 h
ASA VI	A declared brain dead patient whose organs are being removed for donor purposes.

3. Larynx is more anterior and superior.
4. Relative to adult's epiglottis, a child's is longer, more narrow and more floppy.
5. Children younger than 8-years-old have the narrowest part of their upper airway located in the subglottic region at level of cricoid cartilage.
   (a) Any obstruction in this area will significantly impair airflow.
   (b) When supine, care should be taken to ensure the airway remains aligned and excess flexion or extension of the neck does not occur.
6. A roll under the neck and/or shoulders should be utilized to create a "sniffing" position, whereby the horizontal plane of the ears (with patient lying supine) is anterior to the shoulders
7. Mallampati classification may be performed to assist in predicting a difficult intubation but has not been shown to affect risk stratification for children undergoing procedural sedation.

(iii) In addition to the airway assessment, attention should be paid to any physical attributes the child may have that would put him/her at risk for complications, such as:

1. Obesity or failure to thrive.
2. Scoliosis or other skeletal abnormalities.
3. Baseline neurologic impairment.

(iv) A brief examination of the chest with auscultation of lungs and heart should be performed.

(v) After completion of physical exam, assignment of ASA score may be helpful in stratifying the child's relative risk of sedation (Table 5)

4. Intra-procedure:

Table 6 Sedative medications

Drug	Dose	Onset	Duration
Midazolam IV	0.05–0.1 mg/kg	0.5–1 min	30–60 min
Midazolam IM	0.2–0.5 mg/kg	5–15 min	30–120 min
Lorazepam IV	0.03–0.1 mg/kg	2–3 min	2–4 h
Ketamine IV	0.5–2 mg/kg	1–2 min	15–30 min
Ketamine IM	2–5 mg/kg	5–15 min	15–30 min
Propofol IV	0.5–2 mg/kg	0.5–1 min	5–10 min
Dexmedetomidine IV	0.3–1 mcg/kg	10–20 min	30–60 min

(a) Informed consent must be obtained for all sedated procedures.
(b) A time out should occur prior to administering any medications or starting the procedure.

(i) Monitoring and equipment.

1. All children undergoing sedation to facilitate invasive or non-invasive procedures require continuous cardiopulmonary monitoring including pulse oximetry.
2. Capnography, a tool that measures exhaled CO_2, can be very useful in monitoring airway patency and ventilation in the sedated child.
3. The mnemonic SOAPME can help the provider ensure he/she has the appropriate equipment in place prior to starting the procedure.

(a) S (suction).
(b) O (oxygen).
(c) A (airway).
(d) P (pharmacy).
(e) M (monitors).
(f) E (extra equipment).

(ii) Sedative medications (Table 6)

1. There are many sedatives that are currently used for procedural sedation. Some have purely sedative effects while others also provide analgesia. When used in conjunction with a narcotic or another sedative, dosing should be decreased to minimize the chance of over-sedation.
2. Combinations that are often used include versed/fentanyl, versed/ketamine, propofol/fentanyl and propofol/ketamine.
3. Midazolam (Versed) is a benzodiazepine that has sedative, hypnotic, amnestic and anxiolytic properties. It produces its effects by enhancing the inhibitory action of GABA.

Table 7 Analgesic medications

Drug	Dose	Onset	Duration
Morphine IV	0.05–0.1 mg/kg	2–5 min	1–2 h
Morphine IM	0.1–0.2 mg/kg	5–10 min	1–2 h
Fentanyl	0.5–1 mcg/kg	0.5–1 min	15–30 min
Hydromorphone	0.02 mg/kg	10–15 min	4–6 h

(a) Commonly used as intramuscular (IM) injection for the child who either needs a brief procedure where complete stillness is not an issue, or as premed to relieve anxiety for IV placement. Recommended max dosing is 10–15 mg total.
(b) May also be given oral or nasal, but sneezing or coughing will limit its effectiveness and may require redosing.
(c) Flumazenil is a GABA receptor antagonist and can reverse the clinical effects of any benzodiazepine if overdose is suspected.

4. Lorazepam (Ativan) has similar clinical effects as midazolam.

   (a) Rarely used for procedural sedation and cannot be given IM.
   (b) Duration much longer.

5. Ketamine is a PCP derivative that has sedative, amnestic and analgesic effects. It produces its effects by blocking NMDA receptors, leading to inhibition of glutamate mediated transmission.

   (a) Can be used as IM or IV injection alone to achieve both anxiolysis and analgesia, such as is needed for fracture reductions.
   (b) May cause tachycardia and hypertension.
   (c) Can increase airway secretions and cause airway bronchodilation.
   (d) Emergence reaction can occur as hallucinations, confusion and/or agitation. Benzodiazepines in small doses may be given to help minimize this response.

6. Propofol (Diprivan) is an anesthetic medication that has sedative, hypnotic and amnestic effects. It is thought to produce its effect via the GABA pathways.

   (a) Most commonly used sedative medication in procedural sedation.
   (b) Typically used as bolus injections until induction occurs, then as IV infusion to maintain the desired level of sedation.

      (i) Infusion rates for procedural sedation range from 75 to 200 mcg/kg/min.
      (ii) Lidocaine (10 mg in 1 ml) can be left to dwell in PIV for one minute before injection to alleviate local burning sensation.

   (c) Very rapid onset of action.
   (d) May cause hypotension and/or apnea, especially with rapid bolus.

7. Dexmedetomidine (Precedex) is an extremely potent alpha 2 receptor agonist with preference for centrally mediated activity via GABA activity. It produces sedation and analgesia.
   (a) Typically used as a bolus followed by infusion.
   (b) Boluses should be given very slowly and not more frequent than every 10 min.
   (c) Rapid bolus can lead to dramatic bradycardia and hypotension.

(iii) Analgesic medications (Table 7).

1. The majority of analgesics used for procedural sedation are opioid based. Non opioid based analgesics such as non-steroidal anti-inflammatory drugs (NSAIDs) and acetaminophen are typically reserved for use after the procedure.
2. If an adverse effect such as respiratory depression is suspected in the child who received a narcotic, naloxone should be given. This will reverse the analgesic and sedative effect for a brief period of time. Be prepared for the child to awaken and be in obvious pain. Also be prepared to redose as necessary until the symptoms of overdose have resolved.
3. Morphine is the prototypic opiate and acts on the mu receptor to induce analgesia. Higher doses will also cause sedative and hypnotic effects. It can be given PO, PR, SQ, IM or IV. Dosing varies by route and should be verified prior to administration.
   (a) High doses in infants have been associated with prolonged apnea.
   (b) Histamine release may lead to local flushing and pruritus as well as systemic effects like wheezing or hypotension.
4. Fentanyl is a synthetic derivative of meperidine that acts at the mu receptor. It has less hypnotic and sedative effects than morphine.
   (a) As much as 100 times more potent than morphine.
   (b) Has a very rapid onset.
   (c) Should be bolused very slowly (over 2 to 5 min) to minimize the chance of chest wall rigidity and respiratory decompensation.
   (d) May be used in the child with suspected allergy to morphine.
5. Hydromorphone (Dilaudid) is a derivative of morphine that acts at the mu receptor.
   (a) As much as five times more potent than morphine.
   (b) Has the longest duration of the opioids.
   (c) Like fentanyl, causes less pruritus, nausea and dysphoria than morphine.

5. Post-Sedation:

(a) The child should be monitored until they return to their pre-sedation baseline.
(b) A child should not be discharged until certain criteria are met:

    (i) Stable vital signs.
    (ii) Return to baseline mental status.
    (iii) Head control and strength is appropriate to maintain a patent airway.
    (iv) Pain is well controlled.
    (v) Nausea is well controlled.

(c) Special circumstances such as administering a reversal agent or a prolonged adverse event during the sedation may require the child to be monitored longer and/or admitted to the hospital for overnight observation.
(d) Specific instructions should be given to the child's family telling them what to do if the child has any medical problems immediately following the sedation.

6. Adverse events during procedural sedation:

(a) Desaturation may occur from many causes and should be treated immediately. The most common cause is secondary to upper airway obstruction. However, lower airway obstruction, especially in the child with known asthma may occur suddenly and require treatment.

    (i) Check equipment to ensure oxygen is flowing and all tubes are connected and without obstruction.
    (ii) Apnea can be central or obstructive.

        1. Central apnea is usually a result of over-sedation.

            (a) Treatment consists of supporting the child's airway and breathing until the drug has worn off.

                (i) May require bag mask ventilation.

            (b) May consider reversal agent if prolonged.

        2. Obstructive apnea is obvious by the observation that child has abdominal and/or chest movements but no air entry into the lungs.

            (a) Treatment is based on the specific causes that will be discussed below.

    (iii) Upper airway obstruction is the most common cause of desaturation and is usually due to hypotonia of soft palate and/or epiglottis as well as accumulation of secretions.

        1. Audible stridor is likely due to secretions which should immediately be suctioned, using caution not to suction too deep and cause coughing or reflex gagging.
        2. If pharyngeal hypotonia is suspected, reposition the head and neck to ensure midline position with slight extension of neck: Do not overextend.

(a) "Sniffing position" as discussed earlier in the chapter is the desired position to maintain a patent airway.
(b) If midline position is not successful, may try gently rotating child's head to the side while maintaining slight extension of neck.

3. Provide chin lift followed by jaw thrust until obstruction has been relieved.
4. Consider nasal trumpet or oral airway as adjuncts.

(iv) Laryngospasm is a spasm of the glottic structure and can be an emergency if complete; partial obstruction allows some air entry.

1. Typically occurs during induction or emergence but can occur at any time.
2. Stridor may be heard and is often pre-empted by coughing.
3. Child will be attempting to breath but no air entry is occurring.

(a) Prolonged attempts to breath with sudden release of glottic obstruction can cause pulmonary edema.

4. Treatment is multifactorial and stepwise including:

(a) Increase in oxygen supply immediately.
(b) Jaw thrust with suctioning of mouth and oropharynx.
(c) Deepen sedation with bolus of IV sedative.
(d) Pressure on the "laryngospasm notch", an area just anterior to the mastoid process and below the earlobe, can be effective while performing a jaw thrust.
(e) If none of the above are successful within 15–30 s, bag mask ventilation must be applied starting with continuous positive airway pressure (CPAP) and administering positive pressure breaths if necessary.
(f) May require muscle relaxant such as succinylcholine.

(v) Aspiration of oropharyngeal or gastric contents can occur at any time during the procedural sedation.

1. Proactive suctioning of secretions immediately upon hearing airway noises may be preventative.
2. Minor aspiration will not require treatment other than the patient clearing his/her airway with a productive cough.
3. A major aspiration, especially with evident vomitus, will likely require abortion of the procedure with subsequent treatment including:

(a) Increased oxygen supply including bag-valve-mask.
(b) Albuterol treatment may help with airway clearance.
(c) Monitoring after the child has awakened may include hospital admission and observation for signs and symptoms of aspiration pneumonia or pneumonitis.

(vi) Hypotension may occur as a direct result of the sedative medications, as a secondary result of the child's underlying medical condition, or a combination of both. Hypotension may be a result of peripheral vasodilatation, direct myocardial depression or both. Changes in cardiac output from changes in intra-thoracic pressure during sedation may also occur.

1. A drop in BP from baseline is extremely common when using propofol and will be exacerbated by any adjunct medications such as an opioid.
2. Generally responds well to a decrease in the sedative infusion rate and a fluid bolus, i.e. 10–20 mL/kg NSS.

   (a) Re-assessment of the child including palpation of pulses and examination of skin should occur to ensure perfusion is maintained.

(vii) Anaphylaxis or allergic reaction is rare but can be life threatening. Any existing allergies should be identified during the pre-assessment phase.

1. Urticaria with or without redness that was not present prior to the procedure along with any signs or symptoms of respiratory distress, swelling of face and/or tongue should be considered an allergic reaction.
2. Hypotension may not be present but can occur and be very severe due to significant vasodilatation.
3. Treatment consists of the following:

   (a) Abort the procedure and administer 100 % oxygen.
   (b) Stop administering the sedative/analgesic.
   (c) Administer epinephrine 0.01 mg/kg of 1:1,000 concentration IM into the lateral thigh, may need to repeat.
   (d) Give 10–20 mL/kg NSS bolus, may repeat if necessary.
   (e) Consider antihistamines (H1 and H2 blockers).
   (f) Consider IV corticosteroids.
   (g) If not responding immediately, get help and provide cardiopulmonary support until help arrives.

7. Procedural sedation for children can be done safely and with great success. The key is to be thoroughly prepared for any eventuality. Know your equipment, know your medications and know the personnel you are working with. Finally, knowledge about your patient and his/her medical history is a must. Remember, safety first, before, during and after the procedure!

Postoperative Neonatal and Pediatric Critical Care

Edward A. Everett Jr. and Frank A. Maffei

1. Outline:

   (a) Developmental considerations.
   (b) Transition of care from the operative/anesthetic setting to the ICU.
   (c) ICU monitoring of the postoperative patient.
   (d) ICU discharge criteria and transition of care to the ward and home.

2. Developmental considerations.
   Among the most essential tenets of pediatrics is the understanding of the developmental differences that separate the neonate, infant, child and adult. These differences are especially pronounced in the neonate and infant. A brief overview of important physiologic and anatomic differences between the neonate, infant, child and adult will provide a foundation for providing optimal postoperative ICU care.

   (a) Airway.

      (i) There are important anatomic and functional differences between the airway of a small infant and older child. Several characteristics of the neonate and infant airway increase the risk for upper airway obstruction.

         1. A prominent occiput causes flexion of a relatively short neck when lying supine.
         2. The tongue is proportionally larger.
         3. The epiglottis and soft tissues of the upper airway are more compliant.

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4. The larynx is more anterior and cephalad (C3–4 in an infant vs. C4–5 in an adult).
5. Historically the larynx has been thought to be cone shaped and does not assume its cylindrical shape until approximately 8 years of age. However, recent data in neonates using MRI and bronchoscopy reveal a cylindrical larynx with an elliptical cross section and a slightly increased anteroposterior dimension.
6. Incomplete development of the cartilaginous rings throughout the conducting airways increases the potential for airway obstruction along the entirety of the airway.
7. Mucosal edema of the airway is poorly tolerated. The narrowest part of the infant airway is at the cricoid ring in contrast to the vocal cord aperture in older children. Poiseuille's law states that resistance to air flow is inversely proportional to the fourth power of the radius of the airway; consequently, 1 mm of concentric edema at the level of the cricoid ring increases resistance 16 times.
8. Laryngomalacia is a common finding in preterm infants.
9. Infants are obligate nose breathers. Nasal passages may become completely or partially obstructed with mucous, edema or a large nasogastric tube.

(b) Breathing.

(i) The diagnosis and management of postoperative respiratory insufficiency in small infants and neonates requires a clear understanding of the developmental anatomy and physiology of the thorax and lungs.

1. Maintenance of functional residual capacity (FRC) is of vital importance considering that FRC is very close to closing volume (CV) in infants. This is especially true in preterm infants. FRC is the volume of gas within the lung at the end of tidal breathing. It is achieved when the inward forces of the lung balance the outward forces of the chest wall. CV is the volume above residual volume at which the small airways and alveoli begin to collapse. Because CV is very close to FRC, infants have an inherent tendency to have alveolar collapse (atelectasis). This propensity for atelectasis is especially notable during sedation, anaesthesia and in preterm infants at risk for central apnea.
2. Challenges to maintaining functional residual capacity (FRC) in the small infant.

   (a) The chest wall is far more compliant due to a cartilaginous rib cage and poor muscular development both decreasing outward elastic recoil.
   (b) Diminished compliance of the lung due to incomplete conducting airway and alveolar development increasing inward elastic recoil. This is further accentuated in preterm infants by overt or residual surfactant deficiency.

(c) Reduced density of type I muscle fibers when compared to older children and significant respiratory muscle inefficiency at high ventilatory rates increase the risk of early respiratory fatigue.
(d) Unlike adult, infants and neonates lack the large cross-sectional area of the distal airways, and thus, a larger portion of total airway resistance occurs in the peripheral small airways.

3. Compensatory mechanisms during normal tidal breathing targeted at maintaining FRC.

(a) Baseline increased minute ventilation allows short expiratory times resulting in intrinsic positive end expiratory pressure (PEEP).
(b) Laryngeal braking is early glottic closure ceasing expiration prior to reaching closing volume, preventing small airway collapse. An exaggeration of the laryngeal braking phenomenon occurs during states of decreased compliance and is clinically manifested by end expiratory grunting.
(c) Respiratory muscle and diaphragmatic tonicity during exhalation.

4. Central respiratory control of breathing is diminished under anesthesia and is associated with a markedly decreased ventilatory response to hypercarbia. In premature infants, the clearance of anesthetic drugs can be sluggish leading to prolonged recovery and exaggeration of periodic breathing or frank apnea in the postoperative period.

(c) Cardiovascular:

(i) Key events in the transition from the intrauterine to extrauterine cardiovascular function are summarized below:

1. Expansion and oxygenation of the newborn lung causes a gradual reduction in pulmonary vascular resistance (PVR) in the first days after birth. However, PVR does not fall to adult levels until 2–4 months after birth.
2. Postnatal hemodynamic changes cause the flap of the foramen ovale to close against the secundum atrial septum. Concomitantly, rising oxygen tension initiates closure of the ductus arteriosus. The complete anatomic closure of the ductus arteriosus occurs in 98 % of infants by day 4 of age. Persistent patency of the ductus arteriosus, and to a lesser extent the foramen ovale, can lead to clinically significant intracardiac shunting.
3. The neonatal myocardium is composed of poorly organized myocytes that have approximately half the amount of contractile elements (actin, myosin, troponin, and tropomyosin) that are present in older children. Neonates also have a functionally immature sarcoplasmic reticulum that leads to inefficient calcium transport into the myofibrils. Due to this immature myocardium, the infant heart has greater reliance on extracellular calcium. This explains the greater

inotropic response to exogenous calcium and digitalis seen in newborns when compared to adults.

4. Autonomic innervation of the myocardium is also functionally immature. Although the density of beta -adrenergic receptors is high, beta -receptor – adenylase cyclase coupling mechanisms are inefficient. Vagal innervation of the myocardium is complete at birth leading to a state of parasympathetic dominance. This is consistent with the neonate's propensity for bradycardia during stress.
5. The ongoing development of the myocardium results in an infant with a poorly compliant ventricle that has limited responsiveness to increasing preload. The relative inability to increase stroke volume to increase cardiac output leads to a state of chronotropic dependence. Fortunately, infants are generally tolerant of tachycardia during periods of increased metabolic demand. This dependence on heart rate to maintain cardiac output is especially noteworthy when considering the infant's vulnerability to bradycardia secondary to central apnea of prematurity or vagal stimuli. Despite the ongoing development of the cardiovascular system, the infant maintains an impressive baseline cardiac output to meet an already high metabolic demand. Although the absolute cardiac output as measured in L/min is low as compared to an adult, the cardiac output in relation to body size is higher in the neonate, measuring up to 4 L/min/m^2 as compared to 2.5–3.5 L/min/m^2 in the adult.

(d) Central nervous system:

(i) The infant brain is in a rapid phase of cellular development. Although infants possess a similar number of neuronal cells as adults, neurotransmission remains functionally immature due to ongoing axonal and dendritic growth. Glial cells (astrocytes, oligodendrocytes) continue to multiply well into the second year of life; they serve as neuronal support and are important in myelination. Myelination of axons is an ongoing process that begins early in prenatal life and extends to 6 years of age. As myelination advances in a rostrocaudal fashion, axonal impulse transmission becomes more efficient. A major distinction from a gross anatomical basis is the presence of open fontanelles in the small infant.

(ii) The anterior fontanelle remains open until 12–18 months whereas the posterior fontanelle closes by 4 months. A common misconception is that due to the presence of open fontanelles, infants are fully protected from increases in intracranial pressures. This may in part be true in cases of slowly evolving intracranial hypertension (i.e. as may occur in chronic hydrocephalus), but not true during acute increases in intracranial pressure (i.e. as may occur during meningitis or in traumatic brain injury). The dura mater is poorly compliant and does not accommodate acute increases in intracranial pressures; therefore, the infant remains at risk for herniation syndromes during states of increased intracranial pressure.

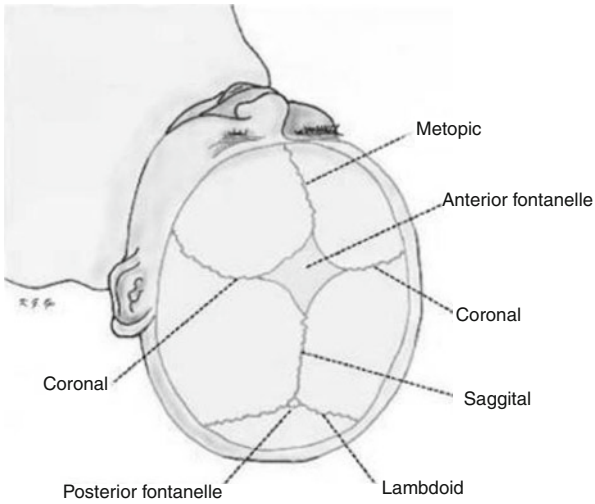

Fig. 1 Cranial sutures and fontanelles. The soft spots between the cranial sutures are the anterior and posterior fontanelles. The fontanelles close by age 18-months-old (*Source*: Adapted from xxjamesxx under Creative Commons Attribution-Share Alike Unported license as published on https://upload.wikimedia.org/wikipedia/commons/5/5f/Sutures_from_top.png. Downloaded 1 Jan 2014)

(f) Temperature control:

(i) Infants and newborns have a large surface area to volume ratio that allows for greater heat loss. They also lack significant amounts of insulating body fat. If proper precautions are not taken, small infants are at profound risk for heat loss from conduction, convection, evaporation, and radiation. Important thermoregulatory mechanisms are immature and inefficient in the preterm and small infant. For example, brown fat which is produced during the third trimester is vital for thermogenesis. This lack of brown fat in infants born preterm impedes the infant's innate ability to mount an adequate response to cold stress. In addition, small infants have a limited or absent ability to shiver. Since infants have immature thermoregulation, close attention to maintaining euthermia should be an essential component in the care of the postoperative patient.

3. Transition of care from the operative /anesthetic setting to the neonatal or pediatric ICU:

(a) The Joint Commission estimates that 80 % of serious medical errors involve miscommunication during care transitions. Miscommunication was the leading root cause of sentinel events reported to the commission between 1995 and 2006. The majority of avoidable adverse events are due to the lack of effective communication. Recent data suggests that standardization of the postoperative handoff process of children undergoing surgery improves outcomes.

(b) Using the acronym SHARE, transitions can become more efficient, informative and safe.

(i) Standardize critical content, including:

1. Providing details of the infant's history to the receiver.
2. Emphasizing key information about the infant when speaking with the receiver.
3. Synthesizing clinical information from separate sources before passing it on to the receiver.

(ii) Hardwire within your system, including:

1. Preparing and stating expectations about how to conduct a successful hand-off.
2. Developing standardized forms, tools and methods, such as checklists.
3. Using a quiet workspace or setting that is conducive to sharing information about a patient.

(iii) Allow opportunities to ask questions, including:

1. Using critical thinking skills and scrutinizing and questioning the data.
2. Sharing and receiving information as an interdisciplinary team.
3. Exchanging contact information in the event there are any additional questions.

(iv) Reinforce quality and measurement, including:

1. Demonstrating leadership commitment to successful hand-offs.
2. Monitoring compliance with use of standardized forms, tools and methods for hand-offs.
3. Using data to determine a systematic approach for improvement.

(v) Educate, including:

1. Teaching staff and standardizing training on how to conduct a hand-off.
2. Providing real-time feedback to staff.

(c) Patient and data hand off:

(i) Patient is transferred from OR to ICU by following personnel:

1. Attending anesthesiologist and or CRNA.
2. Primary surgeon.
3. Respiratory therapy.

(ii) Anesthesia to ICU staff report should include:

1. Nature of airway and how intubation was performed.
2. Anesthetic course.
3. Intraoperative pulmonary function and current respiratory support.
4. Cardiovascular function, rhythm and current hemodynamic support.
5. Intraoperative volume and/or blood product administration.
6. Last sedative/opioid/neuromuscular blocker.

(iii) Surgeon to ICU staff report should include:

1. Review of preoperative lesion.
2. Intraoperative findings.
3. Description of repair.
4. Hemostasis.
5. Expected postoperative course including but not limited to:
   (a) Mechanical ventilation strategy (extubation versus continued support).
   (b) Need for further volume administration.
   (c) Need for further blood product administration.
   (d) Timing of wound closure, when applicable.
   (e) Possible returns to OR (i.e. washouts, closures).
   (f) Specific organ system concerns.
   (g) Perioperative antimicrobial regiment.
6. Current vascular access and drainage tubes.
7. Current medications.
8. Expected lab schedule.

4. ICU monitoring of the postoperative patient:

(a) The postoperative neonatal or pediatric patient requires close and continuous monitoring of vital signs and markers of end organ perfusion. Depending on the preoperative physiologic state, operative intervention and expected postoperative course, monitoring may be noninvasive or invasive. In addition, the intensivist must be capable of increasing the degree of monitoring based on postoperative dynamic changes. Certain physiologic parameters are monitored postoperatively on a continuous basis (i.e. pulse) whereas others are monitored on an interval basis (i.e. capillary refill). The frequency of interval measurements is dependent on the stability of the patient and in very unstable patients interval monitoring may need to be converted to continuous when applicable (i.e. blood pressure).

(b) Standard postoperative monitoring includes:

(i) Cardiac rate and rhythm – continuous.
(ii) Respiratory rate- continuous.
(iii) Pulse oximetry – continuous.
(iv) Capnography – continuous.
(v) Blood pressure via noninvasive oscillometric devices – interval.
(vi) Urine output – continuous via Foley catheter or interval.
(vii) Surgical drain output – continuous or interval.
(viii) Temperature – interval.
(ix) Pain scale – interval.
(x) Blood glucose determinations – interval.
(xi) Weight – daily.

(c) At times, more invasive monitoring may be required and includes:

(i) Central venous waveform and pressure analysis.
(ii) Intra-arterial waveform and pressure analysis.
(iii) Intracranial waveform and pressure analysis.

5. ICU discharge criteria and transition of care to the ward:

(a) Stable gas exchange with decreasing oxygen need and minimal work of breathing.
(b) Stable end organ perfusion as evidenced by normal hemodynamics and mental status, adequate urine output, resolved acidosis and stable intravascular volume status.
(c) Stable electrolytes and acid-base status.
(d) Tolerance of enteral nutrition or plan in place to transition from TPN to enteral nutrition.
(e) No ongoing signs of systemic inflammatory response syndrome (SIRS) or sepsis due to presumed infection.
(f) Stable hemoglobin and coagulation profile.
(g) Surgical drain removed or drains with consistently decreasing output.
(h) Adequate pain control on enteral medication or weaning parenteral medications.

6. Home discharge criteria:

(a) Consistent clinical features of recovery documented (no ongoing SIRS, tolerance of oral intake, pain controlled on enteral medication, wound healing)
(b) Outpatient care plan in place and education / training of the family complete. Family confident in assuming medical care to allow completion of convalescence.
(c) Medication education and administration training.
(d) Discharge equipment ordering and training.
(e) Arrangement of home nursing services if needed.
(f) Follow-up appointment arrangements (subspecialists and primary care).

Disorders of Clotting

Michal A. Miller

1. Evaluation of clotting function.
   (a) Family history, the patient's history of bleeding, including site of bleeding and severity of the abnormal bleeding, age, and prior surgical complications.
   (b) Of note, the possibility of child abuse should be considered in any child with abnormal bleeding and/or bruising. Recurrent bleeding at specific site is generally indicates a local anatomical cause or recurrent local trauma. Bruising over bony prominences such as the pre-tibial areas is common in active children. Large bruises over soft tissue area such the abdomen raise the suspicion of abuse or bleeding disorders.
   (c) Laboratory evaluation:
      (i) Complete blood count (CBC) is the initial laboratory evaluation. A platelet count of >100,000/mcL is usually clinically asymptomatic and frequently does not result in significant intraoperative bleeding. The presence of iron deficiency anemia (microcytic hypochromic anemia) can be a sign of unappreciated chronic bleeding. Pancytopenia signals a need for further evaluation.
      (ii) Coagulations studies: prothrombin time (PT) and partial thromboplastin time (PTT).
      (iii) Platelet Function Assay (PFA) has replaced bleeding time as a screen for platelet function.
      (iv) Fibrin split products (FSP)/Fibrinogen.
      (v) Clotting factor deficiencies other than Factor VIII and Factor IX are rare. A hematologist should be consulted for evaluation of specific factor deficiencies.

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(d) Clinical presentation:

(i) Clotting factor deficiencies: most common presenting signs are bleeding into joints, soft tissues, or muscles. The visible bruising is frequently large, deep palpable, and located on the trunk as well as the extremities. The degree of bruising is out of proportion to the degree of trauma. Frequently the bruising occurs spontaneously without a history of trauma. The increased surgical bleeding is usually more impressive post-operatively than intra-operatively. Small superficial cuts do not tend to bleed excessively.

(ii) Platelet dysfunction and thrombocytopenia: most common presenting signs are petechiae, frequent superficial bruising and mucous membrane bleeding such as epistaxis, menorrhagia, and gingival bleeding. The increased surgical bleeding is usually more impressive intra-operatively than post-operatively. Small superficial cuts tend to bleed excessively. Platelet dysfunction disorders present similar to thrombocytopenia.

(iii) Thrombocytopenia. the degree of bleeding is proportional to the counts. A platelet count of >100,000/mcL is usually not associated with excessive bleeding. 50,000–100,000/mcL results in a moderate increase and 20,000–50,000/mcL in severely increased bleeding complications. Spontaneous bleeding usually does not occur until the count is <20,000/mcL.

2. Hereditary bleeding disorders:

(a) Hemophilia A: factor VIII deficiency. Incidence of hemophilia A in healthy newborns is approximately 1:5,000. It has X-linked inheritance. Approximately one third of severe cases are from spontaneous mutations and have no family history.

(b) Hemophilia B: factor IX deficiency. Incidence of hemophilia B is less common than factor VIII deficiency and comprises approximately 15 % of cases of hemophilia. It has X-linked inheritance.

(c) Hemophilia C: Factor XI deficiency. It is rare and has autosomal recessive inheritance.

(d) Severity of disease:

(i) Severe hemophilia is characterized by a factor level of less than 1 %. Bleeding occurs spontaneously and results in long disability without prophylactic administration of factor VIII.

(ii) Moderate hemophilia is characterized by factor level of 1–5 %. Prolonged bleeding occurs after mild to moderate injuries but does not usually occur spontaneously.

(iii) Mild hemophilia is characterized by factor level of 5–10 %. Mild hemophilia can go undiagnosed until the time of operation.

(e) Von Willebrand disease:

(i) Von Willebrand disease (vWD) is the most common hereditary bleeding disorder. The incidence is reported to be as high as 2 % of the general population. vWD is the result of a deficiency in and/or dysfunction of von Willebrand factor (vWF). vWF is involved in adherence and aggregation of platelets in response to damaged endothelium. It also binds to plasma factor VIII and increases the half-life of plasma Factor VIII. It is synthesized in megakaryocytes and endothelial cells. vWF is stored within the Weibel-Palade bodies in endothelial cells and within the alpha granule in platelets. It is frequently not diagnosed until operation when bleeding occurs in a child who has a history of easy unusual bruising, frequent epistaxis or menorrhagia.

(ii) Classification:

1. Type 1 is a reduction in the amount of vWF. Type 1 is the most common type of vWD and accounts for approximately 75 % of cases. Inheritance is autosomal dominant with variable penetrance. Affected family members of the patient can have different clinical expression. The clinical severity is primarily dependent on the degree of deficiency, but other endogenous as well as exogenous factors can influence the severity of bleeding. The deficiency results in increased bruising, menorrhagia, mucosal bleeding and bleeding during operation.
2. Type 2 is a group of subtypes that consist of dysfunctional vWF activity. This group is uncommon and inheritance can be dominant or recessive.
3. Type 3 is severe reduction or absence of vWF. It is rare. Inheritance is recessive. If VWF is absent or markedly reduced, clinical symptoms are similar to hemophilia A with bleeding into muscles and joints.

3. Acquired pathological bleeding disorders:

(a) Liver disease/injury results in decreased hepatic synthesis of clotting factors and increases fibrinolytic activity.
(b) Kidney disease can lead to loss of coagulation factors and uremia can decrease platelet function.
(c) Gastrointestinal malabsorption results in vitamin K deficiency and deficiency of vitamin K-dependent coagulation factors (II, VII, IX, and X).
(d) Disseminated intravascular coagulation (DIC) results in consumption and activation of multiple coagulation and anticoagulation factors.
(e) Hypersplenism causes moderate thrombocytopenia from sequestration. Conversely, splenectomy will lead to thrombocytosis with platelet counts frequently in the 400,000–600,000/mcL range.
(f) Cyanotic congenital heart disease results in decreased platelet aggregation and shortens platelet survival.

4. Therapy:
   (a) Treatment of hemophilia A and B: factor VIII and IX recombinant factors are widely available for replacement. It is preferable to use the same brand of factor the patient has been using. This limits complications.
   (b) Treatment of vWD:
      (i) Desmopressin (DDAVP) stimulates the release of VWF from storage sites into plasma. Repeated doses deplete the storage pool and thus DDAVP is generally not administered for more than two doses. If a sustained level of vWF is needed for a longer period of time, or if the patient has type 2 or 3 vWD, DDVP will be ineffective and factor replacement is needed. Of note, repeated doses of DDAVP can lead to hyponatremia.
      (ii) vWF replacement: recombinant vWF is not widely available. Currently, anti-hemophilic factor VIII concentrates that also contain vWF are the primary source for vWF replacement.
   (c) Transfusion:
      (i) Platelet Transfusion:
         1. Conservative platelet transfusion indications based on platelet count/scenario:
            (a) <20,000/mcL: immediate prophylactic platelet transfusion.
            (b) <30,000/mcL with minor bleeding (epistaxis, mucosal bleeding menorrhea or persistent "oozing" from incision).
            (c) <50,000/mcL in patients with major trauma, CNS injury, or newly diagnosed cancer .
            (d) <50,000/mcL prior to minor procedures such as a bone marrow aspiration or lumbar puncture.
            (e) <75,000–100,000/mcL prior to a minor operation such as central venous line placement or gastrostomy tube placement.
            (f) <100,000/mcL prior to major operation and neurosurgical operations.
            (g) If the operation is prolonged, additional transfusion may be needed.
         2. Single donor apheresis units are now standard of care. Random donor units can be used if they are all that is available. A transfusion of 10–15 ml/kg (or one unit for an adult-sized child) is expected to increase the platelet count by 50,000–100,000/mcL. Cross-match additional units to transfuse in case bleeding arises intra-operatively.
      (ii) Packed red blood cell (pRBC) transfusion: 10 ml/kg (or one unit for an adult-sized child) is expected to increase the hemoglobin by 1.0–1.5 g/dL. Cross-match additional units to transfuse if bleeding arises intra-operatively.

(iii) The standard of care for type 1 vWD and hemophilia A or B is factor replacement via commercially available factor concentrates. In an emergency when factor replacement is not available, fresh frozen plasma (FFP) can be used. In general, the degree and duration of factor replacement is dependent on the surgical procedure.
(iv) Rare clotting factor deficiencies require FFP.
(v) Platelet dysfunction disorders frequently are not thrombocytopenic but require platelet transfusion to provide functional platelets.
(vi) Special situations: Irradiated leukoreduced blood products should be used for patients with an underlying congenital immunodeficiency, infants <3-months-old, and iatrogenic immunodeficiency such as chemotherapy and transplant patients.
(vii) Patients who have received multiple transfusions can become refractory. Ill patients can consume platelets. Hypersplenic patients will sequester platelets. This occurs in patients with immune thrombocytopenia such as idiopathic thrombocytopenic purpura (ITP).
(viii) FFP contains both pro- and anticoagulation factors. In general, it is the agent of choice to rapidly replace patients with multiple factor deficiencies as occurs in massive hemorrhage or when there is a need for urgent reversal of therapeutic warfarin anticoagulation. FFP is also a source of factors for patient with rare congenital factor deficiencies for which factor concentrates are not yet available.
(ix) Cryoprecipitate most importantly contains fibrinogen. Historically, it was a source of vWF and Factor VIII. Today, patients with hemophilia and vWD should receive commercially available factor concentrates.
(x) Commercially available hemostatic agents:

1. Thrombin.
2. Gelfoam (Pfizer, Inc., New York, NY).
3. Fibrin glue.
4. Surgicel (cellulose polymer, Johnson & Johnson, Piscataway, NJ).
5. QuickClot (Z-Medica Corp., Wallingford, CT).

5. Complications of transfusion:

(a) Hepatitis B transmission: incidence of 1 in 200,000–500,000.
(b) Hepatitis C transmission: incidence of 1 in 500,000.
(c) Human immunodeficiency virus (HIV) 1 and 2: incidence of <1 in one million
(d) Bacterial infection can as high as 1 in 1,000. Usually occurs with platelet transfusion.
(e) Transfusion related acute lung injury (TRALI)
(f) Acute/delayed hemolytic transfusion reactions from antibodies against RBC antigens.
(g) Iron overload.
(h) Fluid overload.

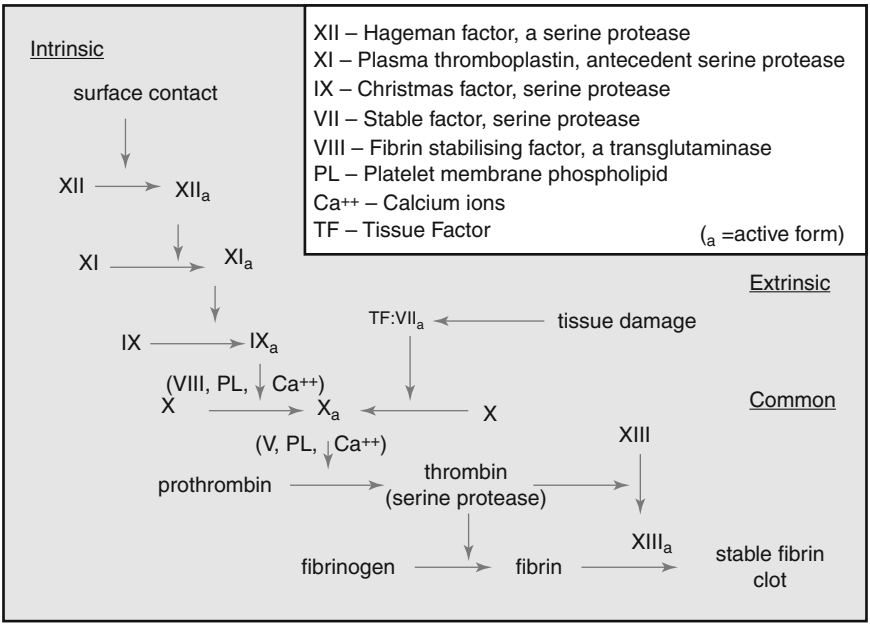

Fig. 1 The clotting cascade: the three pathways that make up the classical blood coagulation pathway are the intrinsic, extrinsic, and common pathways (Adapted from GrahmColm under Creative Commons Attribution-Share Alike 3.0 Unported license as published on https://en.wikipedia.org/wiki/File:Classical_blood_coagulation_pathway.png. Downloaded 28 Dec 2013)

(i) Febrile non-hemolytic transfusion reaction.
(j) Acute onset hemolytic reaction.
(k) Delayed transfusion reaction: two to 14 days after a transfusion.
(l) Alloimmunization.
(m) Sepsis from bacterial contamination.
(n) Allergic reaction.

6. Clotting cascade: (Figure 1).

Suggested Reading

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Part II
Trauma

Epidemiology of Trauma

Bryan S. Walters

Epidemiology of Trauma: Injuries are the leading cause of death for children in the United States. It is the prime reason for years of life lost, and medical costs in the care of children.

1. General:
   (a) 85 % from blunt-force trauma.
   (b) Male > female.
   (c) Leading causes of death by age (excluding non-accidental trauma):
      (i) Less than 1-year-old: Unintentional suffocation.
      (ii) 1–4-years-old: Drowning.
      (iii) 5–18-years-old: Motor vehicle accidents.
   (d) Nonfatal injuries are most commonly falls or being struck by or against an object.
2. Injury by organ system:
   (a) Head trauma:
      (i) A child's brain doubles in size during the first 6 months of life, and reaches 80 % of an adult's brain size by 2-years-old.
      (ii) The three most common mechanisms are:
         1. Motor vehicles.
         2. Bicycle crashes.
         3. Falls.

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(iii) Due to anatomic differences in children, the incidence of head injury is higher in the child. Children are five times more likely to have apnea, hypoventilation, and hypoxia from blunt trauma to the brain than they are to have hypovolemia-associated hypotension. Pulmonary failure is common; cardiac failure is uncommon and is usually secondary to pulmonary failure.

(b) Cervical spine trauma:

(i) Ligamentous injuries are more common than fractures.
(ii) Hyperflexion injuries (anterior wedge fractures and posterior disruption) more common than hyperextension (compression of posterior and anterior disruption of longitudinal ligament).
(iii) Upper cervical spine (C1-C3) is the most vulnerable site in children under 8-years-old.
(iv) Symptoms are delayed in up to 25 % of children with cervical spine injuries.
(v) Pseudo-subluxation of C2 on C3 is a normal finding in 40 % of children under 7-years-old, and in 20 % of children up to 16-years-old.
(vi) Children are more likely than adults to sustain Spinal Cord Injury without Radiographic Abnormality (SCIWORA). A normal cervical spine series may be present in two thirds of children with spinal cord injuries.

(c) Thoracic trauma:

(i) Occurs infrequently in children: Approximately 4–8 % of pediatric trauma patients.
(ii) Around 85 % of thoracic trauma cases are due to blunt forces, with motor vehicle accidents as the most common source. Approximately 8 % are falls and 7 % are abuse.
(iii) Pulmonary contusion is the most common thoracic injury in children.

1. More than 80 % of children with pulmonary contusions also have extrathoracic injuries, thus mortality rates for children with pulmonary contusions can be as high as 22 %.

(d) Abdominal Trauma:

(i) Typically blunt-force trauma: 10–15 % of pediatric trauma patients.
(ii) Isolated injuries are most common.
(iii) Splenic injury is the most common isolated injury.
(iv) Liver, pancreas, and spleen injuries usually occur by one of two mechanisms: direct blow or high-energy injuries (motor vehicle, all-terrain vehicle, and fall from a great height).
(v) Multi-system abdominal injuries carry mortality as high as 12 %.

(e) Pelvic trauma:

(i) Typically high-energy injuries.

(ii) Pelvic fractures occur with less frequency in children (1.6 % of all pediatric trauma admissions).

1. Children have a lower mortality from pelvic hemorrhage than adults due to differences in anatomy.
2. Pedestrian struck by motor vehicle is the most common mechanism (58 %).
3. Pelvic ring disruption and avulsion injuries are commonly reported.

(iii) Common associated injuries:

1. Hemorrhage from other injuries.
   (a) Up to 30 % of patients require blood transfusion due to hemorrhage from associated injury.
2. Head injury: Up to 40 %.
3. Chest/abdomen: 15–33 % association.
4. Genitourinary: 11–12 %.
5. Neurologic: Up to 6 %
6. Extremity: 18 %; femur and tibia injuries most common.

Injury Prevention and Safety

Carol A. Hanson

Injuries are the leading cause of death and disability in the US for people between ages of one and 44-years-old, affecting everyone regardless of race, sex, or economic status. As stated by the Centers for Disease Control, more Americans die from violence and injures - such as motor vehicle crashes, falls, or homicides - than from any other cause, including cancer, HIV, or the flu. Some injuries result in temporary or long-term disability. Survivors are often faced with life-long mental, physical, and financial problems. Injury prevention functions as a primary figure in preventing or reducing the severity of bodily injury and reducing the costs of medical care and loss of productivity.

1. Injury prevention is extremely important due to the fact that 70 % of deaths from injury occur prior to arrival of Emergency Medical Services (EMS).
2. The American College of Surgeons Committee on Trauma describes general principles of injury prevention using the concept of the 4 "E"s.

   (a) Education: educational strategies can be conducted in a variety of ways from bicycle and helmet safety for children to distracted driving awareness for adolescents or violence prevention/conflict resolution for urban populations. Effective programs must be relevant and meaningful to the population targeted.
   (b) Enforcement/enactment: these are opportunities that can be included in legislation for the protection of the population. Such examples include seat belt and car seat regulations, gun safety and wheeled vehicle regulations, railroad crossing and school bus loading and unloading regulations. Although legislative strategies are more time consuming, they can have a greater impact on long term safety concerns than providing educational information to the public alone.
   (c) Engineering: this aspect provides an effective method to reduce the impact of the energy transmission to the victim by structural design of the physical

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equipment or environment. For example, improved design of automobile interiors and restraint systems or development of child safety products can limit the effect of the injury event.

(d) Economic incentives and penalties: in situations where cost creates a barrier to voluntary participation or purchase of the safety improvements, incentives and penalties may motivate the public to provide their loved with greater safety.

3. Injury prevention is most successful with a multi-organization collaborative effort using a public health approach to evaluate the problem, create an action plan, provide for implementation and analyze outcomes and effectiveness. Each mechanism of injury should be assessed for safety needs, programs can be evaluated for effectiveness and analyzed for long term results long term results.
4. Common mechanisms of injury:

   (a) Blunt trauma:

      (i) Falls.
      (ii) Motor vehicle.
      (iii) Pedestrian.
      (iv) Bicycle.
      (v) Watercraft.
      (vi) Assault/abuse.
      (vii) Crush injuries.

   (b) Penetrating:

      (i) Gunshot/stabbing.
      (ii) Animal bites.

   (c) Burns.

5. Prevention Tips:

   (a) Play safely: falls on the playground are a common cause of injury. Check to make sure that the surfaces under playground equipment are safe, soft, and well-maintained (wood chips or sand, not grass/dirt).
   (b) Make your home safer: use home safety devices, such as guards on windows that are above ground level, stair gates, and guard rails. Keep all poisons and flammable materials secure.
   (c) Keep sports safe: make sure your child wears protective gear during sports and recreation. For example, helmets should be worn for any wheeled sports activity.
   (d) Motor vehicle injuries are the leading cause of death among children in the United States.

      (i) In the United States during 2009, 1,314 children ages 14-years-old and younger died as occupants in motor vehicle crashes, and approximately 179,000 were injured.
      (ii) Child safety seats reduce the risk of death in passenger cars by 71 % for infants and by 54 % for toddlers one–four-years-old.

(iii) Age appropriate restraints should be used for every trip in an automobile. The use of previously owned or expired car seats is greatly discouraged due to the fatigue plastic experiences as it ages.
(iv) There is strong evidence that child safety seat laws, safety seat distribution and education programs, community-wide education and enforcement campaigns, and incentive-plus-education programs are effective in increasing child safety seat use.
(v) Child passengers younger than 13-years-old of age should never be seated in front of an airbag. The back seat is the safest. Airbags can injure or kill children in a crash that might otherwise have been survivable. Never place a rear-facing car seat in the front seat or in front of an air bag. Place children in the middle of the back seat when possible, because it is the safest spot in the vehicle.

6. Placing children in age and size appropriate car seats and booster seats reduces serious and fatal injuries by more than half.

(a) Know the stages:

(i) Birth through age two-years-old: rear-facing child safety seat. For the best possible protection, infants and children should be kept in a rear-facing child safety seat, in the back seat buckled with the seat's harness, until they reach the upper weight or height limits of their particular seat. The weight and height limits on rear-facing child safety seats can accommodate most children through age two-years-old, check the seat's owner's manual for details.
(ii) Ages two–four-years-old/up to 40 lbs.: forward-facing child safety seat. When children outgrow their rear-facing seats (the weight and height limits on rear-facing car seats can accommodate most children through age two-years-old) they should ride in forward-facing child safety seats, in the back seat buckled with the seat's harness, until they reach the upper weight or height limit of their particular seat (usually around age four-years-old and 40 lb; many newer seats have higher weight limits-check the seat's owner's manual for details).
(iii) Age four-years-old and older: once children outgrow their forward facing five point car restraint system, they should transition to a belt positioning booster seat (until they are AT LEAST 80 lbs, AT LEAST eight-years-old, AND 4 ft 9 in tall). Remember to keep children in the rear seat for the best possible protection.
(iv) Older than eight-years-old, weight more than 80 lbs, and taller than 4 ft 9 in: seat belts. Children should use booster seats until adult seat belts fit them properly. Seat belts fit properly when the lap belt lays across the upper thighs (not the stomach) and the shoulder belt fits across the chest (not the neck). When adult seat belts fit children properly they can use the adult seat belts without booster seats. For the best possible protection keep children in the back seat and use lap-and-shoulder belts.

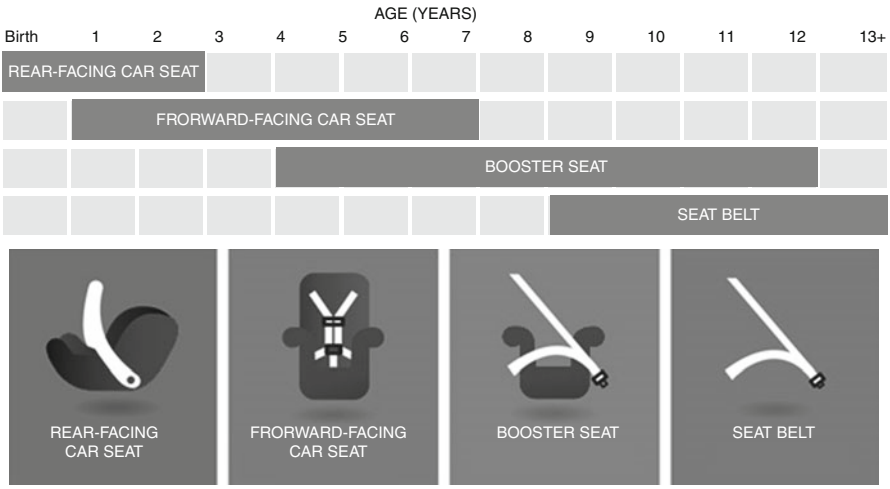

Fig. 1 Appropriate child car seat selection. Children should be seated in a properly installed car seat that is appropriate for their age and size (*Source*: National Highway Traffic Safety administration, Washington, DC. http://www.safercar.gov/parents/RightSeat.htm. Downloaded 8 Jan 2014)

Non-accidental Trauma

Nonette B. Clemens

Child abuse is a major source of injury for children. If not detected, non-accidental trauma is likely to recur and the next event could be fatal. It is the responsibility of any medical provider who detects signs of child abuse to report it and save a life.

1. Types of child abuse:
   (a) Physical.
   (b) Emotional.
   (c) Sexual.
   (d) Neglect.
2. Assessment and history should include:
   (a) History of event, location, time of day, who was present with the child, detail of the injury event, symptoms in the child, response of the caregiver.
   (b) Previous injuries.
   (c) Medical and family history as appropriate to injury: easy bruising, bleeding disorders, recurrent fractures, bone disorders.
3. Features of history and physical suggestive of non-accidental trauma.
   (a) History is inconsistent with mechanism or amount of force required to cause injury.
   (b) History is inconsistent with child's development level.
   (c) History is inconsistent or story changes.
   (d) Delay in seeking medical attention without reasonable explanation.
   (e) Bruising present in a developmentally immobile child "those who don't cruise, rarely bruise".
   (f) Bruising on cheeks, ears or buttocks.

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(g) Multiple bruises of varying types and ages.
(h) Torn upper lip or tongue frenulum in a developmentally immobile child.
(i) Fracture in a developmentally immobile child without plausible explanation.
(j) Fractures of ribs, limb bone metaphyses, scapula, vertebrae, sternum, long bones without osteopenia or bone disorder.
(k) Severe life threatening head injury from reported short fall <5 ft (exception: Epidural hematoma with skull fracture, known to be caused by short fall and can be life threatening).
(l) Acute genital trauma without explanation or history of accidental penetrating injury.

4. Physical exam:

(a) Careful thorough physical exam including all skin areas, ears, genitalia, back and buttocks.
(b) Document any skin markings, location, color, measured size, patterns.
(c) Assess fontanel and perform fundoscopic exam.
(d) Look at upper lip frenulum, palate and tongue frenulum.
(e) Abdominal exam: tenderness and bruising.
(f) Palpate all body areas for pain, swelling, deformity or callus.

5. Laboratory testing:

(a) CBC with differential.
(b) Platelet count.
(c) PT/INR, APTT.
(d) Metabolic/chemistry screens (trauma panel).
(e) Lactate.
(f) Blood gas.
(g) Serum amino acids.
(h) Urine organic acids.
(i) Additional coagulation testing (as recommended by child abuse expert).
(j) Additional metabolic bone disease workup (as recommended by child abuse expert).

6. Diagnostic imaging:

(a) Age less than two-years-old, non-verbal or developmentally immobile should have a full skeletal survey (12+ views), NOT a baby-gram (one–two views).
(b) Age greater than two-years-old and ambulatory: may require x-rays, skeletal survey typically not indicated, should be discussed with child abuse expert for recommendations.
(c) CT head with contrast: indicated with suspected head injury.
(d) CT abdomen/pelvis: indicated with suspected abdominal injury.
(e) MRI, MRV, MRA as recommended by consulting services.
(f) FAST (Focused Abdominal Sonography for Trauma): an abdominal ultrasound is not sufficiently sensitive for abdominal injuries.

7. Additional consults as indicated:
   (a) Pediatric abuse team/pediatric hospitalists: provide guidance for appropriate tests/diagnostics, photographic documentation, court/legal documentation and testimony.
   (b) Ophthalmology: provide evaluation and documentation of retinal hemorrhage/retinoschisis, photographic documentation of retinal injuries.
   (c) Case management/social services: provide guidance and referral to local community services.
   (d) Child protective services/Children and youth services: usually notified by Case management or social services. Appropriate reporting form completion by physician.
   (e) Neurosurgery, orthopaedics, otolaryngology, oral maxillofacial surgery, etc., depending on injury complex.

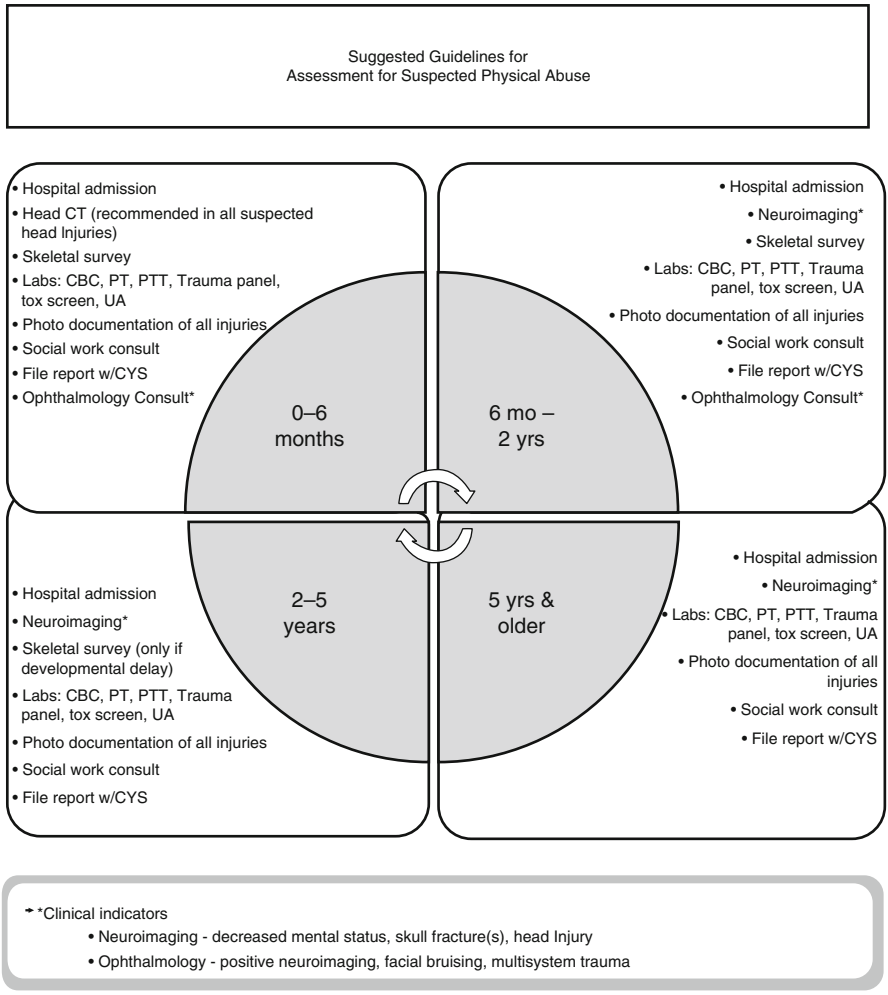

Fig. 1 Suggested guidelines for assessment for suspected physical abuse by age of child (*Source*: Nonette Clemens)

Initial Trauma Assessment

Bryan S. Walters

Assessment of the traumatized child must proceed in a standardized and systemic fashion. Assessment and immediate treatment of life-threatening conditions occur simultaneously.

1. In general, the Advanced Trauma Life Support (ATLS) approach to the evaluation and prioritization of trauma patients is applicable for pediatric patients. A primary survey, followed by the secondary survey, should be completed promptly.
   (a) As one proceeds through these components of trauma resuscitation, thought must be given to the age-specific anatomic and physiologic differences (infant vs. toddler vs. adolescent etc.)
   (b) Ideally, the provider should review the normal vital signs by age prior to the patient's arrival in order to properly assess for the presence of shock upon the patient's arrival.
   (c) Broselow Tape (Armstrong Medical, Lincolnshire, IL) should be laid out on the Emergency Department (ED) bed, preferably prior to the patient's arrival. If given pre-hospital information regarding age and/or weight, the provider should review weight-based dosing and equipment sizes in order to provide expeditious interventions.
2. Pre-hospital personnel and communication:
   (a) Pediatric trauma patients may present via helicopter, ambulance, or "walk-ins" to the ED.
   (b) As in the case of adult trauma, the emphasis in pre-hospital care is:
      (i) Airway.
      (ii) Control of bleeding.

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(iii) Shock management.
(iv) Immobilization.
(v) Minimizing scene time.

(c) Information gathering is crucial to providing appropriate care upon the patient's arrival to the trauma bay. Pre-hospital personnel should relay the mechanism of injury, as this can often suggest the degree of injury, as well as specific injuries for which the child should be evaluated. Other pertinent details should be reported such as time of injury, care received, and patient history.

3. Stabilization of the cervical spine:

(a) As with adults, you should assume that the injured child has a cervical spine injury until proven otherwise.
(b) For infants, there is minimal offset between the occiput and thorax. In addition to airway-positioning considerations, this creates challenges in cervical spine immobilization techniques which vary based on the age, level of cooperation, and habitus of the child.

(i) Unconventional adjuncts are sometimes needed to assist with immobilization of an injured infant or toddler. These may include towel rolls, sheets, pillows, long spine boards, papoose devices, etc.
(ii) The best approach to cervical spine immobilization in the uncooperative pediatric patient is a combination of one or more of the above adjuncts AND a 2-piece rigid cervical collar, such as the Miami J (Ossur Americas, Foothill Ranch, CA) junior sizes for under 12-years-old.

4. "ABCDE" primary survey:

(a) Airway:

(i) Establishing a patent airway is the first priority, always. Failure to maintain a patent airway and subsequent hypoxia is the most common cause of cardiac arrest in children.
(ii) Weight and aged based equipment size references (endotracheal tube, laryngoscope, bag-valve, etc.) are available in the trauma bay, and ideally should be in plain view prior to the patient's arrival.
(iii) A neutral position for the infant and toddler is NOT the "sniffing position". Flexion should be avoided in these patients, and thus the mid-face should be parallel to the spine board. Padding placed beneath the torso will help to compensate for the disproportionate head size and prominent occiput of young children.
(iv) There are distinct and vital differences in the pediatric airway. This includes:

1. Large oropharyngeal soft tissue (tongue, tonsils).
2. Funnel-shaped larynx - allowing secretions to accumulate in the retropharyngeal space.

3. Anterior/cephalad larynx.
4. Shorter trachea (5 cm in infants, 7 cm in 18-month-old) – allowing for easy main-stem bronchus intubation or tube dislodgement.

(b) Breathing:

(i) Again, hypoxia is the most common cause of cardiac arrest in children.
(ii) Normal respiratory rates vary widely based on age. For example, a 1-month-old should have a rate of 30–50 bpm, and a 2-year-old would be 18–30 bpm. Pulse oximetry, capnography, blood gas values, etc. will also assist in monitoring ventilation efforts.
(iii) As with adults, needle decompression, tube thoracostomy, and surgical airways should be considered for the child with inadequate ventilation despite a seemingly patent upper airway.
(iv) Normal spontaneous tidal volumes are 6–8 mL/kg for infants and children; however, the patient will likely require slightly larger volumes (7–10 mL/kg) during assisted ventilation.

(c) Circulation:

(i) Be wary of the pediatric patient's ability to show little sign of impending decompensated shock. Tachycardia and/or diminished skin perfusion may be the only signs of deterioration. Systolic blood pressure alone may not reveal hemorrhage even after a loss of 30 % of circulating blood volume.
(ii) Weight-based fluid resuscitation should be utilized when needed. Blood volume is approximated as 80 mL/kg. Warm isotonic crystalloid should be given as a 20 mL/kg bolus for suspected shock, up to a total of three boluses. Non-responders should be considered for blood transfusion and/or operation.

(d) Disability:

(i) Identify the child's level of consciousness. Almost as a rule in pediatrics, a child who is quiet or stoic in the presence of stress/pain instills a higher level of concern for life-threatening trauma.
(ii) The Glasgow coma scale (GCS) is used in pediatrics just as in adult trauma patients. A pediatric version of the GCS is utilized for younger and/or non-verbal children. The score range is 3–15, same as with adults (Table 1).

(e) Exposure:

(i) Children should be completely exposed and examined, including the removal of all clothing and diaper. Avoid thermal energy loss as the infant/toddler will have a high body surface area to body volume ratio. Warmed blankets should be readily available, and they should be applied as soon as the primary survey is completed.

Table 1 Pediatric and adult Glasgow coma scale

Criteria	Infant	Child	Adult	Score
Eye opening	Spontaneous	Spontaneous	Spontaneous	4
	Verbal stimulus	Verbal stimulus	Verbal stimulus	3
	To pain	To pain	To pain	2
	None	None	None	1
(Criterion total)				(1–4)
Verbal response	Coos, babbles	Oriented, appropriate	Oriented	5
	Irritable cries, consolable	Confused	Confused	4
	Inconsolable, moans	Inappropriate words	Inappropriate words	3
	Inconsolable, agitated	Incomprehensible	Incomprehensible	2
	None	None	None	1
(Criterion total)				(1–5)
Motor response	Spontaneous, purposeful	Obeys command	Obeys command	6
	Withdraw to touch	Localizes pain	Localizes pain	5
	Withdraw to pain	Withdraw to pain	Withdraws to pain	4
	Flexion to pain (decorticate posture)	Flexion to pain (decorticate posture)	Flexion to pain (decorticate posture)	3
	Extension to pain (decerebrate posture)	Extension to pain (decerebrate posture)	Extension to pain (decerebrate posture)	2
	None	None	None	1
(Criterion total)				(1–6)
Total GCS				**3–15**

5. Vital signs and monitoring:

   (a) The Provider should utilize a multi-modal monitoring approach for a child just as they would for an adult. Urine output, vital signs, labs/ABG/i-STAT (Abbott Laboratories, Abbott Park, IL), nasogastric tube, electrocardiogram (ECG), ventilation monitoring, and frequent re-assessment are the mainstay for the monitoring of trauma patients.

   (b) Normal urine output:

      (i) Infants – 2 mL/kg/h.
      (ii) Younger children – 1.5 mL/kg/h.
      (iii) Older children – 1 mL/kg/h.

   (c) Pediatric normal vital signs: (Table 2)

Table 2 Pediatric normal vital signs

Age	Weight (kg)	Heart rate	Systolic BP	Respiratory rate
Preterm	<2.5	120–180	40–60	55–65
Neonate	3	90–170	50–90	40–60
1 month	4	110–180	66–104	30–50
6 months	7	110–180	72–110	25–35
1 year	10	80–160	72–110	20–30
2–3 years	12	90–150	74–124	18–30
4–5 years	17 kg	65–135	79–119	18–30
6–8 years	24 kg	70–115	77–118	18–24
8–10 years	28 kg	65–110	80–118	18–22
10–12 years	35 kg	55–110	84–131	14–22
13–14 years	50 kg	55–110	92–140	12–20
15–18 years	70 kg	55–110	100–140	12–20

6. Life threatening injuries should be promptly identified during the primary survey. Rapid correction of these injuries requires immediate intervention. Some of the injuries which may require immediate intervention while completing the primary survey include:

   (a) Airway obstruction.
   (b) Respiratory arrest.
   (c) Tension pneumothorax.
   (d) Sucking chest wound.
   (e) Flail chest.
   (f) Arrhythmia.
   (g) Ongoing hemorrhage.
   (h) Pericardial tamponade.
   (i) Shock.

7. Secondary survey:

   (a) Obtain further information regarding mechanism of injury.
   (b) AMPLE history.

      (i) Allergies.
      (ii) Medications.
      (iii) Past medical history.
      (iv) Last meal.
      (v) Environment of injury.

   (c) Head to toe evaluation searching for all injuries.
   (d) Includes log-rolling patient to check back.
   (e) Rectal examination to check for blood, tone, and high-riding prostate.
   (f) Examination of all extremities for bony stability, motor, and sensory function.

8. Adjuncts to care:
   (a) IV access:
      (i) Common peripheral sites for IVs in children are the antecubital fossa and saphenous veins, though many other options exist in the extremities, jugular veins (requires: long neck, stable airway, stable c-spine), and scalp (infants). Two large-bore IVs is needed in the case of major trauma.
      (ii) If unsuccessful at gaining peripheral vein access within 90 s or three attempts, then intraosseous (IO) access should be considered, particularly in the unstable patient. This is typically performed using the anterior tibial access.
      (iii) If an IO approach is not an option or is unsuccessful, a femoral venous line may be utilized. The final option, and the least time-efficient, is a venous cut-down (typically the saphenous vein at the ankle).
   (b) Foley catheter:
      (i) Sizes by age are available on the Broselow Tape. Ranges from 5 French to 12 French (Table 3).
      (ii) A relative contraindication exists if blood is present at the urethral meatus, or if there are other reasons to suspect genitourinary injury.
   (c) Laboratory tests:
      (i) Due to lower total blood volumes in children, a minimalist approach should be used when ordering lab work for the pediatric trauma patient.
      (ii) Labs should be requested individually based on suspicion of injury due to signs, symptoms, or mechanism. One should NOT routinely order a full "trauma panel".
      (iii) Trauma panel: CBC, electrolytes, LFT's, amylase/lipase, PT/INR/APTT, ethanol, drugs of abuse, lactic acid, urinalysis, type and hold specimen for blood bank.
9. Radiographic evaluation:
   (a) FAST (Focused Assessment with Sonography for Trauma).
      (i) Rapid ultrasound exam of the RUQ, LUQ, subxiphoid region, and pelvis.
      (ii) This is used routinely for children just as with adults.

Table 3 Foley catheter size estimation

Age group	Foley size (Fr.)
Infant	5–6
Toddler	8
5–11-years-old	10
11–18-years-old	12

(b) Chest and pelvis x-ray.

(i) May be omitted if the child is stable and without apparent injury to these regions, such as in the case of isolated head trauma.
(ii) May be simplified by combining the chest and pelvis with a single "baby-gram" for small children and infants.

(c) CT:

(i) Due to the harmful risks from radiation, a minimalist approach should be employed when ordering CT scans in children.

1. For patients without signs, symptoms, or mechanism for multi-system trauma, order studies by body location rather than panels. Examples:

   (a) Chest versus chest/abdomen/pelvis.
   (b) Head versus head/c-spine.

2. Plan ahead of time for sedation when needed, as younger children and infants may require some degree of sedation for studies to be completed.
3. Obtain CT scans when needed, while balancing the immediate risk of missing an injury against the future risk of a cancer caused by ionizing radiation.

Pathophysiology of Brain Injury

Konstantina A. Svokos and Amir Kershenovich

1. Pediatric Glasgow coma scale (GCS):

	1	2	3	4	5	6
Eyes	Does not open eyes	Opens eyes to pain	Opens eyes to speech	Opens eyes spontaneously	–	–
Verbal	No verbal response	Inconsolable, agitated	Inconsolable, moans	Cries but consolable	Smiles, orients to sounds, interacts	–
Motor	No motor response	Extension to pain (decerebrate)	Abnormal flexion to pain (decorticate)	Withdraws from pain	Withdraws from touch	Moves spontaneously/ purposefully

2. Traumatic brain injury (TBI):
   (a) TBI is the most common cause of pediatric traumatic death. Incidence increased in children <four-years-old and 15–19-years-old.
   (b) Accounts for approximately 37,000 admissions and 2,500 deaths/year in the USA.
   (c) Concussion: mild traumatic brain injury. An alteration of consciousness as a result of non-penetrating traumatic injury to the brain. Findings observed are a vacant stare, delayed verbal or motor responses, difficulty focusing,

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memory deficits, and irritability. CT is negative or significant for mild swelling only.

(d) Severity: minor (GCS 15), mild (GCS 14), moderate (GCS 9–13), severe (GCS 5–8), critical (GCS 3–4).

3. Types of TBI:

(a) Epidural hematoma (EDH): almost always associated with an overlying skull fracture most commonly in temporal and parietal lobes.

(b) Subdural hematoma (SDH):

(i) More common in infants than toddlers and adolescents.

(ii) Large SDH with significant midline shift must be evacuated.

(c) Intracerebral hematoma (ICH):

(i) Due to coup/countercoup injuries from rapid acceleration and deceleration.

(ii) Most commonly in frontal and temporal lobes due to contact of the brain with skull base bony protuberances.

(d) Subarachnoid and Intraventricular hemorrhages (SAH, IVH)

(e) Diffuse multi-lobar injury:

(i) Angular acceleration and deceleration. Shearing injury of axons most commonly at the gray-white matter junction, corpus callosum, and brain stem.

(ii) Can range from minor concussion to severe Diffuse Axonal Injury (DAI) with severe impairment of neurologic function.

(iii) MRI findings: range from normal to deep white matter, corpus callosum or brain stem punctate hemorrhages most evident on gradient-ECHO sequences.

(f) Non-accidental Head injury (NAHI)/Abusive head injury/Shaken-baby syndrome.

(i) Most victims are <3 years old.

(ii) Common acute, subacute, chronic or mixed SDHs and subarachnoid hemorrhages (SAH).

(iii) Best prognosticator factor is the initial patient presentation (GCS).

4. In general for hematomas: small and asymptomatic can be observed; larger ones, especially those causing significant mass effect and or midline shift require surgical evacuation.

5. Skull fractures:

(a) Linear fractures are the most common: most heal on their own and require no treatment.

(b) Closed depressed skull fractures: surgical indications can include cosmetic defects, opened air sinus, compression of dural sinus, presence of dural laceration, compression of eloquent brain area with impaired function.

(c) Open compound depressed fractures may require surgery if grossly contaminated or if a dural laceration is suspected.
(d) Basilar skull fractures: may be associated with carotid artery, middle ear, venous sinus, and cranial nerve injuries. Cerebrospinal fluid (CSF) may leak, due to a dural laceration, via the anterior cranial base through the nose (CSF rhinorrhea) or via the petrous bone through the ear (CSF otorrhea) or through the Eustachian tube into the nose (paradoxical CSF rhinorrhea). These usually resolve spontaneously. No prophylactic antibiotics are indicated to prevent meningitis.

6. Penetrating head injury: gunshot wound to the head account for the majority of penetrating brain injuries. 2/3 of victims die at the scene. Primary injury is caused by injuries to the brain, soft tissues, and skull fractures. The extent of primary injury is related to impact velocity. Secondary injury is caused by cerebral edema and high intracranial pressure. Late complications include: cerebral abscess, traumatic aneurysm, seizures. The best predictor of outcome is the initial GCS and level of consciousness. Patients not in shock and with minimal neurologic function should not be operated on. Goals of surgery is debridement, evacuation of hematomas, removal of accessible bone fragments, hemostasis.
7. Pathophysiology of TBI:

(a) Primary injury: direct glial disruption, axon shearing and vascular injuries.
(b) Secondary injury: due to hypoxia, hypotension, hypovolemia, systemic trauma, electrolyte abnormalities, hyperthermia, infection leading to free radical release. It can double mortality from TBI.

8. Goal of care is to prevent/minimize secondary injury by ensuring adequate cerebral perfusion pressure and oxygenation.

(a) Cerebral auto-regulation: regulates perfusion to the brain over a wide range of mean arterial pressure. Disrupted in TBI leading to large swings in cerebral perfusion pressure (CPP).

(i) CPP = MAP (Mean Arterial Pressure) − ICP (Intracranial Pressure)
(ii) Normal CPP in adults ranges from 50 to 70 mmHg; normal CPP in children is lower than that since they have lower systolic blood pressures, but normal limits have not been well established. ICP should be <20 mmHg. It is normally approximately 15 mmHg in adults and somewhat lower in children and newborns. Based upon a normal ICP <20 mmHg and MAP >60–80 mmHg, depending upon age, normal CPP in children can be calculated to be at least 40–60 mmHg.

(b) Patients with severe TBI (GCS ≤8) are at high risk for intracranial hypertension. The combination of severe TBI and an abnormal head CT suggests high likelihood of raised ICP. Lines of evidence support the use of ICP monitoring in children with severe TBI. Evidence reviewed in the adult guidelines mirrors that for pediatric patients. By contrast, ICP monitoring is not routinely used in children with mild or moderate TBI. Treating physi-

cians may rarely choose to use a monitor in conscious children at high risk of neurologic deterioration due to traumatic lesions or in children whose serial exams are precluded by sedation, anesthesia or pharmacologic paralysis.

(c) Types of ICP monitoring:

(i) Intraventricular catheter (IVC): connected to an external pressure transducer.

1. Advantages: low cost and it allows therapeutic CSF drainage.
2. Disadvantages: may be difficult to insert into distorted/compressed ventricles, it can be a high maintenance system.

(ii) Intraparenchymal monitor: similar to IVC but more expensive. Subject to measurement drift.

(iii) Less accurate:

1. Subarachnoid screw: at high ICP, the surface of the brain may occlude the lumen and give false readings.
2. Subdural.
3. Epidural.
4. In infants, an open anterior fontanelle can be palpated for tension/concavity and convexity to estimate ICP.

9. Measures to decrease intracranial hypertension (IC-HTN)

(a) The goals of therapy are to minimize ICP elevation and maintain adequate cerebral perfusion pressure. General measures include rapid treatment of hypoxia, hypercarbia and hypotension, elevation of the head to 30°, aggressive treatment of fever with antipyretics and cooling blankets because hyperpyrexia increases cerebral metabolism and increases cerebral blood flow increasing ICP. Seizure prophylaxis with anticonvulsants is employed in patients at high risk for seizures. It is also imperative to avoid high positive pressures and end expiratory pressures because they increase intrathoracic pressure and impede venous drainage. Adequate sedation allows controlled ventilation. Neuromuscular blockade may be used if ICP remains elevated despite sedation.

(b) Specific medical measures include:

(i) Mannitol: acts as an osmotic gradient between plasma and parenchyma resulting in net reduction of brain water content. It is indicated in acute herniation, refractory ICP. Goal is to keep serum osmolarity 300–319 mOsm/L.

(ii) Hypertonic saline also acts as an osmotic gradient that reduces brain water content. The optimal form and dose have not been identified. The most common form administered is 3 % saline administered at rates of 0.1–1 mL/kg/h.

(iii) Hyperventilation to a PCO2 30–35 mmHg causes vasoconstriction and decrease in ICP. However, aggressive hyperventilation may decrease cerebral blow flow enough to cause ischemia.
(iv) An intracranial drain can remove CSF and monitor ICP. As ICP increases, the compliance of the brain decreases and small changes in volume can reduce ICP.
(v) Barbiturate coma is used to treat only refractory ICP. Phenobarbital decreases the cerebral metabolic rate which reduces CBF and ICP.

10. Second tier therapy for persistent IC-HTN.

(a) A decompressive craniectomy is a treatment option for control of refractory ICP when conservative therapies have failed. A portion of the calvaria is removed and/or large areas of contused hemorrhagic brain.
(b) According to the 2012 Guidelines for the Acute Management of Severe TBI in Infants, Children and Adolescents, Level II and III evidence suggest:

(i) A CPP threshold 40–50 mmHg may be considered.
(ii) Hypertonic saline 3 % saline should be considered for the treatment of severe TBI with intracranial hypertension.

1. Acute bolus dose: 6.5–10 ml/kg.
2. Continuous infusion of 3 % saline: 0.1–1.0 ml/kg of body weight per hour.
3. Goal is to maintain ICP <20 mmHg and serum osmolarity <360 mOsm/L.
4. Although mannitol is commonly used in the management of TBI, no studies using their inclusion criteria were identified. Traditionally, mannitol has been used in intermittent doses of 0.25–1 g/kg every 4–6 h as needed to control ICP while maintain euvolemia with isotonic fluid.

(iii) Level II data suggests that moderate hypothermia (32–33 C) early after severe TBI should be avoided. Level III evidence suggests that moderate hypothermia beginning early after severe TBI for 48 h may be considered.
(iv) Prophylactic severe hyperventilation to a $PaCO_2$ <30 mmHg should be avoided.
(v) The use of corticosteroids does not improve outcome or reduce ICP.
(vi) Prophylactic treatment with phenytoin may reduce posttraumatic seizures in severe TBI.

11. In addition, serum and urine electrolytes must be balanced in TBI patients. The proper diagnosis is crucial. The most common abnormalities are:

(a) Syndrome of Inappropriate Antidiuretic Hormone (SIADH): treated with fluid restriction to avoid hyponatremia. Demeclocycline is used when fluid restriction does not suffice.

(b) Central diabetes insipidus (DI): treated with cautious fluid replacement and vasopressin
(c) Cerebral salt wasting (CSW): hypovolemia with high urinary sodium treated with saline infusion. Mineralocorticoids are used in cases resistant to therapy.

12. Finally, hyperglycemia is associated with poor outcomes. Tight glycemic control is paramount and associated with improved morbidity and mortality.

Recommended further reading

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Algorithm of Treatment for Head Trauma

Bryan S. Walters

1. Evaluation:
   (a) History of event:
      (i) Height of fall.
         1. 10 feet, 2 stories, 12 steps, mother's arms, etc.
      (ii) Speed of vehicle (MVC/ATV).
      (iii) Rate/method of deceleration.
         1. Concrete wall or floor, trees, carpet, fist, etc.
      (iv) Protective gear.
         1. Helmet, seat belt, car seat, etc.
      (v) Down time prior to being found.
      (vi) Condition and care received pre-hospital.
   (b) History of neurologic function:
      (i) Loss of consciousness.
      (ii) Vomiting.
      (iii) Altered mental status.
      (iv) Incontinent.
      (v) Serial Glasgow Coma Score if available.
      (vi) Seizure.
      (vii) Pre-existing neurologic deficit (congenital).

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(c) Mental status examination

(i) Baseline vs. time of injury vs. now.

(d) Pupillary examination, plus

(i) Extraocular movements.
(ii) Fundoscopic exam.

(e) Neurologic Examination

(i) Glasgow Coma Scale:

1. Mild (GCS 13–15).
2. Moderate (GCS 9–12).
3. Severe (GCS <9).

(ii) Brainstem reflexes (corneal and gag).
(iii) Deep tendon reflex.
(iv) Signs of herniation:

1. Third cranial nerve palsy, followed by hemiplegia.
2. Progressive change in respiration, pupil size, vestibuloocular reflexes, and posture.
3. Cushing's triad:

(a) Hypertension, bradycardia, and slow/irregular respirations.

2. Radiographic imaging:

(a) CT.

(i) Preferred initial imaging modality to identify acute/subacute hemorrhage, edema, herniation, fractures, and hypoxic-ischemic injury.
(ii) A repeat CT scan often shows evolving hematoma and/or secondary injury due to edema.

(b) MRI:

(i) More sensitive for DAI.

(c) Angiography:

(i) Suspected vascular injuries of the neck, face, or head may require computed tomographic angiography.
(ii) Most commonly applied to cases of penetrating trauma, but sometimes indicated for blunt or high-energy (shearing) injuries as well.

3. Treatment:

(a) Algorithm for treatment of child with head injury.

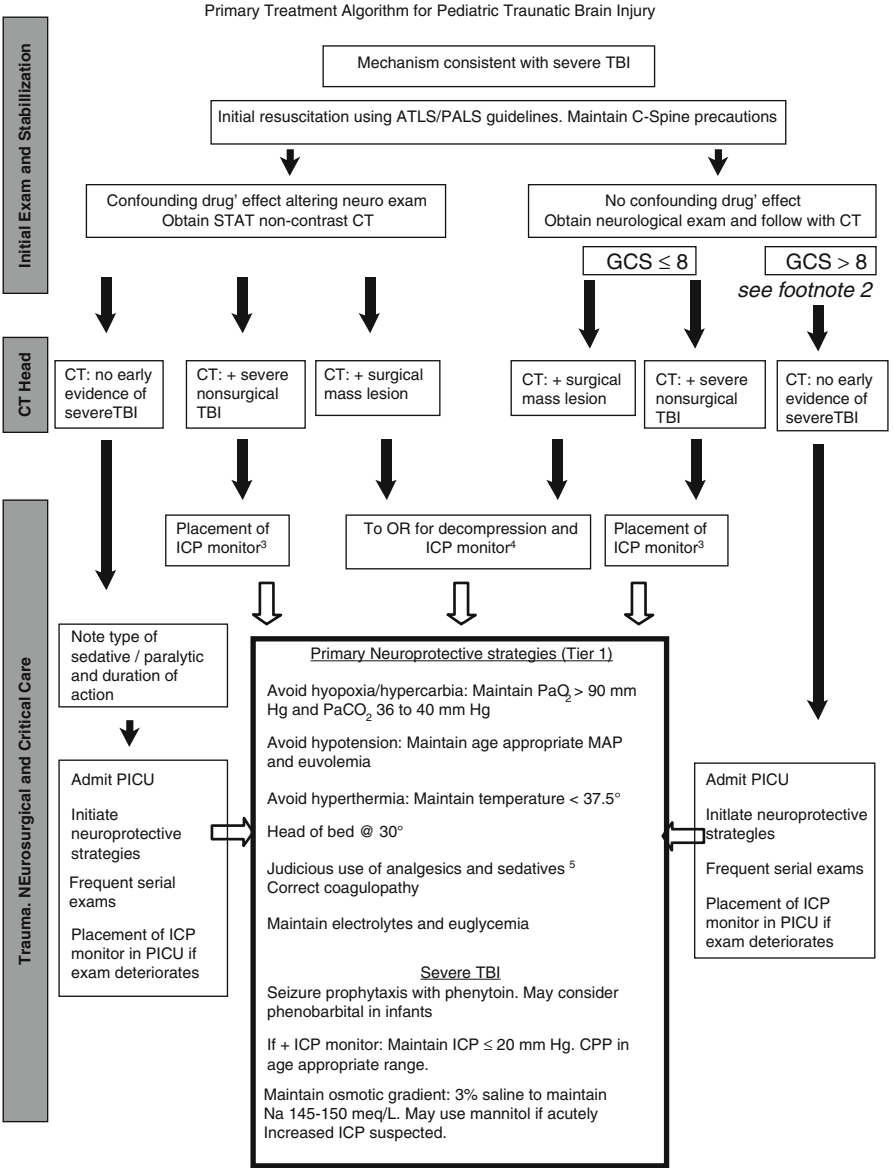
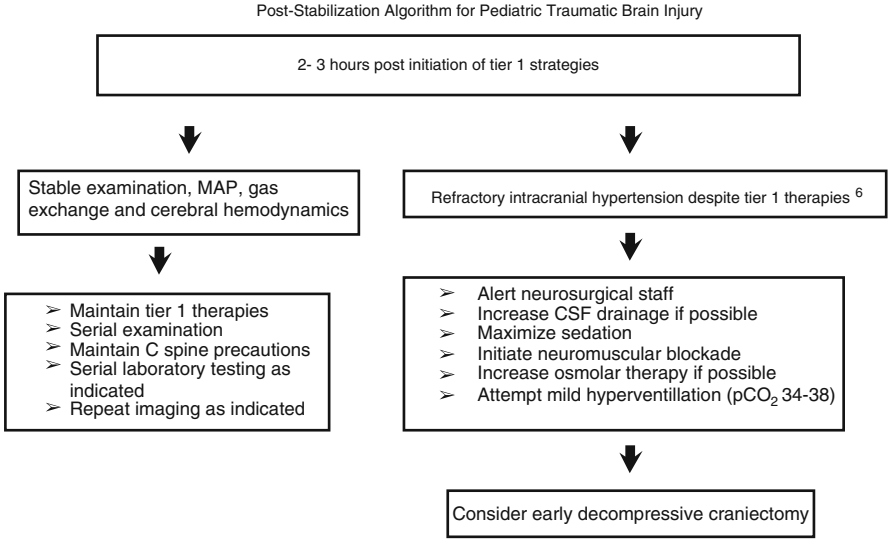


Fig. 1 Primary treatment algorithm for pediatric traumatic brain injury (*Source*: Trauma treatment protocols of the Janet Weis Children's Hospital, Danville, PA)



1. Drugs that may alter neurologic examination include intubation facilitating drugs (sedatives, paralytics) or illcit drugs (ETOH, barbituates, opioids, amphetamines, hallucinogenics etc.)
2. Children with initial GCS > 8 and CT evidence of surgical mass lesion will follow operative pathway. Children with initial GCS > 8 and evidence of nonsurgical TBI may have ICP monitoring discretion of neurosurgery.
3. Type of ICP monitor is at NS discretion, however when possible EVD is preferred. Placement can occur in ED/OR/PICU
4. An ICP monitor may not be necessary after successful evacuation of a space occupying hematoma (i.e. epidural hematoma). Placement in these cases is at the discretion of the neurosurgeon.
5. Sedation regimen dependent upon individual patient and attending discretion.
   If ICP monitor in place and serial awakening unwarranted, combination of benzodiazepine (versed) and opioid (fentanyl) is acceptable. In child with GCS > 8 and no ICP monitoring, short acting and rapidly dissipating agent is desirable (reminfentanil) to allow serial exams. Continuous propofol use is restricted in the PICU. Its use is limited to short term use in adolescents.
6. Refractory intracranial hypertension: ICP consistently > 20 mm Hg for 5 hours or 30 mm Hg for 30 minutes despite tier 1 therapies
7. If using 3% NaCl: 0.1 to 1 mL/kg/h and prn boluses to a maximum serum osmolality of 360 mOsm/L. If using mannitol: 0.25 to 1 gm/kg to a maximum serum osmolality of 320 mOsm/L.

Fig. 2 Post-stabilization algorithm for pediatric traumatic brain injury (*Source*: Trauma treatment protocols of the Janet Weis Children's Hospital, Danville, PA)

(b) Monitoring devices:

(i) Often recommended for children found to have an abnormal CT with a GCS <8.

(ii) Risks:

1. Infection from prolonged use.
2. Hemorrhage from traumatic placement.
3. Excess CSF drainage.
4. Monitor malfunction.

(c) Types:

(i) Bolt (Camino ICP Bolt).

1. Intraparenchymal or subdural.
2. Advantages:

(a) Easier placement.
(b) Lower risk of infection and hemorrhage.

3. Disadvantages:

(a) Inability to drain CSF.
(b) Potential loss of accuracy over several days.

(ii) Ventriculostomy and drain.

1. Intraventricular monitor/drain.
2. Advantages:

(a) Greater accuracy.
(b) Simple measurement.
(c) Ability to drain/sample CSF.

(i) CSF should be removed in very low volumes and slowly. Passive gravitational drainage typically is best.

3. Disadvantages:

(a) Higher rates of infection (up to 20 %).
(b) Higher rates of hemorrhage during placement (approximately 2 %).

(d) Craniotomy and evacuation of hematoma.

(i) Indicated for patients with severe head injury who typically have some or all of the following characteristics:

1. Neurologic deficit.
2. Large hematoma volume (on CT scan).
3. Significant midline shift (generally 5 mm or greater).

(e) Decompressive craniectomy.

(i) Skull is removed to reduce ICP.
(ii) Often done in combination with an evacuation of hematoma.
(iii) Craniectomy alone may reduce ICP 15 %, and with opening of dura may reduce ICP up to 70 %.

4. Adjunctive care: Things to consider in patients requiring more intensive recovery.

(a) Feeding access:

(i) Orogastric/nasogastric feeding tubes (short term).
(ii) Gastrostomy (long term).

(b) Tracheostomy:

(i) Pharyngeal or airway compromise.

(ii) Prolonged intubations.

(c) Stress ulcer prophylaxis.
(d) Nutrition.
(e) DVT prevention:

(i) Most commonly reserved for children >15-years-old.
(ii) Children <15 years old if:

1. Medical/family history consistent with thrombotic risk plus one of the following.

(a) Intubated.
(b) Central line.
(c) Spinal cord injury or severe TBI.
(d) Complex pelvic fracture.
(e) Lower extremity fracture (non-ambulatory).

(f) Prevention of pressure ulcers.
(g) Prevention of contracture/foot drop.

(i) Occupational and physical therapy.

5. Cognitive evaluation and testing:

(a) Pediatric rehabilitation/speech therapy:

(i) Preschool age or younger OR.
(ii) Comatose children of any age OR.
(iii) All children admitted to the hospital for 5 days or more.

1. In conjunction with pediatric neuropsychology.
2. Serial evaluations and cognitive rehabilitation as needed.

(b) Pediatric neuropsychology:

(i) School age.

6. Disability, disposition, and rehabilitation:

(a) Need for medical equipment and rehab disposition (in-patient vs. out-patient) must be addressed as early as possible.
(b) Common issues to address include:

(i) Mobility restrictions and weight-bearing restrictions.
(ii) Nutritional needs (TPN, G-tube feeds, supplemental feeding, etc.)
(iii) Drain care.
(iv) Central venous line care.
(v) Long-term acute care center placement.

(vi) Out-patient rehab referral.
(vii) Medical equipment.

1. Oxygen and supplies.
2. Feeding supplies.
3. Walkers/wheelchairs/shower chairs/bedside commode.

(viii) Follow up appointments with relevant sub-specialties.
(ix) Medications.

Thoracic Trauma

Bryan S. Walters

1. Blunt etiology:
   (a) Most common mechanism is motor vehicle crash, followed by falls and abuse.
   (b) Common blunt trauma injuries in children include:
      (i) Pulmonary contusions (49 %).
      (ii) Pneumothorax/hemothorax (38 %).
      (iii) Rib fracture (35 %).
2. Penetrating etiology:
   (a) Uncommon.
   (b) Most common mechanism is gunshot wound.
   (c) Common penetrating injuries in children include:
      (i) Pneumothorax/hemothorax (64 %).
      (ii) Pulmonary contusion (14 %).
      (iii) Pulmonary laceration (10 %).
      (iv) Vascular injury (10 %).
3. Specific injuries:
   (a) Hemothorax.
      (i) Primary cause is typically lung laceration or laceration of an intercostal vessel.
      (ii) In symptomatic cases, chest tube placement is urgently needed.

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Table 1 Common chest tube sizes for children

Age	Weight (kg)	Chest tube size (Fr.)
Newborn to 1 year	3–5	10–12
≥1–2 years	6–9	12–16
3–4 years	10–11	16–20
	12–14	20–22
5–7 years	15–18	22–24
	19–22	24–28
8–11 years	23–30	28–32
≥12 years	>30	32–42

1. The fifth intercostal space along the midaxillary line is the ideal site of insertion. Tunneling over the rib above the skin incision prior to entering the chest is especially important in children because of a thinner chest wall.
2. Chest tube should be inserted more caudally and more posteriorly for hemothorax than for a pneumothorax.
3. Chest tube size MUST be adequate to evacuate blood. Common tube sizes (Table 1):

(iii) Operation (thoracotomy) is indicated when:

1. The initial thoracostomy tube output is greater than or equal to 20–30 % of the blood volume.
2. The output is greater than 2–3 mL/kg/h over 6 h.
3. Significant hemorrhage occurs.

(b) Pneumothorax, tension:

(i) In most cases, pneumothorax occurs due to small disruptions of lung parenchyma, associated with small to modest air leaks.
(ii) Increased mobility of the mediastinum in children can create a higher risk for physiologic consequence.
(iii) Needle decompression with large-bore over-the-needle catheters should be used with caution in infants and small children due to the longer needle length entering a thinner chest wall.
(iv) Chest tube placement is the definitive treatment for pneumothorax in most cases.

(c) Rib fracture:

(i) Rib fractures are far less common in children versus adults, and serve as an important marker of injury severity. When present, rib fractures in children are associated with a mortality rate of 42 %.
(ii) First and second rib fractures in children are associated with cervical spine injury, pulmonary contusion, and injury to intrathoracic vessels.

(iii) Pulmonary toilet is essential in preventing atelectasis and pneumonia. Adequate analgesia is an important adjunct to facilitate pulmonary toilet, and may include epidural or intercostal nerve blocks.

(d) Sucking chest wound:

(i) Often associated with a lung parenchyma injury as well, and should initially be addressed with a "flutter-valve" occlusive dressing in order to prevent tension pneumothorax. This is accomplished by taping only three sides of a four-sided occlusive dressing.

1. Chest tube placement (at a site other than the open chest wound) should then be performed as soon as possible.

(e) Thoracic vascular injury:

(i) The most common (and lethal) thoracic great vessel injury is traumatic aortic disruption. Work-up and surgical repair (when applicable) should not be delayed.

1. Presentation: midcapsular back pain, unexplained hypotension, upper extremity hypertension, bilateral femoral pulse deficits, and large initial chest tube outputs.
2. Radiographic findings (plain x-ray): widened mediastinum, blurring of the aortic knob, deviation of a nasogastric tube, abnormal paraspinous stripe, rightward tracheal deviation, or upward shift of the left main stem bronchus. The thymus in younger patients can sometimes mask these findings.
3. Diagnostic imaging: Options include helical CT (typically preferred), TEE, CT angiography, and aortography.

(f) Cardiac injury:

(i) Myocardial contusion:

1. Most common cardiac injury in children.
2. May resemble myocardial infarction, supraventricular arrhythmia, or ventricular arrhythmia.
3. Often associated with pulmonary contusion and rib fractures.
4. Diagnosis based on:

(a) High index of suspicion.
(b) ECG.
(c) Labs (CK-MB, Troponin).
(d) Angiography and echocardiography (when needed).

5. The incidence of cardiac sequelae in children with cardiac contusion is low; however, consider other injuries that can be associated with high-energy trauma such as valvular dysfunction from papillary muscle or chordae tendineae rupture, myocardial rupture, or pericardial effusion.

(ii) Pericardial tamponade:

1. Etiology: penetrating trauma > blunt trauma.

(a) Diagnosis:

(i) FAST.
(ii) Beck's triad.

1. Venous pressure elevation.
2. Decline in arterial pressure.
3. Muffled heart tones

(iii) Kussmaul's sign.

1. Rise in venous pressure with inspiration when breathing spontaneously.

(b) Treatment: Evacuation is indicated for patients who do not respond to the usual measures of resuscitation for hemorrhagic shock in whom cardiac tamponade is suspected (including patients with pulseless electrical activity of uncertain origin).
(c) Surgical evacuation (pericardiotomy) is definitive; however pericardiocentesis may be performed as a temporizing maneuver.

(iii) Penetrating cardiac injury:

1. Chest injuries at or below the nipple line in children are better classified as a thoracoabdominal injuries, since intra-abdominal injury must be excluded.
2. Open chest wounds must be treated with an occlusive dressing taped on three sides.
3. Triple contrast CT, ultrasound, and laparoscopy can assist in the evaluation of penetrating chest trauma.

(g) Pulmonary injury:

(i) Pulmonary contusion is the most common thoracic injury in children.

1. Signs and symptoms may include external chest wall abrasions, tachypnea, and abnormal breath sounds.
2. Associated rib fractures occur in up to 32 % of cases.

(a) Serial chest x-ray should be obtained during the initial 48 h after injury to identify other intrathoracic injuries.

3. CT scan may help identify the extent of pulmonary contusion, and thus identify those at greater risk for respiratory failure.
4. Treatment involves supplemental oxygen, pain management, pulmonary toilet, and respiratory support when needed.

(ii) Pulmonary laceration:

1. Mortality rate of 55 %.
2. Can present with hemoptysis, respiratory distress, and hypotension.
3. Management:

(a) Initially, chest tube placement to remove blood and air is typically needed. This is often performed urgently.
(b) IV access and blood products should be immediately available due to risk of ongoing hemorrhage after chest tube placement.
(c) Indications for surgery:

(i) Persistent bleeding.
(ii) Major air leak.
(iii) Air embolism.
(iv) Massive hemoptysis.

(h) Tracheobronchial injury:

(i) Signs and symptoms:

1. Shortness of breath.
2. Subcutaneous emphysema.
3. Hemoptysis.
4. Tension pneumothorax with mediastinal shift.
5. Stridor.

(ii) Radiographic findings:

1. Air within soft tissues or surrounding bronchus.
2. Pneumothorax.
3. Hyoid bone elevation.
4. Obstruction of an air-filled bronchus.

(iii) CT may confirm diagnosis.
(iv) Children may have tracheobronchial injury without an associated chest wall injury due to the flexibility of the thoracic cage.
(v) Injury location is often in close proximity to the carina.
(vi) Management:
1. Endotracheal intubation should be performed in patients who have stable airways, preferably in the operating room with bronchoscopic guidance to avoid worsening an injury.
2. Immediate surgical intervention is required.

(i) Esophageal injury:

(i) Signs and symptoms (often subtle and non-specific):

1. Retrosternal pain which can radiate to the neck or shoulders.
2. Tachycardia.
3. Dyspnea.

4. Subcutaneous emphysema.
5. Abdominal guarding.
6. Left pneumothorax or hemothorax without a rib fracture.
7. History of severe blow to lower sternum or epigastrium and is in pain or shock out of proportion to the apparent injury.
8. Particulate matter in the chest tube after blood begins to clear.
9. Mediastinal air on plain x-ray.

(ii) Diagnosis may be confirmed by contrast study or esophagoscopy.
(iii) Rupture from blunt trauma typically caused by a forceful expulsion of gastric content into the esophagus.
(iv) Empyema often develops following mediastinitis and rupture into the pleural space.
(v) Management varies, but usually involves drainage of pleural space and mediastinum with direct repair via thoracotomy.

(j) Diaphragm rupture:

(i) Left > right.
(ii) Often misinterpreted on plain chest x-ray. May resemble a loculated hemopneumothorax, elevated diaphragm, or gastric distention.
(iii) Diagnostic techniques may include observing plain x-ray following nasogastric tube placement or UGI contrast study. Diagnostic laparoscopy and/or thoracoscopy may be needed for indeterminate cases.
(iv) Treatment is typically direct surgical repair of the defect.

Abdominal Trauma

Bryan S. Walters

1. Blunt injury:
   (a) Blunt force is the most common mechanism of trauma in children.
   (b) Attention should be given to the degree of energy involved, such as in the case of a baseball bat injury versus a motor vehicle accident at a high rate of speed.
2. Penetrating injury:
   (a) Penetrating abdominal injuries are most commonly the result of firearm or stabbing injuries.
   (b) Significant intraperitoneal injuries are present in most children who sustain gunshot wounds, and, as a result, abdominal exploration is often needed. Likewise, stab wounds to the abdomen will almost always require exploratory laparotomy.
3. Specific organ injury:
   (a) Spleen:
      (i) The spleen is the most commonly injured abdominal organ in children.
      (ii) The standard of care in treating hemodynamically stable children with splenic injury is observation alone. Nonoperative management is successful in greater than 90 % of children with splenic injury.
      (iii) Indications for operative intervention are generally limited to persistent hypotension, greater than 50 % blood volume replacement, or additional life-threatening injuries.

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(iv) Intravenous access should be obtained via a large-bore cannula, and the patient should, at least initially, be on bed rest and monitored in the pediatric intensive care unit.
(v) In addition to routine cardiorespiratory monitoring, frequent labs should be obtained over the initial 24–48 h to assess for ongoing hemorrhage.

(b) Liver:

(i) The liver is the second most commonly injured abdominal organ. Similar to splenic injury, liver injuries can be managed non-operatively in the majority of hemodynamically stable children.
(ii) Ecchymosis or abrasion over the right upper quadrant may suggest significant injury.
(iii) Eventual complications of liver injury include hemorrhage, bile peritonitis, abscess, and hemobilia.
(iv) Operative intervention is typically required in the case of disruption of the hepatic vein or retrohepatic caval injuries.
(v) Hepatic vessel embolization is sometimes utilized for select patients.

(c) Pancreas:

(i) Injuries:

1. Trauma to pancreas may require operative intervention, especially if the pancreatic duct is transected.
2. Transection of gland usually occurs where gland crosses spine.
3. Late complication after injury is development of pseudocyst

(ii) History may suggest pancreas injury.

1. Seatbelt injury, particularly when only lap belt is used and is positioned too high above the iliac crests.
2. Handlebar injury.
3. Punch or kick to the epigastrium.

(iii) Clinical signs:

1. Upper abdominal tenderness that may radiate to back.
2. Elevated amylase and lipase levels.
3. Computed tomography revealing gland edema, fluid in lesser sac, transaction or hemorrhage of gland.

(iv) Treatment:

1. Transaction of the pancreas is best treated by early distal pancreatectomy with drainage.
2. If pancreatic duct is intact, non-operative treatment with bowel rest and total parenteral nutrition is likely to work best
3. Pseudocyst, if it occurs, should be allowed 6 weeks for the lining to mature, then is drained into the GI tract. Options include endoscopic transgastric drainage and laparoscopic roux-en-Y pseudocyst-jejunostomy.

(d) Kidney:

(i) Injury:

1. Blunt trauma to the abdomen or flank can result in laceration, hematoma, or vascular injury.
2. Penetrating injury to kidney can cause laceration, hemorrhage, and vascular injury.
3. Late effects can be abscess, urinoma, or scar causing hypertension.

(ii) Clinical:

1. Flank tenderness, mass, pain, and ecchymosis.
2. Hematuria (is not present in all renal injuries).
3. Sonography will detect renal laceration, hematoma, and fluid collections.
4. Computed tomography can diagnose and grade renal injury.

(iii) Treatment:

1. Nonoperative management will usually be successful for renal trauma as long as patient remains hemodynamically stable.
2. Ongoing hemorrhage from a kidney may require open exploration.
3. Renal salvage can be attempted, but often nephrectomy is the safest choice.
4. Before removing a kidney, the surgeon must verify that the opposite kidney is present.
5. If available and patient is stable enough to try the procedure, angiographic embolization can be attempted.
6. Patients with urinary extravasation receive antibiotics.
7. Late intervention may be needed for abscess or infected urinoma.

(e) Duodenum:

(i) Duodenal injuries are uncommon; however they have a higher rate of occurrence then the adult trauma population.

(ii) The majority of duodenal injuries present as duodenal hematoma without intra-luminal perforation. Thickening of the duodenal wall is noted on computed tomography scan. These injuries are treated with observation and parenteral nutrition. For select patients, a nasojejunal feeding tube will allow post-duodenal enteral feeding.

(iii) Full-thickness perforation will typically present with extravasation of oral contrast in the right anterior pararenal space. The majority of these injuries are amenable to debridement and primary closure; however more complex approaches are sometimes needed.

4. Scales for grading organ injury:

Organ/Grade of injury	Grade I	Grade II	Grade III	Grade IV	Grade V	Grade VI
Spleen - Hematoma	Subcapsular, <10% surface area	Subcapsular, <10-50% surface; Intraparenchymal, <5cm diameter	Subcapsular, >50% surface or expanding; Intraparenchymal, <5cm or expanding; ruptured			
Spleen - Laceration	<1cm depth	1-3 cm depth	>3 cm depth or involving vessel	Segmental vessel >25% devascularization	Shattered spleen	
Spleen - Vascular					Hilar injury 100% devascularization	
Liver - Hematoma	Subcapsular, <10% surface area	Subcapsular, <10-50% surface; Intraparenchymal, <10cm diameter	Subcapsular, >50% surface or expanding Intraparenchymal > 10cm or expanding; ruptured			
Liver - Laceration	Capsular tear, <1cm depth	1-3 cm depth, <10cm length	>3 cm depth	Disruption 25-75% of lobe or 1-3 Couinad segments in lobe	Disruption >75% of lobe or >3 Couinad segments in lobe	
Liver - Vascular					Injury to cava or hepatic vein	Avulsion
Duodenum - Hematoma	Single portion	>1 portion				
Duodenum - Laceration	Partial thickness	<50% circumference	50-75% circumference 2nd portion	75% circumference 2nd portion, ampulla or CBD	Injury duodenopancreatic complex	
Duodenum - Vascular					Devascularization of duodenum	
Pancreas - Hematoma	Minor contusion	Major contusion				
Pancreas - Laceration	Superficial	Major	Distal transaction or duct injury	Proximal transaction or ampulla injury	Massive disruption of head	
Kidney - Contusion	Hematuia					
Kidney - Hematoma	Subcapsular, nonexpanding	Confined to retroperitoneum				
Kidney - Laceration		<1 cm depth	>1 cm depth	Through cortex, medulla and collecting system	Shattered kidney	
Kidney - Vascular					Avulsion	

Fig. 1 Organ injury grading scale (*Source of information in Table*: Ernest E. Moore, MD, Thomas H. Cogbill, MD, Mark Malangoni, MD, Gregory J. Jurkovich, MD, and Howard R. Champion, MD. Scaling system for organ specific injuries. The American Association for the Surgery of Trauma. http://www.aast.org/library/traumatools/injuryscoringscales.aspx. Accessed 7 Dec 2013)

5. Common patterns:
   (a) Seat belt trauma:
      (i) Seat belts must contact the body in the proper location. Children less than 6-years-old need a booster suit so that the lap belt crosses over the anterior superior ileac crests.
      (ii) When a lap belt is used without a shoulder strap, it can result in injury.
      (iii) In a head-on motor vehicle crash, if a child is using a lap belt only, the body is forcefully flexed at the waist.
      (iv) The lap belt crushes the viscera against the spine.
      (v) Organs at risk are colon, small bowel, mesentery, duodenum, pancreas, and aorta.
      (vi) Hyperflexion of the spine can result in a Chance fracture: transverse distraction fracture of the posterior portion of the vertebral body.
   (b) Handlebar injury:
      (i) In a bike crash, if the handlebars turn acutely, the child can receive a direct blow to the abdomen with the end of the handlebar.

(ii) This can cause injury to pancreas, perforation of a hollow viscus, or traumatic hernia.

(c) Waddell's triad:

(i) Constellation of injuries when a pedestrian is struck by a car.
(ii) The components of the triad are femur fracture, splenic rupture, and subdural hematoma, but not all three injuries are always present.
(iii) The bumper of the car strikes the left leg and causes a fracture of the femur or the tibia and fibula, depending on the height of the bumper relative to the child.
(iv) When the child is thrown up on the hood of the vehicle, the spleen is ruptured.
(v) When the vehicle brakes, the child is thrown to the ground, resulting in a subdural hematoma.

6. Assessment:

(a) Initial assessment follows Advanced Trauma Life Support assessment with stabilization of C-spine and evaluation of airway, breathing, and circulation.
(b) Physical Examination: Abdomen is examined for ecchymosis, laceration, swelling, masses, and tenderness.
(c) FAST:

(i) Focused Abdominal Sonography for Trauma (FAST) is a rapid, noninvasive, and portable method to evaluate the abdomen.
(ii) Sonographic windows examined are pericardium, Morrison's pouch between liver and right kidney, space between spleen and left kidney, and the retrovesicular space in bladder.
(iii) Right and left pleural spaces are evaluated for pneumothorax.
(iv) The finding of free fluid in abdomen on FAST suggests solid organ injury and need for more diagnostic maneuvers.

(d) Peritoneal lavage:

(i) Used less frequency since FAST became available.
(ii) Peritoneal cavity is entered via a small supraumbilical incision.
(iii) Abdominal fluid is sampled for blood, amylase, or intestinal contents.
(iv) Can diagnose injury to solid organ or perforated viscus.
(v) Will not diagnose retroperitoneal injury with contained bleeding.

(e) CT scan:

(i) Requires ionizing radiation with risk of malignancy.
(ii) Ensure that patient is hemodynamically stable before risking a trip to the scanner.
(iii) Can diagnose and grade injury to solid organs.
(iv) Able to diagnose free fluid and air from hollow viscus injury with 94 % sensitivity, but signs of perforation may require time to develop after injury.

(f) MRI:

(i) Used less frequently to evaluate trauma.
(ii) Requires more time for images, and may require anesthetic for child to stay still.
(iii) Can be used to evaluate for injury to the biliary tree through magnetic resonance cholangiopancreatography (MRCP).

(g) ERCP:

(i) Useful in evaluating pancreas injury.
(ii) Endoscopic ultrasound can be performed at the same time.
(iii) If a pancreatic duct injury is detected, as stent can be placed.

(h) Angiography:

(i) Angiography with embolization offers an intermediate step for patients with hemorrhage from solid organ injury.
(ii) Embolization can be performed to the liver, spleen, kidney, and pelvic vessels.
(iii) Angiography carries the risk of vascular injury and hemorrhage.
(iv) Indications for embolization vary, but usually include blush on computed tomography, ongoing hemorrhage and blood transfusion requirement, and a patient stable enough to undergo the procedure.

7. Treatment:

(a) Correction of shock with intravenous boluses of crystalloid fluid, and transfusion of blood for non-responders.
(b) Non-operative treatment:

(i) The majority of solid organ injuries can be managed non-operatively.
(ii) Patients are admitted and are monitored for vital signs and hemoglobin at regular intervals.
(iii) Patients are kept NPO for possible laparotomy.
(iv) Criteria are established for failure of non-operative therapy such as hemodynamically unstable patient or transfusion requirement greater than six units of blood.
(v) After solid organ injury, children need a period of activity restriction to avoid repeat injury (Table 1).

(c) Operative treatment:

(i) Damage control surgery:

1. Hemodynamically unstable patients who do not respond to volume resuscitation undergo emergent laparotomy.
2. Wide exposure with a midline laparotomy is obtained and sponges are packed into all four quadrants and the pelvis.
3. Volume resuscitation continues through the procedure.
4. Each quadrant is unpacked and efforts are made to diagnose all injuries and control bleeding.

Table 1 American Pediatric Surgical Association recommendations for isolated liver or spleen injury

CT grade	I	II	III	IV
ICU stay (days)	None	None	None	1
Hospital stay (days)	2	3	4	5
Pre-discharge imaging	None	None	None	None
Post-discharge imaging	None	None	None	None
Activity restriction (weeks)[a]	3	4	5	6

[a]Return to full contact, competitive sports (i.e., football, wrestling, hockey, lacrosse, and mountain climbing) should be at the discretion of the individual pediatric trauma surgeon. The proposed guidelines for return to unrestricted activity include "normal" age-appropriate activities (Adapted from: Stylianos S. APSA Trauma Committee. Evidence-based guidelines for resource utilization in children with isolated spleen or liver injury. J Ped Surg. 2000;35:164–9)

5. If bleeding cannot be controlled quickly, the abdomen is left packed with sponges with temporary abdominal closure.
6. The patient is brought to the pediatric intensive care unit (PICU) for rewarming, resuscitation, and correction of abnormalities of coagulation.
7. The patient returns to the OR for washout of abdomen in 24–48 h with more definitive correction of injuries.

(ii) Decompressive laparotomy:

1. Hemorrhage and edema after abdominal trauma can increase the pressure within the peritoneal cavity.
2. Increased pressure will result in abdominal compartment syndrome with reduction in urine output, reduced return of blood to heart, and compromised blood flow to lower extremities.
3. Pressure is measured via a Foley catheter in the bladder. Pressure is normally 12 mmHg or less, and laparotomy must be considered for pressure above 25 mmHg.
4. Decompressive laparotomy is wide midline laparotomy allowing the edematous viscera and retroperitoneum to bulge out, and temporary abdominal closure.
5. Abdomen is washed out every 2–3 days, and fascia is slowly closed as edema decreases.

(iii) Temporary abdominal closure and vacuum assisted closure (VAC): VAC can be used as a temporary dressing when the abdomen is left open after laparotomy. The continuous suction removes fluid collections which could result in bacterial overgrowth and abscess. It also reduced edema fluid of viscera.

(iv) Procedures for liver:

1. Most injuries can be treated non-operatively.
2. If operative intervention is required due to shock or ongoing blood loss, consider damage control surgery with a second look later.

3. Embolization can be employed before or after laparotomy.
4. Bleeding from the vena cava or hepatic veins requires operative control of hemorrhage and has a high incidence of morbidity and mortality.

(v) Splenectomy and splenorrhaphy:

1. Operative intervention for splenic trauma usually results in splenectomy.
2. Attempts can be made at splenic salvage with splenorrhaphy using transfixion suture, omentum, wrapping with mesh, or a combination of techniques.
3. Patients with splenectomy or major injury to spleen require vaccination against infection from pneumococcus, Haemophilus influenzae, and Meningococcus.

(vi) Pyloric exclusion:

1. Minor injuries of the duodenum can be primarily repaired and drained if there is not shock, major contamination, and viable duodenal tissue.
2. Major injuries of the duodenum do better with washout, wide drainage, and pylorus exclusion either by stapling across the distal stomach (without division) or open pylorus exclusion by opening stomach, and closing lumen of distal stomach with running suture placed from the inside.
3. Stomach is drained through a gastrojejunostomy.

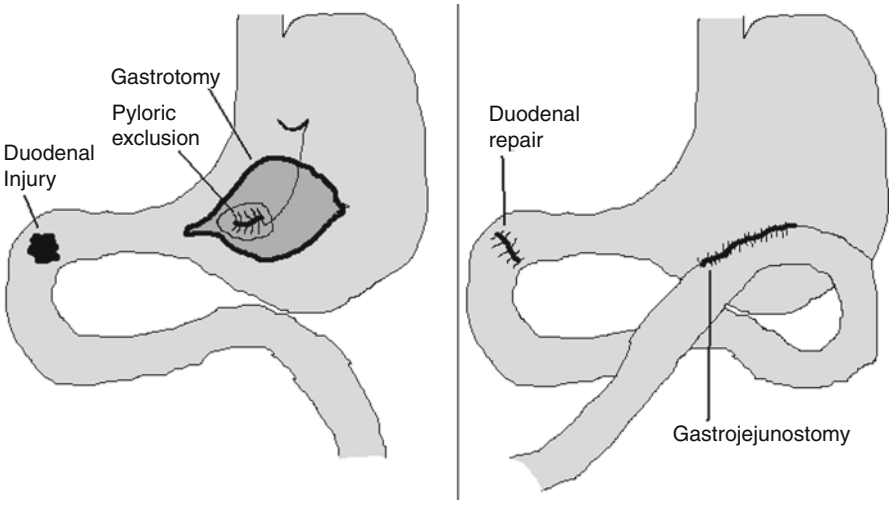

Fig. 2 Pyloric exclusion. Major trauma to the duodenum is best treated with pyloric exclusion. The stomach is opened, and through gastrostomy, pylorus is closed with running suture. The injury to duodenum is debrided, closed, and widely drained. The stomach is drained with a gastrojejunostomy (*Source*: Christopher Coppola)

(vii) Exploration of retroperitoneum:

1. Complete abdominal exploration sometimes includes mobilization of retroperitoneal structures via a medial visceral rotation maneuver.
2. Whether stable or expanding, most hematomas of the retroperitoneum should be explored. Nonexpanding hematomas of the perirenal or retrohepatic region are typically not explored, and instead they are treated with packing.

(viii) Kocher maneuver:

1. Injuries involving a high degree of suspicion for duodenal or pancreatic injury require mobilization of the duodenum and examination of the retroperitoneum.
2. The Kocher maneuver involves dissection of the lateral peritoneal attachments of the duodenum to expose all three segments of the duodenum, as well as the pancreatic head and neck.

(ix) Medial visceral rotation:

1. Rapid exposure of the major abdominal vessels can be obtained by utilizing mobilization maneuvers whereas the viscera are rotated off the midline. There are two common approaches: Left-sided and right sided medial visceral rotation.
2. Left-sided medial visceral rotation (Mattox maneuver) will expose the entire abdominal aorta along with its branches, with exception to the right renal artery. It is carried out by incising the lateral peritoneal attachment (white line of Toldt) about the left colon and sigmoid, then along the splenic flexure, around the posterior spleen, and behind the gastric fundus. The stomach, pancreas, spleen, and left colon are then rotated toward the midline.
3. Right-sided medial visceral rotation (Cattel-Braasch maneuver) is utilized when access is needed to the infrahepatic inferior vena cava. The right colon is mobilized by taking down the hepatic flexure of the colon, then continuing dissection along the paracolic gutter via the peritoneal reflection. Further blunt dissection will allow the colon to be rotated medially toward the aorta. If additional exposure of the vena cava is needed below the liver, then the Kocher maneuver may be utilized with medial mobilization of the duodenum and pancreatic head.

(x) Nephrectomy and ureter repair:

1. Exploration is warranted in children who are hemodynamically unstable, have an expanding hematoma, or have injury to other nearby viscera warranting exploration.
2. Presences of a contralateral kidney should be confirmed prior to proceeding with nephrectomy. Nephrectomy is the safest choice

for children with unstable injuries. Delayed nephrectomy may be warranted in the patients who develop hypertension or symptomatic renal infarction.

3. Ureteral repair may require a staged procedure in patients who are too unstable. For staged repair, the damaged ureter is commonly tied off with long silk ties to assist in localization later on. Children who are exhibit shock are at higher risk of complications due to tissue hypoxia. These complications include stricture, urinoma, fistula, and loss of tissue.
4. Partial transection can typically be repaired primarily, with exception to gunshot wounds.
5. Injuries of the upper ureter are typically repaired with ureteroureterostomy as long as a tension-free anastomosis is achieved. Mobilization of the kidney may be required to reduce tension. Ureteropelvic junction injury is more common in children than adults. Injuries involving ureteral avulsion from the renal pelvis may be anastomosed directly into the renal pelvis as an ureteropyelostomy procedure. Ureterocalicostomy may be performed in situations wh3ere the renal pelvis or ureteropelvic junction is damaged beyond repair.
6. Midureteral injuries are classically repaired with ureteroureterostomy. In extreme cases where there is extensive loss of ureter, transureteroureterostomy via donor ureter may be utilized.
7. Lower ureteral injuries are often accompanied by disruption of blood supply from the iliac vessels. As a result, these injuries are often repaired via ureteroneocystostomy whereas the ureter is re-implanted into the bladder. This repair is typically stented and a suprapubic tube and urethral Foley catheter are placed. For longer defects, the bladder may be mobilized and hitched to the psoas tendon to bridge the defect. Urethral anastomosis is then carried out medial to the hitch.

(xi) Pancreatectomy and trauma Whipple:

1. Operative intervention is typically related to the integrity of the pancreatic duct. In cases of blunt trauma, the pancreas is usually fractured where it crosses the vertebral column. Early distal pancreatectomy and drainage is preferred in cases of pancreatic transection. If an intact duct is confirmed, then non-operative management is an acceptable consideration.
2. Penetrating trauma to the head and neck without duct injury can be treated with simple drainage. When the ductal system is compromised, Roux-en-Y pancreaticojejunostomy is sometimes performed.
3. Severe injuries involving devascularization of the pancreas and duodenum require pyloric exclusion, drainage, and repeat debridement. While rare, children with catastrophic injury of the pancreatic head might require trauma Whipple, or pancreaticoduodenectomy.

Traumatic Spinal Injury in Children

Konstantina A. Svokos and Amir Kershenovich

1. Incidence:
   (a) Less common than in adults.
   (b) Most injuries are cervical (70–80 % of all spine injuries).
   (c) Sixty percent are occiput to C2, then high cervical.
2. Pediatric anatomical considerations:
   (a) Atlas (C1): three ossification centers. Synchondrosis of the spinous process fuses by 3-years-old.
   (b) Axis (C2): four ossification centers; an additional one appears at the summit of the dens between three and six-years-old and fuses by age 12. Synchondroses normally fuse by three to six-years-old.
   (c) C3-7: cervical bodies are normally narrower anteriorly (wedge-shaped).
   (d) Normal synchondroses may be mistaken for fractures, especially in the atlas.
   (e) Pseudospread of the atlas: Results from a discrepancy between the "neural" growth pattern of the atlas and the "somatic" pattern of the axis. More than two mm total overlap of the two C1 lateral masses on C2 on AP open-mouth view. Present in most children three-months-old to four-years-old. Neck rotation can also mimic the appearance of a fracture.
   (f) Pseudosubluxation: anterior displacement of C2 on C3 and/or significant angulation at this level; seen in children up to 10 years of age. Up to two to three mm is normal and does not represent instability.

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3. Injury by age group:
   (a) Neonates: most are birth injuries, especially in breech presentation or caesarean section after attempted vaginal delivery. Newborn spinal column can distract two in. but the spinal cord can only distract a few millimeters before injury.
   (b) Infants to 10-years-old:
      (i) Most common mechanism: motor vehicle collisions, accidents and falls.
      (ii) The upper cervical spine is most commonly injury due to ligamentous laxity.
   (c) Ten-years-old to adolescence
      (i) Most common mechanism: motor vehicle collisions, falls, sports.
      (ii) C3-C7 most commonly injured.
4. Whiplash injury: traumatic injury to the soft tissue structures in the cervical spine including muscles, ligaments, discs, facet joints. Occurs due to hyperflexion, hyperextension or rotational injury in the absence of fractures, dislocations or disc herniation. Symptoms may start immediately or hours to days after the insult.
5. Spinal cord injury without radiographic abnormality (SCIWORA): there is a subgroup of spinal cord injuries in children in which there is no radiographic evidence of bony or ligamentous disruption. This is attributed to the normally increased elasticity of the spinous ligaments and paravertebral soft tissue. Age range 1.5–16-years-old with a much higher incidence in age less or equal to nine-years-old. There is increased risk in children with asymptomatic Chiari I malformations. Workup includes MRI of the cervical spine. Management includes a cervical collar and prohibition from sports, with follow up flexion-extension x-rays at three months.
6. Initial management of suspected spinal injuries:
   (i) Initiate Advanced Trauma Life Support (ATLS) for hemodynamic stability.
   (ii) Immobilize immediately; may need shoulder roll to neutralize cervical spine in young children.
   (iii) Avoid hypotension, hypoxia, hypovolemia, and anemia.
   (iv) Maintain normocarbia. Crucial in upper cervical spine injuries due to diaphragmatic paralysis.
   (v) Perform thorough neurologic exam: motor, sensory, reflexes, clonus, bulbocavernosus reflex and rectal tone.
   (vi) Monitor neurologic exam.
   (vii) Assess for cord compression and cauda equina syndrome which are treated emergently.
   (viii) Determine mechanical instability of the spinal injury and need for surgical stabilization.

(ix) Ensure adequate cord perfusion by maintaining elevated mean arterial pressure.
(x) Maintain systolic blood pressure >90 mmHg.
(xi) Maintain hematocrit >28 %.
(xii) Maintain oxygen saturation >95 %.

7. Spinal cord injury: completeness of lesion

(a) Incomplete: any residual motor or sensory function > three segments below the level of injury.

(i) Signs: sensation or voluntary movement in lower extremities, sacral sparing (sensation around the anus, voluntary rectal sphincter contraction or voluntary toe flexion).
(ii) Types:

1. Central cord syndrome.
2. Brown-Sequard syndrome.
3. Anterior cord syndrome.
4. Posterior cord syndrome.

(b) Complete: no preservation of any motor and/or sensory function > three segments below the level of injury. Approximately three percent of patients will develop some recovery within 24 h. Persistence beyond 24 h indicates that no distal function will recover.

8. Spinal shock: hypotension that follows spinal cord injury due to :

(a) Interruption of sympathetic and loss of vascular tone below level of injury.
(b) Loss of muscle tone due to skeletal muscle paralysis below level of injury.

9. Important reflexes to test:

(a) Rectal tone: external sensation is noted as present or absent. Any sensation felt by the patient means that the injury is sensory incomplete.
(b) Bulbocavernosus reflex: contraction of the anal sphincter in response to pinching penile shaft, or in response to tug on Foley catheter is normal. Its presence used to be indication of an incomplete injury, but its presence alone is no longer considered to have a good prognosis for recovery.
(c) Abdominal cutaneous reflex: scratching one abdominal quadrant with a sharp object causes contraction of underlying abdominal musculature, causing the umbilicus to migrate toward the quadrant. Upper T8-9. Lower T10-12. Its presence indicates an incomplete lesion for cord injuries above the lower thoracic level.
(d) Knee extension/knee jerk: L3-4, quadriceps.
(e) Achilles/ankle jerk/ankle plantar flexion: S1-2, gastrocnemius and soleus.

10. ASIA (American Spinal Injury Association) Impairment scale:

Class	Description
A	Complete: no motor or sensory preserved in sacral segments S4-5
B	Incomplete: sensory but no motor function preserved below neurologic level
C	Incomplete: motor function preserved below the neurologic level (more than half of key muscles below neurologic level have a muscle grade <3)
D	Incomplete: motor function preserved below the neurologic level (more than half of key muscles below the neurologic level have a grade equal to or greater than 3)
E	Normal: sensory and motor function normal

11. Imaging:
    (a) X-rays of cervical spine, antero-posterior (AP) and lateral.
    (b) Computed tomography (CT) of cervical spine if injury is identified on X-rays or if they are inadequate.
    (c) Magnetic resonance imaging (MRI) of cervical spine is recommended if there is injury or neurologic deficit.
12. Cervical spine clearance:
    (a) Requires both radiographic and clinical evaluation.
    (b) Radiographic clearance:
        (i) AP, lateral and open-mouth views.
        (ii) Must be able to visualize superior endplate of T1 for x-ray to be adequate; if not, CT is employed.
        (iii) Consider MRI of cervical spine if there imaging is positive for injury or if there is a neurologic deficit.
    (c) Clinical clearance:
        (i) Per the National Emergency X-radiography Utilization Study (NEXUS) criteria.
        (ii) No current or recent neurologic deficit.
        (iii) Awake patient with no distracting injury.
        (iv) No midline cervical tenderness to palpation and with full range of motion.
        (v) In obtunded patients, long term the cervical collar may lead to skin breakdown or interfere with patient care, in particular in patients with tracheostomy. Consider MRI of cervical spine within 72 h of injury.
13. Fracture types:
    (a) Antlanto-occipital dislocation: twice as common in children than in adults due to ligamentous laxity; can be easily missed; maintain high index of suspicion; highly unstable; require surgical stabilization.
    (b) Atlanto-axial distraction: evaluate the symmetry of the C1-C2 facet joints and the atlanto-dens interval (ADI); some heal in a collar but most require surgery.
        (i) Acceptable ADI:
            1. In children eight-years-old or less: < five mm.
            2. In older children and adults: < three mm.

(c) Jefferson fractures: two fractures of the C1 ring due to axial loading.
   1. Rule of Spence: if the sum total overhang of both C1 lateral masses on C2 is ≥7 mm, the transverse ligament is likely disrupted.
   2. Unstable.
   3. Treated with Halo fixation or surgery.

(d) Odontoid fractures: due to forceful extension.

   1. Type 1: fracture of the odontoid process; usually stable; treated in cervical collar.
   2. Type 2: fracture across the base of the odontoid process; usually unstable; may need Halo, odontoid screw or C1-2 fusion.
   3. Type 3: fracture through the C2 body; stability depends on amount of body involvement; may treat in collar, Halo or surgery.

(e) Hangman's fractures: fracture of bilateral C2 pars interarticularis due to hyperextension and distraction or hyperflexion and distraction. If C2 is not significantly displaced over C3, may be treated in collar; if there is significant displacement, treat in a Halo. If it fails, surgical fixation is needed.
(f) Teardrop fracture: fracture of the anterior inferior corner of the vertebral body due to anterior flexion force; usually unstable, treated in a Halo or with surgical fixation.
(g) Cervical compression fractures: due to anterior flexion, axial loading or both; may be stable or unstable (burst fracture). Stable fractures are treated in a collar; unstable fractures are treated with fusion with or without decompression.
(h) Ligamentous injury without bony abnormality requires either MRI or flexion/extension x-rays for diagnosis; stability and treatment depends on degree of involvement.

14. Three column model: Denis' three column model of the spine attempts to determine instability of thoracolumbar spine fractures.

(a) Anterior column: composed of anterior half of disc and vertebral body (VB) including the anterior longitudinal ligament.
(b) Middle column: posterior half of disc and VB and the posterior longitudinal ligament.
(c) Posterior column: posterior bony complex with interposed supraspinous and interspinous ligament, facet joints and capsule and ligamentum flavum. Injury to this column alone does not cause instability.

15. Thoracolumbar fracture types:

(a) Thoracolumbar Chance fractures: horizontal fracture through the vertebral body often involving the posterior elements; associated with seatbelt injuries; unstable; require surgical stabilization.
(b) Thoracic and lumbar compression fractures: a flexion with or without compression injury; require careful assessment of the posterior elements with CT and MRI. Canal compromise, neurologic deficit, significant angulation or motion on dynamic imaging requires surgery.

(c) Thoracolumbar posterior column fracture: includes spinous process fractures, transverse process fractures and laminar fractures; usually stable if no other associated fractures or significant ligamentous injury is present.

16. Blunt vertebral artery injuries:

(a) Vertebral artery injury may be associated with blunt trauma and may produce vertebrobasilar insufficiency. Fractures through the foramen transversarium, facet fracture dislocation, or vertebral subluxation are frequently seen in patients with blunt vertebral artery injury.
(b) The most common mechanism of injury is motor vehicle collisions. Use conventional angiography or magnetic resonance angiography (MRA) after non-penetrating cervical trauma in patients who have spinal cord injury, fracture through the foramen transversarium, facet dislocation and/or vertebral subluxation.
(c) Treatment:

(i) Anticoagulation with IV heparin with evidence of posterior circulation stroke.
(ii) In cases of posterior circulation ischemia: observation or treatment with anticoagulation for vertebral artery injury.
(iii) If there is no evidence of posterior circulation ischemia, observation is recommended.

17. Gunshot wounds to the spine:

(a) Distribution: cervical 20–37 %, thoracic 50–64 %, lumbar 10–30 %
(b) Indications for surgery: injury to the cauda equina, neurologic deterioration, CSF leak, spinal instability, debridement to reduce risk of infection, vascular injuries.

18. Penetrating trauma to the neck:

(a) Injuries to the neck are divided into three zones:

(i) Zone I: inferiorly from the head of the clavicle to include the thoracic outlet.
(ii) Zone II: from the clavicle to the angle of the mandible to the base of the skull.
(iii) Zone III: from the angle of the mandible to the base of the skull.

(b) Mortality rate is approximately 15 %. Vascular injuries: venous 18 %, arterial 12 %. Most commonly injured vessel is the common carotid artery.
(c) Workup includes angiography, especially in Zone I or III it can demonstrate extravasation of blood, expanding hematomas, pseudoaneurysm, arteriovenous fistula, intimal dissection or occlusion by soft tissue or bone.
(d) Management must first focus on protecting the airway. Immediate intubation is indicated for hemodynamically unstable patients or for airway compromise. Surgical exploration is advocated for all wounds that pierce the

platysma and anterior triangles of the neck. Carotid artery injuries are managed with primary repair, ligation or interposition grafting. Vertebral artery injuries are often managed by ligation than by direct repair. Less urgent conditions require knowledge of the patency of the contralateral vertebral artery.

Suggested further reading

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Burns

Alfred P. Kennedy Jr.

Although there have been tremendous strides in the advancement of burn wound management, thermal injuries still affect approximately one million children in the Unites States per year and are a continued source of major morbidity and mortality. Burn injuries are responsible for nearly 2,500 childhood deaths per year.

1. Mechanisms of injury by age:
   (a) Scald burns most common under five years-old.
   (b) Contact burns more common in toddlers (curling irons, hot stove).
   (c) Flame burns more common in adolescents (fireworks, volatile agents).
      (i) Grease burns almost always represent deep partial thickness or full thickness burns.
2. Initial management:
   (a) Follow ATLS guidelines as for any injury with the following caveats.
   (b) Identify Signs of Inhalational Injury during the process of ABCs. Establish definitive airway.
   (c) Remove any source of ongoing thermal injury.
      (i) Remove all clothing carefully.
      (ii) Brush away any dried chemicals.
      (iii) Rinse involved sites with copious amounts of tap water.
      (iv) Cover burns with dry linens to prevent hypothermia.
   (d) Establish IV access.
      (i) May require placement of IV within an area of burn.
      (ii) May require placement of an intraosseous line.
      (iii) Large bore IV in upper extremity preferable.

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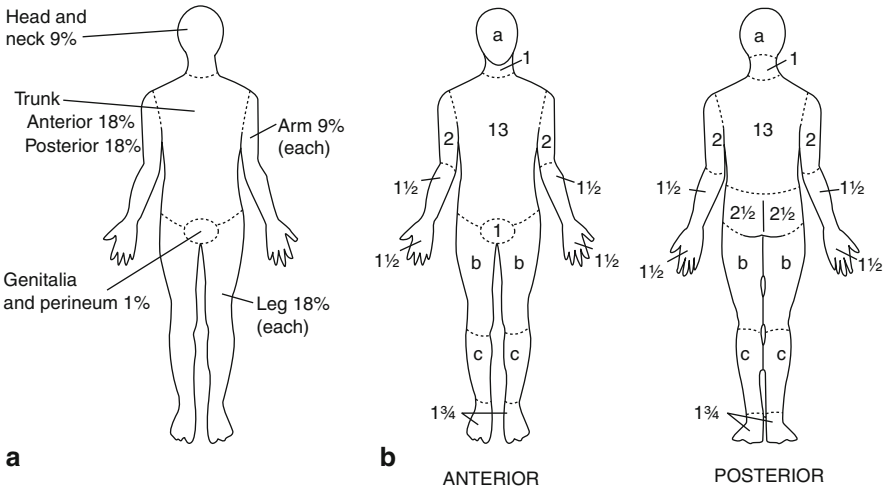
3. Inhalational injury:
   (a) Any burn suffered in confined space is at risk.
   (b) Signs of face or neck burns.
   (c) Singed eyebrows or nasal passages.
   (d) Carbonaceous sputum.
   (e) Hoarseness.
   (f) Impaired mentation (carbon monoxide poisoning)
   (g) Consider early intubation with signs of inhalational injury to prevent loss of airway.
4. Estimation of burn depth:
   (a) Partial thickness: "sunburn appearance", painful – first degree burn (epidermis only).
   (b) Deep partial thickness: "bullae" formation, painful – second degree burn (papillary dermis =/– reticular dermis (deep).
   (c) Full thickness: Pale, sometimes dry, leathery scar, painless – third degree burn (entire dermis and dermal appendages).
   (d) Organ involvement – fourth degree burn.
5. Estimation of % Total Body Surface Area (TBSA) burned.
   (a) "Rule of Nines" – modified for children due to larger head size.
   (b) Lund and Browder chart.
   (c) The size of the child's palm is an estimate of 1 % TBSA.
   (d) Burn % TBSA is re-evaluated after each debridement.
6. Burn resuscitation formulas:
   (a) Parkland: four mL×kg×% TBSA burn.
      (i) Give one half of this volume over the first eight h.
      (ii) Give the second half of this volume over the following 16 h.
      (iii) Lactated Ringer's is the solution of choice.
      (iv) For children <30 kg, give an additional volume of maintenance fluid each hour containing glucose to avoid hypoglycemia.
   (b) Brooke: two mL×kg×% TBSA burn, half given in the first eight h, half given in the following 16 h.
   (c) Initiation time for fluids is set at the time of injury (if starting fluids two h after injury, run the first half of fluid over six h.)
   (d) These formulas provide only estimation, and fluid resuscitation must be continually titrated to individual needs.
   (e) Titrate to ensure urine output of one mL/kg/h.
7. Laboratory examination:
   (a) CBC.
   (b) Type and cross match 10 cc/kg packed cells.
   (c) Serum glucose.



Relative percentage of body surface areas (%BSA) affected by growth

	0*yr*	1*yr*	5*yr*	10*yr*	15*yr*
a — ½ of head	9½	8½	6½	5½	4½
b — ½ of thigh	2¾	3¾	4	4¾	4½
c — ½ of 1 lower leg	2½	2½	2¾	3	3½

Fig. 1 Estimating percent total body surface area in children affected by burns. (**a**) Rule of "nines" for the rapid estimation of total body surface area burned. In infants, relative surface area of the head is larger, and the lower extremities are smaller. (**b**) Lund-Browder diagram for more precise estimation of burn total body surface area (From U.S. Department of Health and Human Services. Downloaded from http://www.remm.nlm.gov;burns.htm. On 7 Dec 2013)

(d) Electrolytes.
(e) ABG including HbCO.

8. Circumferential extremity burns:

(a) Paramount to maintain peripheral circulation.
(b) Remove jewelry.
(c) Assess peripheral perfusion including Doppler flow.
(d) Consider escharotomy for circumferential limb deep burn.
(e) Consider fasciotomy with associated crush injury, associated skeletal trauma, and high voltage electrical injury.

9. NGT insertion is indicated for TBSA >20 % and prior to transfer.
10. Wound care:

(a) If child is to be transferred to a burn center: apply dry linens to allow for later assessment. Wrapping burn provides some pain relief. Do not break blisters or apply or burn cream. Do not apply cold water.

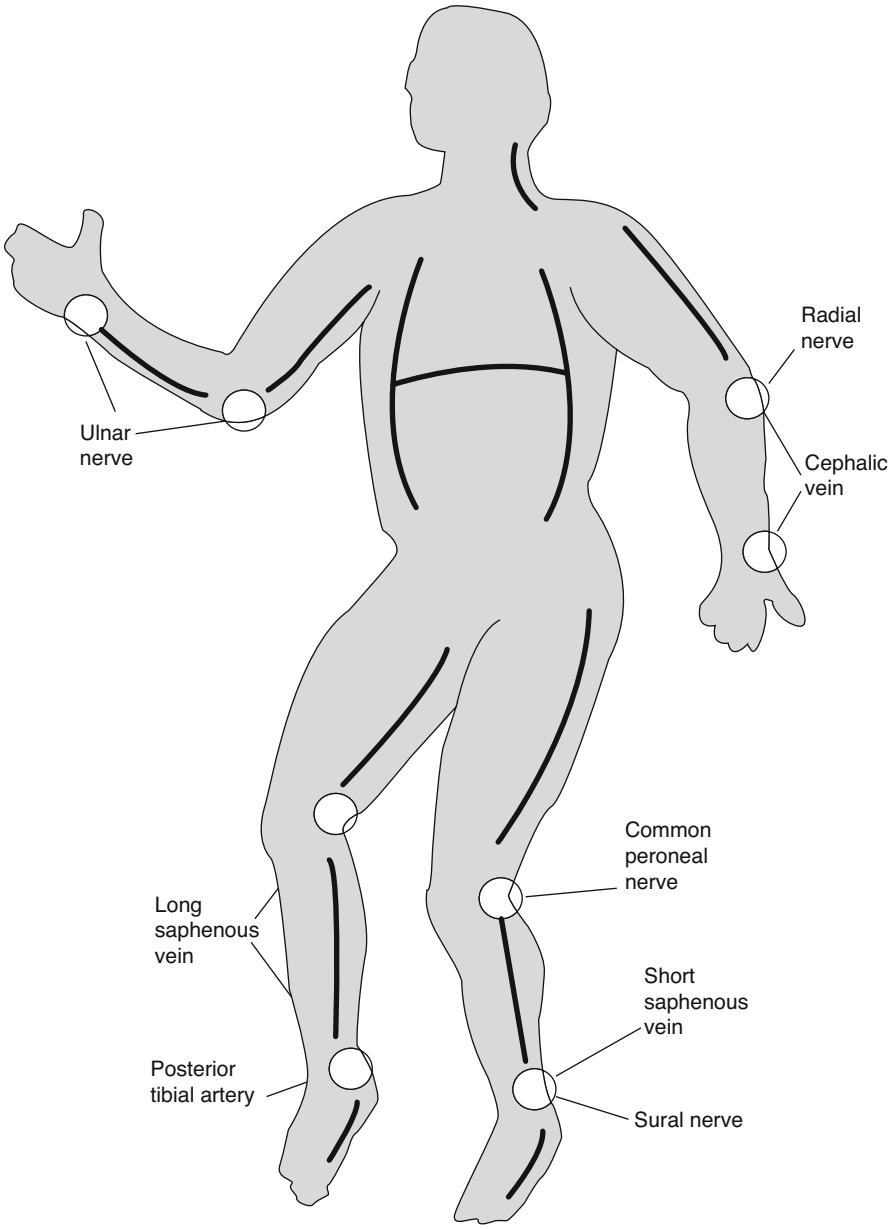


Fig. 2 Escharotomy lines and anatomic structures that need to be protected when performing escharotomy. (*Source*: Christopher Coppola)

(b) If treating burn, first give analgesic, then scrub and clean burn, removing all blisters and necrotic skin. Apply silver sulfadiazine cream and wrap loosely with dry sterile dressings.
(c) Clean burns with warm water bath every day and apply fresh dressing. Analgesics will be required.

11. Systemic antibiotics are contraindicated unless there is biopsy proven burn wound infection on quantitative burn culture.

12. Chemical burns:

(a) Result from exposure to alkali, acids or petroleum products. Alkali burns have deeper penetration of soft tissue and thus create more significant injury.
(b) Removal of chemical product performed with large volume water flush.
(c) Brush away dry powder before irrigation.
(d) Neutralizing agents may cause further injury and are contraindicated.

13. Electrical burns:

(a) Extent of tissue injury usually underestimated. Nervous system most vulnerable.
(b) Primary survey with ABCs.
(c) Evaluate for rhabdomyolysis and subsequent renal insufficiency with serial urinalysis; treat empirically with sufficient resuscitation, consider alkalinization of urine.
(d) Obtain ECG and maintain cardiac monitor for rhythm disturbances.
(e) Most electrical burns require fasciotomy for limb injuries to restore circulation.

14. Criterion for transfer to a burn center from the American Burn Association:

(a) Partial thickness and full thickness burns >10 % TBSA in patients less than 10-years -old or greater than 50-years-old.
(b) Partial thickness and full thickness burns >20 % TBSA.
(c) Full thickness burns > five % TBSA.
(d) Partial thickness and full thickness burns involving the face, eyes, ears, hands, feet, genitalia, perineum or over a major joint.
(e) Significant electrical or chemical burns.
(f) Inhalational injury.
(g) Burn injury in patients with preexisting medical conditions that may complicate treatment and recovery.

Pelvic Fractures

Louis C. Grandizio and Meagan M. Fernandez

Pediatric pelvis and acetabular fractures:

1. Overview:
   (a) Pelvic ring and acetabular fracture are associated with high-energy trauma.
      (i) Motor vehicle collisions, pedestrian versus motor vehicle and falls from height.
      (ii) Association with CNS and abdominal injuries.
   (b) Fracture patterns vary based on status of triradiate cartilage, skeletal maturity.
      (i) Pelvic bones become stronger than ligaments after skeletal maturity.
      (ii) Pediatric patients can have unique fracture patterns.
      (iii) Fractures in adolescent patients often resemble adult type injuries.
   (c) Lower overall mortality compared to adult pelvic fractures.
      (i) Pelvic hemorrhage and vascular injuries are less common in children.
         1. May be due to smaller vessel diameter, increased vasoconstriction.
   (d) Acetabular fractures account for <10 % of all pediatric pelvic fractures.
      (i) Less common than in adults.
      (ii) Can have combined pelvic and acetabular fractures.
   (e) Pelvic avulsion fractures often occur with sports injuries.

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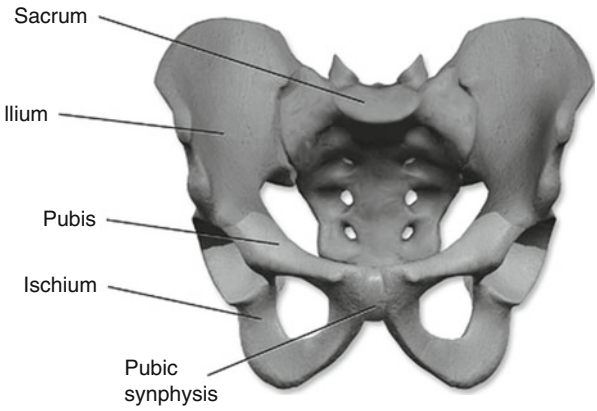

Fig. 1 The pelvis consists of the pubis, the ischium, and the ilium (Adapted from Bruce Blausen, Blausen Medical Communications, under Creative Commons Attribution 3.0 Unported license as published on http://en.wikipedia.org/wiki/File:Blausen_0723_Pelvis.png. Downloaded 28 Dec 2013)

2. Clinical anatomy:
   (a) Pelvic bones:
      (i) Pubis: Superior and inferior rami.
      (ii) Ischium: Body and tuberosity.
      (iii) Ilium: Body and wing.
   (b) Ligaments:
      (i) Pubic symphysis.
      (ii) Sacroiliac (SI).
         1. Posterior SI ligaments are strongest: Important factor for vertical stability of pelvis.
      (iii) Illiolumbar: Avulsion fracture can be sign of unstable ring injury.
   (c) Triradiate cartilage :
      (i) Closes around age 14 in boys, 12 in girls.
      (ii) Adolescent patients with closed triradiate cartilage at greater risk for pelvic ring injuries.
3. History and physical exam:
   (a) ATLS evaluation for life threatening injuries.
   (b) Complete neurovascular examination and secondary survey.
   (c) Compression/palpation of pelvis: assess stability.
   (d) Through urogenital examination.
      (i) Rectal examination required.
      (ii) Retrograde urethrogram required prior to bladder catheter placement if urethral injury is suspected.
      (iii) Assess for Morel-Lavellee lesion: shearing of subcutaneous fat and skin over the fascia.
      (iv) Assess for open pelvic fracture: rare, but may require diverting colostomy.

(e) Look for associated orthopaedic injuries: fractures of long bones, proximal femur and hip dislocations can occur with pelvic and acetabular trauma.

4. Diagnostic imaging:

(a) X-rays:

(i) AP pelvis part of initial trauma evaluation.
(ii) Judet views (45° oblique view of the affected hip): obtained for acetabular fractures.
(iii) Pelvic inlet and outlet radiographs: obtained for pelvic fractures.

(b) Computed tomography (CT):

(i) Improved osseous detail.
(ii) Better delineation fracture pattern: can also use CT three-dimensional reconstructions, when available.
(iii) "Stable" fractures with posterior SI joint tenderness require CT.

(c) MRI/Bone Scan:

(i) Limited value in acute setting.
(ii) Can be useful to diagnose occult fractures or avulsion injuries.

5. Classification of pelvic fractures:

(a) Many classifications systems exist.
(b) Limited clinical utility – variability exists in the ability for classification systems to determine treatment and prognosis.
(c) Pediatric pelvic fracture classification.

(i) Torode and Zieg:

1. I: Avulsion.
2. II: Iliac wing.
3. III: Simple ring.
4. IV: Ring disruption.

(d) Adolescent pelvic fractures often resemble adult patterns.

(i) Tile classification:

1. A: Stable.
2. B: Rotationally unstable, vertically stable.
3. C: Rotationally and vertically unstable.

(e) Acetabular fracture classification:

(i) Salter-Harris classification is used with open triradiate cartilage.
(ii) Salter-Harris classification:

1. I: Fracture through the physis.
2. II: Fracture through physis and metaphysis.

3. III: Fracture through physis and epiphysis.
4. IV: Fracture through physis, metaphysis and epiphysis.
5. V: Crush injury to physis.

(iii) Fractures in adolescents and skeletally mature patients resemble adult patterns.

1. Letournel classification:

(a) Elementary types.

(i) Posterior wall.
(ii) Posterior column.
(iii) Anterior wall.
(iv) Anterior column.
(v) Transverse.

(b) Associated types:

(i) Both columns.
(ii) Transverse and posterior wall.
(iii) T-Type.
(iv) Anterior column and posterior hemitransverse.
(v) Posterior wall and posterior column.

6. Initial treatment principles:

(a) Most pediatric pelvic and acetabular fractures can be managed without surgery.
(b) Life threatening injuries need to be managed first: address hemodynamic stability.
(c) Treatment often dictated by age. Skeletally mature patients have less elastic pubic symphysis and sacroiliac ligaments.
(d) Obtain orthopaedic surgical consult.

7. Management of pelvic avulsion and iliac wing fractures (Torode and Zieg I-II):

(a) Include avulsions, iliac wing and iliac apophyseal separations.

1. Initial protected weight-bearing.
2. Progress to weight-bearing as tolerated with strengthening program.

8. Management of simple ring fractures (Torode and Zieg III):

(a) Include pubic fractures and pubic symphysis separations. Need to rule out genitourinary injury.
(b) Stable fractures.

(i) Compliant patients can bear weight as tolerated.
(ii) Younger patients may benefit from short period of bed rest for pain control.

9. Management of ring disruptions (Torode and Zieg IV):
    (a) Primary concern is hemodynamic stability.
    (b) Unstable fractures.
    (c) "Straddle fractures".
        (i) Bilateral superior and inferior pubic rami fractures.
    (d) Malgaigne fractures.
        (i) Anterior pubic rami or pubic symphysis with posterior fracture or ligamentous injury.
    (e) Initial management of fractures with <2 cm displacement.
        (i) Bed rest.
        (ii) Skeletal traction: often indicated for sacroiliac ligament disruption, vertically unstable fractures.
        (iii) Spica casting.
            1. Can be used for fractures with minimal displacement.
            2. Can convert to spica cast for displaced fractures after period of skeletal traction.
    (f) Initial management of fractures with >2 cm displacement.
            1. Leg length discrepancy <2 cm can be well tolerated.
            2. Bed rest.
            3. Skeletal traction.
            4. Greater than 2 cm displacement often requires operative reduction and fixation.
10. Management of hemorrhage from pelvic fractures.
    (a) Less common than in adults.
    (b) Associated with Torode and Zieg IV fractures (unstable ring disruptions): Iliac vessels often involved.
    (c) When IV fluid boluses and blood transfusions are required, need to rule out other sources of bleeding, such as, abdomen, pelvis, extremity, thorax, and retroperitoneal.
    (d) Pelvic volume can be initially decreased with pelvic binder or sheet.
        (i) Sheet wrapped around greater trochanters.
        (ii) Commercially available pelvic binders.
    (e) Persistent hemodynamic instability requires emergent external fixation, pelvic angiography/embolization. The protocol for treatment will vary by institution and resources available.
11. Management of acetabular fractures:
    (a) Typically stable fractures in children and non-displaced fractures.
    (b) Keep non-weight bearing initially and advance as tolerated.

(c) Immobilization with spica casting may be indicated for younger, non-compliant patients.
(d) Skeletal traction may be indicated for unstable and displaced fractures. Buck's traction is a short term, initial alternative.
(e) Majority can be managed non-operatively, but there is an increasing trend towards operative management in children.
(f) Operative indications:

    (i) Limited evidence.
    (ii) Some authors advocate reduction and fixation with:

        1. Intra-articular displacement >2–5 mm.
        2. Greater than 2 mm of triradiate cartilage displacement.

12. Complications.

(a) Avascular necrosis of femoral head.
(b) Growth disturbance.
(c) Leg length inequality.
(d) Premature physeal closure.
(e) Low back pain.
(f) Post traumatic arthritis.
(g) Peripheral nerve injury.
(h) Myositis ossificans.

Common Pediatric Fractures

Louis C. Grandizio and Meagan M. Fernandez

Diagnosis and initial stabilization of common pediatric fractures:

1. Overview:
   (a) Significant cause of morbidity and mortality in the pediatric population.
   (b) Pediatric fractures have greater remodeling potential than adult fractures.
      (i) Deformity better tolerated closer to physis.
   (c) Majority of pediatric fractures can be managed non-operatively.
   (d) Fractures are more common in boys.
   (e) Open fractures are rare (<5 %).
   (f) Non-accidental trauma needs to be considered in work-up: see non-accidental trauma chapter.
2. Anatomic considerations:
   (a) Physis.
      (i) Growth plate.
         1. Unique to pediatrics.
      (ii) Weaker than cortical bone which pre-disposes to fracture.
         1. Ligamentous injuries are less common in pediatric patients.
   (b) Periosteum:
      (i) Thick fibrous structure that surrounds bone.
      (ii) Can aid in closed reduction of fractures.

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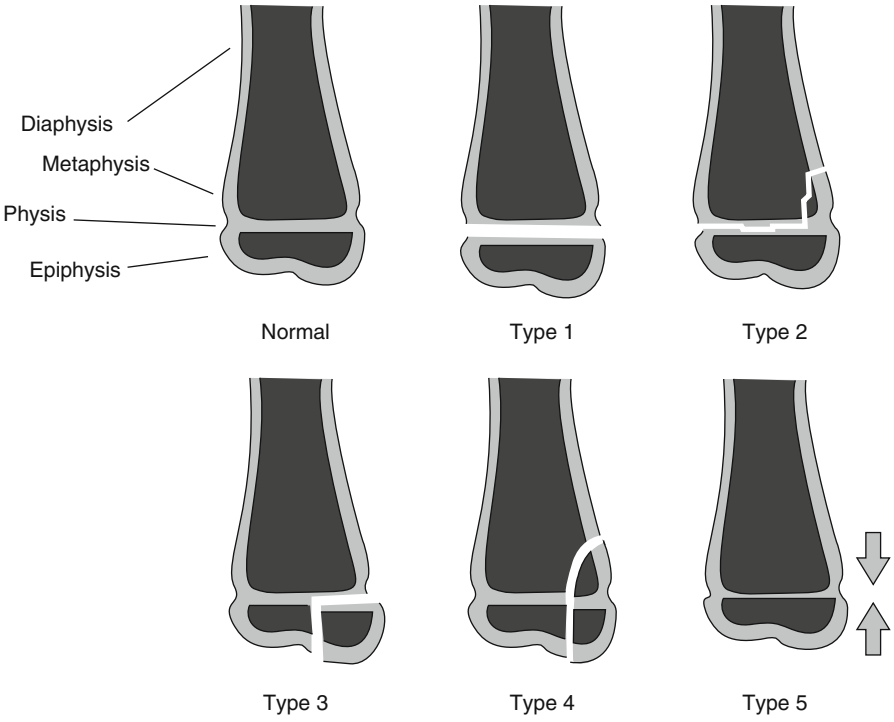

Fig. 1 Salter-Harris classification of fractures (*Source*: Christopher Coppola)

3. Physeal injuries:
   (a) Fractures involving the growth plate can result in growth disturbance.
   (b) Salter-Harris classification:
      (i) I: Fracture through the physis.
      (ii) II: Fracture through physis and metaphysis (most common).
      (iii) III: Fracture through physis and epiphysis.
      (iv) IV: Fracture through physis, metaphysis and epiphysis.
      (v) V: Crush injury to physis.
   (c) As a general rule, higher Salter-Harris grade results in increased incidence of growth disturbance.
4. History:
   (a) Age, mechanism of injury, time of injury, witnesses to event, prior injuries.
   (b) Birth, developmental, family and social history.
      (i) Underlying metabolic bone disorders.
      (ii) Osteogenesis imperfecta.
      (iii) Risk factors for non-accidental trauma.

5. Physical examination:
   (a) Skin:
      (i) Any communication of fracture through skin is an open fracture.
      (ii) Overlying lacerations and wounds may not be at exact level of open fracture.
      (iii) "Impending open" fractures present with skin tenting from underlying bone.
         1. Can erode skin and convert closed fracture to open fracture.
         2. Requires reduction.
      (iv) Open fractures have higher infection risk. They require immediate IV antibiotics and urgent debridement in operating room.
   (b) Soft tissues: assess myofascial compartments for soft tissue swelling.
      (i) Compartment syndrome:
         1. Can occur even without fracture.
            (a) Crush injuries, burns, bleeding.
         2. Assess for 5 P's.
            (a) Pain with passive stretch.
            (b) Parasthesias.
            (c) Pulselessness (late finding).
            (d) Paralysis.
            (e) Poikilothermia.
         3. Exam can be unreliable in children.
            (a) Agitation, anxiety and increasing narcotic analgesia requirement should be monitored in pediatric patients.
         4. Concern for compartment syndrome requires emergent orthopaedic consult.
            (a) Treatment is fasciotomy in the operating room.
            (b) Missed compartment syndrome has devastating neurovascular consequences.
   (c) Neurovascular examination:
      (i) Document complete neurologic examination.
         1. Can be limited by age and patient compliance.
         2. Motor and sensory exam.
      (ii) Vascular exam:
         1. Palpate peripheral pulses.
         2. Duplex ultrasound examination of pulses.
         3. Assess perfusion of extremity.

(a) Capillary refill.
(b) Temperature.
(c) Color.

6. Imaging:
   (a) X-rays:
      (i) Orthogonal views of the fracture.
      (ii) Obtain radiographs of the joint above and below the fracture.
      (iii) Comparative radiographs of contralateral extremity are helpful to distinguish fractures from normal anatomic variants.
      (iv) Descriptive terms:
         1. Location.
            (a) Physeal.
            (b) Metaphyseal.
            (c) Diaphyseal.
         2. Angulation.
         3. Displacement.
         4. Comminution.
         5. Shortening.
   (b) CT:
      (i) Consider for intra-articular fractures, physeal injuries.
      (ii) Increased exposure to radiation compared to plan radiographs.
   (c) MRI:
      (i) Limited use in the acute setting. Can be helpful to assess for physeal injuries and soft tissue injuries.
   (d) Ultrasound:
      (i) Use increasing secondary to availability and low-cost compared to MRI.
      (ii) Useful to assess cartilage in unossified bone.
7. Initial treatment principles:
   (a) Splinting:
      (i) Well-padded with cast padding to avoid soft tissue breakdown.
      (ii) Orthoglass (BSN Medical, Hamburg, Germany) – fiberglass, or plaster splints can be utilized.
      (iii) Immobilization of fracture may require inclusion of joint above and below fracture.
      (iv) Need to immobilize fracture for pain control and maintenance of reduction.
      (v) Tight splints can lead to compartment syndrome.

(b) Reduction: Requires conscious sedation, intra-articular local anesthetic, or hematoma block depending on patient age and compliance.

8. Common upper extremity fractures:

(a) Clavicle fractures:

(i) Midshaft clavicle fractures are common.
(ii) Initial management of closed injuries without skin tenting is sling or shoulder immobilizer.
(iii) Majority can be managed non-operatively.

(b) Proximal humerus fractures:

(i) Overview:

1. Greater than 5 % of all pediatric fractures.
2. Proximal humerus physis closes at age 14 in girls and 16 in boys.
3. Physeal injury: Salter Harris I-II most common.
4. Metaphyseal fractures: Do not involve physis.

(ii) Initial treatment principles:

1. Can be managed with shoulder immobilizer or sling/swathe.
2. Most can be managed non-operatively.
3. Unacceptable angulation treated with closed reduction and percutaneous pinning in the operating room.
4. Angulation better tolerated in patients <12 years old: they are able to remodel.

(c) Humerus shaft fractures:

(i) Overview:

1. Most common long bone fracture seen in non-accidental trauma.
2. Holstein-Lewis fracture.

(a) Mid-distal 1/3 diaphyseal fracture associated with radial nerve palsy.
(b) Radial nerve palsy can be followed with serial exams.

(ii) Initial treatment principles:

1. Rarely require operative intervention.

(a) Angulation <30° is well tolerated.
(b) Varus angulation is most common.

2. Children <4 years old: Sling and swathe.
3. Children >4 years old:

(a) Sarmiento brace.
(b) Coaptation splint in older children and adolescents.

4. Use additional wrist drop splint in presence of radial nerve palsy to avoid contracture.

(d) Management of pediatric fracture about the elbow:

(i) Assessment of neurovascular status is critical.
(ii) Assess for increased compartment pressure.

1. Missed compartment syndrome can result in Volkmann ischemic contracture and devastating functional outcomes.

(a) Emergent orthopaedic consult and compartment pressure measurement with any concern for compartment syndrome

(iii) Well-padded long-arm splints with the elbow at 70° of flexion are suitable for initial stabilization of most elbow fractures.

(e) Supracondylar humerus fractures:

(i) Overview:

1. Most common pediatric elbow fracture.
2. Extension type far more common than flexion type.

(a) Gartland classification:

(i) I: Non-displaced.
(ii) II: Displaced with intact posterior cortex.
(iii) III: Complete displacement.

3. Assess neurovascular status:

(a) Anterior interosseous nerve (AIN) palsy most common in extension-type fracture.
(b) Perfusion of hand is more important than presence of peripheral pulses.

(i) Cold, pulseless hand is a vascular emergency.

(c) Concerning exam findings include antecubital ecchymosis, "button-hole", open fractures.
(d) Frequent monitoring for compartment syndrome.

(ii) Initial treatment principles:

1. Splint in 70° of flexion with a well-padded long arm splint.

(a) Assess pulses and perfusion after splint application.

2. Type I fractures can be managed non-operatively.
3. Type II and III fractures require closed vs. open reduction and percutaneous pinning.
4. When there is risk for neurovascular compromise, absent radial pulses or poor perfusion, a closed reduction attempt should be performed by orthopaedics in the emergency department under conscious sedation.

(a) Vascular compromise in the form of a cold, pulseless hand after reduction attempt requires emergent operative intervention and a vascular surgery consult.

(f) Forearm fractures:

(i) Overview:

1. Can be classified as both bones forearm fractures, isolated radius/ulna fractures, Monteggia fractures or Galeazzi fractures.
2. Both bones forearm fractures can be complete or incomplete (greenstick fracture).
3. Isolated fractures of the radius can be diaphyseal, metaphyseal (most common), or distal physeal.

(a) Closed vs. open treatment depends on patient age, fracture location, shortening, displacement and angulation as well as neurovascular status.

4. Monteggia fractures occur most commonly between ages 4–10-years-old and involve a radial head dislocation with a fracture of the proximal ulna.

(a) Bado classification:

(i) I: Anterior radial head dislocation with apex anterior ulna fracture.
(ii) II: Posterior radial head dislocation with apex posterior proximal ulna fracture.
(iii) III: Lateral radial head dislocation with apex lateral proximal ulna fracture.
(iv) IV: Anterior dislocation of the radial head with proximal radial and ulna fracture at the same level.

5. Galeazzi fracture.

(a) Fracture of the distal radial diaphysis with disruption of the distal radiounlar joint (DRUJ).

(i) Often referred to as a "fracture of necessity" as it often requires operative treatment except in very young patients with anatomic reductions of the radius and the DRUJ.

(ii) Initial treatment principles:

1. Both bones forearm fractures.

(a) Rule out neurovascular injury, associated fracture (floating elbow), and compartment syndrome.
(b) Initial splinting consists of a long-arm sugar-tong up, sugar-tong down splint.

(i) Avoid elbow flexion above 90°.

1. Can "kink" vessels and potentially increase compartment pressures.

(c) Orthopaedic consult to determine need for closed reduction.

(i) Based on age, displacement, angulation, shortening.

2. Isolated radius or ulna fracture:

(a) Rule out neurovascular injury, associated fractures and compartment syndrome.
(b) Stable, incomplete fractures of the distal radial metaphysis without angulation can be initially managed with a short-arm well-padded splint or a sugar-tong splint around the elbow.
(c) Proximal or unstable fractures often require a closed reduction attempt by orthopaedics as well as a long-arm sugar-tong up, sugar-tong down splint.

3. Monteggia fracture:

(a) Require orthopaedic consult
(b) Bado types I-III can be managed with closed reduction of radial head and anatomic reduction of ulna fracture.
(c) Initial pre-reduction management is a long-arm sugar-tong up, sugar-tong down splint.

4. Galeazzi fractures.

(a) Initial splinting includes sugar-tong splint around the elbow or long-arm sugar-tong up, sugar-tong down splint to immobilize the DRUJ.
(b) Adolescent patients or those without anatomic reduction of the radius and DRUJ often require operative intervention.

Part III
Common Neonatal Problems

Necrotizing Enterocolitis

Christopher P. Coppola

Necrotizing enterocolitis (NEC) is the most common disease requiring assistance by a pediatric surgeon in the NICU. It occurs with increased incidence in premature infants and is more prevalent in countries with the capability to keep premature infants alive.

1. Pathophysiology of necrotizing enterocolitis.
   (a) Focal to diffuse ischemia of intestine.
   (b) Likely multifactorial etiology.
   (c) Stress, hypoxia, difficult childbirth, sepsis.
   (d) Can manifest after feeding.
   (e) Pathology ranges from focal disease to pan-necrosis of bowel (known as NEC totalis).
   (f) Terminal ileum most commonly affected.
2. Focal intestinal perforation (FIP), also abbreviated TIP for terminal ileal perforation, is a focal perforation without broad necrosis. It may represent a variant on necrotizing enterocolitis, but has a more favorable outcome. It is seen in premature infants, some who have been treated with corticosteroids, vasopressors, or indomethacin.
3. Incidence:
   (a) One to five percent NICU admissions.
   (b) Two cases/1,000 births in USA.
   (c) Thirty times increased incidence in premature infants.
4. Clinical signs:
   (a) Vomiting.

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(b) Distention.
(c) Vital sign instability.
(d) Tenderness.
(e) Abdominal wall cellulitis.
(f) Blue discoloration of abdomen indicating perforation and spillage of succus.

5. Evaluation:

(a) Examination.
(b) CBC, electrolytes, pH.
(c) X-ray signs of NEC.

(i) Tubular distended loops.
(ii) Thickened bowel wall.
(iii) Pneumatosis coli.
(iv) Portal venous air.
(v) Free air.
(vi) Ground glass background.

(d) Ultrasound.
(e) Paracentesis.

Modified Bell's staging criteria for necrotizing enterocolitis.

Stage	Signs	Abdomen	X-ray	Treatment
I Suspected				
IA	Temperature instability, apnea, bradycardia, lethargy	Gastric distention, emesis, heme-positive stool	Normal or intestinal dilation, mild ileus	NPO, 3 days antibiotics
IB	Same as above	Bloody stool	Dilation or ileus	NPO, 3 days antibiotics
II Definite				
IIA Mildly ill	Same as above	Same as above, absent bowel sounds, +/− tenderness	Dilation, ileus, pneumatosis intestinalis	NPO, 7–10 days antibiotics
IIB Moderately ill	Same as above, metabolic acidosis, thrombocytopenia	Same as above, no bowel sounds, tenderness, +/− cellulitis, mass	Same as above, ascites	NPO, 14 days antibiotics
III Advanced				

Stage	Signs	Abdomen	X-ray	Treatment
IIIA Intact bowel	Same as above, shock, bradycardia, apnea, respiratory acidosis, DIC, neutropenia	Same as above, peritonitis, tenderness, distention	Same as above	NPO, 14 days antibiotics, IV fluid, pressors, ventilator therapy, paracentesis
IIIB Perforation	Same as above	Same as above	Same as above, free air	Laparotomy

Adapted from: Bell ML, Ternberg JL, Feigin RD, et al. Neonatal necrotizing enterocolitis: therapeutic decisions based upon clinical staging. Ann Surg. 1978;187:1–7; Neu J. Necrotizing enterocolitis: the search for unifying pathogenic theory leading for prevention. Pediatr Clin North Am. 1996;43:409–32

6. Treatment:

   (a) Majority treated non-operatively.
   (b) Resuscitation.
   (c) Bowel rest.
   (d) Broad spectrum antibiotics.
   (e) Parenteral nutrition.
   (f) Serial examination.
   (g) Serial x-ray.
   (h) Indications for operative treatment:

      (i) For worsening physiologic status.
      (ii) Respiratory.
      (iii) Sepsis.
      (iv) Coagulopathy.
      (v) Acidosis.
      (vi) Perforation.
      (vii) Suspected gangrene.

   (i) Operative choices:
      (i) Laparotomy.
      (ii) Resection and stoma.
      (iii) Anastomosis.
      (iv) Patch, drain, and wait.
      (v) Skewer (run catheter through discontinuous loops of bowel).
      (vi) Percutaneous peritoneal drain (PPD):

         1. Neonatal peritoneum: less fat than adult, circulation not defined, abdominal wall thin, translocation of bacteria, angiogenesis after NEC, immature immune system.
         2. Technique: intubated, ventilated, anesthetized, keep warm, broad spectrum antibiotics, transfusions, platelets, resuscitation, remain in NICU, avoid wide prep, RLQ (and/or others) incision, irrigation with warm saline, placement of drain, observation and support.

3. Results: need to convert to open surgery: 60 % (NEC totalis: death, worsening ventilatory status, worsening hypotension/acidosis); need for further surgery: 75 % (stricture, fistula, adhesions).

7. Outcome:

   (a) Mortality based on gestational age, as well as cardiac, pulmonary, and neurologic status.
   (b) Prolonged NICU stay.
   (c) Long need for TPN, can result in cholestasis and liver failure.
   (d) Short gut is a risk.

Congenital Diaphragmatic Hernia

Christopher P. Coppola

Congenital diaphragmatic hernia (CDH) – This condition is often severe due to an accompanying pulmonary hypoplasia. Through advanced neonatal intensive care and select application of extracorporeal membrane oxygenation, most children survive to correction.

1. Epidemiology:
   (a) One in 2,000–5,000 births.
   (b) May be higher in stillborn children (1/3 of CDH).
   (c) Higher in females if stillborn children are included.
   (d) Two percent in first-degree relative.
   (e) Eighty percent left-sided.
   (f) Linked to phenmetrazine, thalidomide, quinine, nitrofen, vitamin A deficiency.
2. Associated anomalies are present in 28–50 % of infants with CDH and in 100 % of stillborn infants with CDH. Common defects are neural tube, cardiac, and midline defects, such as pentalogy of Cantrell. CDH is more common in trisomy 13, 18, and 21.
3. Pathophysiology:
   (a) Midgut returns to abdomen week 9–10.
   (b) If diaphragm is open, abdominal viscera herniate to thoracic cavity.
   (c) Occurs by week 12.
   (d) Malrotation occurs.
   (e) Hernia sac present in 10–15 %.
   (f) Left more common than right.
   (g) Bochdalek (posterior lateral) hernia is more common than Morgagni (anterior) hernia.

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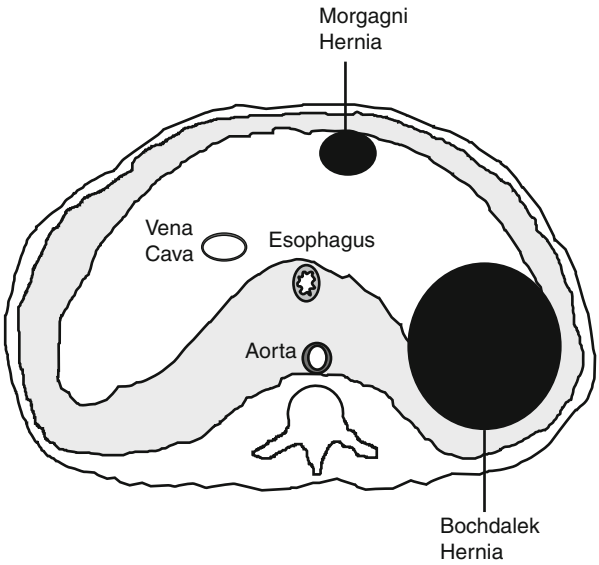

Fig. 1 Types of congenital diaphragm hernias. Morgagni hernias are defects in the anterior diaphragm while the more common Bochdalek hernias are in the posterolateral diaphragm (*Source*: Christopher Coppola)

(h) Can contain liver, spleen, gastrointestinal tract, and kidney.
(i) Associated pulmonary hypoplasia.

(i) Decreased lung mass.
(ii) Decreased bronchial branching.
(iii) Decreased alveoli.
(iv) Decreased total vascular diameter.
(v) Hypertrophied vascular smooth muscle.
(vi) Smooth muscle in alveolar vessels.
(vii) Bilateral lungs affected.

(j) Factors resulting in persistent fetal circulation in CDH.

(i) Pulmonary hypoplasia.
(ii) High pulmonary vascular resistance.
(iii) Hypoxia.
(iv) Acidosis.
(v) Stress.

(k) Diagnosis:

(i) Prenatal ultrasound (CDH, polyhydramnios).
(ii) Respiratory distress after delivery.
(iii) Scaphoid abdomen/funnel chest.
(iv) Abdominal x-ray with nasogastric tube, showing tube in chest.
(v) Ten – 20 percent with delayed presentation.

(l) Predicting severity of CDH:

(i) Prenatally diagnosed CDH as a group has worse prognosis than CDH diagnosed after birth.
(ii) Presence of cardiac defects worsens mortality.
(iii) Smaller pulmonary artery size (which can be quantified by the McGoon Index) carries worse prognosis.
(iv) Defects large enough to allow herniation of the stomach into the chest have worse prognosis and have reported to have survival as low as 30 %.
(v) Right-sided hernia represents more severe disease occurring earlier in gestation.
(vi) After delivery, the Oxygenation Index (OI) = (MAP × FiO2/PaO2) is used to quantify severity of pulmonary hypoplasia.

1. OI <0.06 carries a survival of 98 %.
2. OI >0.175 predicts survival of 0 %.
3. OI can be used to predict need for extracorporeal membrane oxygenation (ECMO).

(m) Prenatal care:

(i) When a patient is prenatally diagnosed with CDH, parents should receive genetic counseling, and karyotyping should be performed on the fetus to detect any associated anomalies or syndromes.
(ii) Prenatal care should be given to support gestation as close as possible to term to minimize the effect of premature delivery on an already compromised pulmonary function.
(iii) When possible, delivery should occur in a center with access to high level NICU care and a plan for access to ECMO therapy if it becomes necessary.

(n) Treatment:

(i) Initial resuscitation:

1. Patient should be assessed by neonatologist and surgeon after deliver. IV fluid should be started and a nasogastric tube is placed to decompress the stomach, which may be in the chest, and to reduce the volume of gas and fluid in the gastrointestinal tract.
2. Pulmonary function should be monitored for signs of pulmonary hypoplasia and failure. The patient may worsen over the course of the initial 48 h “honeymoon” period.
3. Obtain chest x-ray to detect abdominal organs in the chest.
4. When intubation and mechanical ventilation is needed, use a strategy of “gentle ventilation” to avoid barotrauma to the alveoli.

(a) High frequency/low pressure ventilation, with use of an oscillating ventilator when needed.

(b) Permissive hypercapnea.
(c) Medications to reduce pulmonary hypertension.

(i) Inhaled nitric oxide.
(ii) Sildenafil.
(iii) Tolazoline.

(d) When high frequency ventilation and inhaled nitric oxide fail to provide adequate oxygenation, evaluate patient for suitability for ECMO therapy.

(ii) Surgical repair of CDH:

1. Timing of operation: Repair should be delayed until after the initial 48 h "honeymoon period" to allow for stabilization and assessment for extent of pulmonary hypoplasia, rather than add the stress of repair to a child with worsening pulmonary function. When ECMO is needed, it is advantageous to delay repair until after completion of the ECMO run to avoid operation while the child is heparinized and more prone to hemorrhage. Rarely, it is necessary to repair CDH while on ECMO because child is failing to progress.
2. Technique:

(a) Subcostal incision or thoracotomy: On the left, the approach via abdomen allows for convenient reduction of viscera into abdominal cavity. For a right sided CDH, thoracotomy avoids the difficulty of working around the liver that is encountered with an abdominal approach (though a right subcostal incision is a viable option.)

(i) Reduce hernia contents back into abdomen.
(ii) Excise hernia sac when present as it can interfere with visualization of lungs or viscera while closing defect.
(iii) Identify muscular rim of diaphragm circumferentially. Some areas along posterolateral ribs and mediastinal structures may have no diaphragm and require careful placement of sutures.
(iv) Determine if primary closure is possible or if a patch of exogenous material (artificial or biologic mesh) will be needed (approximately 1/3 of cases).
(v) Consider addressing malrotation/non-fixation of bowel, if present, by performing a Ladd's procedure. An inversion appendectomy, rather than an amputation of appendix, will preserve the operative field as clean.
(vi) Close abdomen if possible. If there is insufficient domain in abdomen after reducing viscera from chest to abdomen, a silo or mesh closure of abdomen may be needed on a temporary basis.

(o) Outcome:

(i) Through the twentieth century, CDH has historically had 50 % survival, however advancements in neonatal intensive care unit techniques and select use of ECMO have produced a survival range of 39–95 % (mean, 69 %), depending on setting of care.
(ii) There is a risk of both early and late recurrence. Early recurrence can be due to technical error and increased abdominal pressure. Placement of mesh reduces tension across the repair, but approximately 1/3 of patients will need a late revision of repair due to recurrence of hernia or due to tension on the thoracic wall and deformation of the ribcage.
(iii) Chronic lung disease may result from the accompanying pulmonary hypoplasia.
(iv) Gastroesophageal reflux is common after CDH repair and some patients will require an anti-reflux procedure.
(v) Development/cognitive deficits occur and can be due to associated anomalies or as sequela from ECMO:

Esophageal Atresia and Tracheoesophageal Fistula

Ronald J. Scorpio

Esophageal atresia is an interruption of the normal formation of the esophagus. Classification of esophageal atresia is based on the presence or absence of a fistula to the trachea.

1. Pathophysiology:
   (a) Types of esophageal atresia/tracheoesophageal fistula:

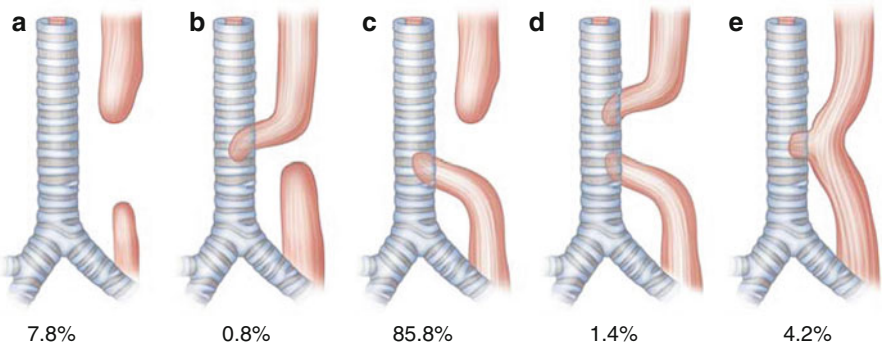

Fig. 1 Types of esophageal atresia and tracheoesophageal fistula. (**a**) Pure esophageal atresia without fistula; (**b**) proximal tracheoesophageal fistula, blind distal pouch; (**c**) distal tracheoesophageal fistula, blind proximal pouch; (**d**) proximal and distal tracheoesophageal fistula; (**e**) intact continuous esophagus with H-type tracheoesophageal fistula (*Source*: Bryan Walters)

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(i) Esophageal atresia with no fistula, 7.8 % (pure esophageal atresia).
(ii) B: Esophageal atresia and a proximal fistula, 0.8 %.
(iii) C: Esophageal atresia and a distal fistula, 85.8 % (most common type).
(iv) D: Esophageal atresia with both a proximal and a distal fistula, 1.4 %.
(v) Fistula to the trachea with no esophageal atresia 4.2 % (H-type fistula).

(b) Embryology:

(i) The esophagus and the trachea appear at three weeks gestation and develop into separate structures up to the level of the larynx by the 4th week of gestation. The defect occurs by the 20th week of gestation with rapid growth of the trachea. A vascular defect accounts for pure esophageal atresia.

(c) Incidence: 1 in 4,000–5,000 live births.
(d) Associated anomalies:

(i) Maternal polyhydramnios: 60 % of patients.
(ii) Cardiac: 30 % with patent ductus arteriosum, ventricular septal defect, or atrial septal defect.
(iii) Gastrointestinal: 12 % have imperforate anus, duodenal atresia or pyloric stenosis.
(iv) VACTERL constellation of anomalies: 20–30 % cases.

1. V: Vertebral.
2. A: Anorectal.
3. C: Cardiac.
4. TE: Tracheal-esophageal.
5. R: Renal.
6. L: Limb, such as absent radius.

2. Clinical presentation:

(a) Prenatal: polyhydramnios, seen more often with pure esophageal atresia.
(b) Symptoms: excessive drooling, choking, coughing, and episodes of cyanosis, especially with attempts to eat.

3. Diagnosis:

(a) Inability to pass a nasogastric tube.
(b) Chest x-ray with nasogastric tube coiled in upper pouch.
(c) Contrast study with either air or 0.5–1 ml of barium via a tube into the upper pouch.

4. Initial management:
   (a) Prevent aspiration: keep patient NPO, keep head up, place nasogastric or orogastric suction catheter in upper pouch.
   (b) Rule out associated abnormalities: physical examination of anus and limbs, sonography of kidneys, and cardiac echo (rule out anatomic abnormalities and determine on which side of chest aorta runs).
5. Treatment:
   (a) Preoperative antibiotics.
   (b) Surgical correction:
      (i) Right lateral thoracotomy, extra plural dissection (if possible), divide azygos vein, divide fistula, close trachea and reanastomosis proximal and distal ends of esophagus, place transanastomotic nasogastric tube, place right retropleural chest tube, close thoracotomy.
      (ii) Operation can be performed thoracoscopically in some centers.
      (iii) Some surgeons omit placement of a chest tube.
   (c) Postoperative care:
      (i) Early extubation, pain control, early enteric feeds via transanastomotic gastric tube and place chest tube to water seal if no air leak.
      (ii) One week after operation, obtain a contrast esophagram, via oral feeding of contrast with a bottle. If no leak is present, the nasogastric tube and the chest tube can be removed. If a leak is present maintain the tubes and repeat the study in 1 week.
6. Complications:
   (a) Gastroesophageal reflux disease. Approximately one third of patients who undergo repair of a tracheoesophageal fistula repair will require a Nissen fundoplication for reflux.
   (b) Tracheomalacia: evidenced by a barking cough present for up to one year after surgical correction of the tracheoesophageal fistula. Severe cases may require aortopexy.
   (c) Stricture: occurs in 30 % of repairs. Treat with dilatation of esophagus using Savary dilators in the operating room.
   (d) Recurrent fistula: treat with fibrin glue plug, cauterization of fistula, or repeat surgery if it fails to close with time and conservative measures.
7. Treatment strategy for pure esophageal atresia without fistula:
   (a) Diagnosis is the same as esophageal atresia with tracheoesophageal fistula; however abdominal x-ray will show a gasless abdomen.
   (b) Treatment:

(i) Maintain suction catheter in upper pouch and operative creation of gastrostomy. Obtain "gapogram": orogastric tube in upper pouch and dilator in lower pouch via gastrostomy. If gap between upper pouch and lower pouch is less than three vertebrae then child is a candidate for repair of esophagus by primary anastomosis. If distance between pouches is greater, than the patient has long gap esophageal atresia, and the optimum treatment is controversial.

(ii) Focker procedure is the placement of traction sutures on both ends of the esophagus and bringing them out through the chest wall. Postoperatively, tension is maintained on these sutures to elongate the esophagus until sufficient lengthening has occurred that primary anastomosis is possible. The esophagus may be also be elongated by attaching it under tension to the spine. The esophageal pouches are elongated in this manner until they can tolerate a primary anastomosis. This may require several operations.

(iii) If esophageal lengthening procedures fail, the esophagus can be operatively replaced with stomach, small bowel or colon.

Pyloric Stenosis

Christopher P. Coppola

Pyloric stenosis is a very common malady of newborns, and is characterized by non-bilious vomiting after meals, which becomes projectile as the stenosis worsens. Before the option of surgical repair, infants with this illness would often die of dehydration and starvation.

1. Hypertrophy of the pylorus in some newborns and infants causes gastric outlet obstruction and vomiting of feeds.
2. In pyloric stenosis the pylorus is thickened and remains in spasm. Hypertrophy develops over several weeks with worsening vomiting. The pyloric lumen is narrowed and eventually remains shut.
3. Pyloric stenosis usually occurs at 4–6-weeks-old, but has been reported in utero and in infants as old as 4-months-old. It affects one in 400 infants and it has a male to female incidence ratio of four to one. It occurs more commonly in first-born children and has a 1–5 % incidence in children of affected individuals.
4. Children with pyloric stenosis present with projectile vomiting of feeds immediately or shortly after meals. Of note, vomitus is non-bilious. (Bilious vomiting requires an emergent investigation for malrotation/volvulus.) Volume and intensity of vomiting increase over a course of 2 weeks. Infants are continuously hungry and may become dehydrated and lethargic.
5. Evaluation:

   (a) Infants with a prolonged course of vomiting may appear dehydrated. The diaper should be checked for clear urine and inguinal hernia. In late stages, there is cachexia. If the child is observed to eat, vigorous gastric succussion waves may be seen or heard in the abdomen and projectile vomiting may be witnessed. However, it is best to avoid feeding so the stomach is empty if an operation is indicated. An enlarged pylorus can be palpated as a firm but

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mobile olive-shaped 1–2 cm mass that lies transversely in the right upper quadrant. Sliding the examining fingertips vertically up or down will best detect this transverse mass. Although sonography has supplanted definitive diagnosis by physical examination, succussion waves can be amplified by emptying the stomach with an 8 Fr. nasogastric tube and letting the child suck on a pacifier or moist sponge dipped in sugar.

(b) Electrolytes must be checked for the common disturbances of hyponatremia, hypochloremia, hypokalemia, and alkalosis. If tested, urine may be concentrated and show paradoxical aciduria as H+ is sacrificed to preserve K+ after hypokalemia develops.

(c) Sonography of the pylorus is the test of choice. It will reveal a single wall muscle thickness of 3.5 mm or greater and a pylorus length of 13 mm or longer. Additionally the pylorus will have a characteristic bowed-outward shape, and little or no succus will be seen to pass. If performed, upper gastrointestinal fluoroscopy shows a narrow or absent pyloric channel and pylorus muscle bulges into the stomach and duodenum.

6. Treatment is operative pyloromyotomy.

(a) Infants must be well hydrated and have all electrolyte abnormalities corrected before undergoing operation. In advanced cases, several days' correction may be needed. Infants are NPO and are given intravenous D5 ½ NS+ 10 meq/L KCl at 1.5 maintenance rate. Repeat electrolytes until normal and ensure normal urine output.

(b) Pyloromyotomy can be performed via laparoscopic or open approach. Laparoscopic approach requires a laparoscope through the umbilicus and two instruments directly through the upper abdominal wall [1]. Open incision can be traditional transverse right upper quadrant or curvilinear supraumbilical with a superior skin flap and midline separation of the recti muscles along the linea alba. The latter incision leaves less of a scar in adulthood, but is avoided when there is omphalitis. The pylorus is delivered out of the wound, which can be the most difficult part of the case. The serosa overlying the pylorus is incised longitudinally, in between the vasculature from lesser and greater curvatures. Incision starts over normal gastric antral muscle and stops just proximal to the first portion of duodenum. The hypertrophic pyloric fibers are split bluntly using the back of a scalpel handle or by spreading, until the submucosa bulges out and the upper and lower cut edges of pylorus slide freely against each other. Care must be taken at the distal margin because the muscle abruptly thins over the duodenum and a full-thickness injury is easily made. If the duodenal mucosa is injured, it can be closed with an absorbable stitch, or the entire anterior pyloromyotomy can be closed with a subsequent pyloromyotomy performed on the posterior surface of the stomach. When pyloromyotomy is complete, pylorus is returned to the abdomen and abdominal wall is closed.

(c) All infants vomit after pyloromyotomy. Intravenous maintenance fluid is given until oral feeds are tolerated. Some practitioners delay feeds several

hours or start with dilute feeds, but allowing usual feeds immediately works equally well. The child can be discharged once two subsequent oral feeds of at least 60 mL each are tolerated. Oral acetaminophen or ibuprofen is used as needed for pain. Infants with sustained fever or peritoneal signs should have a rapid investigation for an unrecognized duodenal injury with water soluble upper gastrointestinal fluoroscopy. Infants who vomit for more than 2 weeks should undergo upper gastrointestinal fluoroscopy to detect incomplete pyloromyotomy.

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Duodenal Atresia

Christopher P. Coppola

Newborns with intolerance of feeds and vomiting may have congenital blockage of the duodenum, a condition strongly associated with Trisomy 21. The characteristic diagnostic finding is a double bubble pattern of intestinal gas on abdominal x-ray.

1. Pathophysiology:

   (a) Normal embryology: development of the duodenum begins in the fourth week of gestation. The primitive digestive tube formed from the endoderm rotates to the right, with the distal portion being drawn to the left as the midgut rotates. During the fifth to seventh weeks of gestation, the duodenum is temporarily obstructed by epithelial proliferation, and it recanalizes by degeneration of epithelial cells during the eighth week.
   (b) The duodenum can be obstructed internally by atresia, a duodenal web, or a stenosis. Duodenal webs, or diaphragms, can vary in thickness, some are long and prolapse distally, like a wind sock, and some webs have an aperture that allows flow of succus, so they do not cause complete obstruction. These partially obstructing webs may not become apparent until later when solid foods are introduced. External obstruction can be caused by an annular pancreas, malrotation and volvulus, or presence of a duplication cyst.
   (c) Duodenal atresia usually occurs near the ampulla of Vater. In 80 % of children it is distal to the ampulla, resulting in bilious vomiting. In 5–10 % of cases it is proximal, with non-bilious vomiting. Some cases will have the atresia between the primary and secondary biliary ducts.
   (d) Classification of duodenal atresia

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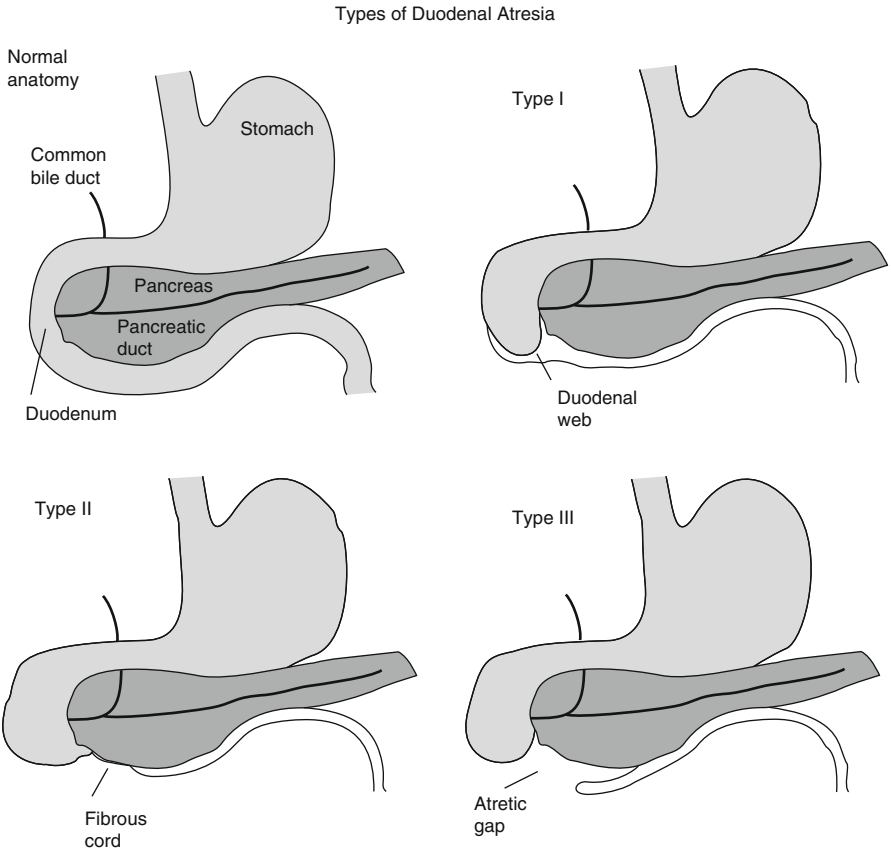

Fig. 1 Classification of duodenal atresia (*Source*: Christopher Coppola)

(i) Type I: This type is most common, with a luminal web or membrane, consisting of mucosa and submucosa.
(ii) Type II: Duodenum is reduced to a fibrous cord connecting the proximal and distal segments
(iii) Type III: There is discontinuity of the duodenal segments with no intervening tissue.

2. Epidemiology: intrinsic duodenal obstruction occurs in 1:7,000 live births. It represents 49 % of small bowel atresia.

   (a) There is a strong association with trisomy 21; if duodenal atresia is detected, a karyotype should be performed.
   (b) Other associated conditions are Hirschsprung's disease, Meckel's diverticulum, esophageal atresia, imperforate anus, congenital heart disease, renal abnormalities and neurologic disorders.

3. Clinical features:
   (a) One third to one half of fetuses with duodenal atresia will have polyhydramnios from inability to pass amniotic fluid through the gastrointestinal tract.
   (b) After birth, newborns with duodenal atresia experience feeding intolerance, distention, discomfort, and vomiting, which is usually bilious.
   (c) Children may pass meconium and have a normal rectal exam.
   (d) Features of trisomy 21 will frequently be present.
4. Assessment:
   (a) Abdominal x-ray will reveal a double bubble sign formed by distention of the stomach proximal to the pylorus, and distention of the obstructed first and second portion of the duodenum, proximal to the duodenal atresia. Distal bowel gas will be present in cases of partial obstruction and rare cases of complete atresia when air can pass from proximal to distal via the papilla of Vater and the secondary papilla.
   (b) Upper gastrointestinal series fluoroscopic study can confirm the diagnosis and differentiate duodenal atresia from malrotation with midgut volvulus.
5. Initial resuscitation:
   (a) Admission to a NICU with IV placement and fluid resuscitation.
   (b) Electrolytes are checked and abnormalities from losses through vomiting are corrected.
   (c) Nasogastric tube is placed for decompression of stomach.
   (d) Evaluation for associated anomalies with echocardiography and renal sonography.

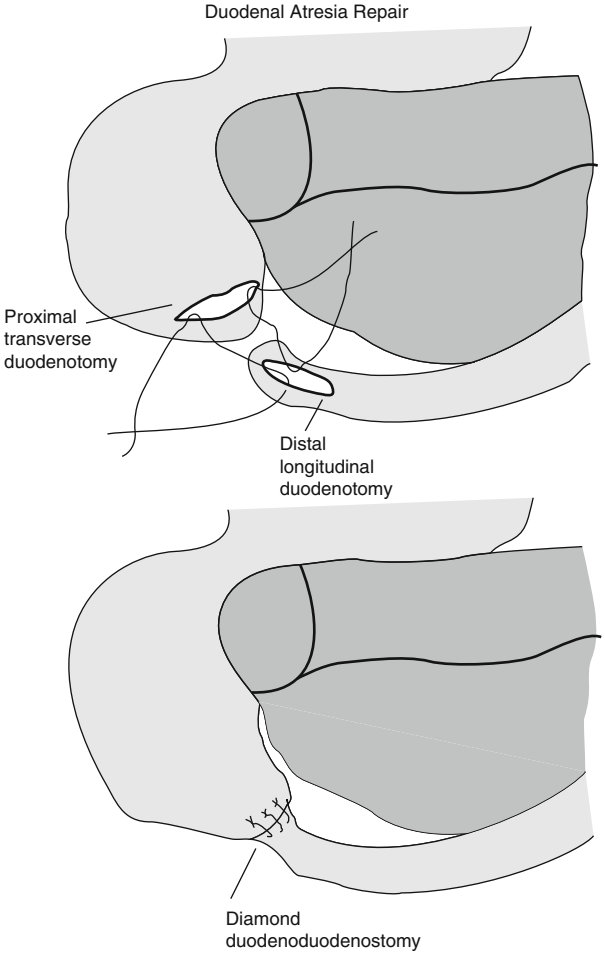

Fig. 2 Diamond duodenoduodenostomy repair of duodenal atresia (*Source*: Christopher Coppola)

6. Treatment – operative repair:

   (a) Repair is performed under general anesthesia after resuscitation. A transverse upper abdominal laparotomy is performed. Bowel is evaluated for malrotation, volvulus, and any other atresias or anomalies, including Meckel's diverticulum. The proximal dilated duodenum is anastomosed to the distal decompressed duodenum. A diamond (Kimura) duodenoduodenostomy is created with a transverse incision on the most dependent portion of the proximal duodenum. This is anastomosed to a longitudinal incision on the distal duodenum with the two incisions oriented perpendicularly so that the aperture is held open to encourage flow of succus into the distal duodenum.

(b) Care is taken to avoid injury of the papilla of Vater by using the portions of duodenum furthest from the interface with the pancreas.
(c) When duodenal web is present, the duodenum is opened and the web is resected, taking care not to injure the ampulla of Vater which may form one portion of the web. If it is not clear that a web is present, the nasogastric tube can be advanced through the duodenum until it stops or causes dimpling of the walls of the duodenum as the web is pulled distally by the tip of the tube.
(d) If annular pancreas is present, a duodenoduodenostomy is performed as a detour around the obstruction, without resection or division of the pancreas.

7. Recovery:

(a) IV hydration is continued until the patient can tolerate enteral feeds.
(b) A tube passed through the anastomosis at the time of surgery can be used to feed the gut distal to the duodenum. If a patient has post-operative vomiting, a nasogastric tube via the other nare can be used to decompress the stomach.
(c) Prolonged postoperative ileus is present, and if the patient cannot tolerate oral feeds within 2 weeks after repair, an upper gastrointestinal series is performed to evaluate the anastomosis.

8. Outcome:

(a) Some patients who fail to empty the stomach through the repair will require revision of anastomosis.
(b) Patients may have long term delayed gastric emptying and gastroesophageal reflux.
(c) In some cases, stasis of succus in a floppy redundant proximal duodenum with bacterial overgrowth will require reoperation for a tapering duodenoplasty. (Some surgeons will perform this at the time of initial operation.)
(d) Mortality is <5 %.

Intestinal Atresia

Christopher P. Coppola

Intestinal atresia is a congenital discontinuity of the gut which results in obstruction and vomiting in neonates. Continuity of the bowel can usually be restored, but the there is a risk of short gut syndrome and inability to survive on oral feeding alone.

1. Epidemiology:
   (a) Incidence: Intestinal atresia occurs in one in every 5,000 live births
   (b) Etiology: Intestinal atresia is thought to be the result of mesenteric vascular occlusion during gestation. The segments of intestine without blood flow during development become atretic.
2. Pathophysiology:
   (a) Location: Any part of the gastrointestinal tract can be affected. The terminal ileum is most common and atresia of the colon is rare.
   (b) Classification:

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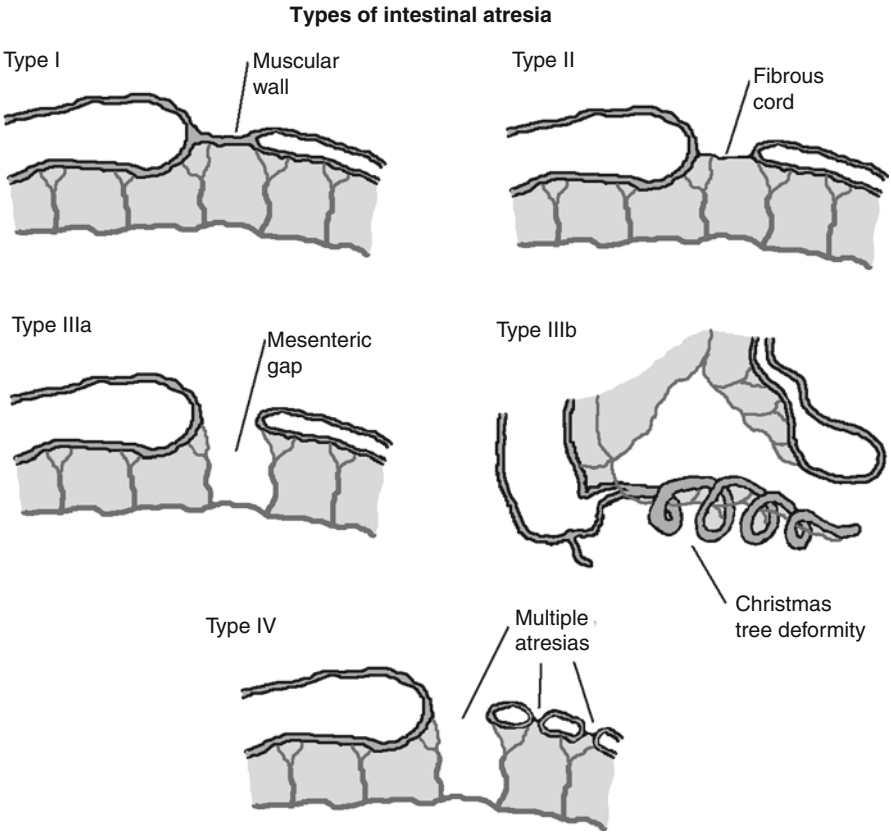

Fig. 1 Types of intestinal atresia. In *Type I*, there is a muscular wall without lumen connecting the two ends of atretic bowel. In *Type II*, they are connected by a fibrous cord. In *Type IIIa*, there is a wedge-shaped gap in the mesentery between the two ends. *Type IIIb* is a severe form of the disease in which the only distal bowel present is spiraled around a branch of the ileocecal artery. *Type IV* is multiple atresias (*Source*: Christopher Coppola)

(i) Type I: Atretic bowel has no lumen, but muscular wall is intact.
(ii) Type II: The atretic segment is a fibrous cord, with no muscular wall.
(iii) Type III: There is absence of the bowel and a wedge-shaped gap in the mesentery.

1. Type IIIa: simple defect of limited length.
2. Type IIIb: This is the dreaded "Christmas tree" or "apple peel" deformity where the bowel ends blindly shortly after the ligament of Treitz. The only distal bowel present is a stenotic spiral wrapped around the ileocecal artery like the coiled peel from an apple. The base of the coil is wider than the tip, giving the cone-shaped appearance of a Christmas tree. Patients with this defect will often have short gut syndrome.

(iv) Type IV: Multiple segments of intestinal atresia.

3. Clinical features: patients will present shortly after birth with distention and bilious vomiting. If the atresia is distal, it may take several meals before the gut fills enough for there to be vomiting. Patients may have oliguria and other signs of dehydration and electrolyte imbalances. Perforation can occur and patients will have signs of sepsis and shock. As the atresia of intestine can happen mid gestation, it is possible for there to be meconium corresponding to succus that passed through the gastrointestinal track before obstruction occurred.
4. Diagnosis:
   (a) Prenatal sonography can reveal distended loops of bowel and polyhydramnios because the fetus is unable to pass swallowed amniotic fluid through the gastrointestinal tract.
   (b) Abdominal x-ray: patients who vomit should be evaluated with x-ray. It will reveal distended loops of bowel and air fluid levels consistent with obstruction. If perforation has occurred, there may be free air.
   (c) Fluoroscopy with contrast can reveal the site of obstruction. When an infant presents with bilious vomiting, it is often a dilemma deciding between upper gastrointestinal series and contrast enema. As the most important lesion to diagnose and treat rapidly is malrotation with midgut volvulus, it is usually better to obtain upper gastrointestinal series first to rule out this condition. Contrast enema can be obtained after upper gastrointestinal series, but it may be limited in interpretation due to contrast in proximal gut.
5. Treatment:
   (a) Patients should be made NPO and a nasogastric tube is placed to decompress the stomach.
   (b) Intravenous access is established, patients are given resuscitative and replacement fluid administration. Any electrolyte abnormalities are corrected.
   (c) Intestinal atresia is repaired with laparotomy and reanastomosis of the two blind-ending loops of bowel.
   (d) Transverse incision superior to the umbilicus will give the best view of the gut from duodenum to rectum.
   (e) Bowel is run to evaluate for location of atresia or possible atresias and to determine if any other intra-abdominal abnormality is present.
   (f) The bowel is anastomosed using an end (of proximal gut) to back (antimesenteric surface) of distal gut. The most dependent portion of proximal gut is used to encourage emptying. Sometimes tapering enteroplasty is needed for extremely dilated gut, but this can be performed at a future operation only if needed. The distal gut is much smaller in diameter, so an antimesenteric slit is made until the aperture matches the diameter of the proximal gut. Prior to closure of anastomosis, a feeding tube is threaded into distal gut to irrigate with saline to prove that there are no additional obstructions downstream.
   (g) There is often a long delay to oral feeding after repair of intestinal atresia, so consideration is made for a central line for parenteral access.
   (h) A nasogastric tube should be placed for postoperative decompression of the stomach while waiting for bowel function.

Hirschsprung's Disease

Christopher P. Coppola

Hirschsprung's disease is a congenital absence of ganglion cells in the distal gastrointestinal tract. The length of affected intestine varies and treatment requires resection of abnormal intestine and pull-through of normal intestine with ganglion cells to the anus.

1. Pathophysiology:

   (a) Hirschsprung's disease is characterized by absence of ganglion cells in distal bowel. The segment without ganglion cells is continuous and ends at the transition zone just above the dentate line. The lack of ganglion cells results in an inability of affected bowel to undergo receptive dilation. Theories of etiology include:

      (i) Failed migration of ganglion cells along the gastrointestinal tract.
      (ii) Hostile micro-environment causing ganglion cell loss.
      (iii) Auto-immune attack of ganglion cells.

   (b) The length of aganglionic colon varies from child to child. Short segment disease from 1 to 3 cm is most common. Length can range up to long segment disease including total colonic aganglionosis, and total intestinal aganglionosis which are more severe and have a higher mortality rate.
   (c) The disease occurs in one in 5,000 births, and has a five to one male to female ratio. It is extremely rare in premature neonates.
   (d) Associated conditions include trisomy 21 (10 %), multiple endocrine neoplasia (MEN) 2A, Intestinal atresia, neuroblastoma, and von Recklinghausen's disease. Genetic defects in Hirschsprung's disease have been traced to the RET and EDN pathways. Six percent of cases of Hirschsprung's disease are familial.

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2. Clinical Presentation:
   (a) Any child who does not pass meconium in the first 24 h of life should be suspected to have Hirschsprung's disease.
   (b) The child may have distention, feeding intolerance, bilious vomiting, fever, and sepsis. In some cases there will be signs of intestinal perforation.
   (c) On rectal examination, there is often a characteristic blast of liquid stool as the examining finger releases the blockage caused by the spasm in the aganglionic segment. This is typical of short segment disease, which is the most common pattern.
   (d) Two thirds of patients with Hirschsprung's disease are diagnosed in the first 3 months of life, while others present late. They often have short segment disease. They have a history of chronic constipation, abdominal distention, recurrent fecal impaction, rectal prolapse, dehydration, malnutrition, failure to thrive anemia, hypoproteinemia. They may have had recurrent bouts of enterocolitis, marked by abdominal pain, fever, and vomiting.

3. Diagnosis: Hirschsprung's disease will be suspected when there is no meconium passage in the first 24 h, but there are many causes of distention and vomiting, so testing will be necessary to differentiate the cause.
   (a) Abdominal x-ray: reveals distended loops of bowel, and if present, free air from perforation.
   (b) Contrast enema: if perforation is suspected, water soluble contrast should be used. Barium gives better image definition. Contrast enema will reveal a transition zone in Hirschsprung's disease: a focus where diameter changes from narrow distally to dilated proximally due to the fact that the affected distal bowel has abnormal spastic contractions. If the patient has undergone a rectal exam, the transition zone may not be apparent. In neonates in particular the transition zone can be difficult to see and is not apparent in 25 %. The involved segment often has irregular spastic contractions. The enema can reveal alternated diagnoses such as meconium plug syndrome and neonatal small left colon syndrome. Contrast retained in the colon longer than 24 h is abnormal.
   (c) Invertogram (not commonly used): Antero-posterior abdominal x-ray with child upside down or transverse x-ray with patient prone reveals failure of air to rise into the rectum in superior position.
   (d) Echocardiogram to rule out associated cardiac anomaly.
   (e) Karyotype to detect genetic anomaly; trisomy 21 is frequently associated.
   (f) Rectal biopsy with the absence of ganglion cells of the Auerbach and Meisner plexi of the muscularis is the definitive diagnosis of Hirschsprung's disease.
      (i) Biopsy should be performed at least 1.5 cm above the dentate line to avoid a biopsy from the normal hypoganglionic region.
      (ii) Biopsy should be performed within 3 cm of the dentate line to avoid missing a very short segment of disease.

(iii) Below age 3-months-old, beside suction rectal biopsy using a suction rectal biopsy tool will give a specimen of adequate depth to include the muscle. Above that age, suction rectal biopsy can be attempted, but it is likely that a full thickness rectal biopsy in the operating room under anesthesia will be required for a specimen of adequate depth.
(iv) After biopsy, patients cannot have rectal exam, temperature, or medication for 24 h.
(v) Complications: hemorrhage requiring transfusion: 1 %, perforation <1 %.

(g) Rectal manometry: Useful in older children to determine if there is a zone of increased pressure and absence of relaxation with proximal distention.

4. Initial management:

(a) Make the patient NPO, start intravenous fluid hydration and resuscitation, and place a nasogastric tube to suction if patient has been vomiting.
(b) If the patient shows signs of enterocolitis, perforation, or sepsis, start broad spectrum antibiotics.
(c) Give rectal irrigations: flush the rectum using an 8 Fr. Feeding tube with 15 mL of saline in three divided doses of 5 mL, attempting to evacuate stool. Older children may benefit from placement of a rectal tube to bypass the affected segment.

5. Treatment: Definitive treatment of Hirschsprung's disease is resection of the aganglionic distal bowel, with pull-through of normal proximal bowel to an anastomosis at the anus.

(a) The surgeon must decide between primary pull-through, as a single operation verses a staged pull-through, following an initial leveling colostomy, proximal to the affected segment. Patients with short segment disease who can be made to stool on a regular basis with the use of rectal irrigations will usually be safe for a primary pull-through. Patients with long segment disease who cannot stool even with rectal irrigations will require a leveling colostomy. Other contraindications to primary pull through are massive dilation, enterocolitis, perforation, malnutrition, and inability to determine transition zone.
(b) Leveling colostomy is performed through a transverse lower laparotomy, and colostomy site is chosen 5 cm above the transition from dilated to narrow colon, usually in the sigmoid colon. Data from visual inspection, and contrast enema can be uses to find the proper level, but biopsy with ganglion cells present is the only way to be certain that normal colon has been chosen for colostomy site.
(c) Timing of operation depends on the length of disease and surgeon preference. Primary pull-through can happen in the first days of life, or can be delayed several weeks or months with parents providing rectal irrigations three times each day at home. Staged pull-through should happen after child has recovered from colostomy and has been able grow, approximately 3 months.

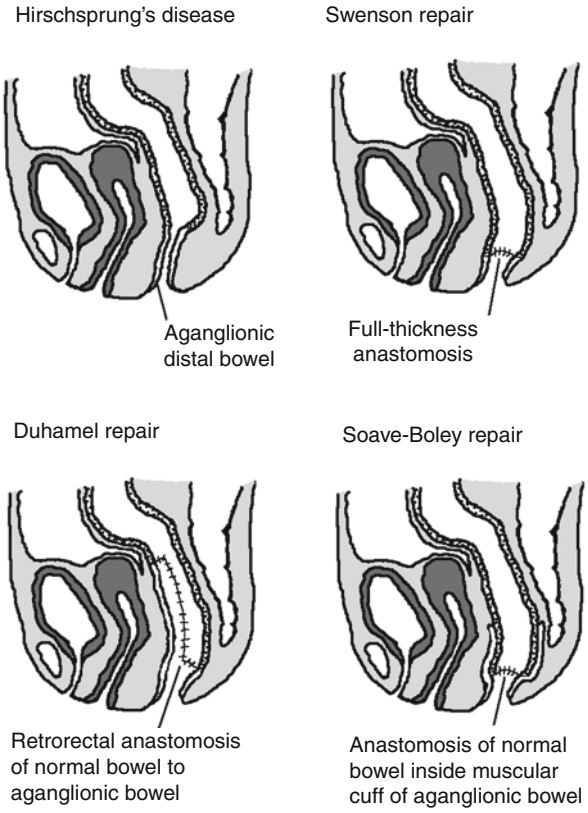

Fig. 1 Types of pull-through repair for Hirschsprung's disease. All repairs pull-through normal bowel with ganglion cells down to the dentate line. In the Swenson repair, a full thickness anastomosis of normal bowel is made. In the Duhamel repair, normal bowel is brought down behind the rectum and an anastomosis is created to the back of aganglionic bowel. In the Soave-Boley repair, mucosa is removed from the aganglionic bowel leaving a muscular cuff, and normal bowel is pulled through this cuff to create an anastomosis (*Source*: Christopher Coppola)

(d) Types of pull-through:

(i) Rectosigmoidectomy (Swenson): Resection of the rectum, eversion of the rectal stump outside anus, and end to end anastomosis between end of normal colon, and rectal stump.

(ii) Retrorectal pull-through (Duhamel): Anastomosis of the normal colon to the posterior surface of the aganglionic rectal stump. This is often done with a stapler inserted in the anus to perform a side to side anastomosis between normal colon and rectum.

(iii) Endorectal pull-through (Soave-Boley): Submucosal dissection is performed via anus to lift the mucosa off of the aganglionic rectum. This leaves behind a cuff of muscle above the anus. The cuff of muscle is splint to the front and back to eliminate constriction. The normal colon is brought through this split cuff and anastomosed to the edge of mucosa above anus.

(iv) Laparoscopic pull-through (Georgeson): Normal colon is identified by laparoscopic muscle biopsy. Abnormal colon is pulled out via the anus, and end to end anastomosis is made via the anus.

(e) Other procedures:

(i) Myomectomy: This can be performed on very short segment disease to incapacitate the spasm of the aganglionic segment.
(ii) Prolapse repair is sometimes needed after pull-through.
(iii) Anal sphincter Botox injection can be used when patients have constipation after pull-through.

6. Outcome:

(a) There is 95 % survival in Hirschsprung's disease; survival is worse in total colonic and total intestinal aganglionosis.
(b) Ninety percent of children have good bowel function, but they may require laxatives or a bowel regimen to achieve this.
(c) One percent will have permanent colostomy.
(d) Symptoms improve with age, and outcome is worse in Trisomy 21.
(e) Complications:

(i) Enterocolitis occurs in 15 % of children and is the leading cause of death in Hirschsprung's disease. It occurs before and after pull-through and is more common in long segment disease. It is due to overgrowth of bacteria in areas of stasis and is characterized by explosive diarrhea, fever, distention, shock. Treatment of enterocolitis involves admission to hospital, fluid resuscitation, broad spectrum intravenous antibiotics, and rectal irrigation. Some patients will require anal dilation, re-do of pull-through or colostomy. When performing colostomy after pull through, care must be taken to preserve blood flow to the distal colon as the hemorrhoidal collaterals are no longer present.
(ii) Other complications are: constipation (10 %), wound infection (10 %), incontinence (5 %), anastomotic leak (5 %), anastomotic stricture (5 %), intestinal obstruction (5 %), and pelvic abscess (5 %).

Malrotation

Christopher P. Coppola

Malrotation is an abnormality of the relative positioning and fixation of organs in the abdomen. It creates the potential for midgut volvulus, a twisting of the intestines which can result in obstruction, ischemia, and catastrophic loss. Volvulus can present at any age, is marked by bilious vomiting, and requires immediate detection and correction.

1. Pathophysiology:

   (a) Epidemiology: malrotation is likely present in many people who never know it. Unless there are symptoms from volvulus or laparotomy for another reason, it will be clinically silent. Most cases of volvulus occur in the very young, but it has been reported in people as old as their 70s.

   (b) Anatomy: with normal rotation and fixation of the midgut, the proximal bowel at the duodenum undergoes a 270° clockwise rotation (from the perspective of looking at the patient) and the colon undergoes a 270° counter clockwise rotation. There is fixation of the duodenum to the retroperitoneum with the ligament of Treitz located in the left upper quadrant of abdomen. There is fixation of the right colon with the cecum in the right lower quadrant and the left colon from the splenic flexure to the left lower quadrant. The mesentery will then arise in an oblique path from the ligament of Treitz to the terminal ileum, in a straight line from the left upper quadrant to the right lower quadrant. This gives the mesentery a broad base which cannot undergo volvulus. In malrotation and non-fixation of the bowel, the mesentery has a narrow base and can easily undergo volvulus around the axis of the superior mesenteric artery. Instead of a ligament of Treitz and fixation of the cecum in the right lower quadrant, abnormal Ladd's bands form, connecting the duodenum to the cecum. When volvulus occurs, it usually happens in a clockwise fashion. With

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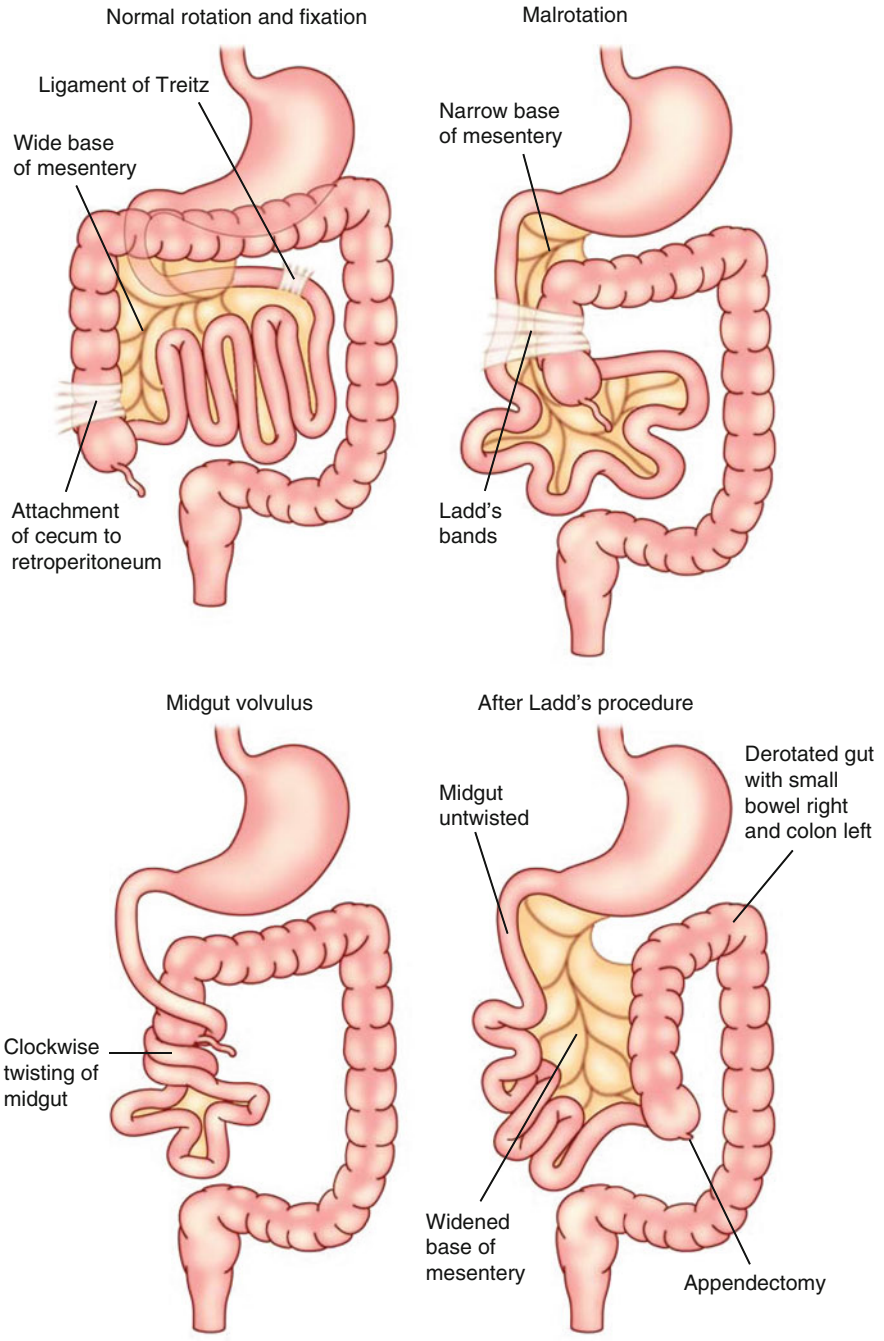

Fig. 1 Malrotation and midgut volvulus. In normal development, the midgut is fixed to the retroperitoneum and the mesentery has a wide base. In malrotation, the narrow base of mesentery can undergo volvulus, and requires repair with the Ladd's procedure (*Source*: Christopher Coppola)

one rotation of the midgut there is obstruction of the lumen, with further twisting, the blood supply is compromised and ischemia of the midgut occurs.

(c) Situs inversus: with suits inversus and heterotaxy, there is often malrotation and non-fixation. Other abnormalities may be present such as asplenia and central globular liver. Of note, when situs inversus is present, volvulus will usually occur with a counter-clockwise rotation of the midgut, which differs from the volvulus when the organs are on the usual side of the body.

(d) There is much variation in malrotation between individual patients. Different segments of the midgut may be free to undergo volvulus and sometimes there is partially complete fixation of the bowel. As an example, even if the cecum is located in the right lower quadrant, it does not guarantee that the proximal gut has undergone normal fixation. Some abnormalities of fixation can cause kinked or tortuous arrangements of the duodenum which can result in obstruction.

(e) Malrotation is often called the "ubiquitous anomaly" because it frequently accompanies other congenital abnormalities. Conditions where the midgut does not return to the abdomen during gestation, such as diaphragmatic hernia, gastroschisis, and omphalocele, will usually involve malrotation and non-fixation of gut.

2. Clinical presentation:

(a) The typical presentation of malrotation is bilious vomiting. It is essential that this history is elicited from the child's parents. They should be asked the color of vomitus, and if it is present on clothing or a towel it can be inspected. Any account of bilious vomiting should raise the possibility of malrotation with volvulus and requires immediate evaluation.

(b) The child may have a scaphoid abdomen resulting from proximal obstruction of the intestine at the duodenum. Inability to pass stomach contents past this point keeps the intestine empty. If vomiting has gone on for a prolonged period, the patient will be dehydrated, anuric, and lethargic or in hypovolemic shock.

3. Diagnosis: there are many causes for bilious vomiting from infectious to neoplastic, but the cause that must be rapidly excluded is malrotation. Upper gastrointestinal series is the most accurate method to determine if it is present.

(a) Upper gastrointestinal series: a radiologist experienced in determining rotation of bowel should be consulted. Barium contrast gives the best definition, but water soluble contrast can be used if there is a chance of perforation, to avoid the irritation caused by extravasated barium. If the child is awake, he or she can drink the contrast from a bottle, otherwise, it can be instilled via a nasogastric tube.

(i) There are three criteria for normal rotation of gut. If any of them are absent, the child has malrotation.

1. Passage of duodenum from left to right, then back again.
2. On transverse view, duodenum is posterior to anterior line of vertebral column.
3. On anterior view, the distal duodenum rises up as high as the level of the greater curvature of stomach.

(b) Abdominal x-ray: may show distended stomach and absence of distal gas, however twisting and untwisting of volvulus can allow some gas to pass distally. If there is a nasojejunal tube present, the abnormal course of the duodenum can be seen.
(c) Sonography: experienced practitioners can identify the reversed position of the superior mesenteric artery and the superior mesenteric vein.
(d) Computed tomography: volvulus can be identified as a swirling of mesenteric blood vessels and proximal intestinal obstruction.

4. Initial management: nothing should delay bringing the child to the operating room. When outpatients are discovered to have malrotation, they should be admitted to the hospital. While waiting for operation, a nasogastric tube should be placed and attached to suction. Intravenous fluid is administered and any electrolyte abnormalities are resuscitated.
5. Treatment of malrotation and volvulus is the Ladd's procedure.

(a) Components of the Ladd's procedure:

(i) Untwist bowel: the bowel usually undergoes volvulus in a clockwise direction (from the perspective of the surgeon.) It is untwisted by rotating it in a counter-clockwise direction, akin to unscrewing a screw, or turning back time on a clock. In situs inversus, the direction of volvulus is usually reversed.
(ii) Divide Ladd's bands: these are clear adhesive bands binding the duodenum and cecum together. With these organs in close proximity, the mesenteric blood supply is contained in a slender stalk which can easily undergo volvulus. There are variations in the Ladd's bands, and they should be carefully retracted and divided to unwind any kinking of the duodenum and spread the cecum far from the duodenum, which widens the base of the mesentery.
(iii) De-rotation of midgut: small bowel is located on the right side of the abdomen and colon is located on the left. Organopexy is not performed: it is not useful. After operation, adhesions develop which keep the loops of intestine in place.
(iv) Appendectomy: Since the appendix will be paradoxically located in the left abdomen after Ladd's procedure, it is removed to eliminate the possibility of future appendicitis with misleading wrong-sided pain. In children younger than 3 months old, an inversion appendectomy can be performed by dividing the mesoappendix with cautery, then inverting the appendix into the cecum with a parotid probe. The last 5 mm of the inverted appendix is ligated, and it auto-amputates into the lumen of the cecum after surgery. This method prevents converting a clean operative field to a contaminated one.

(b) Operative approach: Ladd's procedure can be performed by open or laparoscopic approach. There is some controversy based on the fact that laparoscopic approach may not generate sufficient adhesions to keep the bowel in the de-rotated arrangement after surgery.

(c) Futile cases: if the entire midgut is dead when the abdomen is opened, at the discretion of the surgeon in consultation with the child's family, the abdomen is closed without resecting the necrotic material. Removal of the entire midgut may prolong life for a short while, but it creates a scenario with very poor chances of survival.

6. Outcomes:

(a) Complications:

(i) Loss of major portions of the intestine due to ischemia caused by volvulus results in short gut syndrome and dependence on parenteral nutrition.
(ii) The Ladd's procedure intentionally creates intra-abdominal adhesions to maintain the de-rotated orientation of intestine. Adhesions can result in small bowel obstruction at any point in the child's future life.
(iii) Death: cases of volvulus which are not treated in a timely fashion will result in midgut ischemia, necrosis, sepsis, and death.

Gastroschisis

Alysia A. Agnoni

Gastroschisis, a cleft in the abdominal wall which allows the gut to eviscerate, is frequently diagnosed with prenatal ultrasound. This congenital defect has been increasing in incidence in the past few decades, and newborns with gastroschisis will require the assistance of a pediatric surgeon and a neonatologist.

1. Pathophysiology: Gastroschisis is a defect in the abdominal wall located to the right of the umbilical cord. Protrusion of abdominal contents occurs, usually bowel, and sometimes gonads. Children with gastroschisis always have non-rotation of the gut. There is no peritoneal sac covering abdominal contents, but sometimes a rind of inflammatory tissue will form in response to exposure of the amniotic fluid.
   (a) Embryology of gastroschisis is controversial. The defect may represent a ruptured omphalocele or could be the result of involution of a second umbilical vein.
   (b) Epidemiology: associated with young maternal age and intestinal atresia
2. Diagnosis: usually by fetal sonography after 20 weeks gestation.
3. Treatment:
   (a) Vaginal birth is still possible.
   (b) Stabilize the newborn:
      (i) Examine bowel for perfusion and perforation. Protect exposed bowel and prevent heat and fluid loses; this is usually accomplished by placing a bowel bag up to axillae. Later placement of silastic silo may be needed. The bowel is typically inflamed and matted due to exposure to amniotic fluid.
      (ii) NPO and NGT for gastric decompression and associated ileus.

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Fig. 1 Newborn infant with gastroschisis. The gastroschisis defect in the abdominal wall occurs to the right of the umbilicus and there is no sac covering the viscera (*Source*: Centers for Disease Control and Prevention, Atlanta, GA. http://www.cdc.gov/ncbddd/birthdefects/Gastroschisis-graphic.html. Downloaded 17 Dec 2013)

(iii) IVF resuscitation: may require two to three times usual maintenance due to excessive fluid losses from exposed bowel.
(iv) Broad-spectrum antibiotic coverage.
(v) TPN for nutrition.

(c) Operative repair: Patient and scenario dependent, but primary closure is often successful (50 %).

(i) Some defects may be too large (40–50 %) for reduction into the abdomen. This could result in decreased cardiac output, respiratory distress, renal failure, bowel ischemia, and decreased perfusion to the lower extremities due to abdominal compartment syndrome when reduced.
(ii) Goal should be an abdominal compartment pressure of less than 20 mmHg after reduction of bowel. If this is not immediately possible the newborn will remain in the NICU with a silo protecting the bowel. The top of the silo is suspended perpendicular to the supine infant and secured to the top of the warmer. Gravity and gentle pressure allow the intestine to fill the abdomen over time (usually within two weeks). The bowel should be examined frequently for perfusion and perforation. Antibiotic ointment should be applied around skin edge.
(iii) Reduction and closure:

1. When there is enough intra-abdominal space to allow for complete reduction of intestine.
2. The bowel should be inspected for associated atresia. If identified, atresia can be repaired at the time of primary closure or, more likely, a diverting stoma can be created and takedown and anastomosis undertaken at a later date.
3. The bowel may be too matted to allow for close examination at the time of reduction.
4. A general anesthetic is used for sedation and the abdomen is prepped. The bowel is carefully reduced into the abdominal compartment. The natural opening may need to be enlarged. The fascial edges are then approximated with interrupted sutures. The subcutaneous and skin layers are then closed separately. An umbilicoplasty may be done at this time as well.

(d) Postoperative:

(i) Expect delayed intestinal mobility/ileus.

1. TPN will need to be maintained until the infant tolerates full feeds – this may take more than a month and the infant is at risk for TPN associated cholestasis.
2. Maintain NGT until output decreases.
3. Feeds should begin once clamping of NGT is successful and does not result in emesis. Low volume feeds are started and gradually increased over several days to goal. TPN should be continued until the infant tolerates goal feeds by mouth.

(e) Complications:

(i) Bowel obstruction – atresia, stenosis of anastomosis, adhesions.
(ii) Short gut syndrome.
(iii) Abnormal intestinal motility – especially with associated atresias and dilation of bowel.
(iv) Patients with gastroschisis are at risk for NEC.

(f) Survival rate is 90 %.

Omphalocele

Alysia A. Agnoni

Omphalocele is a congenital defect of the abdominal wall in newborns. Omphalocele is notable for a high frequency of associated congenital anomalies. Outcome is often related to the severity of the associated conditions rather than the omphalocele itself.

1. Pathophysiology: this is a central defect of the umbilicus. Bowel and liver may herniate and are covered in a sac composed of amnion and peritoneum. The umbilical cord inserts into the sac. Defects range from small (2 cm) to very large (>10 cm). A giant omphalocele is one that is large enough to include the liver. This condition is associated with malrotation and Meckel's diverticulum.
   (a) Embryology: defect in umbilical ring occurs during 3rd week of gestation when yolk sac resides on the outside of the embryo.
   (b) Epidemiology: associated with Beckwith-Wiedemann syndrome, Lower midline syndrome, Pentalogy of Cantrell, and trisomies 13–18, and 21. More prevalent in males.
      (i) Beckwith-Wiedemann: gigantism, macroglossia, umbilical defect, visceromegaly, pancreatic islet cell hyperplasia; also associated with tumors: Wilms', neuroblastoma, adrenocortical.
      (ii) Lower midline syndrome: exstrophy of the bladder or cloaca, ambiguous genitalia, vesicointenstinal fissure, colonic atresia, imperforate anus, and sacral vertebral defects.
      (iii) Cantrell's Pentalogy: omphalocele, anterior diaphragmatic hernia, sternal cleft, cardiac defects including ectopia cordis.
2. Diagnosis: usually by fetal sonography at 20 weeks gestation. Amniocentesis can diagnose chromosome abnormalities.

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Fig. 1 Newborn infant with omphalocele. The omphalocele defect occurs within the umbilicus and is covered with a transparent sac (*Source*: Centers for Disease Control and Prevention, Atlanta, GA. http://www.cdc.gov/ncbddd/birthdefects/Omphalocele-graphic.html. Downloaded 17 Dec 2013)

3. Treatment:

   (a) Vaginal delivery is still possible. A cesarean section is considered if the omphalocele is extremely large.
   (b) Stabilize the newborn:

      (i) Orogastric tube to decompress the stomach.
      (ii) Examine sac for rupture and cover with dry plastic sheet or gauze dressing.
      (iii) IV fluid resuscitation.
      (iv) Perioperative antibiotics.
      (v) Monitor respiratory status.
      (vi) Thorough physical exam: may need echo, renal US, CXR.

   (c) Options for repair:

      (i) Small defects can be reduced and closed primarily soon after birth.
      (ii) Medium defects require excision of sac, reduction of contents into abdomen, and closure of fascia and skin.
      (iii) Postoperatively, patients typically do not have a prolonged ileus.
      (iv) Larger defects are patient – situation – dependent:

         1. Some can be reduced with only closure of skin – this results in a large ventral hernia that would need to be repaired in a staged procedure.
         2. Some are so large that the abdominal compartment cannot accommodate complete reduction. In this case it is imperative to preserve the omphalocele sac – this is done with escharotic agents that epithelialize and toughen the sac.

            (a) Silver nitrate, silver sulfadiazine, or povidone-iodine.

         3. Application of a pressure dressing or silo can be used.
         4. If the sac ruptures during this time a biologic dressing may be applied.

5. As the child grows, the omphalocele will gradually reduce into the abdomen. Closure of a large ventral hernia is necessary and can take place months to years after birth.

(d) Outcome:

(i) Complications:

1. Rupture or infection of omphalocele sac.
2. Compartment syndrome, respiratory distress, decreased cardiac output.

(ii) Those with large omphalocele and Cantrell's Pentalogy, trisomies, or midline syndrome have higher mortality.

Imperforate Anus and Cloaca

Christopher P. Coppola

Imperforate anus is a condition affecting males and females, in which the anus fails to form in the correct location. There is wide variation in abnormality requiring individualization of treatment. Often imperforate anus is accompanied by other anomalies.

1. Pathophysiology:
   (a) Epidemiology:
      (i) Incidence is one in 5,000 births.
   (b) Classification
      (i) Male:
         1. Low lesions:
            (a) Anal stenosis (normal location, narrow diameter).
            (b) Ano-cutaneous fistula (a tiny channel which leaks small quantities of meconium) There may be a thin membrane which completely occludes the anal orifice.
            (c) Anterior ectopic anus: anus forms anterior to the normal location through the anal sphincter muscle complex, leading to dysfunction and constipation. When normally located, the anus should be halfway between the tip of the coccyx and the back of the scrotum.
         2. Intermediate:
            (a) Recto-bulbar urethral fistula.

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3. High:

   (a) Recto-prostatic urethral fistula.
   (b) Recto-vesicular fistula.
   (c) Anorectal atresia without fistula.

(ii) Female:

1. Low lesions:

   (a) Anal stenosis.
   (b) Ano-cutaneous fistula.
   (c) Anterior ectopic anus: anus forms anterior to the normal location through the anal sphincter muscle complex, leading to dysfunction and constipation. When normally located, the anus should be halfway between the tip of the coccyx and the posterior fornix of the vagina.
   (d) Ano-vestibular fistula.

2. Intermediate:

   (a) Recto-vestibular fistula.
   (b) Recto-lower vaginal fistula.

3. High:

   (a) Recto-upper vaginal fistula.
   (b) Anorectal atresia without fistula.

(iii) Persistent cloaca: occurs in females, when the urethra, vagina, and rectum all join to form one channel called the persistent cloaca, a term which means "gutter". The vagina can be distended as a hydrocolpos which can compress the trigone of the bladder and cause urinary obstruction. Persistent cloaca can be classified by length of the common channel: longer than 3 cm is considered a long common channel.

(iv) Cloacal exstrophy: A complex combinations of malformations in females where there is a defect of the abdominal wall with an exposed inner surface of two hemi-bladders separated by an intestinal plate with colonic mucosa and two appendices. There is also a defect in the pubic bone.

(c) Associated conditions:

(i) VACTERL constellation of anomalies:

1. Vertebral.
2. Anorectal.
3. Cardiac.
4. Tracheoesophageal fistula.
5. Renal.
6. Limb.

2. Clinical presentation:
   (a) History:
      (i) No passage of meconium.
      (ii) In males, urine may contain stool or bubbles passing from rectum, through fistula, into urethra or bladder.
   (b) Physical examination:
      (i) Anal examination:
         1. No anus present.
         2. Evaluate for sacrum: can be short or absent.
         3. Check for gluteal cleft: flat or "rocker bottom" indicates higher lesion.
         4. May have dimple or puckering at site of muscle sphincter complex under skin.
         5. Low lesion, like perineal fistula, may have meconium "pearls" visible through the thin layer of skin.
         6. "Bucket-handle" flap of skin may be present.
      (ii) Examine abdomen for masses, distended hollow structures (colon, bladder, and vagina) and abdominal wall defects.
      (iii) Evaluate for associated anomalies:
         1. Check limbs for absent radius or other anomalies.
         2. Check spine and sacrum.
         3. Listen for cardiac murmurs.
3. Diagnosis:
   (a) Chest x-ray: check heart, and that there is no esophageal abnormality preventing nasogastric tube from passing into stomach.
   (b) Vertebral x-rays: check for abnormal or missing vertebrae.
   (c) Transverse lateral film with child in prone position: can sometimes show air in the blind ending rectal stump and give information about the height of the defect. In the past this was performed with the child upside down and was called an "invertogram".
   (d) Echocardiography: check for structural heart abnormalities.
   (e) Renal sonography: check for abnormalities.
   (f) Abdominal sonography: to identify obstructed or distended abdominal structures.
   (g) Cystoscopy and vaginoscopy: to identify the mouth and level of the rectal fistula.
4. Treatment:
   (a) IV fluid, nothing by mouth, and nasogastric tube decompression.
   (b) If a fistula is visible in the perineum or vagina, it can be dilated and irrigated to relieve obstruction of colon by thick meconium.
   (c) May need to time treatment around care for other anomalies, such as tracheaesophageal fistula.

(d) Algorithm of treatment: depends upon individual anatomy of defect.
(e) Anal stenosis can be treated with serial dilations.
(f) Anterior ectopic anus can be corrected by surgically re-locating the anus more posteriorly within the anal sphincter muscle complex. This is performed by a midline incision in the perineum and reconstruction of the anus with the two halves of the sphincter complex surrounding it.
(g) The so-called "cutback" operation is to be avoided as it will divide the anterior border of the anal sphincter muscle complex and lead to incontinence.
(h) High lesions will require diverting colostomy.

(i) Use upper sigmoid colon: need to leave enough length of colon below the stoma that it will be able to reach the anus for reconstruction.
(ii) Usually a loop colostomy will suffice, however with a wide open urinary fistula, separated colostomy and mucus fistula will reduce contamination of urine.
(iii) Colostomy can be taken down in a third operation after the anus is reconstructed, any dilation is completed, and the anal canal is retaining a stable diameter, verified with contrast enema or colostogram fluoroscopy.

(i) Low lesions can be repaired with primary repair. If the child can be made to stool through the fistula with dilations and irrigations, the primary repair can be performed after a month when the child has grown and has good nutrition.
(j) Intermediate lesions can sometimes be treated with primary repair, but the safest course is to create a diverting colostomy and construct an anus that will last the child their whole life when he or she is larger, has good nutrition, and with the benefit of a diverting colostomy until the anus has healed.
(k) Posterior sagittal anorectoplasty (PSARP): Pena repair.

(i) Child in prone position, on padding, with hips flexed.
(ii) Locate sphincter muscle complex using Pena stimulator and mark.
(iii) Incise skin in midline and divide sphincter complex in half.
(iv) Locate rectum within ischiorectal fat and levator complex.
(v) Divide fistula with absorbable suture.

1. In males, care must be taken not to narrow the urinary tract. A Foley catheter is used to locate and avoid injury to the urethra and bladder.
2. In females, after closing fistula to back of vulva or vagina, the perineal body is reconstructed as a barrier between vagina and rectum.

(vi) The sphincter muscle complex is reconstructed around the rectum as a newly created anus.

(l) Persistent cloaca:

(i) Less than 3 cm or short lesions: these can often be repaired in a single operation by reconstructing the anal, vaginal, and urethral openings. For very short persistent cloaca, lowering of the urogenital plate to the perineum is sufficient to create the urethral and vaginal openings, and the anus is reconstructed by PSARP.

(ii) Greater than 3 cm or long lesions: these will require a staged repair, and should be treated with a diverting colostomy. In some cases, a colonic conduit will be required to recreate the vagina, and colostomy should be placed proximal enough so that there is enough colon to construct the anus and the vagina.

5. Outcome:

(a) Difficulties with stooling are common.

(i) Patients with absent sacrum, rocker-bottom, and vertebral anomalies are likely to have incontinence.
(ii) Constipation is common in children who have a complete sacrum, a gluteal cleft, and sphincter muscle complex present.
(iii) Children often develop stricture at the reconstructed anus. This is treated with serial dilation. Parents are trained to do this at home. Dilation to a size of 16 mm in infancy, or 19 mm in older children is usually sufficient to allow normal stooling. When anus retains diameter for 2 months, dilation can be stopped.

(b) Children will toilet train later than children who have not had a surgically created anus.
(c) Regular stooling is attainable for most children, but the majority will require some kind of stool regimen, such as daily Miralax (polyethylene glycol, Merk Sharpe, and Dohme, Whitehouse, NJ) and possibly enemas.

Congenital Lung Malformations

Meng-Fey Ferra Lin-Duffy

Congenital lung malformations are a rare but important disorder which can be associated with significant morbidity and mortality. The malformation may be identified during prenatal sonography, presented as respiratory distress in the newborn, or be completely asymptomatic and incidentally discovered in an adult.

1. Pathophysiology:

   (a) Congenital pulmonary airway malformation (CPAM).

      (i) Previously known as cystic adenomatoid malformation (CCAM), CPAM is rare but is one of the most common congenital lung abnormalities. It is the second most common cause (25 %) of newborn respiratory distress.
      (ii) The lesions are hamartomatous proliferation of cysts comprised of adenomatous elements and resemble bronchioles.
      (iii) They have connections with the tracheobronchial tree. The arterial supply and venous drainage from the lesions are typically with pulmonary circulation though they can be from systemic circulation.
      (iv) CPAM are classified into one of five types based on the size and the characteristics of the tissue.

         1. Type 0- These lesions arise from the tracheal or bronchial tissue and are the rarest form. The cysts are small and involve the entire lung. Affected infants die at birth due to severely impaired gas exchange.
         2. Type 1-This type comprises about 60–70 % and is the most common form. It originates from the distal bronchi or proximal bronchioles and consists of well-differentiated tissue. The cysts are usually sin-

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gle but may be multi-loculated. The majority of the cases involve only one lobe of the lung. It has malignant potential.

3. Type 2-This type comprises about 15–20 % of cases and consists of multiple cysts that blend into adjacent normal tissue. The lesions resemble dilated terminal bronchioles. Up to 60 % of cases have other congenital anomalies. It does not have malignant potential.
4. Type 3- These lesions comprise about 5–10 % of cases and are often very large proliferating from the distal airways or airspaces and involving an entire lobe or several lobes. This type has not been associated with malignancy.
5. Type 4-These lesions comprise 10–15 % of cases and have a maximum diameter of 7 cm. They are strongly associated with malignancy.

(v) Previously used classification system for CCAM:

1. Macrocystic: 13 %; has cysts >5 mm in diameter.
2. Microcystic: 73 %; all cysts <5 mm, resemble bronchioles.
3. Solid: 13 %; microscopic cysts.

(vi) Lesions can be multifocal and even bilateral.

(vii) During gestation, compression of heart/central veins by CPAM can lead to hydrops. Compression of GI tract (esophagus) can lead to polyhydramnios.

(viii) Course over time: some prenatally diagnosed CPAM lesions will shrink and disappear on postnatal imaging. If it persists to age 6-months-old it is unlikely to resolve.

(ix) Risk of pneumonia: compression of the lesion on bronchial passages leads to overinflation of distal alveoli, trapping of air and mucus, and recurrent pneumonia, which makes future resection more difficult with increased blood loss.

(x) Risk of Neoplasm: if not treated, certain types of CPAM can develop into malignancy such as broncoalveolar carcinoma, sarcoma, or pulmonary blastoma.

(b) Congenital lobar emphysema (CLE):

(i) CLE is massive overinflation of one of the lobes of lung, usually due to airway obstruction from either intrinsic or extrinsic compression.

(ii) It most commonly affects one of the upper lobes of lung.

(iii) Intrinsic obstruction is more common and often caused by defects in the bronchial wall such as bronchial cartilage deficiency, resulting in airway collapse during expiration. Meconium or mucous plugs, granulomas, or mucosal folds can also cause partial airway obstruction of a lower airway.

(iv) Extrinsic compression may be due to a bronchogenic cyst, an obstructing lymph nodes, or vascular anomalies such as a pulmonary artery sling or anomalous pulmonary venous return.

(v) Bronchial atresia has also been identified as a common finding.

(vi) Overdistention of one or more lobes of the lung leads to compression of the remaining lung tissue and risk of pulmonary compromise with shift of mediastinum.
(vii) The conducting airways are typically normal in size and number, but the alveoli within the gas exchange units are enlarged and increased in number.
(viii) There is a 10 % risk of associated cardiac abnormality.

(c) Pulmonary sequestrations:

(i) These are pieces of parenchymal lung separated from the respiratory tree. The blood supply is from systemic circulation, via an anomalous artery arising from the aorta rather than pulmonary artery. Sometimes the artery will cross the diaphragm.
(ii) They usually occur in the lower lobes.
(iii) Types of sequestration:

1. Intralobar – within the lung parenchyma: (50 %) Intralobar lesions are found within the normal lung and pleura and almost always in the lower lobes. They occur equally on both sides. Venous drainage is usually through the pulmonary vein.
2. Extralobar – outside of the normal lung: (50 %) Extralobar lesions are found outside of the pleural space and the venous drainage is usually into the azygos or hemiazygos veins. Two thirds of the patients may have other congenital abnormalities such as diaphragmatic hernias.
3. Transdiaphragmatic sequestrations or sequestrations with an abdominal component

(iv) Sequestrations can be attached to or communicate with the GI tract (esophagus or stomach).

(d) Bronchogenic cysts:

(i) Arise from an abnormal budding or duplication of the ventral foregut tissue present in the airway.
(ii) 15 % are intrapulmonary and 85 % are mediastinal.
(iii) The cysts may be central or peripheral and may be fill with air, fluid or both.
(iv) Can cause compression of bronchus or trachea.
(v) Can result in pulmonary hemorrhage or infection.

2. Clinical presentation:

(a) Often asymptomatic, diagnosed prenatally by sonography.
(b) Often clinically silent at birth.
(c) Exception is CLE, which may present as emergent pulmonary compromise.
(d) May cause bronchial compression and hyperinflation.
(e) May cause recurring pneumonia.
(f) Bronchogenic cysts can have drainage into bronchial tree.

3. Diagnosis:
   (a) Prenatal sonography: can diagnose lung lesions as cystic or solid, and can track size as the fetus grows.
   (b) Chest x-ray: will reveal fluid filled structures, such as a bronchogenic cyst, can detect hyperinflation of CLE, and if pneumonia is present.
   (c) Chest sonography: useful in tracking size of CPAM lesions as children grow, and can sometimes be used to identify the anomalous systemic blood supply of a sequestration.
   (d) Computed tomography of chest: should be used sparingly, because of the ionizing radiation, but probably offers the best delineation of lung lesions and their relationship to normal structures. With intravenous contrast, vascular supply can be identified.
   (e) Magnetic resonance imaging of chest: gives excellent anatomic definition of lesions without ionizing radiation. Will usually require general anesthesia for small children to remain still for the amount of time necessary for the study. Gadolinium enhanced studies help identify cystic structures, communication with hollow viscus, and vascular supply.
   (f) Echocardiography: to rule out associated cardiac anomaly.
4. Treatment:
   (a) CPAM:
      (i) If CPAM presents prenatally with hydrops, patient should be considered for fetal surgery at a center with that capability. If no hydrops is present, care can be given after delivery.
      (ii) Treatment of pneumonia: often a CPAM will be diagnosed while evaluating for a recurrent pneumonia. The pneumonia should be treated and the patient should have an one-month period without symptoms before undergoing resection. Resection during active infection is difficult, bloody, and dangerous.
      (iii) Period of observation and repeat imaging: Some CPAM lesions will resolve spontaneously. When diagnosed prenatally, the child should undergo imaging after delivery. Sonography is usually sufficient and does not use ionizing radiation. Imaging is repeated at 3 and 6 months of age. If the lesion persists past the age of six months, it should be resected.
   (b) CLE:
      (i) Immediate treatment: some patients will have severe ventilatory distress on birth. This occurs when the emphysematous lobe becomes hyperinflated with first breaths, and causes mediastinal shift. Chest x-ray may have appearance of a tension pneumothorax. If a chest tube is placed into the emphysematous lung, it can help relieve the pressure, but will create a bronchial leak. If the diagnosis of CLE has been made and a child has severe ventilator distress, the best course of treatment would be intubation and emergent thoracotomy.

(ii) When a child has mild or no symptoms, resection of the emphysematous lung should still be performed, but can be done on an elective basis. Resection of CLE will allow the other lobes in the hemithorax to ventilate normally.

(c) Sequestration:

(i) Resection of lesion is recommended whenever it is detected.

1. Thoracoscopic: often lesions can be resected with a non-anatomic wedge resection using laparoscopic stapler.
2. Thoracotomy: open resection may be needed in smaller children, hilar lesions, and when surrounding infection makes removal difficult.
3. Addressing abdominal component: when present, the abdominal component can usually be removed through the diaphragm or behind the diaphragm. When a diaphramatic hernia is also present, it can be repaired after resection of the sequestration. Rarely an additional laparotomy will be needed.

(ii) No matter what approach is used, care must be taken to identify and ligate the systemic blood supply of the sequestration. When possible, this blood supply is identified preoperatively. These vessels are often short and arise directly from the aorta, and can traverse the diaphragm. When not properly ligated, they will retract and result in significant bleeding.

(d) Bronchogenic cyst:

(i) Lesions should be resected when detected.
(ii) If not removed, they can lead to compression of pulmonary, GI or vascular structures.
(iii) Cysts can become infected and be associated with pneumonia.
(iv) Resection should not be performed during active infection/pneumonia because there is increased inflammation and risk of hemorrhage during procedure.
(v) Thoracoscopic or open approach can be used.

5. Outcome: with complete resection of lesion, patients can be expected to have normal pulmonary function.

Meconium Plug Syndrome and Meconium Ileus

Christopher P. Coppola

Meconium plug syndrome, meconium ileus, and meconium ileus equivalent are conditions of gastrointestinal tract obstruction by thickened contents which will not pass.

1. Pathophysiology:
   (a) In meconium plug syndrome, a newborn has obstruction of the colon with thickened plugs of mucus.
   (b) Other terms for meconium plug syndrome are colonic immaturity, small left colon syndrome, and functional colonic obstruction.
   (c) In meconium ileus, thickened meconium blocks the small bowel, rather than the colon, and results in a small diameter colon called microcolon. Meconium ileus can result in perforation, small bowel volvulus, intestinal atresia, and peritonitis. Prenatal perforation can result in formation of a meconium cyst, visible on x-ray as a mass stippled with opacities from calcification.
   (d) Incidence:
      (i) Meconium plug syndrome: one in 1,000 births.
      (ii) Meconium ileus: one in 3,000 births.
   (e) Associated conditions:
      (i) The most common cause of meconium ileus is cystic fibrosis. The abnormally concentrated excretion of epithelial cells results in a dry thick succus. 80 % of infants with meconium ileus have cystic fibrosis.
      (ii) Meconium plug syndrome can be associated with Hirschsprung's disease in approximately 40 % of cases and it is associated with cystic fibrosis in approximately 40 % of cases.

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(iii) Small left colon syndrome, a synonym of meconium plug syndrome, has an association with birth to a diabetic mother.
(iv) Hypotonia.
(v) Hypermagnesiemia.
(vi) Sepsis.

(f) Meconium ileus equivalent is a condition occurring in adolescents with cystic fibrosis in whom succus becomes thickened and difficult to pass resulting in small bowel obstruction. Also called distal intestinal obstruction syndrome (DIOS).

2. Clinical presentation:

(a) No passage of meconium.
(b) Abdominal distention.
(c) Difficulty feeding.
(d) Bilious vomiting.
(e) Patient may have anal stenosis.

3. Diagnosis:

(a) Physical examination will reveal a neonate with softly distended abdomen, sometimes with palpable loops of intestine.
(b) Nasogastric tube aspirate is bilious
(c) Abdominal x-ray: will reveal bowel obstruction, distal colon/bowel not visible
(d) Upper gastrointestinal series is not the best study to reveal the meconium plugs. If performed, it will show normal rotation of bowel which is useful because it will rule out malrotation and volvulus as the cause of the bilious vomiting. Contrast progression will be slow and hindered due to the distal obstruction by the meconium plugs.
(e) Contrast enema fluoroscopy.

(i) This study will be both diagnostic and therapeutic.
(ii) Water-soluble contrast should be used. Barium is avoided for two reasons: if there is a perforation and leak, extravasation of barium will cause peritonitis. Also, barium will thicken in the lumen of the colon and can contribute to the obstruction. If needed by the radiologist, the study can start with water-soluble contrast and can be followed by barium if an alternate diagnosis to meconium plugs is found and the better definition of barium is needed.
(iii) Contrast enema will show thick plugs and ropes of material occluding the colon and distal bowel. Irrigation of the colon with the water-soluble contrast can help wash these plugs free and out of the body.
(iv) Multiple contrast enemas may be necessary.

(f) Chloride sweat testing: Cystic fibrosis is frequently associated with meconium plug syndrome and is diagnosed with elevated chloride concentration in the sweat.

(g) Suction rectal biopsy: Some patients with meconium plug syndrome will have Hirschsprung's disease which is diagnosed by absence of ganglion cells on a rectal biopsy.
(h) Computed tomography of abdomen: in teenagers with cystic fibrosis and bowel obstruction, CT of the abdomen with oral and IV contrast can reveal meconium ileus equivalent: obstruction of small bowel with thick inspissated succus. CT will reveal fecalization of bowel succus and proximal obstruction.

4. Treatment:

(a) Nasogastric tube for gastric decompression.
(b) IV fluid resuscitation and correction of any electrolyte abnormalities.
(c) Malrotation and midgut volvulus should be considered and ruled out if a possibility because missing this diagnosis can lead to a lethal loss of intestine. Malrotation is ruled out with upper gastrointestinal series if suspected. If detected it is immediately corrected.
(d) Water-soluble contrast enema both diagnosis and treats the condition by washing free obstructing plugs.
(e) Children are treated with serial rectal irrigations until gastrointestinal function returns. If no progress is being made, a repeat water-soluble contrast enema can repeated.
(f) Acetylcysteine can be delivered by enema to try and loosen the thickened secretions
(g) Some children will additionally have a bowel atresia which will require operative correction.
(h) In teenagers with meconium ileus equivalent, a nasogastric tube is placed and GoLytely (polyethylene glycol with electrolytes, Braintree Laboratories, Inc., Braintree, MA) are instilled to break up the thickened succus.

Hypospadias

Joel M. Sumfest

1. Pathophysiology:
   (a) Results from arrested penile development causing a proximal urethral meatus and varying degrees of other issues of the penis.
   (b) Underlying cause for most cases of non-syndromic hypospadias is unknown.
   (c) Estimated to have incidence of 1 in 300 live male births.
      (i) Has been increasing past 30 years.
      (ii) Thought to be possibly related to environmental toxins.
   (d) Familial predominance with an index case resulting in an incidence of one in 20 newborns.
   (e) Associated Anomalies:
      (i) Cryptorchidism.
      (ii) Prostatic utricle.
      (iii) Disorders of sexual development (DSD):
         1. Any child with hypospadias and undescended testes, especially if dysmorphic or nonpalpable, should raise concern.
         2. Most common is mixed gonadal dysgenesis, followed by varying degrees of androgen insensitivity.
2. Diagnosis:
   (a) Abnormal appearing penis with proximal meatus and ventrally deficient prepuce.
      (i) Can have penile torsion, bifid or engulfing scrotum, glanular tilt, chordee, or complete ambiguity.

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(b) There is a subset where foreskin is normal and abnormally large meatus is only discovered when foreskin is retracted

(i) Megameatus intact prepuce (MIP).

(c) Can also have a normally positioned meatus and ventral chordee.

(i) Chordee without hypospadias.

(d) Meatus can be located along pathway of normal urethra from glans down to perineum.

(e) If there are many associated issues may be role for karyotyping, but it is not recommended in routine cases.

3. Treatment:

(a) General principles:

(i) Over 200 repairs have been described.

(ii) Suture material is usually per surgeon preference, as is choice of post-op dressings and urinary diversion.

(iii) Each individual surgeon must monitor his or her outcomes in a prospective fashion and adjust if complications reach predetermined levels.

(iv) Should be approached only by a surgeon who has undergone fellowship mentoring.

(v) Use of optical magnification required.

(vi) All repairs should be followed through puberty.

(b) Surgical Repairs:

(i) Most are performed in a single stage as outpatient.

(ii) Rarely very proximal lesions or those associated with severe chordee are corrected in stages.

(iii) All defects must be addressed including repositioning of meatus, ventral chordee, and scrotal abnormalities.

(iv) Re-do repairs and complex redo reconstructions are a special subset.

1. May require use of extragenital skin for urethroplasty.
2. Most common is buccal mucosa.

(a) One- or two-staged approach.

3. Severe shortage of shaft skin is sometimes the limiting factor.

(a) Skin grafting.
(b) Use of tissue expanders.

(c) Glandular hypospadias:

(i) With or without chordee.

(ii) Meatal advancement and glanuloplasty incorporated (MAGPI) variant can work in some as long as repair does not leave a retrusive meatus.

(iii) Division of web separating urethral pit from meatus.

1. Defect usually left open to re-epithelialize.

(iv) Most mild chordee can be corrected with dorsal plication of the tunica albuginea of the corpora in the midline after incision of Buck's fascia.

1. Avoid neurovascular bundle.

(v) Place a voiding urethral splint.

(d) Distal hypospadias:

(i) Includes coronal and distal shaft lesions.
(ii) Must be aware of dysplastic ventral shaft skin.
(iii) Tubularized incised plate (TIP) repair is used for the majority of lesions.

1. Originally described as Thiersch-Duplay urethroplasty.
2. Repopularized by Warren Snodgrass who added incision of the urethral plate.

(iv) Results in a cosmetically and functionally superior glans and urethral meatus than prior repairs.
(v) Can be limited in children with tiny glans caps.
(vi) Very rarely, the urethral plate is poorly developed and cannot be used for reconstruction.

1. May actually be responsible for chordee.
2. May have to be divided.

(vii) Coverage of the neourethra with a dartos fascial flap obtained from the dorsal prepuce is invaluable in reducing the complication of a urethracutaneous fistula.
(viii) Various repairs for chordee:

1. Dorsal plication.
2. Mobilization of urethral plate.

(ix) Typically diverted with urethral drippy tube for 5–7days.

1. Avoid use of balloon Foley catheters.

(x) TIP repair can be extended to some more proximal lesions such as midshaft and penoscrotal meatus.

(e) Proximal Hypospadias:

(i) Includes those at penoscrotal junction and perineum.
(ii) Can be ambiguous.
(iii) More likely to be associated with a bifid scrotum and penoscrotal engulfment of severe chordee.
(iv) Approach in multiple stages (but single stage is an option used by some).
(v) Chordee is addressed first:

1. The penis is degloved and chordee reassessed with an artificial erection.
2. Must be careful that the dorsal plication or formal tuck does not reduce shaft length.
3. If so need to divide or mobilize urethral plate to allow placement of a ventral graft.
    (a) Dermal graft.
    (b) Intestinal submucosa.

(vi) Can then make decision to tubularize urethral plate in a one-stage repair or approach in stages.

1. Must take care not include what will be hair-bearing tissue if performing a proximal TIP.

(vii) Prepuce is transposed ventrally.

1. Preferred method is button-hole technique.

(viii) Delayed urethroplasty usually in 6–9 months via tubularization.

1. Can use TIP repair distally for glans and fossa.

(ix) Dartos coverage of neourethra, if possible, or consider tunica vaginalis flap.

(x) Urinary diversion with drippy tube, or in older boys, suprapubic tube (SPT).

4. Outcome:

(a) Complications:

(i) Best way to manage them is to avoid them.
(ii) Occur in the best of hands.
(iii) Urethrocutaneous fistula (UCF).
(iv) Glanular dehiscence.
(v) Recurrent chordee.
(vi) Postop hematoma or wound infection.
(vii) Rare to have complete loss of repair.

(b) Re-do Repairs:

(i) Include most severe issue classified as hypospadias cripple.
(ii) Can be limited by availability of shaft skin.

1. Creation of neourethra.
2. Shaft skin coverage.
3. Correction chordee.

(iii) Can be simple UCF closure.

1. Try and avoid overlapping suture lines with use dartos interposition.
2. Operating microscope helpful.

(iv) Re-do TIP repair can be used for repair of most glans failures.

(v) The most severe lesions may require skin grafting or buccal mucosa urethroplasty.

1. One or two stages.
2. Aid of plastic surgeon is helpful.
3. SPT diversion.

Disorders of Sexual Differentiation

Joel M. Sumfest

Disorders of sexual differentiation (DSD) are a variety of complex malformations which require a multidisciplinary approach to guide the best therapy for each individual patient.

1. Pathophysiology:
   (a) Incidence is rare.
   (b) Classification (based on genotype).
      (i) XX genotype: most common DSD seen clinically.
         1. Congenital adrenal hyperplasia (CAH).
         2. Complete androgen insensitivity (CAIS).
      (ii) XY genotype:
         1. Partial androgen insensitivity (PAIS).
      (iii) Gonadal dysgenesis.
      (iv) XX/XY genotype:
         1. True hermaphrodite.
         2. Mosaicism.
2. Diagnosis:
   (a) Psychosocial emergency.
   (b) Can be life-threatening in some instances if not recognized and treated as neonate.

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(c) Team approach to diagnosis and treatment:

(i) Pediatric urologist.
(ii) Pediatric endocrinologist.
(iii) Neonatologist.
(iv) Pediatric geneticist.
(v) Social worker.

(d) Must have high index of suspicion.
(e) There may be a family history of sudden death as infant.
(f) There can be an appearance of ambiguous genitalia, however occasionally the phallic structure can appear to be completely normal.
(g) Any male infant with bilateral non-palpable undescended testes (UDTs) should be considered to have a DSD, especially if testes are not palpable.
(h) Genotype:

(i) Can be obtained rapidly.

(i) Pelvic sonography:

(i) Presence of Mullerian structures.

1. Uterus.
2. Fallopian tubes.

(ii) Intra-abdominal gonads.

(j) Scrotal sonography:

(i) Abnormal testes.

1. Streak gonads.
2. Vanishing UDT.

(k) Serial serum electrolytes.
(l) Serum hormones:

(i) Testosterone.
(ii) Luteinizing hormone (LH).
(iii) Follicle stimulating hormone (FSH).
(iv) Mullerian inhibiting substance (MIS).

(m) Beta human chorionic gonadotropin (bHCG) stimulation test is occasionally useful.

3. Treatment:

(a) CAH:

(i) Most common defect responsible for DSD.
(ii) 21-hydroxylasease deficiency is the most common metabolic defect.

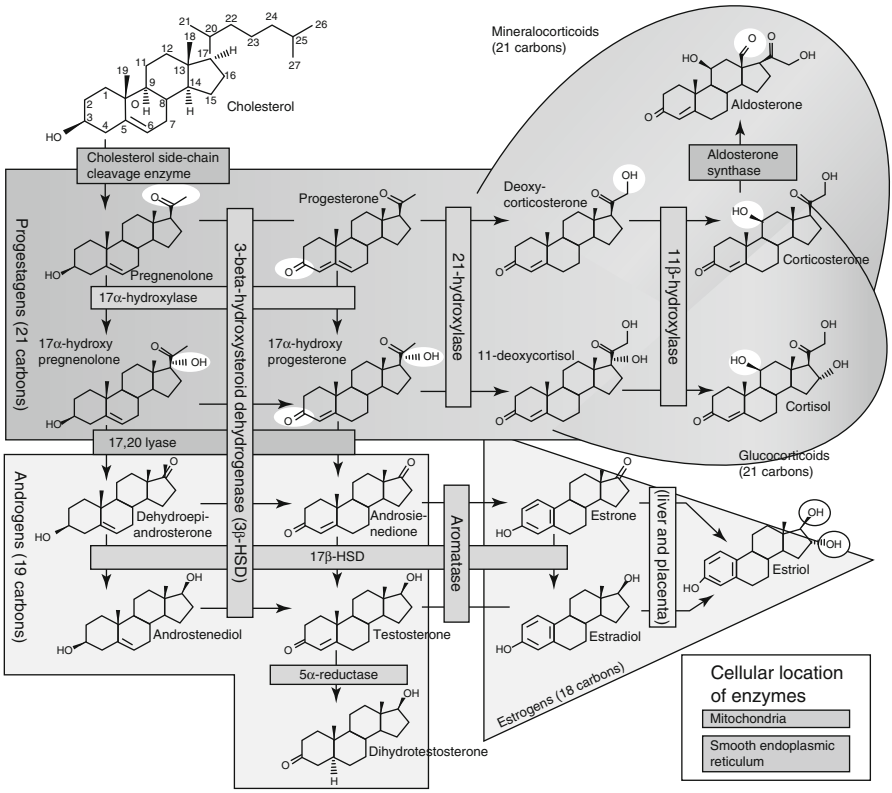

Fig. 1 Cholesterol metabolism. Defects in the steps of cholesterol metabolism can result in surplus or deficiency of androgens, resulting in abnormal sexual differentiation (Adapted from David Richfield and Mikael Haggstrom under Creative Commons Attribution 3.0 Unported license as published on https://upload.wikimedia.org/wikipedia/commons/thumb/1/13/Steroidogenesis.svg/1245px-Steroidogenesis.svg.png. Downloaded 22 Dec 2013)

(iii) Cellular defect in cholesterol pathway.

1. Will cause buildup of testosterone precursors.
2. This is responsible for virilization of the newborn infant.

(iv) The degree of virilization of the external genitalia is variable.

1. Can have massive cliteromegaly with urinary meatus at tip and for all purposes look like a normal male penis.
2. Bilateral non-palpable UDTs.
3. Urogenital (UG) sinus.

   (a) The vaginal and urethra share a common channel.
   (b) It is the length of this common channel which ultimately determines the complexity of surgical reconstruction.

(v) The same cellular defect can cause hypoaldosteronism.

1. Responsible for salt-wasting.
2. If untreated will cause circulatory collapse and sudden death, usually in 7–10 days.

(vi) Pelvic sonography will disclose normal appearing uterus and possibly both ovaries.

(vii) Normal XX genotype.

(viii) Once diagnosis is confirmed must begin treatment with corticosteroids and aldosterone if needed.

1. Lifelong treatment is necessary.

(ix) Autosomal recessive inheritance.

1. Important for genetic counseling.

(x) Surgical reconstruction.

1. Usually early to improve cosmesis.
2. Reduction cliteromegaly.
3. Separation of UG sinus into separate urethral and vaginal openings.
   (a) Can be as simple as cutback vaginoplasty.
   (b) In cases of long channel UG sinus may need urogenital sinus mobilization with interposition flaps.
4. Reconstruction of labia minora.
5. Young woman with this condition will be fertile.
6. Gender identity issues are common.

(b) CAIS:

(i) Old terminology for testicular feminization.

(ii) Cellular defect in dihydrotestosterone (DHT) receptors.

(iii) Normal intra-abdominal UDTs.

(iv) No Mullerian structures.

(v) There will only be a vaginal indentation consistent with the distal 1/3 of the vaginal canal.

(vi) Normal breast development at puberty.

(vii) XY genotype but normal female external genitalia.

(viii) Can present with amenorrhea at puberty.

(ix) All need vaginal reconstruction.

(x) Very high incidence of testicular tumors and bilateral orchiectomy with hormone replacement is recommended.

(xi) Infertile.

(c) PAIS:

(i) XY genotype.
(ii) Partial insensitivity of testosterone receptors or 5-alpha reductase which converts testosterone to DHT, the active metabolite for genital growth and development.
(iii) Varying degrees of virilization possible.
(iv) Usually severe hypospadias.
(v) Unilateral or bilateral UDTs.
(vi) Can improve at puberty but response is variable and difficult to predict.
(vii) Occasionally fertility is possible.

(d) Gonadal Dysgenesis:

(i) XO/XY genotype
(ii) Commonly seen with hypospadias and UDTs.
(iii) As with any DSD and presence of Y chromosome high incidence of malignancy as adult.

1. UDTs should be excised and normal descended testes watched closely.

(iv) Hypospadias reconstructions are commonly necessary.
(v) Infertile.

(e) True Hermaphrodite:

(i) Variable genotype and phenotype.
(ii) Presence of a combined gonad, an ovotestis, is common.
(iii) If Y chromosome is present, there is malignant potential.
(iv) Individualized need for urogenital reconstruction.

Biliary Atresia

Christopher P. Coppola

Biliary atresia is a progressive fibrosing process of the extrahepatic biliary tree in the neonate. It presents with jaundice and is life-threatening if flow of bile is not restored.

1. Pathophysiology:
   (a) Incidence:
      (i) One in 10,000–18,000 births.
      (ii) Higher incidence in females.
      (iii) Higher incidence in Asian and African-American children.
   (b) Fibrosis and obstruction of biliary passages in the liver.
   (c) Progression to extrahepatic biliary tree.
   (d) Types of biliary atresia: classified by how high into the liver the biliary tree is obstructed.
      (i) Type I: (10 % of cases) Obliteration of the common bile duct.
      (ii) Type II: (2 % of cases) Obliteration of the common hepatic duct.
      (iii) Type III: (88 % of cases) Obliteration from the common bile duct up to the right and left hepatic ducts into the porta hepatis (liver hilum).

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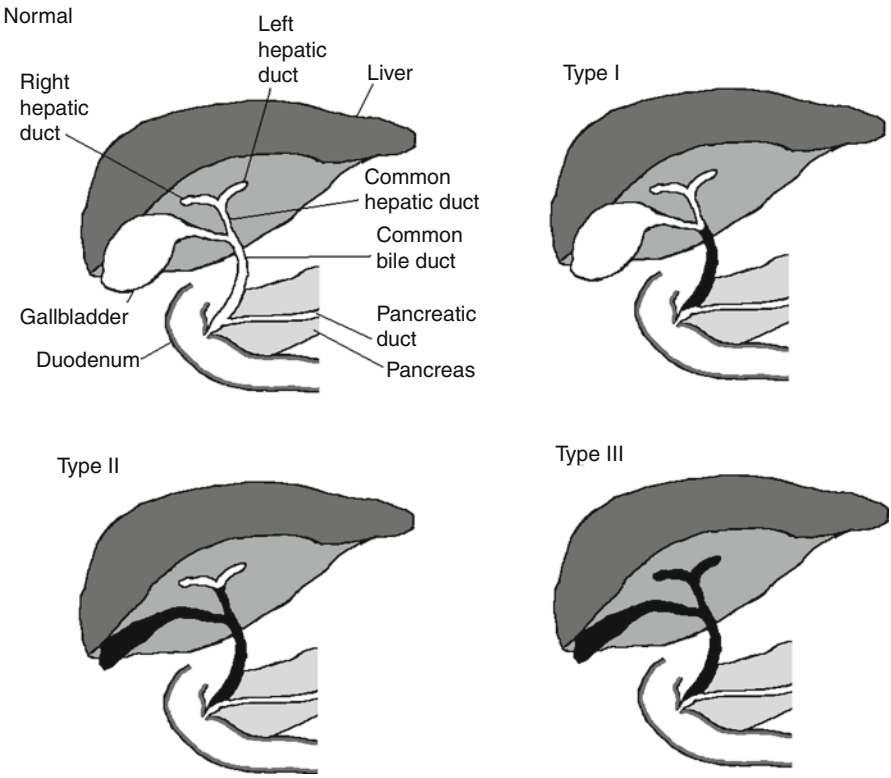

Fig. 1 Classification of biliary atresia (*Source*: Bryan Walters and Christopher Coppola)

(e) No proved etiology, but may be due to prenatal infection initiating a process of inflammation and scarring in the biliary tree.
(f) Associated conditions:

(i) Polysplenia/asplenia.
(ii) Situs inversus.
(iii) Preduodenal portal vein.
(iv) Cardiac anomaly.

(g) Differential diagnosis:

(i) Physiologic jaundice of newborn: should clear by age 2-weeks-old.
(ii) ToRCH infections:

1. Toxoplasmosis.
2. Rubella.
3. Cytomegalovirus.
4. Herpes simplex virus.

(iii) Alagille syndrome: paucity of interlobar bile ducts.

(iv) Choledochal cyst and Caroli disease (intrahepatic ductal dilation).
(v) Alpha 1 anti-trypsin deficiency.
(vi) Cystic fibrosis.

2. Clinical presentation:

(a) History.

(i) Jaundice.
(ii) Dark urine.
(iii) Pale white or grey stools.
(iv) Poor weight gain.
(v) Itching and irritability.

(b) Physical examination:

(i) Jaundice.
(ii) Hepatomegaly/splenomegaly.

3. Diagnosis:

(a) Laboratory workup:

(i) CBC.
(ii) LFTs.
(iii) ToRCH infection titers.
(iv) Alpha 1 anti-trypsin level.
(v) Sweat chloride.

(b) Imaging:

(i) Abdominal sonography: can show presence/absence of gallbladder, dilation of intrahepatic and extrahepatic bile ducts.
(ii) Hepatobiliary iminodiacetic acid (HIDA) scan: should occur after 5 days of phenobarbital treatment for best results. Will show if liver is able to process bile and if there is an intrahepatic or extrahepatic blockage. Normal HIDA scan rules out biliary atresia. Abnormal HIDA scan needs follow-up with liver biopsy and/or cholangiogram.

(c) Liver biopsy: can diagnose the cholestasis of biliary atresia and identify alternate causes such as hepatitis or paucity of interlobar bile ducts (Alagille syndrome).
(d) Intraoperative cholangiogram: when no other cause for the jaundice is identified and HIDA scan is suggestive of biliary atresia, an open or laparoscopic evaluation of liver is performed. If not performed already, a liver biopsy can be done at this time. The gallbladder and extrahepatic bile ducts are examined for patency or obliteration. A cholangiogram is then performed either through the gallbladder or the cystic duct. Biliary atresia is identified by blockage of the extrahepatic bile ducts by obliteration and scarring.
(e) In a few centers, neonatal endoscopic retrograde cholangiopancreatography (ERCP) is available in neonates to determine patency of the biliary tree.

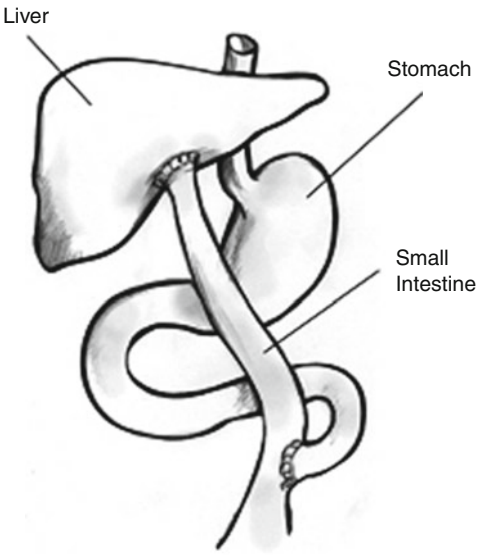

Fig. 2 Kasai procedure for biliary atresia: Hepato-portoenterostomy (Adapted from: National Digestive Diseases Clearinghouse, National Institutes of Health, NIH publication 12–5289, July 2012. http://digestive.niddk.nih.gov/ddiseases/pubs/atresia/#13. Downloaded on 22 Dec 2013)

4. Treatment:

   (a) Phenobarbital.
   (b) Exploratory laparotomy for liver biopsy and cholangiogram.
   (c) Kasai procedure:

      (i) Kasai procedure is hepato-portoenterostomy.
      (ii) The extrahepatic biliary tree (gallbladder, common bile duct, and common hepatic duct) is resected.
      (iii) An anastomosis is formed between the jejunum and the hilum of the liver.
      (iv) A roux limb of jejunum is brought up to the liver hilum, and a down-stream roux-en-y anastomosis is created.
      (v) The level of anastomosis at the liver hilum is determined by how extensive the bile ducts have been scarred. Usually the common hepatic duct is resected, and if bile flows from the right and left hepatic ducts, the anastomosis is made there.
      (vi) If there is no flow of bile, the right and left hepatic ducts are resected up to the level of the hilar plate and the jejunum is anastomosed to the surface of the liver without any formal connection between intestinal wall and biliary ductal wall.
      (vii) Frozen section evaluation of a biopsy from the hilar plate can help by revealing that patent biliary ductules have been reached, but this is no guarantee of successfully postoperative flow of bile.

(viii) Postoperative care:

1. Patients are maintained NPO with IV fluid until gut function returns.
2. LFT's are measured to track drainage of bile.
3. Postoperative antibiotics may reduce the chance of cholangitis.
4. Steroids have been used in the past in an attempt to reduce the ongoing process of scarring and atresia in remaining liver, but they are not universally given after Kasai procedure.
5. Ursodeoxycholic acid aids bile flow.

(d) Liver transplantation: when Kasai has not been performed by age 3-months-old, or if Kasai procedure is unsuccessful in producing bile drainage from liver, the child is a candidate for liver transplantation.

5. Outcome:

(a) Without operation to restore biliary drainage, 50 % of children die by age 5-years-old.
(b) Complications of Kasai procedure:

(i) Failure to drain bile/jaundice. Bile may drain initially and then cease when the process of atresia extends further into the liver.
(ii) Sepsis.
(iii) Cholangitis.
(iv) Bile leak.
(v) Ascites.
(vi) Leak from roux-en-y anastomosis.
(vii) Bowel obstruction.
(viii) Cirrhosis and liver failure.
(ix) Portal hypertension.
(x) Poor growth and malnutrition.
(xi) Deficiency of fat soluble vitamins A, D, E, and K.

(c) Patients treated with Kasai portoenterostomy before age 2-months-old have better prognosis than those treated later.

(i) 1/3 patients have bile drainage with Kasai.
(ii) 1/3 will survive with a liver transplant.
(iii) 1/3 will die.

Part IV
Common Problems of Infancy

Inguinal Hernia/Hydrocele

Alfred P. Kennedy Jr.

Inguinal hernia and hydrocele are the most common problem in children requiring operative correction. Hernia and hydrocele are abnormalities along the same spectrum of incomplete closure of the processus vaginalis after testicular descent.

1. Pathophysiology:
   (a) Types of groin hernias:
      (i) Indirect hernia: Formed by patent processus vaginalis (PPV) and exits lateral to the deep epigastric vessels. This form of hernia is most common in children.
      (ii) Direct hernia: Represents a defect within the inguinal floor (transversalis fascia), medial to the epigastric vessels.
      (iii) Femoral hernia: Also represent a defect in the posterior inguinal floor. The hernia passes thru the femoral ring deep to the inguinal ligament and emerges within the femoral canal at the fossa ovalis.
      (iv) Sliding hernia: Any hernia of which a portion of the sac contains viscera.
      (v) Incarcerated hernia: Any hernia in which a portion of viscera is lodged and represents a surgical urgency.
      (vi) Strangulated hernia: Any hernia in which an incarcerated portion of viscera has lost its blood supply and represents a surgical emergency.

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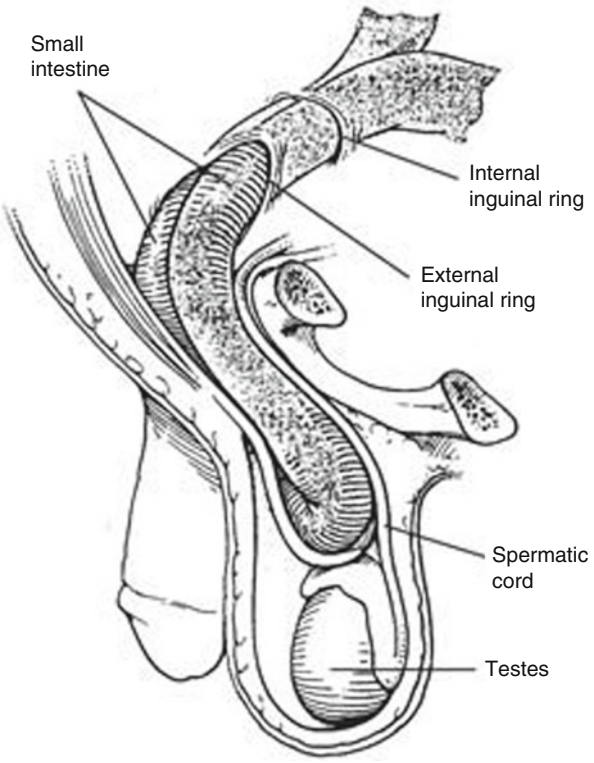

Fig. 1 Indirect inguinal hernia. The indirect inguinal hernia passes first through the internal inguinal ring and then through the external inguinal ring after traversing the inguinal canal (*Source*: National Digestive Diseases Information Clearinghouse, National Institutes of Health, Bethesda, MD. http://digestive.niddk.nih.gov/ddiseases/pubs/dictionary/e-k.aspx. Downloaded 22 Dec 2013)

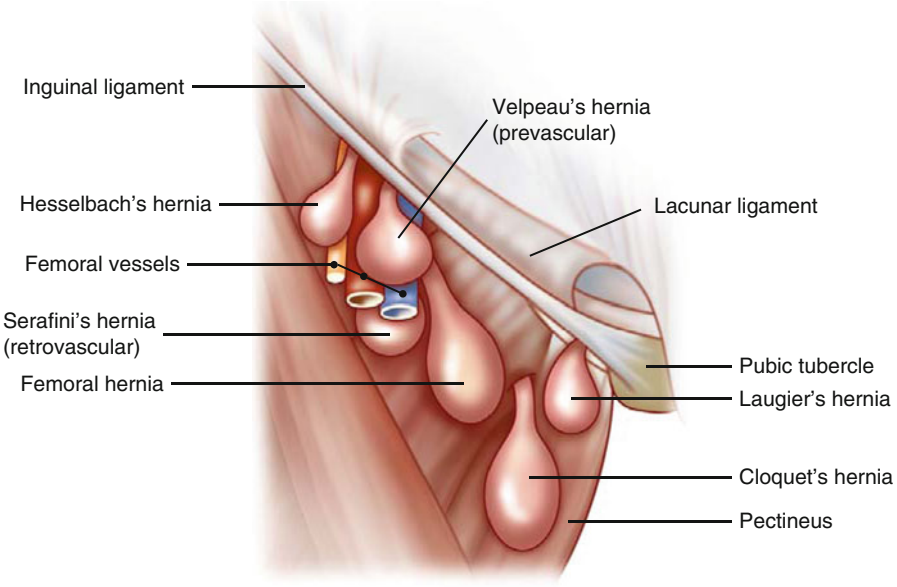

Fig. 2 Femoral hernia types. Femoral hernias are defects in the abdominal wall between the inguinal ligament and the upper pubic ramus (*Source*: Dr. Kumar Nishant, under Creative Commons Attribution Share-Alike 3.0 Unported license, from https://upload.wikimedia.org/wikipedia/commons/6/6e/Femoral_hernia_types.jpg. Downloaded 22 Dec 2013)

(b) Anatomy:

(i) Fascial layers of the abdominal wall contribute to layers of the testis and spermatic cord during testicular descent.

1. Transversalis fascia → internal spermatic fascia.
2. Internal oblique → cremaster.
3. External oblique → external spermatic fascia.
4. Processus vaginalis (peritoneum) → tunica vaginalis.

(ii) Inguinal canal in children:

1. Shorter in infants than adults as the external and internal ring are nearly effaced, which leads to increased risk of incarceration.
2. Scarpa's fascia well developed in children and may be mistaken for external oblique aponeurosis.

(c) Embryology:

(i) Gonads form at 5th week gestation within the retroperitoneal nephrogenic ridges.
(ii) Testis descends to internal ring at seven months gestation.
(iii) Testis descent mediated by calcitonin gene-related peptide (CGRP) released from the genitofemoral nerve.
(iv) Descent of testis proceeds along gubernaculum and "drags" a diverticulum of peritoneum known as the processus vaginalis.
(v) Obliteration of processus vaginalis mediated by CGRP.
(vi) Failure of this obliteration leads to the potential for indirect inguinal hernia formation.
(vii) Ovarian descent arrests within the pelvis.
(viii) Left testicular descent precedes right side therefore right sided hernias are more common as the likelihood of PPV closure decreases after birth.

(d) Incidence:

(i) Undescended testis and PPV are more common in prematurity.
(ii) Approximately 1 in 20 males will develop an inguinal hernia.
(iii) Family history is positive in approximately 10 %.
(iv) Incidence of inguinal herniation increases with prematurity and is estimated within 10–30 %.

(e) Associated Conditions:

(i) Prematurity.
(ii) Cryptorchidism (90 % have PPV).
(iii) Connective tissue disorder.
(iv) Cystic fibrosis.
(v) Ascites.
(vi) Abdominal wall defect.
(vii) Meningomyelocele.
(viii) Ambiguous genitalia.

(ix) Any condition that increases abdominal pressure will likewise increase the chance of a clinical hernia developing e.g. chronic lung disease, ventriculoperitoneal shunt.

2. Clinical presentation:

(a) Asymptomatic inguinal and or scrotal bulge most common and usually discovered by a caregiver.
(b) 60% present on the right side, 25 % on the left.
(c) Diagnosis is clinical and relies squarely on physical examination.
(d) Maneuvers to assist in appreciation include an upright examination and Valsalva maneuver.
(e) "Silk glove sign" involves the sensation of silk rubbing together or a thickened cord upon palpation of the spermatic cord.
(f) Radiography/ultrasonography generally unnecessary. A "negative" groin ultrasound in no way rules out the presence of a hernia.
(g) A "negative" physical exam should prompt caution in groin exploration. May repeat physical exam at a later date.
(h) Treatment of a PPV found during laparoscopy for another condition (e.g. appendicitis) should be deferred until clinical findings are apparent.
(i) Within the female population, a mass is usually found within the labia majora. This often represents an ipsilateral sliding ovary and is the size and shape of an almond.
(j) Incarcerated inguinal hernias present as an inguinoscrotal mass which may be erythematous and tender, are difficult to reduce and often require sedation to do so. The technique involves firm and continuous pressure along the axis of the spermatic cord while stabilizing the ipsilateral testicle. The examiner will note a sudden "pop" as the hernia contents return to the peritoneal cavity.
(k) Strangulated inguinal hernias present with signs of bowel obstruction and or peritonitis. No attempt should be made to reduce the hernia in this situation. Rather, immediate surgical intervention is required.

3. Diagnosis:

(a) Diagnosis is made via detection of the hernia on physical examination.
(b) Parents' description of a bulge that comes and goes at the external inguinal ring is sufficient merit operative repair.

(i) Some conditions may mimic hernia.
(ii) Retractile testicle (able to be displaced into scrotum).
(iii) Lipoma of the cord (empty internal ring).
(iv) Hydrocele (fluid filled sac will transilluminate with a light).
(v) Lymphadenopathy (located below inguinal ligament).
(vi) Neoplasia (testicular mass).

4. Treatment:
   (a) Operative Management:
      (i) Usually performed under general anesthesia.
      (ii) May consider spinal anesthetic for premature infants with BPD to prevent intubation and subsequent prolonged ventilation. Although the incidence of apnea and bradycardia after either form of anesthetic is probably not significantly different.
         1. Postoperative monitoring:
      (iii) Unnecessary for full term infants regardless of gestational age including those receiving a general anesthetic.
      (iv) Risk of apnea and bradycardia increases with prematurity and concomitant lung disease. Our policy is all infants with a history of prematurity and or apnea and bradycardia receive at least 12 h of monitoring if their post conceptual age is less than 56 weeks.
   (b) Timing of operative hernia repair:
      (i) Due to the increased risk of incarceration, premature infants should have their hernia repaired prior to discharge form the NICU, particularly if hernia reduction has been necessary.
      (ii) For outpatients, delay repair until child is at least 56 gestational weeks old.
      (iii) If symptomatic, repair hernia when detected, but plan overnight monitoring in hospital if <56 gestational weeks old.
   (c) Type of repair:
      (i) Indirect Inguinal Hernia:
         1. High ligation of sac at the level of the internal inguinal ring (Marcy repair).
         2. Inguinal incision within Hesselbach's triangle.
         3. Incision of external oblique aponeurosis.
         4. Exposure of spermatic cord.
         5. Exploration of spermatic cord.
         6. High ligation of indirect inguinal hernia.
      (ii) Direct Inguinal Hernia:
         1. McVay or Cooper's ligament repair (approximation of conjoined tendon to ileopubic tract).
         2. Check for concomitant indirect (Pantaloon) hernia.
         3. Consider laparoscopic mesh (Lichtenstein) repair in adolescents.

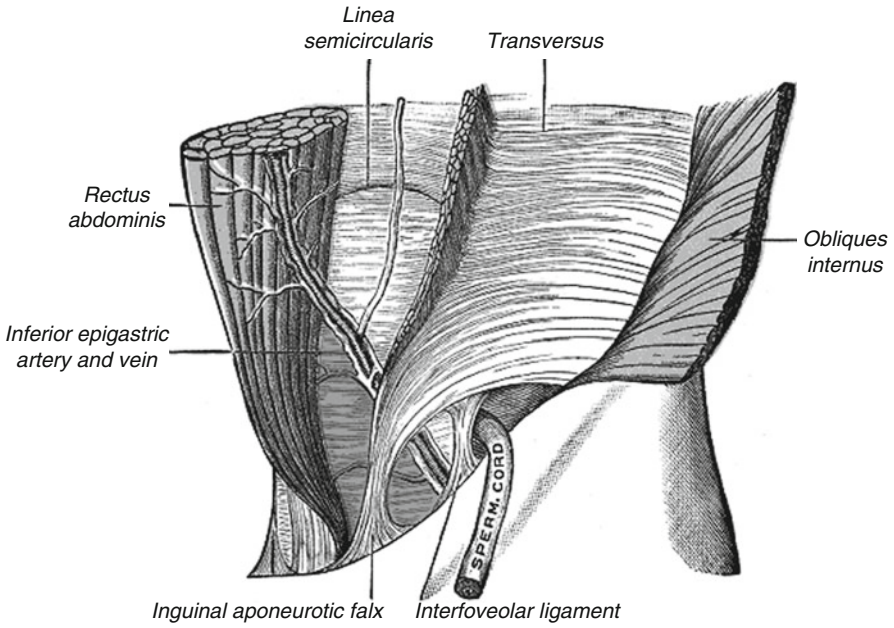

Fig. 3 Hesselbach's triangle from the outside. Hesselbach's triangle is bounded by the lateral margin of the rectus muscle, the inferior epigastric vessels, and the inguinal ligament (*Source*: Gray's Anatomy of the Human Body, 20th U.S. ed., 1918, via public domain reproduction rights)

(iii) Femoral hernia: similar to direct inguinal hernia.
(iv) Incarcerated hernia:

1. If able to reduce, then patient should be admitted for observation of recurrence and subsequent repair within 24–48 h.
2. If unable to reduce, then patient requires emergent repair via inguinal approach.
3. Presence of blood or enteric contents within the hernia sac (strangulation) mandates laparoscopy or laparotomy via La Roque incision.
4. Follow patient yearly for signs of recurrence or testicular atrophy.

(v) Hydrocele:

1. Represent similar pathology as an indirect hernia i.e. PPV.
2. Asymptomatic hydroceles found in a newborn may be observed.
3. Those that fail to resolve by 6-months-old or are noted to vacillate in size (communicating) deserve formal groin exploration.
4. Complete resection of the hydrocele is not necessary and may increase the risk of cord injury.
5. "Hydroceles" that arise in adolescents mandate testicular ultrasound to evaluate for associated lesions such as abdominal tumors.

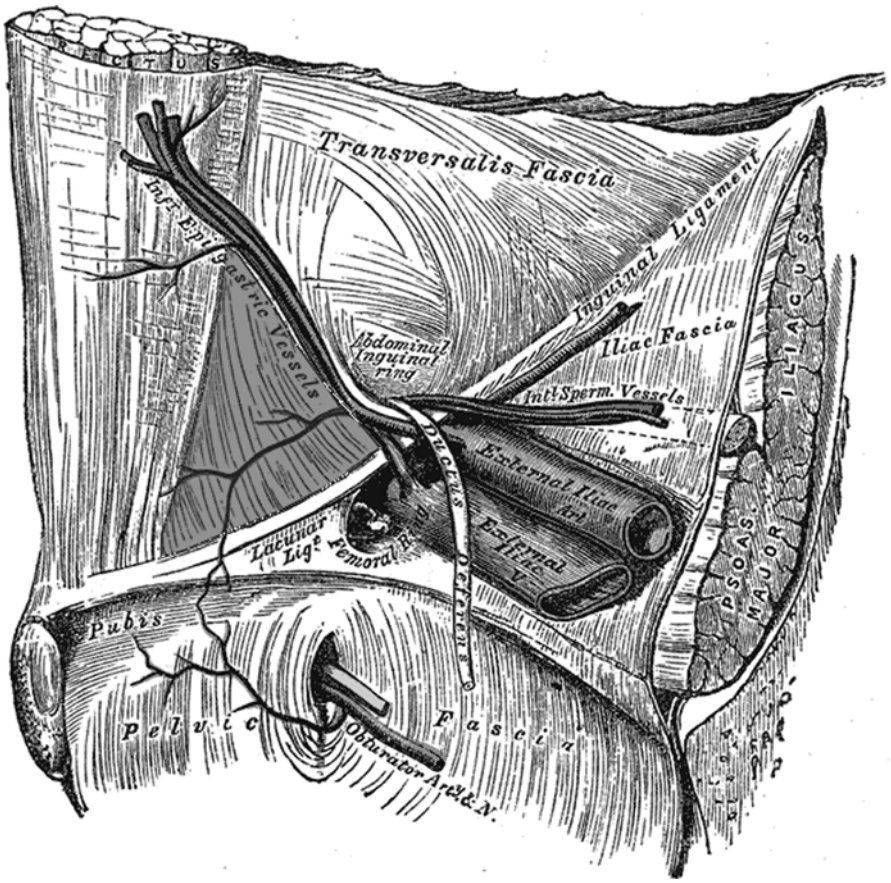

Fig. 4 Hesselbach's triangle from the inside. This is the view seen from the laparoscopic approach (*Source*: Gray's Anatomy of the Human Body, 20th U.S. ed., 1918, via public domain reproduction rights)

5. Outcomes:
   (a) Injury to spermatic cord:
      (i) Incidence approximates 1/1,000 groin explorations.
      (ii) If recognized, perform direct repair of vas deferens with 8-0 absorbable suture, preferably using a microscope.
   (b) Absent vas deferens:
      (i) Consider associated renal abnormality (agenesis) if unilateral.
      (ii) Consider cystic fibrosis if bilateral or atrophic.

(c) Disorders of sexual differentiation:

(i) Suspicion arises when a “testicle” or “epididymis” is found during exploration of the female groin, usually as a sliding indirect hernia.
(ii) Consider laparoscopy and or vaginoscopy to evaluate Mullerian structures (uterus, fallopian tubes, and cervix).
(iii) Transverse biopsy of the gonad should be performed.

Suggested further Reading

Holcomb III GW, Murphy JP. Ashcraft’s pediatric surgery. 5th ed. Philadelphia: Saunders; 2009.
Skandalakis JE, Gray SW, Rowe JS. Anatomical complications in general surgery. New York: McGraw-Hill; 1983.

Umbilical Hernia

Alysia A. Agnoni

Umbilical hernia is a common abdominal abnormality identified in infancy and is usually self-limiting. Only hernias that are very large, symptomatic, or persist past age three will require operative repair.

1. Pathophysiology:
   (a) Etiology: Results from failure of closure of the fascial ring through which the umbilical cord passes. This opening typically closes within a few weeks after the umbilical cord separates.
   (b) Epidemiology: More common in African-American and low birth weight children. Also associated with trisomy 21, congenital hypothyroidism, and Beckwith-Wiedemann syndrome.
   (c) Complications of umbilical hernia are rare.
      (i) Incarceration and/or strangulation.
      (ii) Injury to hernia resulting in rupture or evisceration.
2. Diagnosis:
   (a) Most are asymptomatic and are only identified by visualizing a bulging at the umbilicus.
   (b) Bulge may enlarge with straining and crying.
   (c) Small bowel and/or mesentery typically fill the hernia sac. Most are easily reducible.
   (d) Defect is often small (<1.5 cm) and palpable within the umbilicus.
   (e) There may be redundant skin overlying the umbilicus.

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3. Treatment:
   (a) Reassurance: most spontaneously resolve within the first few years of life.
   (b) Surgical repair:
      (i) Reserved for:
         1. Large defects: >1.5 cm.
         2. Incarceration and/or strangulation.
         3. Children older than age three because resolution is rare after this age
   (c) Technique:
      (i) A general anesthetic is used for sedation. A transverse curvilinear infra-umbilical incision is made. The incision can usually be hidden within a skin fold. The hernia sac is dissected away from the fascia and disposed. Once the edges of the fascial defect are mobilized they are closed using either absorbable or nonabsorbable interrupted sutures. The underside of the umbilical dome is secured to the fascia with a suture. The skin is closed with a running subcuticular absorbable suture. Sometimes a pressure dressing is applied to avoid hematoma.
      (ii) Rarely a 2–3 layer fascial closure (vest-over-pants technique) or mesh repair is needed.
4. Outcomes:
   (a) Complications:
      (i) Wound hematoma.
      (ii) Wound infection.
      (iii) Recurrent hernia.

Intussusception

Alysia A. Agnoni

Intussusception is defined as the invagination of a segment of intestine into the lumen of another segment of intestine causing a bowel obstruction; often referred to as "telescoping" of the bowel. It usually occurs at the ileocolic junction.

1. Etiology: Idiopathic; may be caused by a lead point (Meckel's diverticulum, mesenteric lymph node, hypertrophied Peyer's patch, polyp, or a tumor). Also associated with cystic fibrosis and Henoch-Schonlein Purpura (hematoma of bowel wall can act as lead point). Rarely occurs as a post-operative complication.
2. Epidemiology: Typically occurs before age 1-year-old and most commonly between the ages of 5–10 months. Peak incidence is in the spring-summer and winter months.
3. Pathophysiology: Venous compression at the site of the invaginated bowel causes venous stasis and edema. This can result in a decrease in arterial blood flow to the intestine and eventually necrosis, gangrene, and perforation
4. Clinical Findings:

   (a) History:

      (i) Intermittent severe colicky abdominal pain.
      (ii) Vomiting.
      (iii) Fever.
      (iv) Currant jelly stool.
      (v) Lethargy.
      (vi) Recent upper respiratory infection or viral gastroenteritis.

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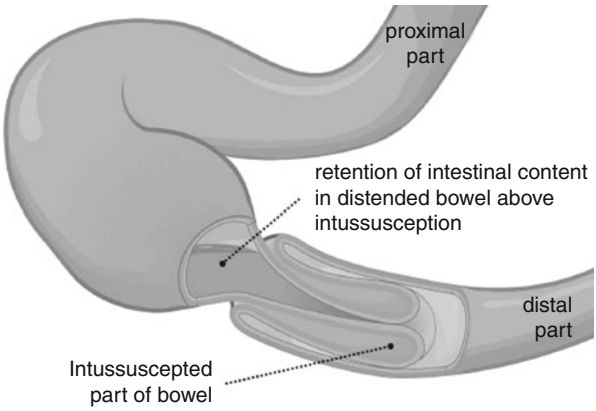

Fig. 1 Intussusception of proximal bowel into distal bowel causing obstruction (*Source*: Aleksander P. Remesz, Creative Commons Share-Alike Unported 3.0 license. https://upload.wikimedia.org/wikipedia/commons/thumb/9/9e/Intussusception_EN.svg/2000px-Intussusception_EN.svg.png. Downloaded 17 Dec 2013)

(b) Physical Exam:

(i) Palpation of a sausage-shaped mass, typically in the right upper quadrant (RUQ).
(ii) Palpation of an empty right lower quadrant (RLQ): Referred to as "Dance's sign".
(iii) Appearance of dehydration.
(iv) If the patient is completely obstructed abdominal distention and/or peritonitis may be present.

5. Diagnosis:

(a) Lab: may have occult blood in stool and leukocytosis.
(b) Plain abdominal films are typically non-diagnostic but may identify an absence of air in the RUQ and evidence of bowel obstruction.
(c) Abdominal sonography: often diagnostic; "target sign" or "pseudokidney sign" is present.
(d) Air or water-soluble enema: this can identify and also reduce an intussusception.

(i) Diatrizoic acid: Gastrografin (Bracco Diagnostics, Princeton, NJ) is the recommended water-soluble contrast and barium should be used cautiously due to the potential for perforation.

(e) Abdominal CT: Should be avoided and is not necessary for diagnosis.

(i) An asymptomatic transient small bowel into small bowel intussusception is commonly an incidental finding on abdominal CT during work-up for another condition and does not necessitate treatment.

6. Treatment:

(a) Nasogastric tube and IV fluid may be needed if excessive vomiting is present.

(b) Antibiotics if perforation is suspected.
(c) Attempting reduction using an air or water-soluble enema is successful in 85 % of children with ileocolic intussusception. If the intussusception cannot be reduced in 1–3 attempts, operative reduction is required.
(d) Older children with intussusception are more likely to have small bowel into small bowel intussusception from a pathologic lead point and attempts at reduction via air or water-soluble enema are typically unsuccessful.
(e) The presence of free-air or peritonitis necessitates surgical exploration and often segmental bowel resection.
(f) Patients older than age three should be considered for surgical reduction because older children and adults often have a surgical lead point.
(g) Surgical technique:

    (i) Usually requires a right-sided infra-umbilical or RLQ incision. The intussusception is delivered through the wound and manually reduced, much like squeezing toothpaste from a tube. Gentle pressure must be applied to avoid iatrogenic perforation. Appendectomy can also be performed at this time.
    (ii) Laparoscopic reduction of intussusception is becoming more common and is usually successful. The terminal ileum is gently pulled out of the colon using atraumatic graspers.
    (iii) If manual reduction is unsuccessful, a bowel resection is performed. An end-to-end anastomosis is often possible but in cases with perforation and spillage of stool into the abdomen an ileostomy may be necessary.
    (iv) Close examination to search for a lead point should also be done during operation.

Foreign Body Ingestion and Aspiration

Christopher P. Coppola

Ingestion of foreign objects and substances is a danger to children once they become mobile in their environment through crawling and before they develop the knowledge to avoid putting non-food objects in their mouths. Some ingestions cause serious and lasting damage to the aerodigestive tract.

1. Pathophysiology:

   (a) Demographics and risk factors:

      (i) Uncommon in age less than 6-months-old because children usually cannot find objects to swallow until they are able to crawl around. An exception to this rule is when older siblings provide objects for younger children to ingest.
      (ii) The behavior is uncommon after age 2-years-old; however there are exceptions due to children acting out or taking dares with peers at older ages.
      (iii) In households where cleaning products or other caustic substances are stored in containers intended for food or drink, there is an increased risk of ingestion by children.

   (b) Ingestion of foreign objects:

      (i) Coins are the most commonly swallowed objects.
      (ii) Button batteries are particularly dangerous. They can cause tissue damage within a few hours. Potential injuries are burn, stricture, perforation, and fistula to vocal cord, trachea, esophagus, and arterial structures. Death is possible, in particular with aortoenteric fistula.

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All swallowed objects must be evaluated to determine if they are a button battery or similar injurious object.

(iii) Swallowed magnets can be a danger. A solitary magnet will usually pass easily if it is small enough, but when multiple magnets are swallowed, they can cause perforation when magnets in neighboring loops of bowel cause pressure necrosis of the bowel walls between them. Magnets are present in household objects, jewelry, and some toys.

(iv) Food impactions can lodge in the esophagus. This may be due to poor chewing, motility disorder or anatomic obstructions such as webs or strictures.

(v) Children can swallow narcotic packets either through error when they are left in reach or through intentional exploitation of the child for drug smuggling.

(vi) Any object that passes the lower esophageal sphincter will usually pass through the entire gastrointestinal tract.

(vii) Sharp objects such as fish bones, pins, needles, and screws can lodge in the wall or cause perforation of the esophagus. If they pass to the stomach, most will pass through the gastrointestinal tract. Rarely can lodge in the appendix.

(c) Ingestion of caustic substances:

(i) Poison control should be contacted to determine components of ingested substances, determine potential for injury, and discuss treatment options.

(ii) American Association of Poison Control Centers: 1-(800)222-1222.

(iii) Acids:

1. Battery acid, cleaning products.
2. Cause coagulation necrosis of mucosa.
3. Appears hemorrhagic or erythematous.
4. Usually injure stomach more than esophagus.

(iv) Alkalis:

1. Lye, drain cleaner.
2. Cause liquefaction necrosis.
3. Injure esophagus more than stomach.
4. Cause deeper injuries and are prone to scarring and stricture.
5. Appear as pale tissue, sometimes with denuded ulcers.

(d) Aspiration of foreign object:

(i) Objects commonly aspirated are seeds, nuts, small toys, and coins.

(ii) Represent a true emergency, and small movements in the position or orientation of the object can turn a partial airway obstruction into a complete asphyxiation.

(iii) Blockage of the trachea is immediately apparent; blockage of a bronchial branch may not become apparent until pneumonia develops.
(iv) Some objects with natural oils or chemicals can cause a chemical or allergic pneumonitis.

2. Clinical presentation:

(a) Parents will usually present after witnessing child swallowing object.
(b) Patient may present with difficulty eating, swallowing, or breathing.
(c) Patient may present with recurrent pneumonias.
(d) Perforation of a hollow viscus will cause presentation with abdominal pain, vomiting, and sepsis.

3. Diagnosis:

(a) History:

(i) Parents may have witnessed child swallowing object/substance.
(ii) Difficulty eating, swallowing and copious saliva.
(iii) Vomiting, hematemesis.
(iv) Coughing, difficulty breathing, chest pain.
(v) If perforation, abdominal pain.
(vi) There may be a history of chronic, recurrent pneumonias.

(b) Physical examination:

(i) Burns or cuts on lips and mouth.
(ii) Changes in breath sounds, unequal breath sounds, wheeze.
(iii) Unable to speak or hoarse voice/cry.
(iv) Abdominal tenderness.
(v) Fever.

(c) Diagnostic imaging:

(i) Chest x-ray:

1. Can detect radio opaque foreign objects and their position.

(a) Antero-posterior and lateral views allow the object location to be determined in two planes.
(b) Consideration must be given to determine if the foreign object is a button battery as this will make removal of the object an urgent necessity.

2. X-ray cannot detect a radiolucent foreign object.

(a) Presence of object in bronchus can be suspected if air trapping in bronchus is present.
(b) Decubitus antero-posterior x-rays of the chest for both right and left decubitus position can make air trapping more apparent.

(c) If a child has multiple episodes of lobar pneumonia in the same lobe of lung, an aspirated foreign object should be suspected.

3. Can reveal perforation and mediastinal or neck air:
4. Foreign objects in the esophagus will bulge out to the sides of the esophagus because of the hard spine behind it and the stiff rings of the trachea anteriorly. This will cause coins in the esophagus to be positioned in a coronal plane.
5. Foreign objects in the trachea will bulge out posterior toward the soft muscular wall of the trachea in the in the direction of the esophagus. This will cause coins in the trachea to be positioned in a sagittal plane.

(ii) Contrast esophagram:

1. Will demonstrate perforation if present
2. Can detect fistula between esophagus and trachea.
3. Can show if an acute or chronic stricture is present.

(iii) Computed tomography:

1. Computed tomography can identify the location of swallowed or aspirated radiopaque foreign objects.
2. It can detect perforation and leak of air or succus from a loop of bowel.

(d) Endoscopy:

(i) Rigid esophagoscopy: useful for both diagnosis and treatment of foreign objects. The wider channel of rigid esophagoscopy allows use of larger instruments for retrieval of foreign objects. Rigid esophagoscopy also allows visualization of the wall of the esophagus without the necessity of air insufflation which can worsen a perforation of the esophagus.

(ii) Flexible endoscopy: Useful for seeing past the length of a rigid scope, such as into the stomach or the duodenum. Small flexible graspers can be inserted through the suction port of the instrument.

(iii) Bronchoscopy: Both rigid and flexible available. Use of a rigid bronchoscope can be set up so that the patient can be ventilated through the bronchoscope during the procedure, and optical grasper can be inserted to retrieve aspirated foreign objects through the bronchoscope.

4. Treatment:

(a) Initial assessment:

(i) If patient has respiratory or cardiac arrest, provide basic life support with maneuvers for airway, breathing, and circulation.

(ii) Make child NPO.

(iii) Assist child to remain calm.
(iv) Provide humidified oxygen if child has difficulty breathing.
(v) Maintain child in seated position, or if more comfortable, lying down on side, so that an object coughed or vomited out does not fall back into airway.
(vi) Start IV for access to provide rapid medication and anesthesia.

(b) Treatment of specific swallowed objects:

(i) Button batteries should be immediately removed if in the esophagus, and within 24 h if in the stomach or intestine and do not pass spontaneously per anus.
(ii) Coins which lodge in the esophagus are removed endoscopically, and allowed to pass if they progress distal to the lower esophageal sphincter. If there is any possibility that the object is a battery, it is removed immediately.
(iii) Sharp objects are removed if in the esophagus or stomach, and allowed to pass if distal, unless they cause perforation or fail to pass. When removed, they are drawn out with the sharp end trailing.
(iv) Objects longer than 5 cm are prone to become lodged in the GI tract and should be removed.
(v) Food impactions in the esophagus are pulled out piecemeal, or pushed into the stomach. If past lower esophageal sphincter, no treatment necessary unless large enough to be a bezoar.
(vi) Ingestion of one magnet is safe, but ingestion of more than one magnet can result in perforation and requires immediate removal.
(vii) Narcotic packets will usually pass spontaneously, but if rupture of packet is suspected, surgical removal is necessary.

(c) Retrieval of foreign object from esophagus:

(i) Perform in OR under general anesthesia.
(ii) Rigid esophagoscopy provides best view, greatest range of instrumentation, largest diameter for extraction.
(iii) Position patient with shoulder roll and neck extended.
(iv) Take care not to injure mouth or throat.
(v) An alternative to esophagoscopy is use of a Foley catheter.

1. Procedure is monitored with fluoroscopy.
2. Foley catheter is passed distal to esophageal foreign object.
3. Balloon is inflated.
4. Catheter is pulled up and out of esophagus, pulling foreign object before it.
5. Care is taken to grasp the foreign object in the mouth, and prevent it from entering the trachea.

(d) Foreign object in stomach or more distal:

(i) Flexible esophagoscopy/gastroscopy has reach to a greater distance.
(ii) Graspers, biopsy forceps, shares, and baskets are smaller, flexible, and must fit through port of scope.
(iii) Provide better magnification and lighting for view.

(e) Retrieval of foreign object from trachea:

(i) Procedure is emergent, delicate, and high risk.
(ii) Team must be ready for rapid delivery of CPR and other emergency support.
(iii) Prepare instruments for tracheostomy and sternotomy, in case these maneuvers are needed.
(iv) Take care not to convert a partial obstruction to a complete obstruction, for example, by tipping a coin in the trachea from a position that allows air passage to one that allows none.
(v) Rigid bronchoscopy is best as it allows viewing of the bronchial tree and instrumentation for removal of foreign objects.
(vi) Use a ventilating bronchoscope which can be hooked up to the circuit of the anesthesia cart.
(vii) Particularly helpful is a lighted grasper, which gives a close-up view from just behind the jaws of the grasper.
(viii) Take care not to crush and fracture objects such as nuts into multiple fragments.
(ix) Suction cannulae and balloon catheters can be used to remove some foreign objects from the trachea.

(f) Care of a caustic ingestion:

(i) Patient is evaluated for aspiration and perforation with chest x-ray and abdominal x-ray.
(ii) Bronchoscopy is used to evaluate for chemical burn to trachea and bronchi.
(iii) Rigid esophagoscopy is used to evaluate extent of burn in esophagus: it is less traumatic than flexible esophagoscopy because insufflation is not needed.
(iv) Flexible endoscopy is used to evaluate stomach after ensuring there is no esophageal perforation.
(v) Patients are evaluated with fluoroscopic swallow study to ensure there is no perforation.
(vi) If perforation is present, patients are maintained NPO and given broad spectrum antibiotics.
(vii) If there is perforation with extensive leaking and soilage in chest, thoracotomy is performed for drainage, and repair if possible.
(viii) Perforation in the abdomen is treated with laparotomy and repair.
(ix) If there is no leakage, clear liquids are given, and diet is slowly advanced.

(x) Patients are monitored for development of esophageal stricture.
(xi) Some patients will require late dilation of stricture and when damage to the esophagus is extensive, repair or replacement of the esophagus may become necessary.

5. Outcome:

(a) Most objects pass through the entire gastrointestinal tract without assistance.
(b) Button batteries and multiple magnets are a particular danger for perforation, fistula, hemorrhage, and sepsis.
(c) Ingestion of caustic substances, particularly alkalis, has the potential to cause extensive and chronic scarring and stricture of the esophagus requiring operative treatment.

Branchial Cleft Remnants

Alysia A. Agnoni

Branchial fistulas, sinuses, and/or cysts result from incomplete obliteration and resorption of a branchial cleft. They are most often identified in childhood.

Branchial Cleft Remnant Fig. 1:

1. Pathophysiology:
   (a) Embryology: Four branchial clefts/arches develop externally along the head and neck bilaterally in the fourth week of gestation. These cover the internal branchial pouches that form structures of the head and neck.
      (i) 1st branchial pouch becomes: eustachian tube, middle-ear cavity, and mastoid air cells.
      (ii) 2nd branchial pouch becomes: palatine tonsil and supratonsillar fossa.
      (iii) 3rd branchial pouch becomes: inferior parathyroid.
      (iv) 4th branchial pouch becomes: superior parathyroid and thymus.
   (b) The majority (75 %) of branchial abnormalities occur from the 2nd cleft. About 20 % occur from the 1st and the remaining occur from the 3rd and 4th.
   (c) Branchial fistula:
      (i) 2nd branchial cleft: Opens along lower third of the neck along the anterior border of the sternocleidomastoid. The opening appears as a very small skin dimple and can secrete mucus. The fistula travels deep through the tissue beneath the platysma and then turns medially and enters the pharynx at the tonsillar fossa.
      (ii) 1st branchial cleft: tract extends to the external auditory canal.

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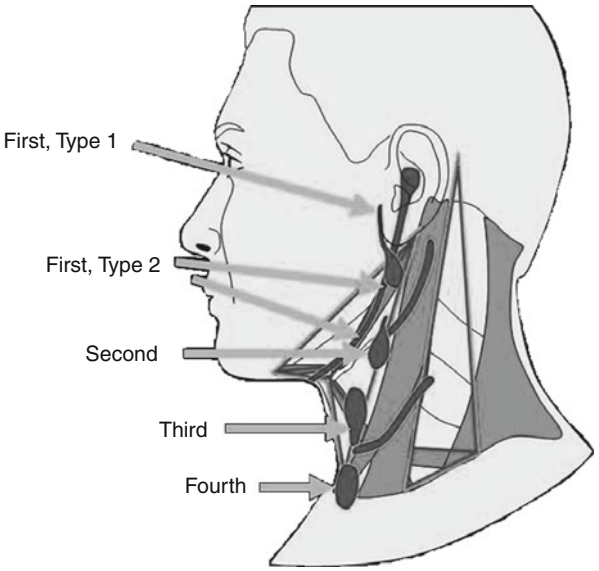

Fig. 1 Location of branchial cleft remnants. Branchial cleft cysts and sinuses are classified based upon the level of branchial cleft remnant from which they arise (*Source*: Adapted from Dr. James Smirniotopoulos, with permission, via http://rad.usuhs.edu/medpix/cow_image.html?pt_id=13954&imid=56175&quiz=no&week=590#top. Downloaded 27 Dec 2014)

(d) Branchial sinus: These only extend a short distance from the skin opening and do not connect to the pharynx.
(e) Branchial cyst: Often palpated along the anterior border of the sternocleidomastoid (2nd branchial cleft) or along the posterior border of the parotid (1st branchial cleft). These may connect to a fistula. They also have potential to become very large and develop infection.

2. Clinical Findings: Diagnosis is usually based on history and physical exam.
   (a) History:
      (i) Lump in neck, may increase in size.
      (ii) Intermittent clear drainage from neck.
   (b) Physical:
      (i) Small sinus opening or cyst palpable along the anterior border of the sternocleidomastoid or posterior border of parotid.
      (ii) Non-tender, unless currently infected.
      (iii) Expression of mucus upon gentle palpation.
   (c) Sonography can help identify the extent of branchial cleft remnants as well as relationship to nearby structures.
3. Treatment:
   (a) Surgical excision:
      (i) A general anesthetic is used and the patient is prepped in the usual sterile fashion.

(ii) Gentle injection of methylene blue dye through an open sinus can be used to identify and highlight a fistulous tract. Another option includes gently probing the opening with a small lacrimal duct probe or stiff suture.
(iii) An elliptical incision over the sinus or cyst is made.

1. Cysts should be bluntly dissected from the surrounding tissue and care should be taken to identify a connection to a fistulous tract.

   (a) If infected, the cyst should be incised, drained, and operative removal done on a separate occasion.

2. Sinuses and fistulas require a more extensive dissection and a deep understanding of the surrounding anatomy.

   (a) 2nd branchial fistulas run over the hypoglossal and hypopharyngeal nerves and between the bifurcation of the carotid artery.
   (b) 1st branchial fistulas run in close proximity to the facial nerve branches.

(iv) Depending on how deep the tract extends a second incision may need to be made superior to the first in a "stepladder" fashion. This allows for better exposure of the tract as it travels superiorly.
(v) Incomplete resection of the tract may result in recurrence.
(vi) Tissues are closed in layers with absorbable suture.

Choledochal Cyst

Christopher P. Coppola

Choledochal cyst is a malformation of the extrahepatic biliary tree. It may be clinically silent in childhood, but frequently it will present with abdominal mass and jaundice. If untreated, it can result in cholangitis or malignancy.

1. Pathophysiology:
   (a) Demographics:
      (i) Increased incidence with Asian heritage: seven times more common.
      (ii) Female to male ratio is four to one.
   (b) Etiology: May be associated with abnormal junction of the common bile duct and pancreatic duct which could result in increased pressure and retrograde flow of pancreatic secretions into the common bile duct.
   (c) Types of choledochal cyst (Classification by Todani, Alonzo-Lej, and others)
      (i) Type I: Cystic dilation of the common bile duct (fusiform) 90 %.
      (ii) Type II: Diverticular malformation of the common bile duct (saccular).
      (iii) Type III: Choledochocele within the wall of the duodenum.
      (iv) Type IV: Intrahepatic and extrahepatic multiple cysts.
      (v) Type V: Multiple intrahepatic cysts.
      (vi) Type VI: Rare variant – cystic dilation of cystic duct.

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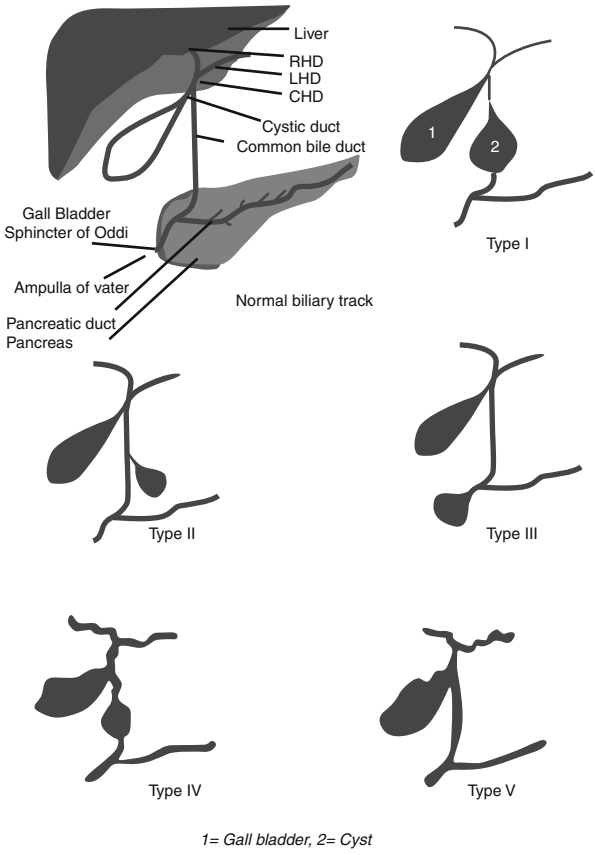

Fig. 1 Types of choledochal cyst. *Type I:* fusiform dilation of the common bile duct. *Type II:* saccular diverticulum of the common bile duct. *Type III:* choledochocele within wall of duodenum. *Type IV:* intrahepatic and extrahepatic biliary dilation, *Type V:* intrahepatic biliary dilation. *CHD* common hepatic duct, *LHD* left hepatic duct, *RHD* right hepatic duct. *1* Gallbladder, *2* Choledochal cyst (*Source*: Drriad, Creative Commons Attribution-Share Alike 3.0 Unported License via https://en.wikipedia.org/wiki/File:Choledochal_cysts.svg. Downloaded 23 Dec 2013)

(vii) Caroli disease:

1. Caroli disease is multiple intrahepatic cysts associated with hepatic fibrosis.
2. Cysts in Caroli disease are bile collections that do not communicate with biliary tree.

(d) Risk of malignancy if left untreated.

2. Clinical presentation:

(a) Prenatal diagnosis with sonography: has become a more common mode of presentation.
(b) Newborns: present with obstructive jaundice.
(c) Infants: present with abdominal pain, abdominal mass, and jaundice.
(d) Adolescents and adults: vague chronic abdominal pain, mass, and occasional pancreatitis.

3. Diagnosis:
   (a) History:
      (i) Jaundice.
      (ii) Abdominal mass.
      (iii) Poor feeding history and delayed growth.
      (iv) Pale color of stools.
   (b) Physical examination: Check for jaundice and abdominal mass.
   (c) Laboratory tests:
      (i) Complete blood count, electrolytes, liver function tests, pancreatic enzymes
      (ii) If liver disease is a possibility, check for viral hepatitis and alpha 1 antitrypsin level
   (d) Imaging:
      (i) Prenatal sonography: a common way in which choledochal cysts are discovered.
      (ii) Sonography: will detect cyst, location of cyst, and if intrahepatic biliary tree is dilated. Sonography is often the only test needed to identify choledochal cyst.
      (iii) Hepatobiliary iminodiacetic acid (HIDA) scan: will reveal if biliary obstruction is present and can differentiate from other causes of neonatal jaundice.
      (iv) Endoscopic retrograde cholangiopancreatography: Useful when biliary obstruction is present, can diagnosis cause and level of obstruction and obstruction can be relieved with sphincterotomy or stent. Will provide an anatomic map of choledochal cyst to help plan operation.
      (v) Intraoperative cholangiography: can aid in identification and dissection of branches of the biliary tree during excision of cyst.
4. Treatment:
   (a) Choledochal cyst is treated with excision.
      (i) Cyst must be excised to address or prevent biliary obstruction.
      (ii) If not resected, cyst carries a risk of future malignancy.
      (iii) Total cyst excision is possible in most cases, however in some, chronic inflammation around the cyst has caused scarring, and it is safer to resect the cyst lining via an intramural plane, to avoid injury to surrounding organs.
      (iv) After excision, drainage of bile from the liver must be re-established
      (v) Method of drainage depends on anatomy of cyst.
   (b) Methods of reconstruction of biliary tree after cyst excision:
      (i) Choledochojejunostomy, with roux-en-y creation.
      (ii) Hepatojejunostomy, with roux-en-y creation.

(iii) Choledochoduodenostomy (less preferable: has increased incidence of cholangitis when compared to roux-en-y drainage).

5. Outcome:

(a) Complications:

(i) Cholangitis.
(ii) Biliary obstruction.
(iii) Anastomotic leak.

(b) Most patients do well with early cyst excision.
(c) Some patients with intrahepatic disease will have progression of intrahepatic disease, ductal ectasia.
(d) If not resected, the epithelial lining of the cyst can undergo malignant degeneration and develop cholangiocarcinoma. There are also reports of cholangiocarcinoma after a cyst has been excised.

Thyroglossal Duct Cyst

Alysia A. Agnoni

Thyroglossal duct cysts are one of the most common causes of midline anterior neck masses in children. These are remnants of the thyroglossal duct. Most thyroglossal duct anomalies present as cysts but draining sinuses can also occur. Although they are congenital, most do not present until childhood or early adolescence.

1. Pathophysiology:
   (a) Embryology: At week four of gestation the thyroid diverticulum forms at the foramen cecum (base of the tongue) and descends forming the thyroglossal duct. As the thyroid reaches its destination in the neck, the thyroglossal tract should atrophy and obliterate.
   (b) Remnants may persist anywhere along the tract from the base of the tongue to the thyroid; however, most occur at the level of hyoid bone.
   (c) Some thyroglossal duct cysts contain thyroid tissue.
2. Clinical findings:
   (a) History:
      (i) Painless lump which may change in size.
      (ii) If the cyst has been infected there may have been drainage.
      (iii) Rarely, hypothyroidism.
   (b) Physical:
      (i) Well-defined, solitary mass: Usually 1–3 cm.
      (ii) Located midline anterior neck, most commonly at the level of the hyoid; some may be higher or lower in the neck.

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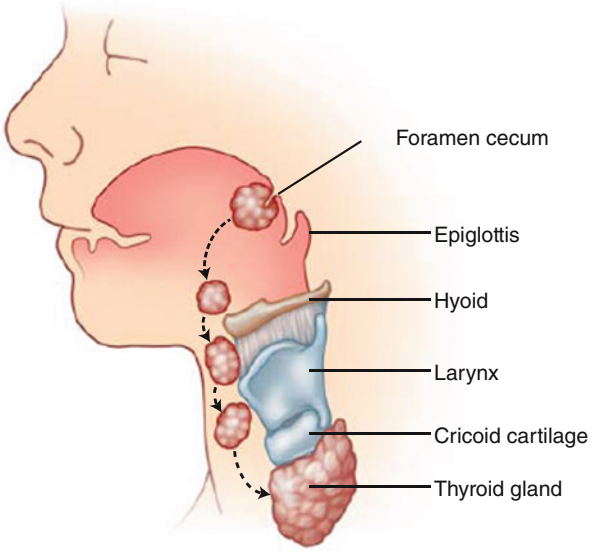

Fig. 1 Potential location of a thyroglossal duct cyst (*Source*: Bryan Walters)

(iii) Mass moves with swallowing.
(iv) Palpation of a normal thyroid gland: may not be present.

3. Diagnosis:

(a) Ultrasound: May help distinguish between dermoid cyst, lymph node, and thyroglossal cyst.
(b) Thyroid scan: In rare cases, ectopic thyroid can present much the same as a thyroglossal duct cyst. If this is the child's only functioning thyroid tissue, removal will leave the patient hypothyroid. This would be a contraindication for excision.
(c) Thyroid function tests.
(d) Fine needle aspiration (FNA): rarely used to confirm diagnosis.

4. Treatment: Surgical excision.

(a) Indications:

(i) High rate of infection if not excised.
(ii) 1–2 % of thyroglossal duct cysts can become malignant.

(b) Sistrunk procedure: resection of the cyst, portion of the hyoid bone, and suture ligation of the tract at the foramen cecum.

(i) The patient is put under general anesthesia with the neck slightly hyperextended. The area is prepped in the usual sterile fashion.
(ii) A transverse incision over the cyst is made; if a draining sinus is present an elliptical incision is made over the opening.

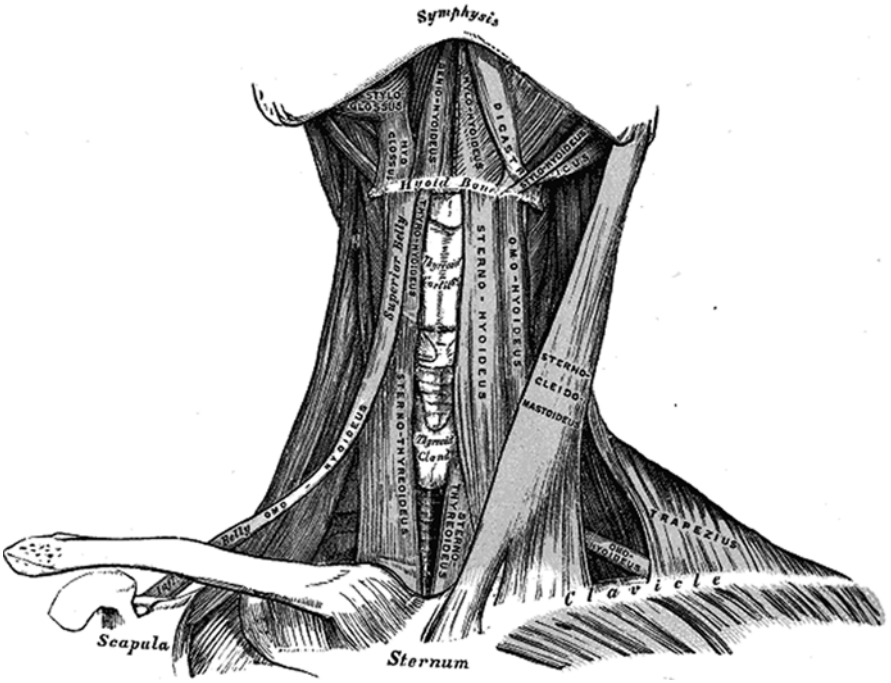

Fig. 2 Muscles of the anterior neck. Resection of a thyroglossal duct cyst involves resection of the central portion of the hyoid bone which has attachments to the strap muscles of the neck (*Source*: Gray's Anatomy of the Human Body, 20th U.S. ed., 1918, via public domain reproduction rights. https://en.wikipedia.org/wiki/File:Gray386.png. Downloaded 23 Dec 2013)

(iii) The cyst and duct remnant are dissected down to the hyoid where the sternohyoid and thyrohyoid muscle attachments are divided enough to see the path of the thyroglossal duct. About 1 cm of the central part of the hyoid, through which the duct remnant passes, is removed. The myohyoid and geniohyoid muscle attachments are divided and dissection is carried out up to the foramen cecum at the base of the tongue. The end of the duct remnant is suture ligated.
(iv) The hyoid is left in discontinuity.
(v) The wound is closed in layers with absorbable suture.
(vi) Cyst and duct remnant should be sent to pathology for evaluation of thyroid tissue and malignancy.

(c) An infected cyst should be treated with a course of antibiotics prior to surgical excision.
(d) Recurrence after complete excision of the duct remnant is low.
(e) Incomplete resection or excision of only the cyst has a higher recurrence rate.

Renal Duplication Anomalies

Joel M. Sumfest

Renal duplication anomalies are common in the pediatric population, but are frequently clinically silent. They will come to the attention of a medical provider if they develop symptoms of infection or obstruction.

1. Pathophysiology:
   (a) Epidemiology:
      (i) Very common.
      (ii) Estimate 1/50 population has a duplicate collecting system.
      (iii) Only come to medical attention if there is associated pathology.
   (b) Definition:
      (i) Renal duplication:
         1. Complete: two separate openings for drainage into bladder.
         2. Incomplete: join together with one ureter entering bladder.
      (ii) Ureterocele:
         1. Cystic dilation of distal ureter.
         2. Can be associated with a single system: described as orthotopic.
         3. Duplex system:
            (i) Ectopic.
            (ii) If extend down urethra submucosally known as cecoureterocele.

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(iii) Ureteral ectopia.

1. Lateral ectopia within bladder results in a short submucosal tunnel of the ureter and usually vesico-ureteral reflux (VUR).
2. Caudal ectopia results in ureteral atresia and obstruction.
3. Possible exit outside bladder and incontinence in girls.

(c) Weigert-Meyer law:

(i) Governs the relationship of duplication and ectopia.
(ii) The ureter from lower pole moiety kidney always is lateral ectopic and refluxes.

1. Can also have concomitant ureteropelvic junction obstruction (UPJO).

(iii) The upper pole ureter terminates caudal and medial and is obstructive.

(d) Ureterocele.
(e) Bladder neck.
(f) Outside of bladder possible in girls.

(i) Continuous incontinence.

(g) In boys can terminate in Wolffian system.

(i) Rare problems with epididymitis.

2. Clinical features:

(a) Historically these children presented with pyelonephritis and/or recurrent urinary tract infections (UTIs) or flank pain.
(b) Most are now discovered on prenatal ultrasound: can create dilemma of how aggressive to be with surgical intervention.
(c) The most abnormal termination of the ureter, i.e. severe VUR or obstruction, the more likely that moiety of the kidney will be dysplastic and poorly functioning: greatly affects surgical approach.

3. Diagnosis:

(a) Evaluate with renal/bladder sonography and voiding cystourethrogram (VCUG).
(b) Mercaptoacetyltriglycine (MAG3) furosemide renal scan, if required, to assess function and/or obstruction.

4. Treatment:

(a) Many options for repair of this diverse spectrum of disorders.

(i) Primarily use a top down approach.
(ii) Do not recommend complete one-stage renal and bladder repair.

(b) Management of the neonate with duplication anomaly:

(i) Common prenatal detection.
(ii) Use antibiotic prophylaxis if significant VUR or hydronephrosis.

(c) Early surgical intervention usually focuses on relief of obstruction.

(i) Primarily from ectopic ureterocele.
(ii) Usually approached endoscopically.

(d) Transurethral puncture of an ectopic ureterocele.

(i) Used to relieve obstruction.

1. Usually of upper pole ureter and kidney.
2. Occasionally of the bladder outlet.

(ii) Preferred method is holmium laser with multiple small punctures.
(iii) Can create iatrogenic VUR.
(iv) In cases of severe VUR other moieties can be more aggressive.
(v) Must be careful in cases of cecoureterocele not to create a flap which can obstruct flow of urine and create bladder outlet obstruction.

(e) Surgical management of the child with duplication abnormality:

(i) Primarily associated with correction of hydronephrosis and stasis of urine responsible for recurrent UTIs and possible renal scarring.
(ii) Must have a high index of suspicion for the ectopic ureter in girls that could result in continuous incontinence once toilet-trained.

1. In the past, children typically presented around age 5 to 9-years-old.
2. MRI is helpful in delineating this complex anatomy: Must inform pediatric radiologist of suspected anatomy so study can be tailored.

(iii) Preferred technique is the top down approach when intervention is required.
(iv) Preserve renal parenchyma if functioning well; however, dysplasia is common and most often involves excision of the poorly functioning segment.
(v) Most commonly this is the upper pole moiety, but occasionally the lower pole if associated with severe VUR.
(vi) Do not address bladder and VUR initially, as in a total reconstruction

(f) Renal reconstruction:

(i) Most often upper-pole heminephroureterctomy is performed: Leave relatively normally functioning lower pole moiety and ureter intact.
(ii) Can be approached open via flank extraperitoneal incision or laparoscopically: Use of robotic assistance is helpful.
(iii) Pre-stent normal ureter to aid in identification and preservation.
(iv) Great care mobilizing the upper pole megaureter under the renal vessels.

1. Undue tension on these small vessels can result in spasm or thrombosis.
2. Less likely with laparoscopic approach.

(v) Usually small branch to upper pole segment, which once divided shows clear demarcation for excision.
(vi) Must be aware that lower pole segment can be injured if not careful and result in bleeding and/or entry into the collecting system.
(vii) Upper pole ureter is excised down to pelvis where the two ureters tend to share a common wall.
(viii) Stent is removed at end of case.
(ix) If the upper pole segment is thought to have good function can consider upper to lower pole ureteropyelostomy (preferred for stenting).
(x) Both of these approaches will resolve issue of ureteral ectopia and in cases of massive hydroureteronephrosis take care of urinary stasis.
(xi) Estimated that 66 % duplication anomalies can be handled with renal approach alone.
(xii) Additional third will need bladder reconstruction, primarily for severe VUR.

(g) Bladder reconstruction in duplication anomalies:

(i) Must address severe VUR and or ureteroceles.
(ii) The ureterocele is excised or marsupialized.
(iii) Ipsilateral and even the contralateral ureters may need to be reimplanted.
(iv) Remove remaining stump of upper pole ureter at this time.
(v) Can result in a large defect in the base of the bladder.

1. This must be closed.
2. Take care to avoid injury to underlying vagina.

(vi) Use of double-J stenting is helpful.
(vii) Usually can manage with Foley catheterization postoperatively.
(viii) Close follow-up for ureterovesical junction obstruction and/or persistent VUR.

Cryptorchidism

Joel M. Sumfest and Alfred P. Kennedy Jr.

Cryptorchidism or undescended testis is one of the most common congenital abnormalities seen in pediatric urology. It has been studied extensively but etiology still remains unclear.

1. Pathophysiology:
   (a) Incidence:
      (i) Up to 45 % incidence in pre-term males.
      (ii) Approximately 1–4 % incidence in full-term infants.
      (iii) At 6-months-old, incidence is approximately 1 %.
   (b) Most UDT will descend within the first year of life; those that descend will not require operative treatment.
   (c) UDT is usually unilateral.
   (d) Primary or congenital non-syndromic variant most common.
   (e) Secondary UDT is after prior inguinal surgery.
   (f) Anatomic location: may be located anywhere along the in-utero path of migration from the retroperitoneum to the high scrotum.
      (i) Intraabdominal (nonpalpable UTD).
      (ii) Annular: at the level of the internal ring.
      (iii) Cannilicular: within the inguinal canal (most common).

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(iv) High scrotal or retractile testis.
(v) Ectopic locations:

1. Perineal.
2. Femoral.
3. Penopubic.

(g) Pathogenesis:

(i) Unilateral UDT:

1. Usually developmentally abnormal testicles.
2. May never achieve spermatogenesis if not treated operatively early in life. However, the paternity rate of men with unilateral UDT approaches the normal population.
3. Have a shortened vascular supply.
4. More prone to develop malignancy later in life. Risk of malignancy of the UDT is upward of 60 times greater than normal. Orchidopexy does not prevent this risk, but allows for easier testicular examination and potentially earlier detection of a testicular mass. Intraabdominal (non-palpable) UDT has the highest risk of premalignant degeneration. The most frequent tumor type is a seminoma. Non-seminomatous germ cell tumors are most common in those UDT that have undergone orchidopexy. Interestingly, as many as 15 % of cancers that arise in patients with UDT do so in the normally descended testicle.

(ii) Bilateral UDT:

1. Likely due to a hormonal deficiency preventing in-utero descent.
2. May be associated with an anatomic anomaly that is responsible for the UDT.
3. Patients with bilateral UDT have fertility rates of only 50–65 %, even after early orchidopexy.

(h) Associated conditions:

(i) Defects of androgen synthesis or receptors.
(ii) Disorders of sexual differentiation.
(iii) Klinefelter's syndrome.
(iv) Down's syndrome.
(v) Prune belly syndrome (Eagle-Barrett syndrome).
(vi) Omphalocele and gastroschisis.
(vii) Bladder exstrophy.
(viii) Hypospadias.
(ix) Posterior urethral valves.
(x) Upper tract genitourinary anomalies, such as renal agenesis.

2. Diagnosis:
   (a) Infants and boys should be examined supine, frog-legged, and if possible standing.
   (b) Use of liquid soap helpful to palpate UDT under prominent fat pad.
   (c) Retractile testes are the most common diagnosis in differential.
      (i) Does not represent a true UDT.
      (ii) May be manipulated into the scrotum whereas an UDT can never be manipulated to the confines of the scrotum.
      (iii) Sometimes a difficult distinction, especially in a "ticklish" or hyperactive child.
      (iv) Cremasteric reflex responsible for retraction is diminished with frog-legged position.
      (v) May without manipulation normally lie within the inguinal canal.
      (vi) Ipsilateral hemiscrotum is normally developed.
      (vii) Is normal with respect to spermatogenesis.
      (viii) Has no increased risk of malignant degeneration.
      (ix) If unclear, repeat examination another setting.
      (x) Requires no other treatment than parental reassurance.
   (d) Classify palpable vs. non-palpable UDT.
      (i) Guides surgical approach.
   (e) Palpable UDT:
      (i) Majority of UDTs are palpable.
      (ii) Most are low or present in inguinal canal. Some can be "peeping" in and out of internal ring.
      (iii) Almost all can be approached surgically in standard fashion with inguinal orchidopexy.
   (f) Non-palpable UDTs:
      (i) Approximately 15 % all UDTs.
      (ii) In morbid obese boys consider ultrasound to localize testis.
      (iii) No indication for routine imaging of UDT.
      (iv) In large series, half of the UDTs are intra-abdominal and the remainder has undergone prenatal torsion and can be classified as "vanishing" testes.
      (v) Bilateral nonpalpable UDT must be distinguished from anorchia. This may be accomplished with a beta human chorionic gonadotropin (bHCG) stimulation test including baseline measurements of serum testosterone, follicle stimulating hormone (FSH) and luteinizing hormone (LH). Laparoscopy is also useful to determine if testes are present.

3. Treatment:

   (a) Orchidopexy is indicated to maximize spermatogenic potential, reduce or aid in diagnosis of testicular malignancy, provide cosmetic benefit, and treat associated hernia if present.
   (b) Infants should be followed for spontaneous descent until 6 months of age (taking into account prematurity). There is no indication for observation after this time point.
   (c) There is no current indication for hormonal treatment.
   (d) When a palpable UDT presents in adolescence, orchiectomy may be the best choice for an abnormally small testicle, especially if the contralateral descended testicle is normal on examination.
   (e) For a unilateral nonpalpable UDT found after puberty, orchiectomy is recommended due to its malignant potential and unlikely potential for spermatogenesis.
   (f) Inguinal orchidopexy:

      (i) Inguinal orchidopexy is the standard approach for palpable UDT.
      (ii) Small transverse incision is made in the inguinal skin crease. If UDT is high in the canal, center incision closer to the internal ring.
      (iii) The gubernaculum is divided with cautery and the spermatic cord is skeletonized back to internal ring.
      (iv) If UDT is in the inguinal canal, may need to open external oblique fascia, but this is not routinely necessary.
      (v) If a patent processus vaginalis is present, it must be carefully mobilized off the cord structures and a high ligation of hernia sac is performed.
      (vi) All lateral spermatic attachments are divided. If necessary, in cases of higher UDT, formal division of all retroperitoneal attachment is accomplished by following the cord up under the peritoneum.
      (vii) May involve opening the inguinal floor to allow sufficient mobilization.
      (viii) Placement of testis in subdartos pouch:

         1. Creation of a subdartos pouch through a separate mid scrotal incision.
         2. Two point fixation: Sutures are placed between the tunica vaginalis and dartos fascia to secure the testicle in place. No sutures are placed thru the tunica albuginea of the testicle.
         3. Take care to avoid undue tension or torsion of the spermatic cord.

   (g) Surgical approach for non-palpable UDT:

      (i) Top down vs. bottom up approach: Top-down approach with diagnostic laparoscopy is preferable.
      (ii) Careful examination under anesthesia for remnant of testis in scrotum. If scrotal remnant present, scrotal exploration will suffice.

(iii) On laparoscopy, occasionally blind-ending vessels will be found; indicating absence of the testis, and no further treatment is needed.
(iv) Spermatic vessels must be located. Occasionally, the vas is seen, and the UDT can be completely separate in a much higher position.
(v) If UDT is intraabdominal and thought to be capable of being mobilized into the scrotum, a standard laparoscopic orchidopexy is performed.
(vi) If bilateral intra-abdominal UDTs are present, or the UDT is very high in position, consider ligation of the spermatic vessels in a staged Fowler-Stephens procedure. Ligation of spermatic vessels must be done primarily, and not after mobilization of the cord.

(h) Laparoscopic orchidopexy:

(i) Incise peritoneum lateral and medial to internal spermatic vessels and then mobilize vessels as high as feasible.
(ii) Beware underlying iliac vessels, ureter, and bowel.
(iii) Leave triangle of peritoneum between vas, UDT and vessels.
(iv) Divide gubernaculum.
(v) If open internal ring, perform hernia repair.
(vi) Develop subdartos pouch.
(vii) Place laparoscopic trocar through scrotum and pouch into abdominal cavity.

1. Enter peritoneum through, or just lateral to median umbilical ligament.
2. Bladder must be empty.

(viii) Fixation in pouch:
(ix) If there is tension on the cord, sometimes additional length can be gained with further mobilization superior to vessels.

(i) Staged laparoscopic Fowler-Stephens orchidopexy.

(i) Usually 6–9 month interval from initial ligation of vessels to second stage of operation.
(ii) UDT mobilized on vas with the new large collateralization of vas arteries, which hopefully has formed.
(iii) Success only 60–70 % long-term.

4. Outcomes:

(a) Successful placement of UDT in intra-scrotal position via standard inguinal orchidopexy without atrophy is possible in over 98 % of cases in experienced hands.
(b) Up to 80–90 % success rate is possible after laparoscopic orchidopexy.
(c) Scrotal location of UDT does not change potential for malignant changes later in life. Testis must be monitored for masses with regular examination. Self-examination is vital in adults who have undergone orchidopexy as a child.

Vesicoureteral Reflux

Steven V. Kheyfets and Joel M. Sumfest

Vesicoureteral reflux (VUR) is significant in that an incompetent ureterovesical junction (UVJ) allows the retrograde flow of urine from the bladder into the ureter which may result in significant upper urinary tract pathology.

1. Epidemiology:
   (a) Overall incidence: Affects about 1–2 % of the general healthy population.
   (b) Found in up to 50 % of children with history of symptomatic urinary tract infection (UTI).
   (c) Incidence is inversely related with age.
   (d) Affects Caucasian children more commonly.
   (e) Genetics:
      (i) Primary VUR is familial.
      (ii) Autosomal dominant inheritance with variable penetrance.
      (iii) Increased familial prevalence.
         1. 32% of siblings of children with VUR.
         2. Affects roughly 2/3 offspring of parents with VUR.
   (f) Gender:
   (g) Male children are affected earlier in life and often with higher grades of VUR.
   (h) Febrile UTI more commonly leads to diagnosis of VUR in female children.

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2. Pathophysiology:
   (a) Primary VUR is related to a congenital anatomic defect within the ureterovesical junction.
   (b) Secondary VUR refers to a separate pathology leading to retrograde flow of urine across the ureter primary due to increased bladder pressures.
      1. Neuropathic bladder dysfunction.
      2. Posterior urethral valves.
      3. Dysfunctional voiding.
      4. Iatrogenic.
3. Clinical signs:
   (a) Primarily related to symptomatic UTI or renal failure.
      (i) Failure to thrive.
      (ii) Isolated fever: Can be a sign of pyelonephritis.
      (iii) Abdominal pain.
      (iv) Urinary frequency/urgency.
      (v) Urinary incontinence.
      (vi) Flank pain.
4. Evaluation:
   (a) Physical examination:
      (i) Flank tenderness.
      (ii) Suprapubic distention.
      (iii) Lower back inspection for signs of spina bifida, such as a tuft of hair.
   (b) Urinalysis and urine culture.
   (c) Renal function.
   (d) Imaging of lower urinary tract.
      (i) Ultrasound.
      (ii) Voiding cystourethrogram (VCUG).
      (iii) Radionucleotide cystogram.
      (iv) Magnetic resonance voiding cystography.
      (v) Imaging of the upper urinary tract.
         1. Renal ultrasound: May reveal scarring, hydronephrosis, duplication anomalies or renal growth retardation, however, a normal study does not exclude the presence of VUR.
         2. Dimercaptosuccinic acid (DMSA) renal scan may identify acute pyelonephritis or scarring.
         3. Excretory magnetic resonance urography (MRU) may detect ureteral duplication and ectopia.
5. Screening the siblings of children with VUR.
   (a) VCUG is recommended if there is evidence of renal cortical abnormalities or in cases of a previous UTI which has not been assessed.

6. Screening a neonate with history of prenatal hydronephrosis.
   (a) VCUG is recommended for neonates with high-grade hydronephrosis.
7. Grading of VUR according to International Reflux Study Committee system.
   (a) Grade I: Reflux occurs part way up the ureter.
   (b) Grade II: Reflux involving the ureter and renal pelvis.
   (c) Grade III: Similar to Grade II with additional findings of mild tortuosity of ureter and mild calyceal blunting.
   (d) Grade IV: Moderate dilatation and tortuosity of the ureter and collecting system with blunting of fornices.
   (e) Grade V: Severe dilatation and tortuosity of the ureter and collecting system with absence of papillary impressions in the calyces.
8. Medical management.
   (a) Intervention goals include prevention of kidney damage, which may otherwise lead to hypertension and renal insufficiency.
   (b) According to the American Urologic Association (AUA) guidelines, antibiotic prophylaxis should be instituted in children with severe reflux (grades IV and V) who are less than 1-year-old.
      (i) Amoxicillin, cephalexin or trimethoprim may be used in children less than 2 months of age.
         1. Trimethoprim-sulfamethoxazole or nitrofurantoin may be used in children greater than 2-months-old; its use may result in kernicterus in children less than 2-months-old and, thus, should be avoided.
         2. Antibiotic suppression also remains an option in lower grades of VUR in children less than 1-year-old without the presence of a febrile UTI.
   (c) As per the AUA guidelines, in children with VUR over the age of 1 year without bladder/bowel dysfunction, recurrent febrile UTI or renal cortical abnormalities, antibiotic suppression or observation both remain options.
   (d) As per the AUA guidelines, in children with VUR over the age of 1-year-old with bladder/bowel dysfunction, recurrent febrile UTI or renal cortical abnormalities, antibiotic suppression is recommended.
   (e) As per the AUA guidelines, if a breakthrough UTI (BT-UTI) occurs, a change in therapy is recommended.
      (i) In children not receiving antibiotic suppression who develop a febrile UTI, initiation of antibiotic suppression is recommended.
      (ii) In children not receiving antibiotic suppression who develop a non-febrile UTI, initiation of antibiotic suppression is an option.
      (iii) In children receiving antibiotic suppression who develop febrile UTI, surgical treatment is recommended, however, initiating an alternative antibiotic agent in children with a single BT-UTI and without evidence of previous or new renal cortical abnormalities prior to surgical intervention remains an option.

9. Surgical Management:
   (a) Ureteral reimplantation surgery:
      (i) Open surgical techniques aim is to create a flap-valve mechanism of the UVJ via establishing a submucosal tunnel with an ideal ratio of 5:1 between the tunnel length and ureteral diameter.
         1. Open surgical approaches include extravesical reimplantation (e.g. Lich-Gregoir) and intravesical reimplantation (e.g. Cohen cross-trigonal, Politano-Leadbetter); these approaches have a success rate of 98 %.
      (ii) Endoscopic treatment is a minimally invasive option that involves injection of a bulking agent into the submucosal tunnel below the ureteral orifice; Deflux (dextranomer microspheres, Salix Pharmaceuticals, Raleigh, NC) is FDA approved, and the most commonly used agent.
      (iii) Laparoscopic and robotic ureteral reimplantation is also an option.
10. Outcome:
   (a) The natural history of VUR often involves spontaneous resolution.
      (i) Spontaneous resolution is more common in younger children, in cases of lower grades of VUR and in unilateral VUR.
   (b) Open surgical resolution rate is 97 % in children with and without bladder/bowel dysfunction while endoscopic resolution rate is 50 % in children with bladder/bowel dysfunction and 89 % in children without bladder/bowel dysfunction.
   (c) Complications of surgery include persistent reflux and obstruction.
11. Follow-up after resolution of VUR (spontaneous or surgical):
   (a) Obtaining an ultrasound following surgery to rule out presence of obstruction is standard of care.
   (b) Obtain blood pressure, height and weight and urinalysis annually through adolescence.
   (c) Evaluate for bladder/bowel dysfunction or recurrent VUR if child experiences a febrile UTI.
   (d) A family discussion should convey the long-term concerns of hypertension (especially during pregnancy), renal insufficiency, recurrent urinary tract infection, and familial VUR in the child's siblings and offspring.

Megaureter

Steven V. Kheyfets and Joel M. Sumfest

Megaureter refers to a ureteral diameter that exceeds 5 mm in childhood.

1. Epidemiology:
   (a) Primary obstructed megaureter: 1 per 10,000.
   (b) Megaureter more common in males and has predilection to the left side.
2. Pathophysiology:
   (a) Megaureter is either of primary or secondary origin.
      (i) Primary megaureter is thought to be a consequence of an adynamic portion of distal ureter usually found at the ureterovesical junction (UVJ), resulting in deficient peristalsis and proximal obstruction.
         1. Pathology reveals increased deposition of collagen type I and III along with decreased smooth muscle fibers in the adynamic ureteral segment.
      (ii) Secondary megaureter is caused by a different process.
         1. Urinary tract infection.
         2. Bladder stone.
         3. Neurogenic bladder.
         4. Bladder mass.
         5. Bladder outlet obstruction.

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(b) Megaureter can be further divided into the following types:

(i) Obstructed.
(ii) Refluxing.

3. Associated Conditions:

(a) Primary refluxing megaureter is associated with prune belly syndrome.
(b) Secondary refluxing megaureter is associated with posterior urethral valves.

4. Diagnosis:

(a) 50 % of cases are detected prenatally, and 50 % are discovered after delivery. Infants are usually asymptomatic, while children may have symptoms of failure to thrive, nausea and emesis.

(i) Physical exam seldom reveals a palpable abdominal mass.
(ii) Urinalysis, urine culture.
(iii) Electrolytes, renal function tests.

(b) Imaging:

(i) Renal ultrasound.
(ii) Bladder ultrasound.
(iii) Voiding cystourethrogram (VCUG).
(iv) Renal scan.
(v) MRI urogram.

(c) Diagnostic procedures:

(i) Urodynamics: Evaluate for voiding dysfunction.

5. Treatment:

(a) Spontaneous resolution of primary refluxing megaureter of infancy can occur.

(i) Observe with imaging studies and antibiotic prophylaxis (optimal).

(b) Indications for surgery include the following:

(i) High grade reflux without improvement.
(ii) Worsening renal function with observation.
(iii) Recurrent breakthrough febrile urinary tract infections.

6. Surgical management:

(a) Intravesical, extravesical or combined approaches may be used for ureteral reimplantation. A ratio of at least 5:1 between the submucosal ureteral tunnel and ureteral diameter needs to be maintained.

(i) Tapered repair includes plication: Starr or Kalicinski techniques.

1. Excisional tapering: Hendren technique used for an extremely dilated ureter.

(ii) Endoscopic subureteric injection is not widely used.

7. Complications of surgery:
   (a) Reflux, ureteral obstruction, ureteral stricture, diverticula.
8. Results:
   (a) 90% success rate with operative management.
9. Follow up:
   (a) Medical management:
      (i) Urinalysis, urine culture, renal function and renal/bladder ultrasound every 3–6 months.
      (ii) Interval nuclear renal scan.
   (b) Surgical management:
      (i) Renal/bladder ultrasound 3–6 months post-surgery.
      (ii) VCUG at 6 months post-surgery.

Ureteropelvic Junction Obstruction

Alison M. Rasper and Joel M. Sumfest

Pediatric ureteropelvic junction (UPJ) obstruction can occur in all age groups; increased detection in neonates due to prenatal hydronephrosis can be seen on ultrasonography. It is more common in boys than girls and more common on the left side.

1. Pathophysiology:
   (a) Causes of UPJ obstruction:
      (i) Intrinsic: interruption in development of circular musculature of the UPJ causing fibers to be widely separated and leading to functional discontinuity of muscular contractions, poor emptying; other causes include valvular mucosal folds or upper ureteral polyps.
      (ii) Extrinsic: aberrant, accessory, or early-branching lower pole vessel passing anteriorly to the UPJ.
      (iii) Secondary UPJ obstruction: severe VUR with tortuous course causing proximal kinking of ureter due to fixation causing obstruction.
   (b) Associated anomalies:
      (i) Renal dysplasia.
      (ii) Multicystic kidney.
      (iii) Duplicated collecting system: more typically lower segment.
      (iv) Vesicoureteral reflux.
      (v) Horseshoe kidney or ectopic kidney.
      (vi) VACTERL constellation of anomalies.

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2. Clinical presentation:
   (a) Infants: Asymptomatic; previously found due to palpable masses but now found on prenatal ultrasonography; occasionally due to failure to thrive, feeding difficulties, urosepsis, presence of nephrolithiasis.
   (b) Older children: Episodic flank pain, cyclic vomiting; occasionally present with hypertension due to reduced blood flow leading to renin-mediated hypertension.
3. Diagnosis:
   (a) VCUG: rule out VUR.
   (b) Renal scan: degree of obstruction and renal function.
   (c) Consider operative repair if decline in ipsilateral decline in renal function > 10 % or symptomatic.
   (d) If <10 % relative renal function of affected side, may consider nephrectomy.
   (e) If no intervention indicated, surveillance with renal ultrasound or renal scan every 3–6 months.
4. Treatment: surgical repair by open, endoscopic, laparoscopic, and robotic-assisted approaches with the three main groups being flap type, incisional-intubated type, and dismembered type.
   (a) Open approaches (open pyeloplasty):
      (i) Typically dismembered pyeloplasty: preserves lower pole or crossing vessels, excision of pathologic UPJ with repositioning, and successful reduction.
      (ii) Approach can be subcostal, flank or posterior lumbotomy.
      (iii) Problematic if inadequate ureteral length: can use spiral flap if sufficient extra-renal pelvis to accommodate this gap.
      (iv) Most common is Anderson-Hynes pyeloplasty: identify the UPJ, traction sutures placed, transect the UPJ, ureter spatulated on lateral side, pelvis opened medially or excision of redundant pelvis, align the pelvis and spatulated ureter to allow for dependent drainage, close with anastomotic suture; can be done with or without a stent.
      (v) Other options:
         1. Foley Y-V plasty: good for high insertion of the ureter.
         2. Spiral flap: long gap with adequate extrarenal pelvis.
         3. Vertical flap: dependent junction with large box-shaped extrarenal pelvis.
      (vi) Outcomes: dependent on minimal handling of ureter at time of repair, use of internal stenting or nephrostomy tube drainage.
         1. Early complications: uncommon, prolonged urinary leak from drain which can usually be managed conservatively or may require ureteral stenting.

(b) Endoscopic approaches (endopyelotomy):

(i) Antegrade or retrograde.
(ii) Balloon dilation, laser incision, or Acucise ureteral cutting balloon device (Applied Medical, Rancho Santa Margarita, CA).
(iii) Requires stenting for at least six weeks typically.
(iv) Higher rates of recurrence.

(c) Laparoscopic approaches (including robotic-assisted pyeloplasty):

(i) Typically the dismembered approach as discussed previously: allows for excision of stenotic segment and preservation of vessels.
(ii) Non-dismembered pyeloplasty technically easier as allows for easier tension free suturing.
(iii) Robotics allows for improved anastomotic suturing.
(iv) Transabdominal approach as compared to retroperitoneal open approaches.
(v) Minimally invasive, smaller scars.
(vi) Patient needs to be large enough to allow for adequate pneumoperitoneum and work space to complete repair: typically older patients.
(vii) Intraoperative complications: bleeding, trocar damage to other structures including bowel or vessels, conversion to open.
(viii) Postoperative complications: wound infection, persistent urine leak, UTI.

Gastroesophageal Reflux Disease

Amber Batool

Gastroesophageal reflux (GER) is defined as a normal passage of gastric contents into the esophagus or oropharynx through a transient relaxation of lower esophageal sphincter. GER is a normal physiologic occurrence in children and occurs in two thirds of infants for the first year of life. Gastroesophageal reflux disease (GERD) consists of symptoms and complications secondary to persistent GER.

1. Pathophysiology: The causes and risk factors of GERD are multifactorial and increase in physiologic reflux is attributed to the following factors:

   (a) Motility disorders.
   (b) Hiatal hernia.
   (c) Obtuse angle of His.
   (d) Neurodevelopmental disabilities, such as Down's syndrome and cerebral palsy.
   (e) Premature birth.
   (f) Hypotonia.
   (g) H. pylori infection.
   (h) Atopic disease.
   (i) Medications, e.g. diazepam, theophylline.
   (j) Supine position.
   (k) Poor dietary habits: High frequency of feeds in infants.
   (l) Smoking, alcohol.
   (m) Obesity.
   (n) Psychological factors: Stress, anxiety, depression.

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2. Complication of GERD can be categorized into three systems:
   (a) Respiratory: Asthma, chronic cough, nocturnal cough, apnea and acute life-threatening events (ALTEs), recurrent aspiration pneumonia.
   (b) Ear Nose and Throat: Hoarseness, laryngitis, sinusitis, dental erosions, recurrent otitis media.
   (c) Gastrointestinal: Esophagitis, esophageal ulcers and strictures, Barrett's esophagus.
3. Clinical features:
   (a) Infantile GERD can present as excessive crying, irritability, arching of back during feeds, recurrent vomiting, feeding refusal, poor weight gain, and sleep disturbances.
   (b) In older children and adolescents, GERD usually presents as heartburn or retrosternal chest pain and regurgitation.
4. Diagnosis:
   (a) In most cases of GERD, a thorough history and physical examination is sufficient for diagnosis. Diagnostic imaging should be utilized as needed for atypical presentation and to rule out complications, predisposing conditions, or alternative diagnoses.
   (b) The proton pump inhibitor (PPI) test: A trial of empiric treatment with PPI can be utilized to confirm the diagnosis with resolution of symptoms.
   (c) The following studies can be utilized to diagnose GERD and associated complications:
      (i) The upper gastrointestinal series (UGI) is nonspecific for GER but is useful to detect anatomic abnormalities including vascular rings, achalasia, esophageal and intestinal webs, strictures, hiatal hernia, pyloric stenosis, and malrotation.
      (ii) Gastroesophageal scintigraphy provides information about GER, aspiration, and gastric emptying.
      (iii) Esophageal pH monitoring measures the frequency and duration of acid esophageal reflux episodes.
      (iv) Multichannel intraluminal esophageal impedance helps detect acid and non-acid reflux by measuring retrograde flow of fluids, solids, and air in the esophagus.
      (v) Impedance measurement in conjunction with 24 h pH monitoring helps correlate between reflux episodes and apnea, cough, and other respiratory symptoms.
      (vi) Esophagogastroduodenoscopy with biopsy can be utilized for patients whose GERD is refractory to medical management. It can help diagnose H. pylori infection, peptic ulcer disease, strictures, and esophagitis.
5. Treatment:
   (a) The primary goal of treating GERD is reducing symptoms, improving quality of life, and preventing complications of GERD. In uncomplicated infantile GERD, conservative measures such as smaller and more frequent feeds,

thickened formula, and post-prandial prone positioning may be sufficient to diminish regurgitation. If these conservative measures fail, pharmacological therapy with gastric acid buffers, mucosal surface barriers or gastric anti-secretory agents can be instituted. PPIs are efficacious for symptomatic relief and mucosal healing, and are safe for long-term use. In older children and adolescents, dietary modifications, avoidance of alcohol, weight loss, positional changes, and cessation of smoking should be advocated.

(b) Operative therapy:

(i) Anti-reflux surgery is reserved for children with severe GERD refractory to medical management, patients who develop complications such as failure to thrive and respiratory symptoms, patients with neurological impairment, and occasionally children who require gastrostomy tube placement.

(ii) Fundoplication is the most common anti-reflux procedure and involves partial or complete wrapping of fundus of the stomach around the esophagus above the gastroesophageal junction. Fundoplication prevents reflux episodes by decreasing the number of the transient lower esophageal sphincter relaxations, increasing the lower esophageal sphincter pressure, accentuating the angle of His, and reducing the hiatal hernia.

(iii) This procedure can be performed open or laparoscopically. Laparoscopic Nissen fundoplication (360 ° wrap) is the preferred surgical option due to decreased morbidity, shortened hospital stay, and fewer perioperative complications. Major complications following fundoplication include breakdown of the wrap, intrathoracic herniation of the wrap, bowel obstruction, gas bloat syndrome, dysphagia, early satiety, dumping syndrome, and postoperative retching and gagging.

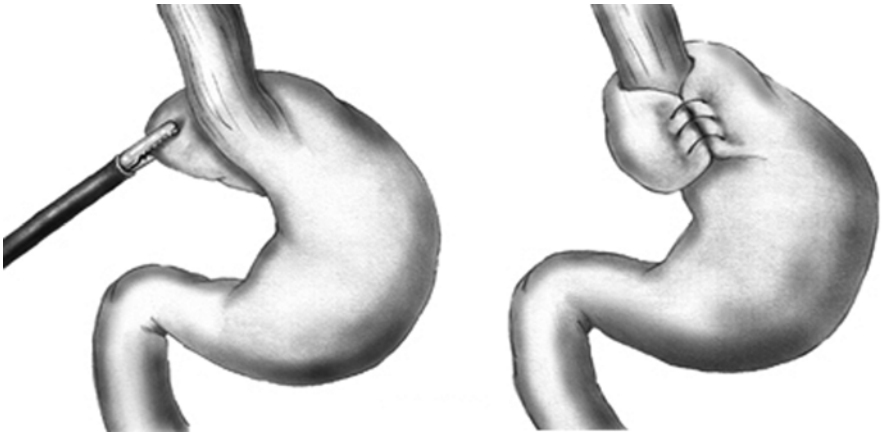

Fig. 1 Nissen Fundoplication. In the Nissen fundoplication procedure, the fundus of the stomach is used to create a 360 degree wrap around the lower esophagus (*Source*: Dr. James P. Gray, released to public domain, via https://upload.wikimedia.org/wikipedia/commons/d/dc/Nissen_fundoplication.png. Downloaded on 24 Dec 2013)

Gastrointestinal Bleeding

Amber Batool

Gastrointestinal (GI) bleeding in infants and children is a fairly common problem; however it tends to be minor and self-limited. Occasionally, it can be severe and life threatening.

1. Clinical presentation: The evaluation of any child with GI bleeding should begin with a thorough history and physical examination, assessment of hemodynamic parameters, diagnosis, and treatment of the source. It is important to differentiate between an upper GI bleed vs. lower GI bleed. Upper GI bleeding usually arises proximal to the Ligament of Treitz and presents as hematemesis (vomiting of coffee-ground like material or blood) or melena (stools that are jet black and tarry). In comparison, lower GI bleeding occurs anywhere from Ligament of Treitz to the anus and usually presents as hematochezia (bright red or maroon blood in stool).
2. The etiology of lower gastrointestinal bleeding varies by age and geographic location. The following discussion will focus on the most common causes of the lower GI bleeding in the United States.
3. Specific etiologies of GI bleeding:

   (a) Anal Fissures:

      (i) Pathophysiology: Anal fissures are the most common cause of rectal bleeding in neonates, infants and can be seen in up to approximately 30 % of older children with retentive constipation. Anal fissures are slit-like tears of the anal canal, usually located in the posterior midline and are caused by painful passage of hard stool or prolonged diarrhea. Presence of multiple anal fissures or aberrant location should prompt

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evaluation for Crohn's disease, perianal dermatitis, crypt abscess, or traumatic injury (e.g., sexual abuse).

(ii) Clinical presentation: The most common symptom is burning or tearing pain upon defecation and it may be accompanied with bright red blood seen on the surface of the stool.

(iii) Diagnosis: Diagnosis can be made by careful history and physical examination. Examination of anus with gentle spreading of the buttocks would reveal disruption of anoderm in the midline, distal to dentate line. In case of a chronic fissure, a sentinel skin tag or hypertrophied papillae may be seen at the anal verge. Digital rectal examination will reveal an internal sphincter spasm. Occasionally, proctoscopic examination is necessary to identify fissures in the anal canal.

(iv) Treatment: Most fissures heal rapidly with conservative management. Initial management should involve treatment of constipation with dietary modifications and stool softeners. Acute pain can be relieved with warm sitz baths and lubrication of the anal skin. Anal dilation (digital or by dilator) can sometimes be utilized to relieve anal pain and spasms and helps promote healing. On rare occasions, silver nitrate cauterization or perianal injection of botulinum toxin A can be employed. Lateral internal anal sphincterotomy is the procedure of choice if conservative management has failed. This procedure can be performed in an open or closed fashion and usually involves division of internal anal sphincter muscle from distal end to the dentate line or to the level of the apex of the fissure. It helps provide symptomatic relief and expedites the fissure healing process. Incontinence is a risk of sphincterotomy so conservative measures should be tried first.

(v) Note: It is a priority of treatment to avoid anal fissure surgery in patients with Crohn's disease due to high risk of recurrence, poor healing, and progression after surgery.

(b) Perianal abscess:

(i) Pathophysiology: Perianal abscess usually results from infection of the anal glands (cryptoglandular infection) found in the intersphincteric plane due to obstruction of anal crypts. The severity and depth of the abscess tends to be variable and may be associated with fistulous tract formation.

(ii) Most common organisms implicated in abscess formation include E. coli and Enterococcus species. Recently, there has been an increase in Staphylococcus and methicillin resistant staphylococcus aureus (MRSA) being causative organisms.

(iii) Clinical presentation: Patients with perianal abscess usually present with severe anal pain, anal pruritus or burning, and scant rectal bleeding. Pain is usually exacerbated by movement, coughing, or straining. Patients may also experience systemic fevers, chills, or urinary retention.

(iv) Diagnosis: Diagnosis is made by careful rectal examination that usually reveals a tender and erythematous palpable mass accompanied with occasional purulent drainage. Digital rectal examination tends to be painful because of sphincter spasms. Optimal physical examination may require an exam under anesthesia. It is important to evaluate for any fistulous tracts that connect the abscess to the anal gland. A sigmoidoscopic examination would reveal deep, red, edematous anal columns/crypts of Morgagni. Imaging studies are rarely required for diagnosis of perianal abscess.

(v) Treatment: Immediate incision and drainage. Primary antibiotic therapy is insufficient at resolving the underlying problem. Delayed or inadequate treatment may result in extension of infection and septicemia.

1. Drainage of perianal abscess can be accomplished in office or emergency department under local anesthesia. A cruciate or elliptical skin incision is usually made over the most prominent part of the abscess and wide drainage is ensured. Blunt dissection is utilized to break all the loculations. It is important to excise dog ears to prevent premature closure. Usually, iodoform gauze is utilized to pack the wound; however, packing is not necessary. Gauze is removed after 24 h and sitz baths are started. Postoperative antibiotics are used as adjuncts based on the results of the wound culture.
2. In case of recurrent abscesses or chronic drainage, it is imperative to identify the fistula and surgically excise the fistula and the crypt of origin. Perianal abscess or fistula may be the presenting signs of Crohn's disease in older children.

(c) Juvenile polyps:

(i) Pathophysiology: Juvenile polyps are usually benign, pedunculated, rounded masses that are composed of hamartomatous overgrowth of tissue that is highly vascular. These are the most common gastrointestinal tumors that frequently occur in school-aged children.

(ii) Clinical presentation: Polyps usually present as intermittent, painless bleeding with normal or constipated stool. Polyps may also prolapse during defecation and can be felt as a small, mobile mass on a stalk. These may also present with colic-like abdominal pain, diarrhea, or unexplained iron deficiency anemia.

(iii) Diagnosis: Diagnosis is typically made via sigmoidoscopy or colonoscopy. Most polyps are solitary and are usually localized in rectosigmoid region. Most children have only one or two juvenile polyps. Presence of multiple polyps should raise suspicion for polyposis syndrome and warrant further testing.

(iv) Treatment: All polyps should be removed. Most polyps can be usually excised by snare and cautery during colonoscopic evaluation if the

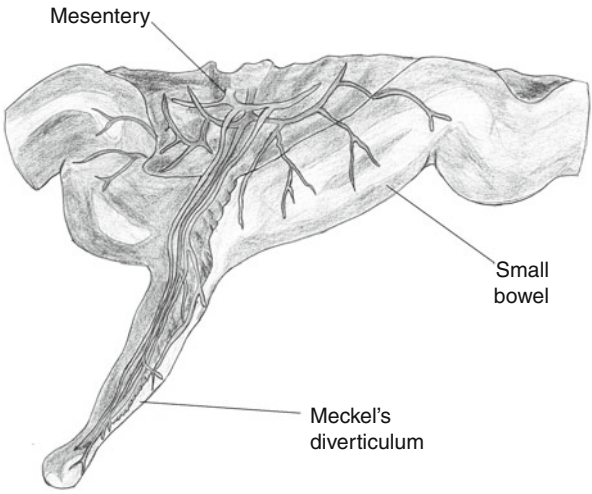

Fig. 1 Meckel's Diverticulum. The Meckel's diverticulum is an outpouching on the antimesenteric side of the small bowel. It may contain ectopic tissue and be a source of gastrointestinal bleeding (Adapted from: Raziel, under Creative Commons Share-Alike 1.0 license, via https://commons.wikimedia.org/wiki/File:Diverticule_de_Meckel.jpg. Downloaded 24 Dec 2013)

polyp size is small. Endoscopic polypectomy of a large polyp is contraindicated due to an increased risk of perforation. Polyps may also autoamputate due to torsion and pass with stool.

(d) Meckel's diverticulum:

(i) Pathophysiology: Meckel's diverticulum is the most common congenital malformation and it is a remnant of the omphalomesenteric duct located on the antimesenteric border of the distal ileum. It usually follows the rule of twos: present in 2 % of the population, most commonly located within 2 ft from ileocecal valve; usually 2 in. long, mostly presents before age 2 years; two types of ectopic tissue are found within the diverticulum (pancreatic and gastric tissue); and 2 % develop symptoms.

(ii) Clinical presentation: The most common presenting symptom is painless rectal bleeding. Bleeding is due to deep ulceration of adjacent mucosa caused by acid secretion from ectopic gastric tissue. Meckel's diverticulum may also present as intestinal volvulus around the fibrous band attaching the diverticulum to the umbilicus, intestinal obstruction due to ileocolonic intussusception or Meckel's diverticulitis, which usually presents with abdominal pain, fever, and leukocytosis.

(iii) Diagnosis: Diagnosis of Meckel's diverticulum is usually made with a Meckel's scan, where Technetium-99 (^{99m}Tc)-pertechnetate is taken up by the gastric mucosa in the diverticulum. Giving pentagastrin or H2 blocker before administering the radionuclide can increase the sensitivity of the test by increasing the uptake and retention of ^{99m}Tc-pertechnetate by the heterotopic gastric mucosa. Ultrasonography or computed tomography of the abdomen and pelvis may be helpful and usually demonstrate an edematous, inflamed, blind ending segment of bowel in the right lower quadrant of the abdomen.

(iv) Treatment: A symptomatic Meckel's diverticulum requires surgical resection. In case of an asymptomatic Meckel's diverticulum incidentally discovered during laparotomy in children, surgical resection should be pursued; however, resection of asymptomatic lesion in adults remains a controversial subject.

1. Options for surgical resection include an open or laparoscopic diverticulectomy, wedge resection, or partial small bowel resection with primary anastomosis.
2. A diverticulectomy can be performed if the diverticulum is narrow-based or if gastric tissue is localized at the tip of the diverticulum. One must ensure that resecting the diverticulum at its base will not compromise the ileal lumen. In presence of diverticulitis, the zone of resection should be free of any inflammation.
3. A simple diverticulectomy is contraindicated if there is evidence of diverticulitis, presence of ectopic gastric tissues at the diverticular-intestinal junction, or if there is associated perforation, ischemia, or ulceration in the adjacent tissue. In these circumstances, a segmental ileal resection with primary end-to-end anastomosis is preferred.

Vascular Rings and Slings

Laura A. Masters and Anastasios C. Polimenakos

Vascular rings and slings are abnormal formations of the great vessels which completely or partially encircle the trachea and/or esophagus is a vascular ring. Vascular rings are a rare occurrence and account for 1–2 % of all congenital heart defects.

1. Embryology: Early in fetal life the vascular system is structured with six pairs of aortic arches between the dorsal aorta and ventral roots. During normal embryologic development fetal left fourth aortic arch will become the aortic arch and the right fourth arch the innominate artery. The fetal sixth arches become the pulmonary arteries. The patent ductus arteriosus (PDA) originates from the fetal left sixth aortic arch. The left dorsal aorta becomes the descending aorta. From the two pairs of aortic arch formations; the right (posterior) and the left (anterior) encircle the developing esophagus and trachea. Usually, the right regresses and the left (anterior) will form the aorta and its branches at birth. If the involution of the fetal right arch structure fails, an abnormal vascular formation can be developed with various levels and degree of interference to the normal growth of the trachea and/or esophagus.
2. Anatomy, pathology, and associated anomalies:
   (a) Complete vascular rings: form a complete encirclement of the trachea and/or esophagus.

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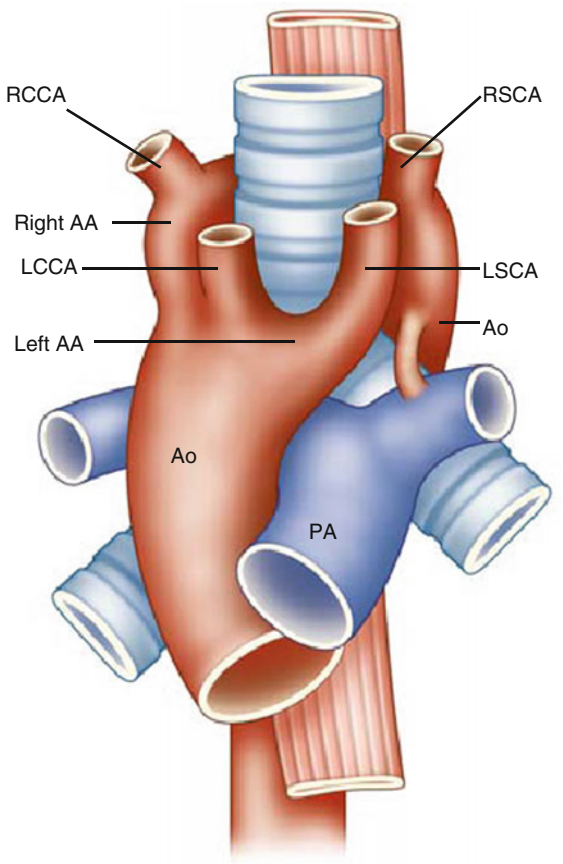

Fig. 1 Double aortic arch. Esophagus and trachea are encircled by the dominant right (*Right AA*) and the hypoplastic left (*Left AA*) aortic arches. The left subclavian (*LSCA*) and common carotid artery (*LCCA*) originate from the left aortic arch. The right common carotid (*RCCA*) and subclavian (*RSCA*) arteries originate from the dominant right aortic arch. *Ao* aorta, *PA* pulmonary artery, *AA* ascending aorta (*Source*: Dr. Anastasios Polimenakos)

(i) Double aortic arch:

1. Among the most common forms of vascular rings.
2. Occurs when the left or right fourth fetal aortic arch fails to regress during fetal development.
3. In 75 % the right aortic arch is the dominant, in 20 % the left and in 5 % both.
4. Usually an isolated cardiac anomaly rarely (<1 %) associated with other congenital cardiac defects such as Tetralogy of Fallot (TOF) or atrioventricular septal defects.
5. The Kommerell's diverticulum represents an important component of a vascular ring (associated with the descending thoracic aorta and the ligamentum arteriosum) that need to be resected during the repair to prevent recurrence of compressing symptoms to the adjacent structures.
6. Associated extracardiac malformations such as cerebral, oropharyngeal or genitourinary are rarely present but need to be considered during the diagnostic work-up.

(ii) Right aortic arch with aberrant left retroesophageal subclavian artery and left-sided ligamentum arteriosum.

1. Second most common complete vascular ring.
2. Almost always presents as an isolated cardiac anomaly.

(iii) Right aortic arch with mirror-image branching, left-sided ligamentum arteriosum (coming off the descending thoracic aorta) and Kommerell's diverticulum. In 10–15 % is associated with other congenital cardiac defects such as TOF (in 25 % of TOF), truncus arteriosus (in 30 % of truncus arteriosus), or atrioventricular septal defects.

(iv) Left aortic arch with mirror-image branching and right descending thoracic aorta (rare).

(v) Left aortic arch with right descending thoracic aorta and right-sided ligamentum arteriosum (rare).

(b) Incomplete vascular rings: form an incomplete encirclement of the trachea and/or esophagus with/or without compression. These include:

(i) Anomalous origin of the innominate artery. Anterior compression of the trachea can be developed if the takeoff from the aorta is more posterior or to the left.

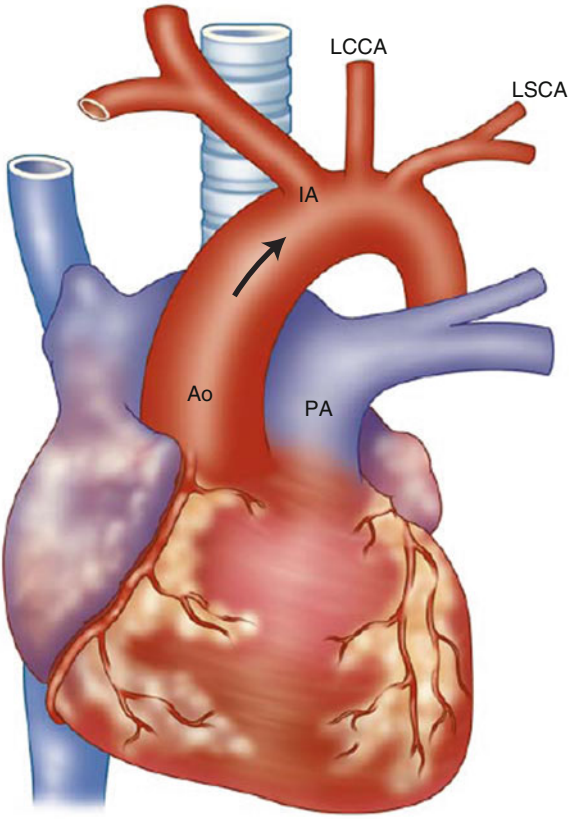

Fig. 2 Anomalous origin of the innominate artery. The *arrow* shows the abnormal posterior and lateral origin of the innominate artery (*IA*) with anterior compression of the trachea. (*Ao* aorta, *LCCA* left common carotid artery, *LSCA* left subclavian artery, *PA* pulmonary artery) (*Source*: Dr. Anastasios Polimenakos)

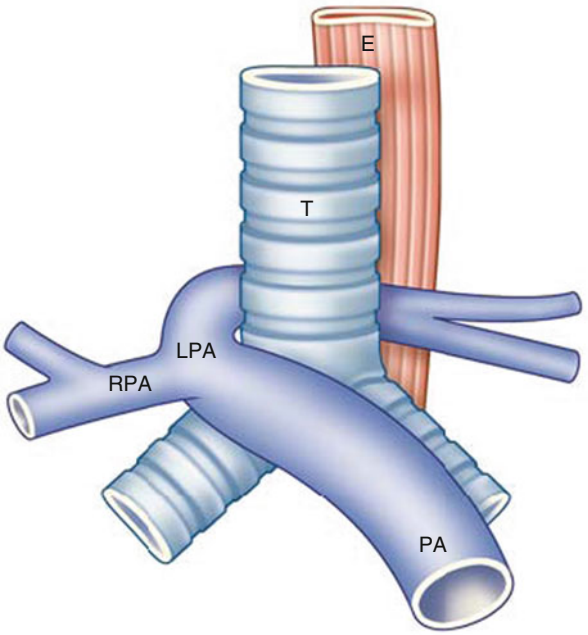

Fig. 3 Pulmonary artery sling. The left pulmonary artery (*LPA*) originates from the right pulmonary artery (*RPA*) and travels between the trachea (*T*) and the esophagus (*E*) (PA pulmonary artery) (*Source*: Dr. Anastasios Polimenakos)

(ii) Aberrant right retroesophageal subclavian artery.

1. Occurs in 0.5 % of general population but it is often underdiagnosed due to absence of symptoms.
2. Commonly encountered as incidental finding.
3. Often associated with coarctation of the aorta.

(iii) Right aortic arch with mirror-image branching and left ligamentum arteriosum (coming off the innominate artery instead of the descending thoracic aorta).

(iv) Pulmonary artery sling.

1. Left arises from the right pulmonary artery instead of the main pulmonary artery; courses behind the trachea and in front of the esophagus which causes the compression.
2. Often associated with tracheal stenosis (up to 50 %, "ring-sling complex") with formation of complete cartilaginous rings; tracheomalacia is commonly present.

3. Clinical presentation:

(a) General Manifestations:

(i) Vascular rings may remain asymptomatic for life.

(ii) 75 % demonstrate early symptoms within the first month of life.

(iii) Signs and symptoms are often related to extrinsic compression on the trachea and/or esophagus. When trachea is compressed, symptoms may be present early in life (80–90 %); stridor or noisy breathing (that worsens with feeding), recurrent respiratory infections, respiratory distress or wheezing are, often, the clinical manifestations. If esophageal compression is present (70–80 % in older children) choking episodes, dysphagia, or gastroesophageal reflux are the dominant symptoms.
(iv) Physical exam can be often inconclusive; most patients have normal growth; rhonchi or wheezing are frequently present; severe upper respiratory distress with typical posture (breathing pattern with hyperextended neck) is rather uncommon finding.

(b) Specific Manifestations (of most common vascular rings):

(i) Double aortic arch is usually asymptomatic but it, often, causes minor or major obstructive symptoms related to tracheal (mainly recurrent respiratory tract infections or stridor) and/or esophageal (various degrees of difficulty of swallowing) compression. If present, symptoms appear during infancy or early childhood.
(ii) Anomalous origin of innominate artery causes symptoms due to anterior compression of the trachea. The symptoms are rather infrequent (30 %) and often present later during childhood.
(iii) Aberrant right subclavian artery is rarely symptomatic and when compression on the posterior esophageal wall is significant manifests as "dysphagia lusoria" (older patients).
(iv) Pulmonary artery sling is often symptomatic due to anterior tracheal compression (respiratory distress, stridor); Combined esophageal and tracheal clinical manifestation are less frequent.

4. Evaluation and diagnostic studies:

(a) Chest x-ray:

(i) May show pulmonary infiltrates, atelectasis or hyperinflation (air trapping) of the lungs.
(ii) Lateral tracheal indentation may be visible.

(b) Bronchoscopy:

(i) Shows external compression of the trachea.
(ii) Helpful in delineating the degree of severity and extent of stenosis.
(iii) Documents complete cartilaginous tracheal rings in pulmonary artery sling.
(iv) Valuable perioperatively in all patients with vascular rings.

(c) Barium swallow:

(i) Indentations of the esophagus might guide diagnostic steps.

(ii) The locations of the indentations can be used to help determine the type of vascular ring:

1. Bilateral indentations indicate double aortic arch.
2. Posterior indentation might indicate an aberrant retroesophageal subclavian artery or double aortic arch.
3. An anterior indentation is usually associated with pulmonary artery sling.

(iii) The precision of advanced imaging (such as CT or MRI), readily available to pediatric cardiac centers, led to diminished importance of barium swallow as a preoperative diagnostic tool.

(d) Echocardiogram:

(i) Complements CT/MRI in defining the diagnosis of the vascular ring anatomy.
(ii) Supports detection of associated intracardiac congenital lesions.

(e) MRI/CT scan:

(i) The preferred diagnostic methods (near 100 % accuracy).
(ii) Provide precise delineation of vascular ring anatomy.
(iii) Serve as preoperative "road-map" and to outline topography of surrounding structures (especially three dimensional imaging).

(f) Cardiac catheterization: Invasive study that is rarely used.

5. Management strategy:

(a) When feeding difficulties are present, aspiration precautions should be taken.
(b) Recurrent respiratory infections occur frequently in patients with complete vascular rings and should be treated aggressively.
(c) Surgical intervention is recommended:

(i) When complete vascular rings are diagnosed:

1. During early infancy (highly likelihood to become symptomatic during early childhood) regardless if symptoms are present.
2. As early as symptoms are detected (even mild).

(ii) When diagnosis of pulmonary artery sling is established regardless if symptoms are present.
(iii) For other incomplete vascular rings intractable symptoms should be treated with surgical correction; otherwise mild symptomatology early during infancy can be managed conservatively considering that maturation and growth of trachea and esophagus can withstand deterioration of the clinical picture in early childhood.

6. Medical treatment and therapeutic interventions:
   (a) Watchful waiting: Patients with vascular rings do not always require surgical repair. For those who surgery is not recommended should be selectively followed (as described above) for monitoring of early onset of symptoms.
   (b) Surgical repair strategy (most common vascular rings):
      (i) Double aortic arch: Double aortic arch repairs are performed through a left posterolateral thoracotomy incision. Careful dissection is performed to reveal the two aortic arches. The smaller of the two arches is divided and the largest is preserved. The ligamentum arteriosum is ligated.
      (ii) Right aortic arch with aberrant retroesophageal left subclavian artery and left ligamentum arteriosum: Repair is performed through a left posterolateral thoracotomy incision. The ligamentum arteriosum is identified and ligated. If a Kommerell's diverticulum is present resection is recommended to prevent future recurrent compression of surrounding structures or regional aneurysmal formation. If resection of the diverticulum is performed the left subclavian artery origin might need to be divided and re-implanted to the left carotid artery or aorta.
      (iii) Anomalous origin of innominate artery: Repair is performed through a right anterolateral thoracotomy or median sternotomy incision. The innominate artery is identified but not dissected away from the trachea. The artery is then suspended and sutured to the posterior sternum (innominopexy) to relieve anterior tracheal wall compression. Division of proximal end and reimplantation of innominate artery to the proximal ascending aorta may be considered in selective cases.
      (iv) Aberrant retroesophageal right subclavian artery: Repair is performed through a left thoracotomy incision. The right subclavian artery origin is to be divided and re-implanted to the common carotid or ascending aorta to relieve posterior esophageal wall compression.
      (v) Pulmonary artery sling: Repair is performed through a median sternotomy incision. The repair requires cardiopulmonary bypass with cannulation of the right atrium and aorta. For patients without need for tracheal repair, the origin of the left pulmonary artery is divided off the right pulmonary artery and re-implanted anteriorly to the trachea on the left lateral wall of the main pulmonary artery. When tracheal stenosis is present (up to 50 % of the pulmonary artery slings), tracheal repair will be necessary. If the stenotic tracheal component is short (less than 30 % of tracheal length, up to seven rings) the narrowed part is entirely resected with end-to-end anastomosis repair. If the stenotic tracheal component is long (greater than 30 % of tracheal length, eight rings or more), then an extensive tracheal reconstruction is necessary (slide tracheoplasty or tracheal autograft repair). After tracheal repair, the divided origin of the left pulmonary artery is, then, re-implanted anteriorly to the trachea on the left lateral wall of the main pulmonary artery.

(c) Surgical technique:

(i) An open (more frequently) or a minimally invasive approach technique can be utilized as indicated.

(ii) In open approach, when a thoracotomy approach is required, a small left or right thoracotomy via third or fourth intercostal space is performed. The serratus muscle is usually spared. The lung parenchymal is gently retracted and high frequency/low tidal volume ventilation is introduced to optimize exposure. The parietal pleura is entered. Care must be practiced to identify and spare the left recurrent laryngeal nerve around ligamentum arteriosum. The vascular ring is repaired as described above. Ligation and division of ligamentum arteriosum accompanies the repair in complete vascular rings. It is critical to dissect all paraesophageal and paratracheal fibrous bands.

(iii) In minimally invasive approach, three small (3 mm) ports are inserted via the fourth or fifth intercostal space between anterior and posterior axillary lines. The same principles followed in open approach should be honored for video-assisted thoracoscopy (VATS) surgical repair. VATS can be particularly successful in patients with complete vascular rings (most commonly right aortic arch with aberrant left subclavian and left ligamentum arteriosum). The use of VATS approach in infants smaller than 1 kg might not be feasible. This approach is associated with superior cosmetic results and minimal discomfort.

(iv) Complications are infrequent following vascular ring repair. Respiratory infections or failure, injury of the recurrent laryngeal nerve and chylothorax are the most noteworthy. Mortality rate is extremely low unless extensive tracheal reconstruction is required. Depending on the severity of tracheal compression, resolution of breathing issues may take months. Esophageal related symptoms are usually relieved within few days to several weeks.

Suggested Further Reading

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Part V
Common Problems in School Age Children

Lymphadenopathy

Alysia A. Agnoni

Children are frequently referred to a pediatric surgeon for evaluation of enlarged lymph nodes. In the majority of cases, this is a natural response to infection and no operation is needed.

1. Pathophysiology: Lymph nodes, along with the spleen, tonsils, adenoids, and Peyer's patches, are composed of immune cells. Within the lymph node these immune cells, lymphocytes, work to filter antigens from extravascular fluid that travels through the lymphatic system. Lymph nodes enlarge under the following circumstances: response to infective organisms (most common cause of enlargement and usually benign and self-limiting), inflammation due to infection within the node, staphylococcus, metastatic infiltration of neoplastic cells, localized neoplastic proliferation, or infiltration of macrophages (storage disorders).
   (a) Anatomy:
      (i) Approximately 600 lymph nodes in body.
      (ii) Distribution of commonly found enlarged nodes:
         1. Head/Neck: Cervical, occipital, submental, submandibular, pre- and postauricular, clavicular.
         2. Upper extremities: Axillary, epitrochlear.
         3. Chest/Abdomen: Mediastinal, mesenteric, inguinal.
         4. Lower extremities: Femoral, popliteal.
   (b) Epidemiology: Although lymphadenopathy may present at any age, it is most common among the pediatric population. Most are benign reactive lymph nodes.
2. Clinical features:

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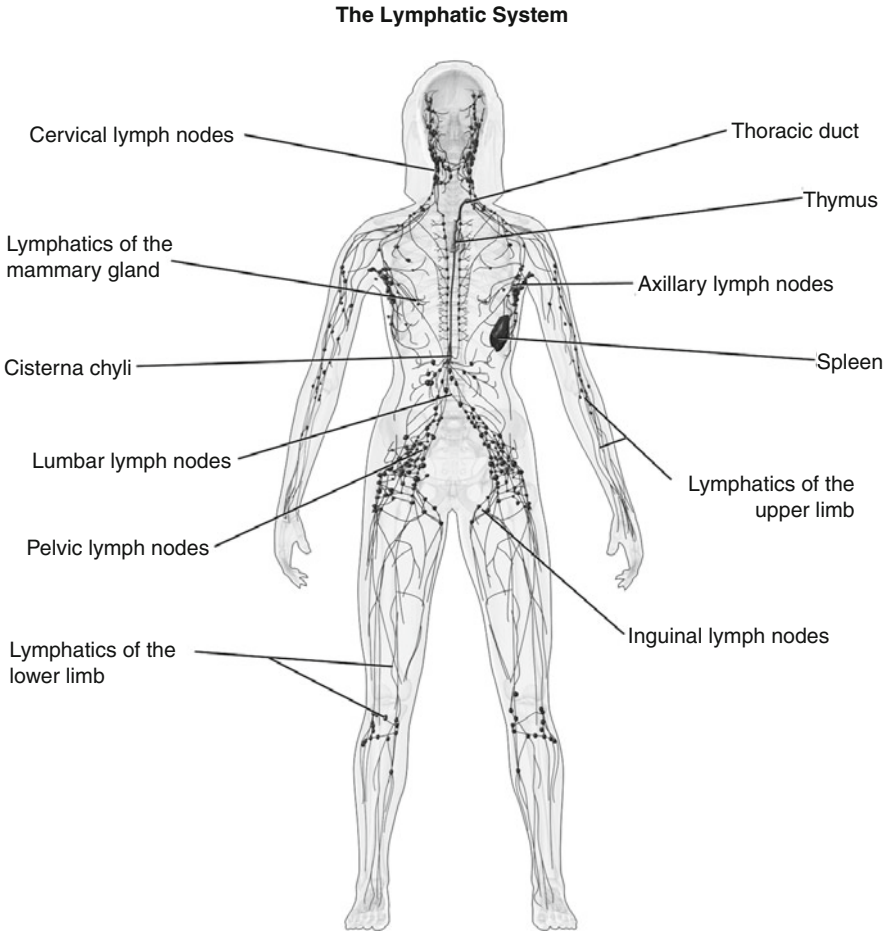

Fig. 1 The lymphatic system (*Source*: Bruce Blausen, Blausen Medical Communications, via Creative Commons Attributions Unported 3.0 license. https://upload.wikimedia.org/wikipedia/commons/f/f4/Blausen_0623_LymphaticSystem_Female.png. Downloaded 17 Dec 2013)

(a) History:

(i) Recent illness, infections, bites, trauma, exposure to tuberculosis (TB) or new animals.
(ii) Constitutional symptoms: fever, unintentional weight loss, night sweats, fatigue.

(b) Physical examination: Full body examination for local or generalized enlargement of nodes.

(i) Normal small nodes are palpable in most children.
(ii) Examination includes palpation of spleen to detect splenomegaly and abdomen to detect enlarged abdominal lymph nodes.

(iii) Enlarged nodes may be tender or painless, mobile or fixed, discrete, matted or, shoddy, and firm or rubbery.
(iv) Examination of the skin surrounding the enlarged nodes may show signs of infection or trauma.

3. Diagnosis:

(a) In most cases, diagnosis of reactive lymphadenopathy is established through a complete history and physical examination.

(b) Further work-up is needed only if there is suspicion for a cause that requires further treatment (malignancy, TB, suppurative).

(i) Labs:

1. CBC with differential if suspicious of leukemia, lymphoma, Epstein-Barr virus (EBV), cytomegalovirus (CMV), or toxoplasmosis.
2. Lactate dehydrogenase (LDH) if suspicious of leukemia or lymphoma.
3. TB skin test.
4. Monospot heterophile antibody test for EBV.
5. EBV and CMV, cat scratch disease, or toxoplasmosis titers.

(ii) Imaging:

1. Chest x-ray if suspicious of TB or hilar adenopathy.
2. Sonography: When there is a question of whether a lump is an enlarged lymph node, or if it represents an alternate diagnosis, such as dermoid cyst, abscess, or ectopic testis.
3. CT: Can detect deep lymph nodes and may detect a source of malignancy if this is suspected.
4. Positron emission tomography (PET): Used if malignancy is suspected.

4. Treatment:

(a) Incision and drainage of suppurative node: Antibiotics may be used to prevent need for drainage. The node may be aspirated and cultured or incised, cultured, drained, and excised.
(b) Excisional biopsy: May be completed with or without labs or imaging. This is diagnostic and done as an outpatient procedure. If many nodes are enlarged, the largest is typically chosen for biopsy.

(i) The patient is taken to the operating room and put under a general anesthetic. The surgical field is prepped in the usual sterile fashion. An incision is made to the skin overlying the node in question. Blunt dissection with hemostats or electrocautery is often used to separate the node from surrounding structures. Care to avoid vital structures such as nerves and vessels, especially in the neck, are made during the procedure. Interrupted absorbable sutures are used to close the subcutaneous tissue

and the skin is closed with absorbable suture in a running subcuticular fashion.

(c) Pathology and further treatment:

(i) Reactive lymphadenopathy: Inflammatory node due to exposure to viral or bacterial organisms.

1. Viral: Most often in the presence of recent or recurrent upper respiratory infections. Can also be due to EBV or CMV. Treatment is supportive.
2. Bacterial: Most commonly associated with staphylococcus aureus and group B streptococcus. History of recent upper respiratory infection, pharyngitis, tonsillitis, or otitis media. The patient may need antibiotic treatment.
3. Atypical mycobacterium: Treatment is excision of affected lymph nodes.
4. Mycobacterium tuberculosis: Should have a chest x-ray and tuberculin skin test. Treatment is rifampin and isoniazid per CDC recommendations.
5. Cat scratch disease: Infection with Bartonella henselae, a rickettsial organism. Results from animal scratches, most often a cat or kitten. This is a self-limiting infection and antibiotic treatment is no longer indicated.

(ii) Malignancies: Lymphoma (Hodgkin's and Non-Hodgkin's), leukemia, or metastatic tumors need extensive work-up and involvement of a pediatric oncologist.

(iii) Other rare causes: Kawasaki's disease, sarcoidosis, Langerhans cell histiocytosis, systemic lupus erythematosus.

(d) Fine needle aspiration: Predominantly used in adults, much less frequently in children.

5. Outcomes:

(a) No further treatment if node is reactive.
(b) Nodes may take weeks to months to decrease in size and may never shrink back to former size.

Appendicitis

Christopher P. Coppola

Appendicitis has a peak incidence of 11-years-old and is one of the most common reasons an otherwise healthy child would require an operation. Most children with appendicitis recover quickly after appendectomy, but the condition has a wide spectrum of severity and in rare cases can cause death.

1. Epidemiology:

   (a) There are approximately 600,000 pediatric cases of appendicitis in the United States each year, of which 20,000 are perforated. There are approximately 100 deaths each year.
   (b) Appendicitis can affect a wide range of ages from toddlers to the elderly. There is a bimodal distribution of incidence with peaks at 11-years-old and in the early twenties. The very young, the very old, and patients with neurologic disability can experience more severe cases of appendicitis because the early symptoms often go unrecognized.
   (c) Appendicitis has a seasonal variation thought to be related to incidence of viral illness.

2. Pathophysiology:

   (a) The appendix is a blind ending tubular structure arising from the dependent portion of the cecum which can be intraperitoneal or retroperitoneal, behind the cecum. The function of the appendix is unclear, but it may serve as a reservoir for non-irritating gut flora from which the colon can be repopulated after an episode of infectious enteritis or colitis.
   (b) When the mouth of the appendix becomes occluded, pressure increases until vascular flow is impeded, which leads to suppuration, gangrene, and perforation. The appendix can be occluded by inflammation and swelling

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of lymphatic tissue at the mouth of the appendix, or by a hard plug of stool called an appendicolith or fecalith. Other rare causes of appendiceal obstruction are parasitic worms or swallowed foreign objects.

3. Clinical features:

   (a) Appendicitis is a very common disease that can present in a multitude of uncommon ways. The most reproducible feature is a steady, unrelenting, progression in severity over a course of 1–3 days. This process starts as vague malaise, becomes a focal right lower quadrant pain, and develops into diffuse abdominal peritonitis. Along this time course, the patient will develop anorexia. In the later stages, there may be nausea, vomiting, and diarrhea. When the presenting symptoms are vomiting and diarrhea, if the pain intermittent, the diagnosis is more likely to be infectious gastroenteritis than appendicitis.
   (b) The location of pain informs the progress of the illness. Appendiceal distention is felt as periumbilical pain. Transmural inflammation causes irritation of the parietal peritoneum and becomes right lower quadrant pain. After perforation, diffuse peritoneal contamination is manifest as diffuse peritonitis and abdominal pain.
   (c) Several eponymous clinical signs can be elicited in the physical examination of a patient with appendicitis.

      (i) McBurney's sign: pain and tenderness at a point 2/3 the distance from umbilicus to the right anterior superior iliac spine.
      (ii) Rovsing's sign: palpation of the left lower quadrant causes pain in the right lower quadrant.
      (iii) Blumburg's sign: rebound tenderness resulting when a hand compressing the abdomen is abruptly removed.
      (iv) Psoas sign: extension of the right hip, moving the thigh posteriorly results in right lower quadrant pain as the extending psoas muscle irritates the appendix.
      (v) Obturator sign: With right knee bent, the lower leg is rotated laterally, as the obturator internus is stretched; it irritates the appendix and causes pain.

4. Diagnosis:

   (a) A careful history and physical by an experienced surgeon is the most accurate diagnostic maneuver, and an observant parent can usually provide an account of the recognizable progression of symptoms in appendicitis.
   (b) Vital signs usually are marked by fever, and there may be evidence of dehydration, sepsis, or shock.
   (c) Blood is drawn for blood count, electrolytes, and when indicated liver function tests and pancreatic enzymes. Blood count commonly shows a left shift in differential and leukocytosis.

(d) Urinalysis is checked for blood from renal lithiasis or evidence of urinary tract infection, possible alternate diagnoses.
(e) Sonography of the right lower quadrant can diagnose appendicitis when a distended/edematous appendix greater than 6 mm in diameter is seen, as well as the bull's-eye target-like appearance of the appendix, and increased periappendiceal vascularity. Accurate sonography can be hampered by obesity, overlying bowel gas, variable location of appendix, and operator inexperience with the examination.
(f) Magnetic resonance imaging of the abdomen can diagnosis appendicitis.
(g) When the diagnosis cannot be made by other means and a period of hospital admission with serial abdominal exam is not an acceptable option, computed tomography can be considered. However, the ionizing radiation required for computed tomography carries the risk of a future neoplasm. When employed, computed tomography should be performed with intravenous and oral contrast to give the best chance of diagnosing the possibility of appendicitis.

5. Treatment:

(a) Initial resuscitation:

(i) Bolus 20 mL/kg normal saline.
(ii) Pain medication should not be withheld. Morphine, 0.05 mg/kg can be given to provide temporary relief. Signs of appendicitis will return if it is present. If a patient has pain severe enough to require narcotic medication, they should not be discharged home until the cause of pain is determined.
(iii) If a patient demonstrates shock, severe dehydration, or electrolyte abnormalities, these should be corrected before undertaking an operation.

(b) Observation: if the diagnosis of appendicitis is uncertain, an excellent option is to admit the patient for serial abdominal examinations. If this course is chosen, the patient can receive pain medication, which can be intermittently held to repeat the abdominal medication. The patient should not receive antibiotics as they can mask the symptoms of appendicitis.
(c) Appendectomy: before operation, child should be hemodynamically stable and is given a broad spectrum antibiotic which covers gut flora. (Choices include cefoxitin, piperacillin/tazobactam, ampicillin/sulbactam, and ampicillin/gentamicin/metronidazole. For patients allergic to these choices, clindamycin is an alternative). Appendectomy can be safely performed by open approach, laparoscopic approach, or single incision laparoscopy. In any approach, care is taken to completely excise the appendix, securely close the appendiceal orifice on the cecum, avoid narrowing of the terminal ileum, securely obtain hemostasis of the mesoappendix, break up any phlegmon in the right lower quadrant or pelvis, and break up and irrigate any interloop abscesses between lengths of small intestine.

(d) Advantages of laparoscopy: The laparoscopic approach may bestow an advantage in obese patients, when the diagnosis is in doubt, in female patients past menarche where ovarian pathology is possible, when the appendix is perforated with spread of contamination beyond the right lower quadrant, and when cosmesis is of concern to the child's family.

(e) Encountering a normal appendix: In approximately 4 % of explorations for suspected appendicitis, a normal appendix will be encountered. An appendectomy should still be performed to avoid the need to repeat the procedure in the future, and because there will sometimes be microscopic inflammation of the appendix detected by the pathologist even if the gross appearance is normal. When a normal appendix is removed, the surgeon must search for an alternate diagnosis causing the pain, such as ovarian cyst, ovarian torsion, pelvic inflammatory disease, inflammatory bowel disease, Meckel's diverticulitis, mesenteric adenitis, intestinal parasites, and others.

(f) Postoperative antibiotics: If a nonperforated appendix is removed, no further antibiotics are needed. For perforated appendicitis, a postoperative course of antibiotics will reduce the risk of infection (intraabdominal abscess or suppurative wound infection) from approximately 15–5 %. Many antibiotic options are available, but the choice should cover gut flora, and lengths of therapy used run from 7 to 14 days. An acceptable choice is 12 days of antibiotics, piperacillin/tazobactam while an inpatient, with conversion to oral amoxicillin/clavulanate and metronidazole when well enough to be discharged from the hospital (Table 1).

(g) Presentation with abdominal abscess: In cases of advanced appendicitis with abscess formation, there is the option of treating the child with percutaneous drain placement in the abscess, guided by computed tomography or sonography. Patient should also receive antibiotics. Drain is removed when drainage decreases and abscess is resolved, usually 4–7 days. Antibiotics should be given for 2 weeks, longer if patient demonstrates continued signs of active infection. Placement of a percutaneously inserted central catheter (PICC) enables the child to receive therapy at home. The patient undergoes a scheduled interval appendectomy after an interval of 6 weeks.

Table 1 Antibiotic regiment for appendicitis

Preoperative:	One dose of cefoxitin or piperacillin/tazobactam
Postoperative:	
Non-perforated:	No further antibiotics
Gangrenous:	Two additional doses of piperacillin/tazobactam, or ertapenem if allergic
Perforated:	12-day total course of postoperative antibiotics: piperacillin/ tazobactam while an inpatient; amoxicillin/ clavulanate and metronidazole once discharged home
Percutaneously drained abscess:	14-day course of piperacillin/tazobactam; ertapenem if allergic. Drain is removed when output is <15 mL/24 h

6. Outcome:

   (a) Survival is > 99 %, deaths are rare, but possible, and usually occur in advanced cases with severe sepsis.
   (b) After appendectomy, most patients with non-perforated appendicitis can go home within 24 h, and within a week for cases of perforated appendicitis
   (c) Patients who are tolerating oral intake, afebrile, ambulating, and have pain controlled by oral analgesics are safe to discharge home.
   (d) Complications include: intraabdominal abscess, suppurative wound infection, ileus, appendiceal stump leak, Clostridium difficile colitis, adhesion, small bowel obstruction, and enterocutaneous fistula.

Hereditary Spherocytosis

Jeffrey S. Taylor

Hereditary spherocytosis (HS) is a common inherited hemolytic anemia caused by abnormalities in spectrin, or in other proteins involved in the structural integrity and function of spectrin, such as ankyrin and protein 4.2, among others.

1. Pathophysiology:

   (a) These protein defects lead to a weakened osmotically fragile cell membrane in the patient's red blood cells (RBCs). "Conditioning" of RBCs occurs in the spleen where damaged RBCs are trapped and decreased oxygen and limited glucose availability contributes to their demise.
   (b) The deficiency leads to a remodeling of the usual biconcave disk into cells which resemble spheres. These abnormal cells have a shortened life span which contributes to anemia.
   (c) The spherical shape of patient RBCs results from a reduction in the amount of membrane material available thus producing a reduced surface to volume ratio and increase in cell density. As more membrane is lost, the limit of the RBC membrane stretching is reached and lysis occurs
   (d) Prevalence and inheritance:

      (i) Occurs in all racial groups but is most common in northern Europeans.
      (ii) 75 % of cases are of dominant inheritance and 25 % are recessive with new mutations common.
      (iii) Many mutations may cause phenotypic HS, with mutations often unique to each family

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2. Clinical Presentation

   (a) Patients present with varying degrees of anemia, jaundice (may wax and wane), increased reticulocyte count, variable splenomegaly, and often evidence of gall stones.
   (b) The CBC with differential shows spherocytes and a positive osmotic fragility test in conjunction with a family history is confirmatory. Patients red blood cell (RBC) indices are also helpful with the combination of elevated red blood cell distribution width (RDW) and increased mean corpuscular hemoglobin concentration (MCHC) are strongly suggestive of HS.
   (c) HS can be classified as mild, moderate and severe based on baseline hemoglobin, degree of reticulocytosis, and resting bilirubin levels. These clinical classifications correlate with degree of osmotic fragility (severe has more fragile RBCs) and spectrin content (mild having near normal spectrin, severe with less than 50 % of normal value).
   (d) In mild cases of HS there is a compensated hemolytic anemia with normal or near hemoglobin and mildly increased reticulocyte counts. These patients are asymptomatic at steady state and often only come to light during an illness (such as infectious mononucleosis), pregnancy or physical stress (military training).
   (e) Severely affected patients have significant anemia and are transfusion dependent. Peripheral smears in these patients show spherocytes as well as bizarrely shaped cells (poikilocytes). Growth failure, and delayed sexual maturation are common and these patients are at risk for severe aplastic crises (later).
   (f) Moderately affected patients usually do well but may become symptomatic when stressed (i.e. illness, significant physical exertion).
   (g) Neonates with HS often have prolonged jaundice. Anemia during the first year of life may necessitate transfusions. There may be a role for erythropoietin in these infants. The osmotic fragility test, if done during the newborn period, should have newborn cells as controls.

3. Diagnosis:

   (a) Laboratory evaluation:

      (i) Hereditary spherocytosis is a hemolytic anemia and thus laboratory values are consistent with this and include: reticulocytosis, indirect hyperbilirubinemia, and variable anemia. The hemolysis is mainly extravascular and thus haptoglobin and lactate dehydrogenase (LDH) are poor indicators of hemolysis. The peripheral smear is diagnostic and shows spherocytes with RBC indices remarkable for an elevated RDW and MCHC. Other important causes of spherocytic anemias such as autoimmune hemolytic anemia can be distinguished by the negative coombs test in HS.
      (ii) Osmotic fragility (OF) testing: this test is performed by exposing control and suspected HS cells to increasingly hypotonic sodium chloride concentrations. Normal cells are able to swell and increase their volume

since they are starting as a normal biconcave disc. HS cells already have a decreased surface to volume ratio (spheres) and thus reach their limit at a higher concentration of sodium chloride compared to normal cells. The incubated OF test is the gold standard and consists of a period of incubation (24 h at 37 °C) prior to exposure to the saline concentrations which metabolically depletes the cells and making the test more sensitive.

(b) Differential diagnosis:

(i) Blood group ABO incompatibility and other immunohemolytic anemias: morphology of the RBCs is similar but HS lacks a positive coombs test. It is important to include a C3 Coombs test (as a test for a positive IgM antibody) in the evaluation of a suspected immune mediated hemolytic anemia if the IgG Coombs test is negative.
(ii) Heinz body anemias: spherocytes may be seen but are usually not the predominant cell. Rather, so called bite or blister cells are more common.
(iii) Undiagnosed patients presenting in aplastic crises- this may be confusing since the reticulocyte count will be low and the bilirubin level may also be falsely low.

4. Treatment:

(a) Routine care:

(i) Patients should be followed by a hematologist or a pediatrician familiar with this disease and its complications.
(ii) Baseline (several) hemoglobin levels, bilirubin levels and reticulocyte counts are helpful to document a significant change in the event of illness.
(iii) Counseling concerning the various crises must be discussed, especially aplastic crisis with parvovirus B19 (this particular variety of crises is not obvious and signs and symptoms must be understood by caretakers).
(iv) Counseling concerning indications, risks, and benefits of splenectomy must be discussed and understood in patients who are candidates for this procedure.
(v) Palpation of the spleen and avoidance of contact sports during periods of significant enlargement must be taught to families. The use of abdominal padding/protection is an option in patients who strongly desire to participate in contact sports.
(vi) Folic acid should be prescribed and reasons for its use understood.
(vii) Patients who have undergone splenectomy must be educated and reminded of the risks for sepsis and thrombosis. Penicillin prophylaxis is indicated immediately following splenectomy but the length of administration is under debate. Immunizations should be administered.
(viii) Signs and symptoms of gallstones and gallbladder disease should be discussed as well as indications for cholecystectomy.

(b) Splenectomy

(i) Splenectomy ameliorates the anemia, hyperbilirubinemia and reticulocytosis of most patients with HS. Spherocytes and a positive osmotic testing result are still present. The risk of post splenectomy sepsis must be considered and if splenectomy is to be done it should not occur till after age 5-years-old.

(ii) Postsplenectomy sepsis is a serious complication and may be fatal. Streptococcus pneumonia, Neisseria meningitidis and Haemophilus influenza type b are the major organisms and vaccination should be done before splenectomy occurs. It is unclear if the risk of overwhelming sepsis lessens over time.

(iii) Thrombosis and thromboembolism: the overall risk after splenectomy is between 1.5 and 55 %. Portal vein thrombosis incidence after splenectomy is believed to be between 6.3 and 10 % while that of stroke or ischemic heart disease is increased in HS patients who underwent splenectomy as compared to HS patients who did not.

(iv) The cause of this is not well understood but may relate to thrombocytosis which occurs after splenectomy, the presence of an increased number of abnormal RBCs in circulation triggering platelet aggregation, and release of free hemoglobin into the vasculature leading to decreased nitric oxide.

(v) Further studies are needed to understand this significant post splenectomy complication.

(vi) Indications for splenectomy:

1. Indications for splenectomy must be clear and the cost benefit ratio must be understood by all involved in the decision making.
2. Patients with severe disease who are transfusion dependent and showing growth disturbances should undergo splenectomy.
3. Other compelling but not absolute indications include physical limitations due to anemia, leg ulcers, or when extramedullary hematopoiesis is significant.

(vii) A laparoscopic approach is the method of choice and partial splenectomy is an increasingly considered and used option, especially in light of the increased risk for thrombosis and sepsis after total splenectomy.

(viii) Partial splenectomy: near total splenectomy by removing all splenic pedicles except the left gastroepiploic vessel.

(ix) Embolization has also been used: regrowth of the spleen with increased hemolysis may occur.

5. Complications:

(a) Gallstones: the presence of bilirubin containing gallstones is common. Ultrasound is a non-invasive method for diagnosis and should be done every 5 years to access the presence of stones. If stones are detected cholecystectomy is indicated for recurrent symptoms or obstruction.

(b) Increased hemolysis: these may occur in the setting of acute infection and are marked by increasing bilirubinemia, enlargement of the spleen, anemia and increased reticulocytosis. Management is conservative and transfusions may be indicated.
(c) Aplastic crises: these occur as a result of viral infections including parvovirus B19 (fifth disease) and are marked by anemia (may be severe) and reticulocytopenia. Symptoms may be severe and transfusions may be necessary to treat/prevent congestive heart failure. The etiology of this complication lies in the shortened life span of RBCs in patients with HS. Cessation of RBC production for 10 days in patients with normal RBC lifespan (120) days results in an 8–10 % drop in hemoglobin. RBCs from HS patients may have a life span of 20 days resulting in a 50 % reduction in hemoglobin often starting from a lower baseline hemoglobin level.
(d) Megaloblastic crises: these may occur during periods of time of increased RBC production which outstrips the body's supply of folate (such as pregnancy). These may be easily prevented by the administration of folic acid.

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Idiopathic Thrombocytopenic Purpura

Jeffrey S. Taylor

Idiopathic thrombocytopenic purpura (ITP) is marked by thrombocytopenia (often profound) occurring due to destruction of antibody sensitized platelets by the reticuloendothelial system (RES).

1. Pathophysiology:
   (a) Incidence:
      (i) Most common autoimmune disorder affecting blood elements.
      (ii) Affects 1 in 10,000 children. This may be an underestimate: Asymptomatic patients may not come to medical attention.
      (iii) Acute ITP: duration of thrombocytopenia <6 months (some experts prefer using a 12 month cutoff for acute ITP).
      (iv) Chronic ITP: duration of thrombocytopenia >6 months (or 12 months, according to some).
   (b) Autoantibodies interact with membrane glycoproteins on platelets or megakaryocytes which are then cleared by the reticuloendothelial system.
   (c) Process is not well understood:
      (i) Viral immunocomplexes attach to platelet membrane.
      (ii) Antiviral antibodies cross react with platelet antigens.
      (iii) Absorption of viral particles on to platelet membranes.
      (iv) This process results in increased destruction and possibly decreased production of platelets.
   (d) Thrombopoietin levels are normal or only slightly elevated.

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2. Clinical presentation:
   (a) Classical presentation is that of a healthy child who presents with bruising, petechiae, and isolated thrombocytopenia. It is important to rule out artifact or laboratory error and "spurious" thrombocytopenia secondary to EDTA anticoagulant.
   (b) Often preceded by immunization (measles mumps rubella) or viral syndrome.
   (c) Bleeding is usually milder than expected for the degree of thrombocytopenia compared to other thrombocytopenic states such as aplastic anemia or leukemia. ITP platelets are young and hemostatically active.
   (d) Spleen may be palpable in 10 % of patients.
   (e) Systemic signs and symptoms are absent, i.e. bone pain, night sweats, malaise, adenopathy, and if these signs are present it suggests another disorder.
   (f) Peak age is 3–6-years-old. Infants and adolescents have an increased risk of having chronic ITP and having other immune disorders present such as lupus.
   (g) In younger children, the female to male ratio is equal. In adolescents, the female to male ratio is two to one.
3. Diagnosis:
   (a) White blood cells and hemoglobin are normal (unless patient is anemic secondary to bleeding).
   (b) Bone marrow shows normal or increased megakaryocytes.
   (c) Laboratory testing: complete blood count with differential, blood type and Coombs test, quantitative immunoglobulins, anti-nuclear antibody, and/or anti double stranded DNA if lupus is a possibility, and HIV, hepatitis C serology in appropriate circumstances.
   (d) Other etiologies for thrombocytopenia to be considered include Wiskott-Aldrich syndrome, thrombocytopenia-absent radii (TAR) syndrome, congenital amegakaryocytopenia, type 2B von Willebrand disease, and Bernard-Soulier syndrome among others. These other possibilities should be considered especially if the patient does not respond to usual ITP therapies.
4. Treatment:
   (a) Medical therapy of acute ITP does not alter the natural history of the disease.
   (b) Whom to treat:
      (i) Therapy is indicated in patients with platelet count less than 20,000/mcL and mucosal bleeding (wet purpura), or platelet count <10,000/mcL (although some hematologist will not treat at this level).
      (ii) Must consider the behavioral characteristics of the child (e.g. rough and tumble boy), parents coping skills and ability to monitor the child, and other members in the family.

(iii) Decision to treat is based upon concern over intracranial hemorrhage risk (0.1–0.9 %); although there is little evidence that treatment alters this risk.
(iv) Unlikely that a randomized clinical trial will be done (because of low risk of event and ethical concerns).

(c) Aspirin is withheld, parents should be educated, and limits placed on child's activities.
(d) Various medical treatments are available:

(i) Glucocorticoids: easy to give, may be done at home, side effect profile is extensive. There is continued debate on need for a bone marrow aspiration prior to giving steroids due to the risk of undiagnosed leukemia. Numerous variations of dose and schedule are available including high dose brief therapy, or longer term therapies with weans.
(ii) Intravenous immune globulin (IVIG): thought to interact with the FC receptors thus blocking platelet phagocytic cells of the RES. Usually given at 1 g/kg daily × 2 days as a slow infusion. IVIG is expensive, may cause allergic reactions especially in IgA deficient individuals, and 10 % of individuals may experience aseptic meningitis which may result in unnecessary CT scans due to concern over a possible intracranial hemorrhage.
(iii) Anti-D preparation: may be used in Rh + individuals. Anti-D coats red blood cells which then block access of platelets to the RES. Usually results in a slight reduction of hemoglobin. Side effects include headaches, chills, fevers, hemolysis (usually less than one gram). Occasional patients may have severe hemolysis with renal toxicity.

(e) Chronic ITP: 20 % of patient will have thrombocytopenia >6 months. The primary goal of therapy is to prevent bleeding and it may be difficult to maintain a normal platelet count due to side effects and toxicity of medical management.
(f) Splenectomy: the spleen is the major site of platelet destruction and antibody production. Splenectomy should be reserved for patients with significant risk of bleeding where medical therapy has either failed or the patient has developed unacceptable toxicity and should be delayed until after age 5-years-old due to risk of post-splenectomy sepsis. American Society of Hematology 1996 guidelines suggest that ITP must persist for >1 year, bleeding symptoms must be present, and platelets <10,000/mcL for ages 3–12-years-old before splenectomy is considered.
(g) Laparoscopic splenectomy is preferred.
(h) Attempts to raise the platelet count prior to surgery should occur.
(i) Splenectomy produces prolonged remission in 72 % of children and correlates with post-splenectomy platelet counts of >500,000/mcL and earlier response to glucocorticoids. In non-responders an accessory spleen should be sought.

(j) Patients should be immunized prior to procedure against Haemophilus influenzae type B, pneumococcus, and meningococcus. Penicillin prophylaxis should be started but the length of therapy is controversial.
(k) New treatments may decrease the need for splenectomy and include Rituximab (anti-CD 20), alemtuzumab (anti CD 52), and eltrombopag (thrombopoiesis stimulating agent).

5. Outcome:

(a) Prognosis: most children eventually recover normal platelet counts.
(b) 50 % recover counts within 8 weeks, 76 % within 6 months.
(c) Treatment or lack of treatment does not affect the platelet recovery outcome.
(d) Patients who become chronic tend to be >10-years-old, female and have a more insidious onset of symptoms.
(e) Overall prognosis is excellent with appropriate care.

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Thrombotic Thrombocytopenic Purpura

Jeffrey S. Taylor

Thrombotic Thrombocytopenic Purpura (TTP) is a syndrome marked by waxing and waning episodes of organ and tissue ischemia secondary to reversible platelet aggregation in the microvasculature. It often presents with thrombocytopenia, microangiopathic hemolytic anemia, neurologic symptoms, fever and renal abnormalities. Without treatment the mortality rate may reach 90 %. TTP is usually acquired although inherited chronically relapsing forms also occur.

1. Pathophysiology:
   (a) Characterized by thrombi containing aggregated platelets with little inflammatory response.
   (b) Lesions usually involve small vessels (arterioles and capillaries).
   (c) Brain, abdominal organs and the heart are most commonly involved.
   (d) Schistocytes are often present due to microthrombi present in the small vessels. These thrombi contain large amounts of von Willebrand factor (VWF).
   (e) An abnormal von Willebrand protein is thought to be directly involved in the pathology.
      (i) Von Willebrand protein comes in small, medium, and large multimers with differing biologic effects. Patients with TTP often have ultralarge (UL) multimers present. These multimers are biologically active and cause increased platelet aggregation.
      (ii) UL von Willebrand multimers are usually processed by the VWF-cleaving metalloproteinase ADAMTS13.
      (iii) Loss of function or deficiency in ADAMTS13 leads to increased levels of UL von Willebrand protein. Acquired TTP is often secondary to an autoantibody against ADAMTS13 which disrupts its function.

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2. Clinical presentation:
   (a) Incidence is one to four per million with a slight female predominance (female to male ratio of three to two), and peaks between ages 30–40-years-old.
   (b) Most cases are idiopathic, 15 % of patients have an underlying condition such as infection, pregnancy, collagen vascular disorders, and drugs have been implicated including cyclosporine, tacrolimus, quinine and clopidogrel.
   (c) Presentation is usually acute with bleeding and rapidly progressive neurologic symptoms including headache, coma, stupor, seizures, and hemiparesis.
   (d) Fever, anemia, bleeding, renal abnormalities, jaundice, fatigue, nausea and vomiting, abdominal pain, chest pain, cardiac arrhythmias and joint or muscle pain are variably present. The diagnosis is clinically based and therapy should not wait for confirmatory lab testing (i.e. ADAMTS13 testing).
   (e) Lactate dehydrogenase (LDH) is usually high due to tissue ischemia and hemolysis.
3. Diagnosis:
   (a) Confirmed by measurement of ADAMTS13 activity or antibodies against it.
   (b) Autosomal recessive TTP (Upshaw-Schulman syndrome) is secondary to mutations in the ADAMTS13 gene and may present early in life and be relapsing in nature.
4. Treatment:
   (a) Congenital TTP is treated with fresh frozen plasma (FFP) to replace the deficient enzyme. The number and schedule of infusions varies as to the severity of the deficiency, clinical response and is individualized.
   (b) Acquired TTP is treated with plasma exchange. This serves to both replete the deficient enzyme and to remove any anti-ADAMTS13 antibodies present.
   (c) Glucocorticoids are often used but there is little evidence that they help.
   (d) Recovery is variable and often one group of symptoms abates before another. Mental status and central nervous system problems may rapidly resolve while renal disease may have a slow recovery.
   (e) The use of rituximab may result in remissions in patients with poor response to plasma exchange.
   (f) Laparoscopic splenectomy is an option for refractory or relapsing TTP and has good results.

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Sickle Cell Anemia

Jeffrey S. Taylor

Sickle cell anemia is one of the first diseases understood at the molecular level. Valine substituted for glutamic acid in the beta globin subunit leads to decreased solubility in the de-oxygenated state. It may partially protect infants from cerebral falciparum malaria in the heterozygous state (sickle trait), and geographic distribution of the disease parallels the incidence of malaria.

1. Pathophysiology:

   (a) Hemoglobin S in the deoxygenated state polymerizes. This produces a change in the red blood cell from the biconcave disk to long rigid shapes: Polymerization and shape change is the disease and produces all the complications.
   (b) Acidosis, fever and dehydration increase the rapidity of polymerization/shape change and thus occlusion as well as the need to be treated aggressively in patients with sickle cell disease.
   (c) Sickle cells are damaged by repeated deoxygenation/shape changes and have a shortened life span. Sickle cell disease is one of the congenital hemolytic anemias with elevated reticulocyte count.
   (d) Disease severity is modulated by other hemoglobins:

      (i) Hemoglobin F (fetal) is elevated early in life which abolishes most complications.
      (ii) Hemoglobin C with hemoglobin S (HGB SC)- slightly milder form of disease
      (iii) Beta-thalassemia: presence of HGB A1 lessens severity of sickle cell disease complications. This effect is dependent on what percentage of hemoglobin is HGB A1.
      (iv) Sickle cell trait: HGB A1 > HGB S- patients are essentially asymptomatic.

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2. Diagnosis:
   (a) Hemoglobin electrophoresis is the gold standard; most new patients are diagnosed through newborn screening.
      (i) Newborn screening pattern depicts hemoglobin percentage in decreasing order.
         1. FA (normal): hemoglobin F percentage exceeds that of HGB A.
         2. FAS (sickle cell trait): Hemoglobin F is greater than HGB A, which is greater than HGB S.
         3. FSA (sickle cell/beta-thalassemia +): hemoglobin F is greater than HGB S, which is greater than HGB A.
         4. FSC (hemoglobin SC disease): hemoglobin F is greater than HGB S which is greater than HGB C.
         5. FS (hemoglobin SS or hemoglobin S/beta-thalassemia 0): hemoglobin F is greater than HGB S.
         6. These values will change as the infant ages; specifically the relative percentage of HGB F will decrease significantly to less than a few percent of the total hemoglobin.
3. Clinical manifestations are highly variable:
   (a) Marked by the "sickle crisis".
   (b) There are three major types:
      (i) Vaso-occlusive crisis (VOC): acute often painful episodes caused by intravascular sickling with tissue infarction.
         1. Painful crisis: this is the major clinical manifestation of sickle cell disease. It affects bones, brain, liver, lungs, spleen, and penis. Pain is dull, throbbing, may or may not produce heat or redness. Patients often can differentiate sickle cell pain from pain secondary to other sources. Infants may present with hand-foot syndrome. Early onset (<age 1-year-old) correlates with increased later disease severity. Treatment includes analgesics, IV fluid, and transfusions.
         2. Acute chest syndrome (ACS): VOC affecting lungs. ACS is the leading cause of death over age 10-years-old. Hypoxia leads to further sickling which leads to worsening hypoxia. ACS treatment includes bronchodilators, incentive spirometry, pulmonary toilette, oxygen, transfusions, antibiotics including macrolides, and maintenance fluid.
         3. Abdominal pain: may mimic acute abdomen (patients may be able to differentiate acute abdomen from usual sickle cell abdominal pain). Common etiologies include gallstones and mesenteric sickling. Treatment: Fluids, analgesia, and antibiotics to "cool down" the gallbladder prior to cholecystectomy.

4. Central nervous system/stroke: eventually affect up to 7 % of patients. Middle cerebral and anterior cerebral arteries are most common sites. Treatment includes hydration and transfusions with exchange transfusion, where possible, preferred over simple transfusions, followed by long term chronic transfusions.
5. Priapism: occurs in all age groups, recurrences may lead to sexual dysfunction. Treatment: Hydration and analgesics. The role for urologic intervention is controversial.

(ii) Splenic sequestration: sudden rapid enlargement of the spleen in which a significant portion of the RBC mass is entrapped.

1. Patient may present in shock with immediate need for blood volume restoration. In some cases the sequestration proceeds so rapidly that the patient dies before getting to the hospital.
2. Approximately 50 % of children will experience a second event.
3. The decision to undergo a splenectomy should be made with the family and hematology team taking into account proximity of family to a facility capable of transfusion, reliability of parents, and their skill in recognizing this complication. Many children are already functionally asplenic and thus the decision to undergo splenectomy is easier.
4. The early use of hydroxyurea may change the length of time that splenic function is preserved thus making the decision more difficult. All patients should be immunized against encapsulated organisms (including Streptococcus pneumononiae, Haemophilus influenzae type b, and Meningococcus) prior to splenectomy.

(iii) Aplastic crises: RBCs in patients with sickle cell anemia have a shortened life span, as short as 8 days (normal is 120 days). An interruption in production will lead to rapidly decreasing hemoglobin levels.

1. May be associated with viral or bacterial infections including parvovirus B-19.
2. Treatment is supportive with transfusions as needed.

(c) Infections in sickle cell anemia:

(i) All patients should be given Penicillin: 125 mg BID age 0–3-years-old, then 250 mg BID until age 5-years-old.
(ii) Immunizations are effective and are available for Streptococcus pneumononiae, Haemophilus influenzae type b, and Meningococcus.
(iii) Infections are not due to unusual organisms; they just behave more virulently due to splenic dysfunction. The presence of Howell-Jolly bodies (small blue/purple inclusions in RBCs) on the peripheral smear supports the conclusion that the patient is functionally asplenic.

(iv) All patients with a temperature greater than 101 °F once or greater than 100.4 °F on more than one occasion need evaluation. The decision to admit is individualized. If the temperature is <39.9 °C, child appears well and labs, chest x-ray are reassuring, long acting antibiotics such as ceftriaxone may be given with assured follow up the next day. Otherwise the child should be admitted.

(d) Other organ systems:

(i) Heart: patients with sickle cell disease experience very few myocardial infarctions due to very rapid transit of blood through the coronary circulation. Hypertension is not tolerated well due to relative left ventricular hypertrophy from chronic anemia. Hypertension should be managed aggressively.

(ii) Kidneys: ability to concentrate urine is compromised early due to hyperosmolar renal medulla which predisposes to sickling. Many patients develop hematuria with a small percentage progressing to renal failure. Patients with sickle cell trait may have an increased risk of developing renal cell carcinoma.

(iii) Eye: anterior chamber of the eye is hypertonic. Eye trauma with hyphema may result in acute obstructive glaucoma: A true ophthalmologic emergency. May also occur in patients with sickle cell trait.

(iv) Hepatobiliary: patients are predisposed to gallstones. If the decision to do a cholecystectomy is made, the gallbladder should be allowed to "cool down" with antibiotics prior to surgery. The patient also needs correction of anemia and preoperative hydration.

(v) Skeletal: avascular necrosis of the femoral head and humeral head should be considered and evaluated in any patient with limp or long standing complaints. Codfish vertebrae (exaggeration of the concavity of vertebral surface due to osteoporosis) may be seen due to frequent cycles of VOC in vertebral bodies resulting in collapse and remodeling.

(vi) Pulmonary: repeated episodes of ACS result in pulmonary fibrosis/hypertension, which is an ominous development. Sleep apnea should be treated aggressively to prevent prolonged hypoxia at night.

4. Treatment:

(a) Routine care:

(i) Patients should be followed by a hematologist or a pediatrician familiar and comfortable with the care necessary.

(ii) Crises care and any identifiable precipitating causes should be discussed.

(iii) Infection and fever management, penicillin adherence and immunizations.

(iv) Discussion concerning medication adherence including folic acid, hydroxyurea, and pain medications.

(v) Referral to appropriate subspecialists where indicated, i.e. pulmonary, dentistry, cardiology, ophthalmology, orthopaedic surgery, and pediatric surgery, as needed.
(vi) Psychological/social support including school, physical limitations (e.g. activity restriction secondary to avascular necrosis of femur), and family dynamics.
(vii) Central nervous system imaging and assessment of stroke risk.
(viii) Baseline laboratory studies: hemoglobin, reticulocyte count, platelet count, pulse oximetry, proteinuria, WBC, liver function tests, bilirubin level, and kidney function testing.
(ix) Spleen size and assessment of gall bladder, where indicated.

(b) Management of painful crises:

(i) Most painful crises are managed at home. Clinicians see only the tip of the iceberg.
(ii) "Drug seeking" behavior is no more common in patients with sickle cell disease than in the general population.
(iii) Doses must be titrated to effect, not standard doses. Frequent evaluations are necessary to adjust dosages. Decreased ventilation from splinting due to insufficient pain control is just as damaging as sedation from too much. Both result in hypoxia and increase the risk of pulmonary complications.
(iv) Oxygen is helpful if hypoxic but should not be used if patient is not hypoxic. The overuse of oxygen may result in a decrease in the patient's reticulocyte count and resultant reticulocyte rebound which may cause a new crisis.

(c) Long term therapy:

(i) Bone marrow transplantation: very toxic and the decision as to who to transplant is difficult. Ideally candidates should be healthy, although it may be difficult to justify a toxic therapy in a healthy child. Children with complications obviously need the transplant but are at higher risk.
(ii) Hydroxyurea: use is increasing as safety concerns have been addressed. Mechanism: increases hemoglobin F, increases hydration of sickle cells, decreases reticulocyte count as well as platelet count and WBCs: all positive side effects. Ability to preserve splenic function is under active investigation.

(d) Surgical preparation:
(i) Transfusion to hemoglobin of 10 g/dL is advisable prior to surgical procedure. In general, 5 mL per kg of pRBCs will raise hemoglobin by 1 g/dL. Alternatively the formula, $4 \times$ delta (desired HGB $-$ pre transfusion HGB) $\times$ weight in kg $=$ mL of pRBCs to be transfused, may be used for smaller patients.

(ii) The use of simple transfusions vs. exchange transfusions is controversial. There are more complications associated with exchange transfusions. Additionally vascular access and time availability are also considerations. The present consensus is that there is likely no increase in surgical complications by the use of simple transfusions as compared to exchange transfusions.
(iii) Most common surgical complication is acute chest syndrome.
(iv) Anesthesia care is critical: patient should be warm, appropriately oxygenated and hydration provided to insure circulatory optimization.

5. Other sickle syndromes:

(a) Sickle cell trait: HGB A1 > HGB S. Tends to be benign.

(i) Complications include increased VOC risk of unpressurized high altitude flying; loss of urine concentrating ability due to hyperosmolar kidney medulla causing sickling, and hyphema due to high tonicity of anterior chamber of eye.
(ii) Likely increased risk of renal cell carcinoma.
(iii) The incidence of sickle cell trait in the National Football League parallels that of the general population.
(iv) Patients participating in sports should have access to fluids at all times and remain well hydrated.

(b) Hemoglobin SC disease: tends to be milder than HGB SS, but may be severe.

(i) Beta globin has HGB S, the other has HGB C.
(ii) Some complications are more common, i.e. avascular necrosis of femur.
(iii) Hemoglobin is usually in the 10 g/dL range with target cells present on smear as well as some sickle cells.

(c) Sickle beta-thalassemia: severity depends on the output of the beta-thalassemia gene.

(i) If no normal hemoglobin A1 present (Hemoglobin S/beta-thalassemia 0) then the course is similar to hemoglobin SS. Patients with some HGB A1 production may have a milder course (hemoglobin S/beta-thalassemia +).
(ii) Microcytosis and an increase in hemoglobin A2 help make the diagnosis.

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Part VI
Common Adolescent Problems

Gynecomastia

Alysia A. Agnoni

Gynecomastia is the proliferation of glandular breast tissue in men and is common in males of all ages. In infancy, gynecomastia is caused from excess estrogen during pregnancy and regresses over the first month of life. Pseudogynecomastia is the proliferation of fatty tissue around the breast in obese men and is often mistaken for gynecomastia.

1. Etiology:
   (a) Many cases of gynecomastia have no detectable cause.
   (b) Other causes include:
      (i) Puberty: Usually resolves but some persist into adulthood.
      (ii) Drugs: Anti-androgens and hormones, antibiotics (ketoconazole, metronidazole, isoniazid), antireflux (cimetidine, ranitidine, omeprazole), some chemotherapeutic drugs, cardiovascular (spironolactone, angiotensin-converting enzyme inhibitors, amiodarone, calcium channel blockers, digoxin, methyldopa, reserpine), psychoactive (diazepam, haloperidol, phenothiazines, tricyclic antidepressants), recreational (alcohol, amphetamines, heroin, marijuana, methadone), herbals (tea tree and lavender oil), and others (highly active antiretroviral therapy, metoclopramide, theophylline).
      (iii) Tumors, hypogonadism, malnutrition, hyperthyroidism, and renal insufficiency are other less common causes of gynecomastia.
2. Pathophysiology: A decrease in androgen production and an increase in estrogen production cause glandular proliferation, ductal epithelial hyperplasia, ductal elongation and branching, and proliferation of the periductal fibroblasts.

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3. Clinical findings:
   (a) History:
      (i) When did symptoms develop: birth, puberty, weight gain
      (ii) Medications
   (b) Physical exam:
      (i) Well-defined glandular tissue is palpated directly beneath the nipple/areola and is firm and rubbery.
         1. Pseudogynecomastia does not present as a discrete mass.
         2. Tumor/mass: Most often do not present directly under the areola.
      (ii) May be tender.
      (iii) Nipple sensitivity.
      (iv) GU: Examination of scrotal contents: Hypogonadism or testicular mass.
   (c) Lab results:
      (i) Consider obtaining human chorionic gonadotropin, luteinizing hormone, testosterone, estradiol, and thyroid stimulating hormone to evaluate for potential causes (tumor, hypogonadism, or hyperthyroidism).
4. Treatment:
   (a) Pubertal: Due to an increase of estradiol prior to an increase in testosterone. This usually resolves within 6 months to 2 years and observation is recommended. Persistence beyond age 17 is uncommon and results in fibrotic breast tissue that will likely need surgical excision.
      (i) Drug treatment: Reasonable option in a pubertal boy with significant breast enlargement, tenderness, and embarrassment that affects quality of life. Will decrease tenderness and breast tissue but typically does not cause complete regression
         1. Androgens (testosterone): Not suggested for use in eugonadal men; may help in hypogonadal men.
         2. Selective estrogen receptor modulators (tamoxifen and raloxifene): Some breast regression with relief of tenderness.
         3. Aromatase inhibitors (anastrozole).
      (ii) Operative therapy: Reserved for patients with gynecomastia who have had no regression of breast tissue with observation or medication and remain with considerable tenderness and psychological distress.
         1. Regrowth may occur if still undergoing puberty.
         2. Techniques may differ but a common approach is direct excision of glandular tissue via a semicircular incision inferior to the areola.
         3. Complications include: Compromised blood supply to the areola and nipple, unevenness of breast tissue, and seroma or hematoma.
   (b) Medication: Discontinue any drugs with gynecomastia side effects.

Pectus Deformity

Christopher P. Coppola

Pectus excavatum is a common abnormality of the chest wall that ranges from a mild cosmetic deformity to a severe condition with crippling effects on cardiac and pulmonary function coupled with associated anomalies. As the condition often changes in severity during periods of growth, it is best to wait until late adolescence when operative repair is indicated, if the clinical status of the child allows a period of observation.

1. Classification of pectus deformity
   (a) Pectus excavatum: "funnel chest" – sunken/concave appearance of the sternum and ribs with a "soup bowl" or "Death Valley" appearance. Approximate occurrence is one in every 400 births.
   (b) Pectus carinatum: "pigeon chest" – outward protrusion of the sternum and ribs with a bird-like appearance of the chest.
   (c) Combined deformity: asymmetric or mixed deformity with tilt of the sternum off of the coronal plane, flaring of the ribs, and disordered alignment of the ribs.
   (d) Chest wall deficits: missing components of the rib cage including missing vertebrae, hemivertibrae, missing ribs, fused ribs, missing clavicles, and can also include deficits in the soft tissue of the chest.
      (i) Poland syndrome is characterized by unilateral chest wall deficit and can extend to missing pectoral muscle, breast, and limb component deficits.
      (ii) Jeune's asphyxiating thoracic dystrophy is characterized by failure of rib growth and a bell-shaped chest with a narrow apex. Patients often have difficulty handling secretions and require tracheostomy.

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(iii) Jarcho-Levin syndrome (spondylothoracic dysplasia) is characterized by shortened deformed thoracic vertebrae and associated pulmonary, cardiac, and renal anomalies and a corresponding high mortality.
(iv) Pentalogy of Cantrell is a condition with 75 % mortality due to the sequelae of five components:

1. Pericardial defect.
2. Ectopia cordis.
3. Sternal deficit.
4. Anterior diaphragm hernia.
5. Abdominal wall defect.

(e) Pectus excavatum can be associated with other disorders:

(i) Marfan syndrome may be present. Individuals often have an earlier presentation and greater severity of defect. Individuals with Marfan syndrome must be evaluated for the presence of aortic root dilation as it carries a risk of aortic dissection.
(ii) Ehlers-Danlos syndromes: soft tissue defect with hyperextensible joints and loose skin.

2. Etiology of pectus excavatum is thought to be related to disordered or rapid growth of costal cartilages during periods of rapid growth, such as adolescence.
3. Clinical effects of pectus excavatum:

(a) Pain: disordered costal-chondral joints at edge of sternum can hurt with activity.
(b) Cardiac: patient may experience palpitations, fainting, or mitral valve prolapse with murmur.
(c) Pulmonary: Shortness of breath, exercise intolerance, and restrictive defect on pulmonary function testing.
(d) Scoliosis is present in 15 % of patients with pectus excavatum.
(e) Psychosocial: Embarrassment when chest is exposed, avoidance of activities such as swimming, use of baggy or loose clothing to hide defect, behavioral changes, teasing in school or sports, depression, and suicidal ideation may be present, and may not be elicited unless specifically queried.
(f) Cosmetic component: This is often erroneously thought to be the only aspect of the disease and can introduce problems for patients when they seek medical insurance approval for treatment.

4. Evaluation of pectus excavatum:

(a) Physical examination:

(i) Examination of chest including ribs, sternum, clavicles, breath sounds, and heart tones/rhythm.
(ii) Look for signs of Marfan syndrome: lens abnormality, high arched palate, tall height, long thin fingers, hyperextensible joints, heart murmur.
(iii) Evaluate for scoliosis.

(b) Other studies:

(i) Chest x-ray: Check cardiac silhouette, ribs and vertebrae
(ii) Spine series: Evaluate for scoliosis.
(iii) Pulmonary function tests: Evaluate for restrictive defect, can be required for insurance approval of repair.
(iv) Echocardiography: Evaluate for dilated aortic root and mitral valve prolapse.
(v) Computed tomography chest:

1. Non-contrast.
2. Calculate Haller index: Measure widest transverse distance across thoracic cavity (inside ribcage) and divide by smallest antero-posterior distance across thoracic cavity (from back of sternum to front of spine). Normal value is 2.5. Haller index greater than 3.2 is considered severe pectus excavatum, and this value is sometimes required for insurance approval of repair. Value less than 2 is abnormal and indicated pectus excavatum. There is a wide crossover between normal and abnormal.
3. Calculate correction index: Measure the antero-posterior distance from most anterior costal ridge to deepest portion of sternal depression and divide by antero-posterior depth of chest, including ribs, as a percentage. This newer index better discriminates between normal and pectus excavatum. Normal value is <10 %.
4. Aortic root diameter can also be measured.

5. Treatment of pectus excavatum.

(a) When no cardiac or pulmonary disturbance or pain is present, patients and families may choose observation only, and live with the thoracic defect, returning for care if the condition worsens or physiologic symptoms develop.

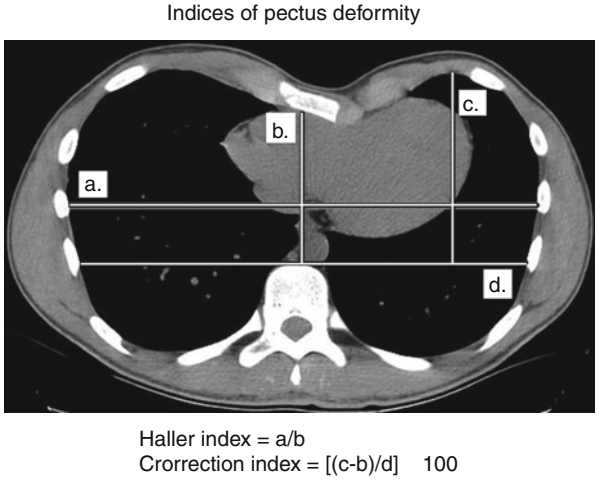

Fig. 1 Indices of pectus deformity. The Haller index is obtained by dividing the width of the thoracic cavity by the depth. The Correction index is obtained by dividing the difference between the highest costal ridge and the lowest depth of sternal depression by the width of the thoracic cavity at the anterior vertebral margin. Source: Christopher Coppola

(b) Surgical repair of defect, when chosen, is best deferred until age 15-years-old or older, because there is a higher incidence of recurrence in children who are repaired younger.
(c) Ravitch repair: This method has been available longer. The anterior chest is incised either transversely or midline. The cartilaginous segments connecting bilateral ribs five to eight (with adjustment for individual cases) are resected. The sternum is fractured transversely between the fourth and fifth ribs, and it is repositioned anteriorly to correct the central depression. A short bar spanning the severed ends of the seventh ribs is used to support the sternum in this position.
(d) Nuss repair: bilateral axillary incisions are made at the same level of the deepest depression of sternum. Using a thoracoscope for guidance, a curved bar is guided in one incision and out the other, passing beneath the sternum to elevate it into a more anterior position. The ends of the bar are secured to the intercostal muscles bilaterally. The bar is removed after 3 years when the corrected shape of chest has stabilized.
(e) Patient may require a chest tube, and a chest x-ray must be obtained after operation.
(f) Recovery: Patients are hospitalized 5–7 days after repair. To avoid bar dislodgement, patients must avoid lying on the side, twisting the torso, or lifting themselves up with the arms. An epidural catheter with local anesthetic improves pain control.
(g) Patients will usually miss school for 3 weeks, no lifting or gym class for 6 weeks, and full impact sports, such as football, are allowed after 3 months. Most patients require oral analgesics for 6 weeks.
(h) Repair may be complicated by hemorrhage, pulmonary injury, cardiac injury, arrhythmia, pneumothorax, infection, effusion, seroma, chronic pain, bar dislodgement, allergy to metal in bar, and recurrence of defect.

6. Treatment of pectus carinatum

(a) Patients can be treated an orthotic brace which places pressure on the protrusion of the sternum. If the child can tolerate wearing the brace for 12 h each day for a year, there is a good success rate. The brace must be individually fit to the patient and adjusted as progress is made. Side effects are pain and damage to skin at the brace contact points.
(b) Pectus carinatum can be surgically repaired by the open Ravitch method.

Imperforate Hymen

Meng-Fey Ferra Lin-Duffy

The hymen membrane consists of fibrous connective tissue connecting to the vaginal wall and separates the vaginal lumen from the urogenital tract.

1. Pathophysiology:
   (a) Anomalies of the hymen: Result from incomplete degeneration of the central portion of the hymen.
      (i) Imperforate.
      (ii) Microperforate.
      (iii) Septate.
      (iv) Cribriform.
   (b) Imperforate hymen is one of the most frequent obstructive anomalies in the female genital tract.
   (c) It is usually sporadic but familial occurrence has been reported.
   (d) Urologic anomalies are not associated.
2. Clinical presentation:
   (a) Infants may present with a bulging introitus at birth due to vaginal secretions from maternal hormonal stimulation. Secretion is usually reabsorbed by the child's body and she will be asymptomatic until menarche.
   (b) Routine examination of the genitalia by primary pediatrician is essential to rule out any congenital anomalies.
   (c) Adolescents may present with cyclic abdominal pain or pelvic pain.
   (d) The typical presenting symptom is primary amenorrhea which would be a late finding.
   (e) Abdominal or pelvic mass may also be present.

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3. Diagnosis:
   (a) Pelvic ultrasound is the initial diagnostic test and it often shows a hematocolpos, hematometra and possibly hematosalpinges when diagnosis is made after the onset of endometrial shedding.
   (b) MRI should be obtained if ultrasound showed equivocal result.
   (c) Pelvic exam under anesthesia may aid in confirmation of diagnosis and surgical planning.
4. Treatment:
   (a) Optimal timing for surgical repair is after the onset of puberty but before menarche. The presence of estrogen assists in surgical healing.
   (b) Surgery should not be performed under emergent presentation.
   (c) Oral contraceptive and non-steroidal anti-inflammatory agents can provide symptomatic relief.
   (d) Surgical repair consists of an ellipse incision on the membrane close to the ring of the hymen with extra tissue excised to create a normal size orifice. The vaginal mucosa is sutured to the hymen ring to prevent adhesion or recurrence.

Inflammatory Bowel Disease

Filip Moshkovsky

Inflammatory bowel disease (IBD) encompasses two similar but distinct disease processes that cause idiopathic inflammation of the gastrointestinal tract. IBD disease process may involve an altered immune system, genetic predisposition and environmental factors to varying degrees. Crohn's disease (CD) has no known cure but does have complicated treatment options which span the medical and surgical expertise. Total colectomy will allow the only option for cure in ulcerative colitis (UC). Treatment is best offered in a multidisciplinary fashion with the goal of therapy to control disease and maintain patients in remission.

Crohn's Disease

1. Pathophysiology:
   (a) An abnormal inflammatory response to the intestinal mucosa leads to ulceration (aphthous ulcers) and form in linear fashion. The surrounding mucosa becomes edematous and forms a distinctive appearance of "cobblestone".
      (i) Histologically the bowel wall reveals neutrophilic as well as eosinophilic inflammatory cells
      (ii) In nearly half the cases a non-caseating granuloma will be seen and is pathognomonic for CD.
   (b) As the inflammation progresses it affects all the layers of the bowel and will lead to the hallmark "creeping fat" as the mesentery surrounding the affected bowel thickens and proliferates.

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(c) Submucosa is edematous and fibrotic.
(d) If the inflammation progresses it may lead to transmural edema with sinus creation and fistula formation. Chronic inflammation may form strictures and rarely perforation.
(e) Crohn's disease may involve the entire gastrointestinal tract and may be difficult to distinguish from UC. Most commonly the ileum and cecum are affected together. The disease can be isolated just to the small bowel, most likely the ileum or more rarely isolated to the colon.

2. Clinical features:

(a) Most common presenting signs and symptoms:

(i) Crampy abdominal pain.

1. Focal peritonitis, mass in RLQ and fever may indicate perforation.

(ii) Weight loss/failure to thrive: Common to see child "fall off" the growth chart.

1. Decreased appetite.
2. Growth and sexual maturation delay.

(iii) Diarrhea and/or multiple bowel movements.

1. Occult blood present in stool. Frank blood may not be present.

(b) Other signs and symptoms:

(i) Perianal disease.

1. Anal fissures.
2. Perianal fistulas.
3. Perianal abscess.

(ii) Erythema nodosum.
(iii) Arthritis and/or arthralgia.
(iv) Episcleritis and uveitis.
(v) Nephrolithiasis.
(vi) Hepatobiliary symptoms.

1. Primary sclerosing cholangitis.
2. Cholelithiasis.
3. Hepatitis.
4. Steatosis.

3. Diagnosis:

(a) History and physical: Due to the multiple body systems involved a thorough exam is very important.

(i) Trend child's weight and recent complaints/hospitalizations.
(ii) Family history.
(iii) Special attention to the abdominal exam which may reveal a subtle mass in the right lower quadrant.

(b) Laboratory tests:

(i) Complete blood count (CBC): For anemia.
(ii) Basic metabolic panel (BMP): Electrolyte disturbances.
(iii) Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP): Inflammatory markers may be elevated during acute flare-ups.
(iv) Nutritional studies and hepatic function tests: May be abnormal.
(v) IgA and IgG anti-saccharomyces cerevisiae antibody is highly specific for CD in children.
(vi) Send stool to be checked for blood and infectious agents.

(c) Rectal exam:

(i) In a younger child or a very anxious patient examination under anesthesia may be advantageous.

1. Anorectal endoscopic ultrasound which may help in detecting an abscess.

(d) Imaging:

(i) Upper gastrointestinal (UGI) contrast study with small bowel follow through (SBFT) may show strictures.
(ii) Computed tomography (CT) imaging may show mucosal nodularity, area of inflammation, abscess, creeping fat.
(iii) Magnetic resonance imaging (MRI) with gadolinium enhancement is sensitive to identifying proximal small bowel disease.

(e) Endoscopy:

(i) Ileocolonoscopy: be cautious if there is concern for perforation.

1. Direct observation:

(a) Skip lesions: Cobblestone appearance.
(b) Aphthous ulcers.
(c) Inflamed terminal ileum.

2. Terminal ileum biopsy.

(ii) EGD: May have lesions in the upper gastrointestinal tract as well.
(iii) Capsule endoscopy may have a role in certain patients.

4. Initial management:

(a) If no sign of perforation or obstruction then medical treatment is first line.

(i) Mesalamine (an aminosalicylate) is the first-line therapy for mild to moderate active CD.
(ii) Intravenous fluids and correction of electrolyte abnormalities.
(iii) Immunosuppressant medication:

1. Steroids.
2. Azathioprine.
3. 6-mercaptopurine.

(b) Concern for perforation requires exploratory laparotomy and resection.

(i) May need a temporary ostomy while the inflammation subsides.

(c) Obstruction requires operative exploration and as minimal resection as possible.
(d) Intraabdominal abscess is best treated with image guided drainage and antibiotics.
(e) Fistulas between bladder, uterus, and other viscera are treated with limited resection.
(f) Perianal abscess and complex fistulas are treated surgically with as minimal disruption to the sphincter as possible.

(i) Complex fistulas may require a diverting colostomy.
(ii) Use a noncutting seton: Avoid reactive sutures or seton as this may lead to further inflammatory response.
(iii) Fibrin glue may also be attempted.

5. Treatment:

(a) Multidisciplinary team approach.

(i) Gastroenterologist.
(ii) Pediatrician.
(iii) Pediatric surgeon.
(iv) Nutritionist.
(v) Social worker.

(b) Maintaining patient in remission is goal of treatment.

(i) Infliximab:

1. Test for latent tuberculosis before infliximab therapy is started.

(ii) Metronidazole and ciprofloxacin in mild to moderate disease:

1. May help delay recurrence at surgical anastomosis.
2. Aid in healing of perianal fistula.

(iii) Dietary change/enteral therapy:

1. Liquid diet - advance as tolerated.
2. If not obstructed - enteral nutrition:

(a) Enteral formulas with transforming growth factor-β (TGF-β) and omega-3 fatty acids.

(iv) Surgical treatment:

1. If medical therapy fails to control the flare-up then resection of the affected area is indicated.

2. Ileocecectomy is the treatment of choice for patients with isolated ileocecal disease.
3. Stricturoplasty for strictures causing near obstruction.
   (a) Heineke-Mikulicz for 1–10 cm strictures.
   (b) Finney stricturoplasty for longer strictures.
   (c) Michelassi technique: For longer strictures (side-to-side isoperistaltic stricturoplasty).

6. Outcome:

(a) There is no cure for Crohn's disease at present.
(b) Remission is goal.
(c) Minimize bowel resection if surgical intervention is required.
(d) Colorectal and small bowel cancer is increased in CD.
   (i) Screening for colorectal cancer should start 10 years after start of symptoms.

Ulcerative Colitis

1. Pathophysiology:

(a) Chronic inflammatory response confined to mucosa and does not extend into the muscularis propria. The inflammation usually involves the rectum and extends proximally in a contiguous fashion without skip lesions.
   (i) Microscopically: Crypt abscesses and mucosal ulceration.
   (ii) Undermining of mucosa by ulcerations leads to mucosal bridging and characteristic pseudopolyp formation in circumferential pattern.

(b) The acute phase of the disease significantly affects the colon.
   (i) Distention, decreased peristalsis, muscularis propria thins and bleeds.
   (ii) If the acute phase is not treated it may progress to toxic megacolon: severe colonic dilatation, peritonitis and even perforation.

(c) In chronic disease a characteristic lead pipe appearance is observed on imaging.
   (i) The muscularis propria thickens and haustral folds are flattened with decreased peristalsis.
   (ii) Mucosal surface becomes atrophic and may develop dysplasia
   (iii) Mesentery may shorten and serosal surface increase in vascularity as well as adipose tissue.

2. Clinical features: presentation can range from mild to severe.
   (a) Most common presenting signs and symptoms:
      (i) Multiple daily loose bowel movements.
         1. The diarrhea may have blood, mucus and pus.
         2. Significant disease may cause frank blood per rectum necessitating blood transfusions.
      (ii) Abdominal pain: intermittent and crampy.
         1. Decreased appetite or anorexia.
         2. Rapid weight loss in severe disease.
      (iii) Tenesmus.
      (iv) In severe cases patient may present in sepsis: Toxic megacolon.
      (v) Aphthous stomatitis often present during active disease.
   (b) Other signs and symptoms:
      (i) Anemia.
         1. Secondary to bloody diarrhea.
         2. Anemia of chronic disease.
      (ii) Fevers.
      (iii) Hemorrhoids.
      (iv) Erythema nodosum.
      (v) Arthralgia.
      (vi) Pyoderma gangrenosum.
         1. Most common in the lower extremities and occasionally the trunk.
      (vii) Uveitis.
      (viii) Nephrolithiasis.
      (ix) Delayed sexual maturation.
      (x) Primary sclerosing cholangitis.
3. Diagnosis:
   (a) History and physical: due to the multiple body systems involved a thorough exam is very important.
      (i) Signs of anemia.
      (ii) Signs of dehydration.
      (iii) Signs of sepsis and/or shock in severe cases.
   (b) Lab work:
      (i) CBC: For anemia.
      (ii) BMP: Electrolyte disturbance with persistent diarrhea.

(iii) ESR and CRP: Inflammatory markers may be elevated in acute phase.
(iv) Nutritional studies:

1. Hypoalbuminemia secondary to protein loss from ulcerated colonic mucosa.

(v) Send stool to be checked for leukocytosis, blood and for infectious etiology.

1. Clostridium difficile infection is possible and can significantly complicate the diagnosis.

(c) Rectal exam:

(i) External hemorrhoids are common.
(ii) Absence of fistulas, fissures, and abscess. If seen may suggest CD.

(d) Imaging:

(i) UGI contrast study with SBFT helps to identify if there is small bowel disease which would suggest CD rather than UC.
(ii) Abdominal plain film x-ray.

1. If toxic megacolon suspected, get plain film first.

(a) Quickly identify if free air is present.

(iii) CT imaging may show mucosal thickening of the large bowel with sparing of the small bowel.
(iv) MRI with gadolinium enhancement is sensitive to identifying proximal small bowel disease and less sensitive for large bowel. May be a better choice due to lack of ionizing radiation and the multiple imaging the child will most likely receive in their life time.
(v) Barium enema (BE): Not commonly used due to concern of barium stimulating acute manifestations of the disease.

1. Classic lead pipe appearance due to loss of the haustral folds.
2. Pseudopolyps may be visible.

(e) Endoscopy:

(i) Ileocolonoscopy may be very helpful in establishing diagnosis between CD and UC.
(ii) Flexible sigmoidoscopy with biopsy:

1. Direct observation:

(a) Friable and edematous colonic mucosa with a thin, purulent exudate.

(iii) Upped endoscopy: Use if unsure between UC and CD.

4. Initial management:
   (a) Toxic megacolon presentation especially with perforation will require immediate surgical intervention.
      (i) Total colectomy with end ileostomy.
   (b) Severe rectal bleeding and severe symptoms refractory to medical treatment also require urgent surgical intervention.
   (c) If no sign of perforation or shock then:
      (i) 5-aminosalicylic acid, 5-ASA is first line of therapy in mild to moderate disease.
      (ii) IV fluids and correction of electrolyte abnormalities.
      (iii) In severe disease, IV steroids.
         1. If disease refractory to other treatment options Cyclosporine may be given.
5. Treatment:
   (a) Multidisciplinary team approach:
      (i) Gastroenterologist.
      (ii) Pediatrician.
      (iii) Pediatric surgeon.
      (iv) Nutritionist.
      (v) Social worker.
   (b) Maintaining patient in remission or potential cure is goal of care.
      (i) Long term steroids and other immunosuppressant lead to higher complication rates after surgery.
         1. The sooner the colectomy is done the less likely complications to follow.
         2. Due to the potential cure of disease elective colectomy should be considered.
      (ii) Total colectomy with ileoanal pouch procedure is now the preferred surgical option for elective colectomy secondary to UC.
         1. Distal rectal mucosectomy (lower 4 cm) performed in order to spare the pelvic nerves and sphincter muscles.
         2. J-shaped pouch is most common. S-shaped pouch is also commonly constructed. Key is to keep pouch length approximately 8 cm in length.
         3. A temporary loop ileostomy is created to protect the anastomosis.
         4. A water-soluble contrast enema is performed 2 months after pouch creation to ensure that the ileal reservoir has no leaks or sinus tracts at which time the loop ileostomy is usually reversed.

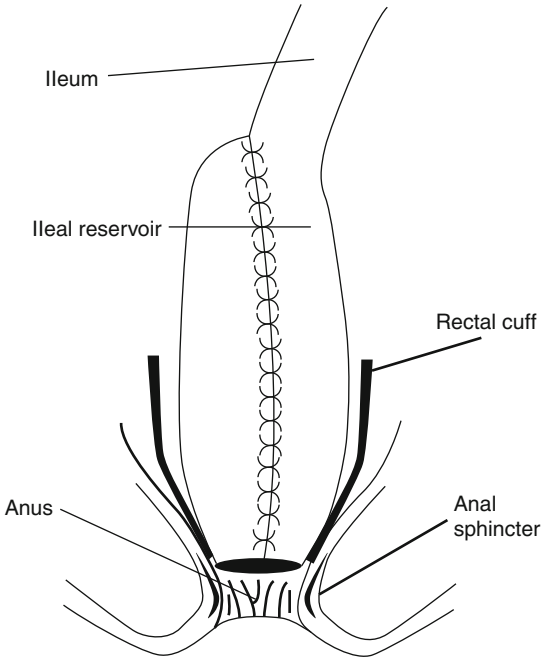

Fig. 1 J-pouch with ileoanal anastomosis. After proctocolectomy for ulcerative colitis, a J-pouch is created and an anastomosis made to the rectal cuff creating an ileoanal reservoir for stool. Usually a protective diverting loop ileostomy is created proximal to the J-pouch to allow it to heal (Adapted from: National Digestive Diseases Clearinghouse, National Institutes of Health, NIH, May 2012. http://digestive.niddk.nih.gov/ddiseases/pubs/dictionary/e-k.aspx. Downloaded on 6 Jan 2014)

5. Significantly increased stooling frequency is common which typically gets better in several months.
   (a) Multiple stools each day is common lifelong.
      (i) Bulking agents and anti-diarrhea medication may help to decrease frequency.
   (b) Pouchitis is the most frequent complication.
      (i) Crampy lower abdominal pain and increased frequency of stooling, often with watery diarrhea.
      (ii) Initiate metronidazole or ciprofloxacin. Barium enema to confirm pouch is open. Consider daily pouch washout with tap water. If needed diverting ileostomy
      (iii) Probiotics may decrease the recurrence of pouchitis.
   (c) In cases of pull-through failures a permanent ileostomy is required.
6. Outcome:
   (a) Medical management of UC with goal of remission.
   (b) Surgical treatment with goal of cure.
      (i) Patient and family need to be well informed of what to expect after surgery and the possible complications.

(c) Colorectal cancer is increased in UC.

(i) Three percent risk of colorectal cancer in first decade with 10–15 % increased risk each subsequent decade after the diagnosis.
(ii) Evidence of dysplasia on biopsy: Increased risk of developing a carcinoma.
(iii) Screening should start 10 years after diagnosis.

1. May consider sooner if dysplasia initially seen on biopsy.

Multiple Endocrine Neoplasia Syndromes

Luiz G. Foernges

1. Multiple endocrine neoplasia type 1 (MEN 1), Wermer syndrome: Pathophysiology:
   (a) Autosomal dominant pattern of inheritance. Caused by mutation in the *MEN1* gene. This gene encodes *MENIN* protein, *MENIN* acts as a tumor suppressor. Affects 1 in 30,000 people in the United States.
2. Clinical diagnosis:
   (a) Parathyroid tumors: Hypercalcemia, altered mental status, lethargy, confusion, anorexia, constipation, nausea, vomiting, dehydration, hypercalciuria, kidney stones, increased bone resorption, hypertension, shortened QT interval.
   (b) Pituitary tumors:
      (i) Prolactinoma: Oligomenorrhea or amenorrhea and galactorrhea in females, sexual dysfunction and gynecomastia in males, headache, nerve compression.
      (ii) ACTH-secreting tumors: Cushing's disease.
      (iii) GH-secreting tumors: Gigantism and acromegaly.
   (c) Neuroendocrine tumors:
      (i) Zollinger-Ellison (gastrinoma): Peptic ulcer and chronic diarrhea.
      (ii) Insulinoma: Hypoglycemia.
      (iii) Glucagonoma: Hyperglycemia, anorexia, glossitis, anemia, diarrhea, venous thrombosis, necrolytic migratory erythema.
      (iv) Vasoactive intestinal peptide secreting tumor (VIPoma): Watery diarrhea, hypokalemia and achlorhydria.

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(d) Other tumors:

(i) Facial angiofibromas.
(ii) Collagenomas.
(iii) Lipomas.
(iv) Meningiomas.
(v) Ependymomas.
(vi) Leiomyomas.

3. Laboratory testing:

(a) Molecular genetic testing: *MEN1* gene mutations.
(b) Primary hyperparathyroidism: Increased serum parathyroid hormone and calcium.
(c) Prolactinoma: Increased serum prolactin.
(d) Neuroendocrine tumors:

(i) Gastrinoma: Increased serum gastrin, secretin stimulation test.
(ii) Insulinoma: Fasting hypoglycemia, high serum insulin, C-peptide or proinsulin.
(iii) Glucagonoma: Increased serum glucagon level.
(iv) VIPoma: High serum VIP.

4. Imaging:

(a) Prolactinoma: MRI.
(b) Gastrinoma: Endoscopic ultrasound, somatostatin receptor scintigraphy, CT, MRI.

5. Natural history:

(a) Hyperparathyroidism: onset between 20 and 25-years-old, 100 % incidence by age of 50-years-old.
(b) Prolactinoma: First manifestation of MEN syndrome in 10 % of the cases, incidence 10–60 %.
(c) Gastrinoma: 40 % incidence, frequent multiple and malignant.

6. Treatment:

(a) Hyperparathyroidism: Subtotal parathyroidectomy or total parathyroidectomy with autotransplantation.
(b) Prolactinoma: Dopamine agonists.
(c) Gastrinoma: Proton pump inhibitors and surgical resection.

7. Surveillance:

(a) Hyperparathyroidism: Fasting total serum calcium and/or ionized calcium concentration, serum PTH, yearly after 8-years-old.
(b) Prolactinoma: Serum prolactin, yearly after 5-years-old; head MRI every 3–5 years.
(c) Gastrinoma: Fasting serum gastrin, yearly after 20-years-old; abdominal CT or MRI every 3–5 years.

8. Multiple endocrine neoplasia type 2: Pathophysiology:
   (a) Caused by mutation in the *RET* proto-oncogene. Affects 1 in 30,000–50,000 people in the United States. Three subtypes MEN 2A or Sipple syndrome (70–80 % of the MEN 2 cases), FMTC- familial medullary thyroid carcinoma (10–20 % of the MEN 2 cases), and MEN 2B (5 % of the MEN 2 cases).
9. Clinical diagnosis:
   (a) MEN 2A: Medullary thyroid carcinoma, pheochromocytoma and parathyroid adenoma or hyperplasia.
   (b) MEN 2B: Mucosal neuromas, medullary thyroid carcinoma, medullated corneal nerve fibers, and Marfanoid body habitus.
10. Laboratory testing:
   (a) Molecular genetic testing: *RET* gene mutations.
   (b) Medullary thyroid carcinoma: Elevated plasma calcitonin.
   (c) Pheochromocytoma: Elevated norepinephrine, epinephrine, metanephrine and vanillylmandelic acid in plasma or 24-hour urine.
   (d) Parathyroid adenoma or hyperplasia: Elevated serum calcium or PTH.
11. Imaging:
   (a) Pheochromocytoma: CT, MRI, 18F-FDG PET and MIBG scintigraphy.
   (b) Parathyroid: ^{99m}Tc-sestamibi scintigraphy.
12. Natural history:
   (a) MEN 2A: 95 % medullary thyroid carcinoma, 50 % pheochromocytoma and 20–30 % parathyroid disease.
   (b) MEN 2B: 100 % medullary thyroid carcinoma, 50 % pheochromocytoma.
13. Treatment:
   (a) Medullary thyroid carcinoma: Total thyroidectomy with lymph node dissection before first year of life for high risk patients.
   (b) Pheochromocytoma: Adrenalectomy.
   (c) Parathyroid adenoma or hyperplasia: Parathyroidectomy, subtotal parathyroidectomy or total parathyroidectomy with autotransplantation.
14. Surveillance:
   (a) Medullary thyroid carcinoma: Annual serum calcitonin after prophylactic thyroidectomy.
   (b) Pheochromocytoma: Annual biochemical screening at age of 8 years for MEN 2B patients and MEN 2A with mutation of codons 630 and 634, and by age of 20 for all other codons; MRI and CT if abnormal laboratory results.
   (c) Parathyroid adenoma or hyperplasia: Annual biochemical screening at age of 8 for MEN.

Thyroid Nodules

Amber Batool

1. Pathophysiology:
   (a) A thyroid nodule is an abnormal growth of tissue that forms a lump within the thyroid gland. Thyroid nodules are rare in children and tend to be more malignant. Prevalence of thyroid nodules in children varies anywhere from 0.2 to 5.1 % and approximately 25 % of nodules tend to be malignant.
   (b) Risk factors for the development of malignant thyroid nodules include:
      (i) Family history of thyroid cancer (MEN syndromes, medullary thyroid cancer).
      (ii) Head and neck radiation exposure.
      (iii) Iodine deficiency.
2. Clinical presentation:
   (a) Children with thyroid nodules are more likely to present with a palpable neck mass. In addition, symptoms of hypo- or hyperthyroidisms such as weight loss or gain, palpitations, fatigue, and nervousness may be present. On rare occasions, symptoms of compression, shortness of breath, coughing, swallowing disturbances, or permanent change in voice may be evident.
3. Diagnosis:
   (a) History and physical examination:
      (i) A clinician should focus on eliciting information about symptoms of hypo- or hyperthyroidism, co-existence of thyroid disorders, history of malignancy, family history (MEN syndromes), and exposure to radiation. It is also important to inquire about the duration, growth, and

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signs of previous infection i.e. erythema, pain, and edema, because a nodule that varies in size over a period of time may be indicative of a cyst. Physical evaluation should focus on complete examination of the head and neck. A clinician should palpate the thyroid gland and assess for presence of nodules that should be further characterized as mobile or fixed, soft or firm, and for tenderness to palpation.

(b) Laboratory testing:

(i) Laboratory evaluation should include thyroid laboratory panel (TSH, T4, T3).
(ii) If medullary thyroid cancer is suspected, calcitonin and calcium levels should be obtained.

(c) Neck sonography: The thyroid ultrasound (US) is the modality of choice for assessing thyroid nodules. It is employed to measure the thyroid gland's size, characterize nodules based on presence of micro-calcification, echogenicity, vascular flow. In addition, it is important to identify and characterize any adenopathy that may be present since children with thyroid malignancies carry a higher risk of nodal metastases than adults.
(d) Fine needle aspiration biopsy (FNAB): Nodules that are greater than 1 cm, with cystic features, or those with suspicious US findings should be biopsied by US guided fine needle aspiration. Cystic fluid should be also sent for cytology.

4. Treatment:

(a) Cystic nodules: If FNAB reveals benign process, the patient should undergo US examination in 6 months and after that, yearly for 3 years. If the thyroid cyst recurs, diagnostic thyroid lobectomy with isthmusectomy should be pursued.
(b) Toxic nodules: In case of a solitary toxic nodule, thyroid lobectomy with isthmusectomy should be performed, whereas, patients with toxic multinodular goiter should undergo total thyroidectomy due to involvement of both lobes. Radioactive iodine therapy and anti-thyroid medications can also be employed to treat toxic nodules; however, it is associated with a higher failure rate and greater chance of hypothyroidism.
(c) Benign solitary nodule: Benign nodules based on US findings and/or FNA results usually do not require surgical therapy unless there are compressive symptoms or a history of head & neck irradiation. These patients should be followed with a neck US exam at 6 months and an annual US thereafter for 3–5 years.
(d) Malignant nodules: Thyroid nodules with features of malignant cytology usually require total thyroidectomy. In case of suspicious adenopathy, neck dissection is indicated. Serum TG and TG antibodies can be used to monitor for recurrence in patients after total thyroidectomy.

5. Outcomes: Complications of thyroid surgery include bleeding, infection, vocal cord paralysis secondary to injury to the recurrent laryngeal nerve, hypoparathyroidism, thyrotoxic storm, injury to superior laryngeal nerve, and hypothyroidism.

Breast Nodules

Meng-Fey Ferra Lin-Duffy

Breast nodules: The majority of the breast masses in children are benign. However, when a breast mass is found in a child or adolescent, it often causes anxiety in patients and their parents. In prepubertal children, most common cause is unilateral or bilateral thelarche, which does not require any treatment. Breast nodules in adolescents will be discussed here.

1. Pathophysiology:
   (a) Fibrocystic change:
      (i) More than 50 % of women in reproductive age experience fibrocystic changes.
      (ii) It is thought to result from an imbalance between estrogen and progesterone.
   (b) Fibroadenoma:
      (i) These are the most common breast lesions in adolescents.
      (ii) They are composed of fibrous and glandular tissue.
      (iii) It is very rare that fibroadenomas progress to malignancy.
   (c) Mastitis:
      (i) Mastitis and breast abscesses are more common in infants younger than two months old than in children or adolescents.
      (ii) Mastitis occurs in the same frequency in girls and boys in their first two weeks of age.
      (iii) Mastitis is twice as common in girls than in boys after two weeks of age.

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(iv) This is possibly due to the longer duration of physiologic breast hypertrophy in females.
(v) In older children or adolescents, it occurs less frequently.
(vi) Possible causes are trauma (nipple piercing, plucking periareolar hair, shaving or manipulation of breast tissue during sexual activity), obesity, skin infection, mammary ectasia or epidermoid cysts.
(vii) Staphylococcus aureus is the most common pathogen.

(d) Phyllodes tumor:

(i) These are rare tumors of the breasts. They are mostly found in middle-aged women but can be found in children as young as 10-years-old.
(ii) It contains two types of breast tissue: Stromal (connective) tissue and glandular (lobule and duct) tissue.
(iii) It can be classified as benign (85–90 %), malignant (10–15 %) or borderline.
(iv) Benign tumors have a tendency to grow aggressively and can recur locally.
(v) Less than 5 % metastasizes and can metastasize to the lung, mediastinum and skeleton.

2. Clinical presentation:

(a) Fibrocystic change:

(i) It is painful breast tissue before menses and improves during menstruation.
(ii) Fibrotic tissue can be palpated and most commonly found in the upper outer quadrant of the breasts.

(b) Fibroadenoma:

(i) They are typically asymptomatic but may cause discomfort before menses.
(ii) On examination, they are rubbery, mobile and well-circumscribed.
(iii) The average size is 2–3 cm.
(iv) Most frequent location is upper outer quadrant but can be anywhere.
(v) They can be recurrent or multiple in 10–25 % of cases.

(c) Mastitis:

(i) Pain and fever.
(ii) Swelling, erythema, warmth, tenderness, induration and/or fluctuance of the breast.

(d) Phyllodes tumor:

(i) Patients usually present with a large painless breast mass that grows rapidly.
(ii) The overlying skin may be stretched from the rapid growth.
(iii) On physical examination, it presents as a firm, mobile, well-circumscribed and nontender mass.

3. Diagnosis:

   (a) Although malignant tumors in the breasts are extremely rare in adolescents, careful history and physical examination is necessary to rule out any life threatening processes. Most adolescents with a breast mass have normal physiologic breast tissue or fibrocystic changes. Once malignancy is ruled out, the mass can be observed through one or two menstrual cycles for any physiological changes.

   (b) History:

      (i) Onset and duration of the mass.
      (ii) Size and changes in size of the mass.
      (iii) Menstrual history.
      (iv) Associated symptoms including pain, nipple discharge or skin changes.
      (v) Past medical history including previous breast disease or malignancy and history of chest radiation.
      (vi) Pregnancy history.
      (vii) Medication history including oral contraceptive medications.
      (viii) Family history.

   (c) Examination:

      (i) Size: Should be monitored through the menstrual cycle.
      (ii) Consistency: cystic vs. solid.
      (iii) Mobility.
      (iv) Tenderness.
      (v) Skin changes.
      (vi) Nipple discharge.
      (vii) Appearance of the nipple: nipple retraction can be congenital or due to underlying malignancy.
      (viii) Lymphadenopathy.
      (ix) Hepatosplenomegaly

   (d) Imaging:

      (i) Ultrasonography is preferred due to dense breast tissue in adolescents. It may be necessary if the mass persists for more than one or two menstrual cycles.
      (ii) Ultrasound is helpful in the diagnosis of fibrocystic change.
      (iii) Ultrasound of fibroadenoma reveals a solid, avascular and well circumscribed mass.
      (iv) Ultrasound findings suggestive of phyllodes tumors include lobulations, a heterogeneous echo pattern, and an absence of microcalcifications.

   (e) If mastitis is suspected, Gram stain and culture confirm the diagnosis if aspiration or incision and drainage performed. In infants less than two months old, complete blood count with differential and blood culture should be obtained prior to start of antimicrobial therapy.

(f) It is hard to differentiate phyllodes tumor from fibroadenomas on imaging studies, fine needle biopsy or even core biopsy.
(g) Excisional biopsy for smaller lesions and incisional biopsy for larger lesions are the definitive method for diagnosis.

1. Treatment:

(a) General guidelines:

(i) Cystic lesions typically resolve spontaneously over weeks to months.
(ii) Aspiration can be performed for persistent cystic lesions. Fluid other than clear should be sent for cytology. A follow-up ultrasound in three months should be obtained for re-evaluation.
(iii) Suspicious solid lesions or cyst lesions that do not resolve with aspiration require excisional biopsy.

(b) Fibrocystic change:

(i) Mild analgesia such as nonsteroidal anti-inflammatory agents relieves pain.
(ii) Oral contraceptives with low dose ethinyl estradiol can improve symptoms in 70–90 % of patients.
(iii) Decreased caffeine intake might improve symptoms in some patients.

(c) Fibroadenoma:

(i) Treatment is reassurance and careful follow-up initially as most fibroadenomas decrease in size in adolescents.
(ii) Fibroadenomas less than 5 cm without concerning features can be observed every 1–2 month for growth or regression. If it is regressing, the interval can be lengthened.
(iii) The decision to excise depends on the patient's age, level of anxiety and family history of breast cancer. When the patient approaches adulthood, the risk of breast cancer increases and therefore excisional biopsy is recommended.
(iv) If there's growth of the lesion, size bigger than 5 cm, lesion persists into adulthood or has any concerning features, such as nonmobile, hard, associated skin changes or adenopathy, excisional biopsy should be performed.

(d) Mastitis

(i) Infants with mastitis should be treated with parenteral antibiotics initially regardless of temperature or white blood cell count as simple mastitis tend to progress to abscess when infants are treated with oral antibiotics initially.
(ii) Older children and adolescents without systemic symptoms can be treated with oral antibiotics. Empiric antibiotics with coverage for Staphylococcus and Streptococcus should be administered for

7–10 days. Clindamycin or trimethoprim-sulfamethoxazole may be necessary for areas with high incidence of MRSA.

(iii) Breast abscesses may require aspiration or incision and drainage unless spontaneous drainage occurs. Extra care should be taken to avoid damaging the breast bud as it can create a defect in the future.

(e) Phyllodes tumor:

(i) Complete excision with clear margin is required as recurrence rate is high.
(ii) In recurrent diseases, further local excision or total mastectomy is usually curative.
(iii) There is no proven role for adjuvant chemotherapy or radiation therapy.
(iv) Prognosis is excellent in benign tumors after adequate local excision.
(v) Regular short-interval follow-ups should be done to detect possible local recurrences.

Adrenal Tumors

Alysia A. Agnoni

In children, adrenocortical tumors of 1 cm or larger are rare but can be incidentally found during evaluation for a non-related issue. For this reason, they have earned the nickname "adrenal incidentalomas".

1. Pathophysiology:
   (a) Anatomy and physiology: The adrenal glands are small golden endocrine organs that reside along the superior medial aspect of the kidneys. They are composed of an outer cortex and an inner medulla.
      (i) The adrenal cortex is comprised of three separate zones that each function independently
         1. Zona glomerulosa: Secretes aldosterone
         2. Zona fasciculata: Secretes cortisol
         3. Zona reticularis: Secretes androgens
      (ii) The adrenal medulla secretes catecholamines: Epinephrine, norepinephrine.
   (b) Causes of adrenal masses include: Neuroblastoma, adrenal adenoma, adrenocortical carcinoma, pheochromocytoma, adrenal metastases, adrenal hyperplasia, adrenal cysts, and adrenal hemorrhage.
      (i) Adrenal masses other than neuroblastoma are very rare in the pediatric population.

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(c) The majority of adrenal incidentalomas in children are not hormonally active. The adrenal mass that is hormonally active likely produces symptoms which leads to its discovery on CT scan. These patients may present, to varying degrees, with signs of virilization, hypercortisolism, or subclinical Cushing's syndrome identified through lab work-up.
(d) Adrenal tumors have been associated with the following syndromes:

(i) Beckwith-Weidemann, Li-Fraumeni, familial adenomatous polyposis.

2. Diagnosis:

(a) History and physical examination:

(i) History: Palpitations, sweating, headaches.

(ii) Physical: Hirsutism, penile or clitoral enlargement, Cushingoid features, abdominal pain, hypertension, hypokalemia.

(b) Labs:

(i) 24 hour urine cortisol, midnight cortisol level, baseline corticotropin, high dose dexamethasone suppression.
(ii) Plasma aldosterone and renin.
(iii) 24 hour urinary metanephrines and catecholamines.
(iv) Estradiol, testosterone, urinary and plasma DHEA, and 17-ketosteroid.

(c) Imaging:

(i) CT scan: Most commonly identifies an adrenal mass during work-up for an unrelated problem. CT, in many cases, cannot differentiate an adrenal carcinoma from a benign adrenal adenoma. General features of adrenal masses on CT imaging are listed below but are typically not diagnostic:

1. Benign adrenocortical adenomas: Low unenhanced CT attenuation value. Appear round and homogenous, often unilateral, and are usually less than 4 cm.
2. Adrenocortical carcinoma: Increased unenhanced CT attenuation value. Irregular shape and heterogeneous density due to necrosis and calcifications, often unilateral, and can be greater than 4 cm.
3. Adrenal metastases: Increased unenhanced CT attenuation value. Appear like adrenocortical carcinoma in the presence of known or newly discovered distant disease and may be bilateral.
4. Pheochromocytomas: Increased unenhanced CT attenuation value with increased vascularity.

(ii) MRI: May be used to further differentiate a benign adrenal mass from a malignancy.
(iii) PET: Can be used in those patients with known history of malignancy to identify new malignancy.

3. Treatment:
   (a) Imaging and lab evaluation do not always indicate a definite diagnosis and clinical judgment regarding surgical excision is important.
   (b) Indications for observation:
      (i) Tumors small in size, uniform, and hypodense can be considered for observation. There are no studies indicating the optimal duration and frequency of follow-up for adrenal incidentalomas.
         1. Repeat imaging can be done at 3–6 months after diagnosis to assess for growth/change.
   (c) Indications for surgical excision:
      (i) Documented pheochromocytoma, hormone-secreting tumors, adrenocortical carcinoma, and metastatic disease in patients with known malignancy.
      (ii) Any lesions suspicious for adrenocortical carcinoma or greater than 4 cm.
   (d) Adrenalectomy: Despite lack of defined protocols for management of an adrenal incidentaloma, the general consensus is that if there is suspicion for malignancy, adrenalectomy is the standard of care.
      (i) Laparoscopic: mostly commonly done transabdominally although the more technically demanding retroperitoneoscopic adrenalectomy is becoming more common.
         1. Indications: Small tumors.
         2. Laparoscopic transabdominal adrenalectomy:
            (a) Patient is placed in the lateral decubitus position
            (b) Three to four port sites are required, depending on which side is being operated on, and these are placed subcostally and midline.
            (c) Right adrenalectomy:
               (i) The liver and colon are retracted medially to allow exposure.
               (ii) Gerota's fascia is incised and dissected superiorly to the diaphragm.
               (iii) Dissection through the perirenal adipose tissue is carried on medially until the IVC is identified. The right adrenal vein is along the lateral border of the IVC – this is clipped or suture ligated. Care is taken to avoid injury to the IVC.
               (iv) Ligation of the adrenal arteries arising from the aorta, phrenic, and renal arteries.
               (v) Dissection of the adrenal gland from the kidney is taken on at this point: care is again taken not to injure the IVC, kidney, and tumor capsule.
               (vi) Adrenal gland is placed in the endoscopic bag and removed.

(d) Left adrenalectomy:

(i) Mobilization of the splenic flexure and spleen. In some cases the body and tail of the pancreas can be mobilized superiorly.
(ii) Gerota's fascia is incised.
(iii) The left adrenal vein is identified and ligated. The small adrenal branches of the left suprarenal and phrenic arteries are ligated.
(iv) Dissection of the adrenal gland from the medial edge of the kidney and diaphragm, ligating any small vessels.
(v) Place the adrenal gland in the endoscopic bag and remove.

3. Retroperitoneal laparoscopic approach:

(a) Patient is positioned prone or slightly turned onto the contralateral side.
(b) Three port sides are placed, all inferior to the 12th rib.
(c) Exposure of the superior pole of the kidney is undertaken next by carefully opening Gerota's fascia and dissecting the adrenal gland and perinephric fat from the kidney.
(d) Ligation of the adrenal arteries and veins is carried out next using clips and electrocautery.
(e) The adrenal gland is then dissected from the peritoneum and extracted.

(ii) Open: The transabdominal approach is the most common technique although open retroperitoneal approach is an alternative.

1. Indications: High suspicion of or confirmed adrenocortical carcinoma, large tumor, invasion of mass into surrounding structures, or bilateral masses.
2. Open transabdominal adrenalectomy:

(a) Patient is placed in the supine position with the operative flank side elevated with a cushion.
(b) An upper midline or subcostal incision is made.
(c) The mobilization of abdominal organs, dissection, and removal of the adrenal gland is carried out in much the same manner as discussed above in the description of the laparoscopic technique.

4. Outcome:

(a) For benign adenomas surgical resection is curative.
(b) Postoperative endocrine hormonal and steroid management may be necessary in cases of bilateral adrenalectomy or after excision of hormonally active tumors.

Pancreatitis

Amber Batool

Pancreatitis is characterized by inflammation of the pancreas. It is classified as acute, chronic, necrotic, or hemorrhagic. Acute pancreatitis results from ectopic activation of pancreatic enzymes. Chronic pancreatitis is defined as a continuing inflammatory process of the pancreas, characterized by irreversible morphologic changes (e.g. calcifications, fibrosis, ductal stricture, dilatation), which may lead to exocrine and endocrine insufficiency.

1. Epidemiology:

   (a) Pancreatitis is an uncommon disease in pediatric population and the etiology tends to be more diverse compared to Adults. Males and females are equally affected. Most cases of acute and chronic pancreatitis are idiopathic. The most common identifiable etiologies of acute pancreatitis are listed below:

      (i) Abdominal trauma.
      (ii) Anomalies of pancreaticobiliary system: Pancreas divisum, Annular pancreas, Choledochal cyst.
      (iii) Biliary disease: Gallstone pancreatitis.
      (iv) Multisystem disease.
      (v) Drugs and toxins: L-asparginase, Valproic acid, Azathioprine, Mercaptopurine, Mesalamine.
      (vi) Viral infection.
      (vii) Hereditary disorders: Cystic fibrosis transmembrane conductance regulator (CFTR) mutation, Hereditary pancreatitis.
      (viii) Autoimmune pancreatitis.
      (ix) Metabolic disorders

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2. Clinical features: Classically, pancreatitis presents as an acute onset of epigastric pain with radiation to the back. It may be accompanied with nausea, vomiting, abdominal distention, ileus, anorexia, jaundice (with gallstones or obstruction), fever, tachycardia, and hypotension.
   (a) Upon physical examination, one may find signs of abdominal trauma, tenderness, distention, or peritoneal signs. In a case of hemorrhagic pancreatitis, Grey Turner's sign (bluish discoloration of flanks) and/or Cullen's sign (bluish discoloration in the peri-umbilical region) may be present.
   (b) Chronic pancreatitis may mimic acute pancreatitis. It usually presents as chronic abdominal pain that is difficult to treat. It may be accompanied with steatorrhea, growth failure, and pancreatic exocrine and endocrine insufficiency.
3. Diagnosis:
   (a) If pancreatitis is suspected based on history and physical examination, amylase and lipase should be measured. Serum amylase and lipase (more specific) tend to be at least three times greater than the upper limit of the normal. Other laboratory findings include leukocytosis, hyperglycemia, glycosuria, hypocalcaemia, hypertriglyceridemia, acidosis, hypoalbuminemia, hyperbilirubinemia, and elevated LFT's.
   (b) Imaging studies provide evidence of structural changes in the glands or ducts. Trans-abdominal ultrasonography, CT, MRCP, ERCP, and EUS may be utilized and provide evidence of acute vs. chronic changes.
   (c) US is a primary screening tool and its findings may reveal a diffusely enlarged or edematous pancreas with dilated pancreatic ducts, peri-pancreatic fluid collection, an abscess or a pseudocyst.
   (d) CT scan may be utilized for evaluation of abnormalities on US, chronic pancreatitis, pancreatic necrosis, complications, pancreatic trauma, or neoplastic disease process. CT scan findings include an enlarged gland with ill-defined margins, peripancreatic fluid, areas of decreased or enhanced density, or pseudocysts.
   (e) Magnetic resonance cholangiopancreatography (MRCP) is a non-invasive imaging study of biliary tree and surrounding structures that helps delineate an anatomical lesion. It is more sensitive than CT scan.
   (f) Endoscopic retrograde cholangiopancreatography (ERCP) can help diagnose various pancreatic and biliary anomalies, ductal abnormalities or obstructions and also serve as a therapeutic intervention, i.e. sphincterotomy or stent placement.
4. Treatment:
   (a) The management of acute pancreatitis usually consists of supportive medical therapy with IVF hydration, pain control, and bowel rest. The initial management is directed at stabilizing the patient and aggressive hydration to help decrease the risk of multi-organ failure. It is important to optimize

nutritional status with early institution of nutrition. Oral feeding can be started within 24–48 h after admission. In cases of severe pancreatitis, enteral feeding or TPN can be employed. Jejunal feeds are preferred over TPN due to lower complication rates and lower cost. In case of chronic pancreatitis, low-fat meals, pancreatic enzyme supplements may help restore exocrine enzyme deficiencies.

(b) Surgical management is rarely required and it is reserved for chronic, relapsing pancreatitis, unsuccessful conservative medical therapy, intractable pain, impaired nutrition, and narcotic addiction. Surgical options include distal pancreatectomy with Roux-en-Y pancreaticojejunostomy (i.e., Duval procedure), lateral pancreaticojejunostomy (i.e., Puestow procedure), or ERCP sphincteroplasty.

5. Complications: Pancreatitis is associated with a variety of complications. The most common are pseudocyst formation and mechanical obstruction of the duodenum and common bile duct. Less frequent complications include pancreatic ascites, pleural effusion, splenic vein thrombosis with portal hypertension, and splenic artery pseudoaneurysms.

Gallbladder Disease

Christopher P. Coppola

Laparoscopic cholecystectomy for pediatric patients is increasing in incidence. A major reason for this is detection of biliary dyskinesia through use of HIDA scan. Children have gallstones less frequently than adults, but when they do, pigment stones, rather than cholesterol stones, are more common.

1. Pathophysiology:

   (a) Epidemiology: pediatric biliary disease in increasing in incidence [1]. Approximately 2 % of children have gallstones, and a greater number have biliary disease without stones. After puberty females have quadruple the incidence of gallstones, compared to males.
   (b) Cholelithiasis: presence of gallstones can be asymptomatic or associated with abdominal pain. Bile is usually liquid, but unbalanced proportions of cholesterol, bile salts, and lecithin lead to precipitation of gallstones. Stress, shock, and dehydration can lead to concentration of bile and gallstones.
   (c) Types of gallstones: the color reflects content, and usually there will be one type of stone present. Cholesterol stones are most common in adults, but children have greater incidence of pigment stones.

      (i) Black (48 %): pigment stones (calcium bilirubinate) present in hemolysis and parenteral nutrition.
      (ii) Yellow (21 %): from supersaturation of bile with cholesterol.
      (iii) Protein stones (5 %).
      (iv) Calcium carbonate (4 %).
      (v) Brown (3 %): associated with cholangitis and bile stasis, made of bilirubin, fatty acids, and calcium.

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(d) Cholecystitis (inflammation of the gallbladder): occurs when gallstone (or other process) obstructs the cystic duct, creating pressure, stasis, distention, vascular congestion, edema, and ischemia of gallbladder.
(e) Gallstone pancreatitis: occurs when stones from gallbladder pass through common bile duct and obstruct pancreatic duct.
(f) Biliary dyskinesia: occurs when there is impaired or painful emptying of the gallbladder after a meal without the presence of gallstones. The etiology of this process is not known, but may be due to a long cystic duct, tortuous valves in duct, low junction of the cystic duct and common bile duct, or a visceral hypersensitivity of the gallbladder.
(g) Associated conditions:
    (i) Increased incidence of cholelithiasis is seen in hemolytic disease, obesity, use of parenteral nutrition, after ileal resection, Crohn's disease, shock/sepsis, and pregnancy [2].
    (ii) Patients with sickle cell disease have double the incidence of gallstones. Half of children with sickle cell disease have gallstones by the time they are adults. The gallstones seen in sickle cell disease are pigmented stones.
    (iii) Spherocytosis with increased turnover of red blood cells leads to pigment stones.
    (iv) Inherited conditions predisposing to cholelithiasis (cholesterol stones) are progressive familiar intrahepatic cholestasis, Type 3 and defect in ABCB4 gene.

2. Clinical presentation:

(a) Biliary colic: a sharp stabbing pain in the right upper quadrant that comes and goes. Pain may be related to dietary intake. It can be associated with nausea and symptoms of reflux. Biliary colic can accompany gallstones and biliary dyskinesia. An attack of pain lasting greater than 6 h is suggestive of cholecystitis.
(b) Patients may also have avoidance of fatty foods, anorexia, and weight loss as they learn that certain foods worsen their symptoms.
(c) Many patients with cholesterol stones are obese.
(d) Charcot's triad: jaundice, fever, and right upper quadrant pain: suggests choledocholithiasis.
(e) Reynolds' pentad: components of Charcot's triad plus shock and altered mental status, suggesting ascending cholangitis.
(f) Murphy's sign: this physical exam finding is elicited by palpating the liver edge under the right costal margin. The patient is asked to inhale deeply, and if this causes right upper quadrant pain, Murphy's sign is present. A sonographic Murphy's sign is when the sonographer compresses the gallbladder with the ultrasound probe and elicits pain.

3. Diagnosis

(a) Abdominal sonography (specifically the right upper quadrant): can detect gallstones and choledocholithiasis, can detect cholecystitis in the form of pericholecystic fluid and thickening of the gallbladder wall, can detect

enlargement of the common bile duct or the intrahepatic bile ducts, which indicates distal obstruction. Sonography is best when patients are NPO because there is less bowel gas present to interfere with ultrasound transmission. Sonography can also detect alternate pathology.

(b) Lab tests: patients presenting with right upper quadrant pain should undergo complete blood count to detect leukocytosis, electrolytes to detect dehydration and electrolyte abnormalities, liver function tests and gamma glutamyl transferase to detect hyperbilirubinemia, hepatitis, and elevated alkaline phosphatase. Testing amylase and lipase will detect pancreatitis, and a urinalysis will differentiate a urinary tract infection or urinary stone from biliary disease.

(c) Abdominal x-ray is useful to discern bowel gas pattern when patients are vomiting, and rarely gallstones with high calcium content will be visible.

(d) Magnetic resonance cholangiopancreatography (MRCP) can identify biliary ductal obstruction, when lab tests show hyperbilirubinemia or pancreatitis. Younger children will require sedation or an anesthetic to accomplish MRCP.

(e) Hepatobiliary iminodiacetic acid (HIDA) scan with gallbladder ejection fraction (GBEF): HIDA scan will demonstrate ability of the liver to conjugate bile, patency of the cystic duct, and patency of the common bile duct. Additionally, a gallbladder ejection fraction (percentage of tracer in gallbladder excreted 1 h after gallbladder is stimulated with cholecystokinin analog) can be measured. Values less than 40 % are considered diagnostic of biliary dyskinesia. Values >75 % associated with pain may indicate a hyperkinetic gallbladder, and even if the GBEF is normal, if the patient has pain with cholecystokinin analog administration that mimics pain they feel with meals, they may benefit from cholecystectomy.

(f) Computed tomography: not typically used to diagnose biliary disease and carries the risk of ionizing radiation. However, gallstones can be visible on CT scan.

4. Initial stabilization:

(a) Patients with cholangitis and or pancreatitis are in danger of sepsis and shock so need immediate fluid resuscitation and antibiotics. Some may require intensive care unit monitoring.

(b) Patients with cholecystitis need fluid and antibiotics.

(c) Pain control is given for the biliary colic.

(d) Patients who have evidence of biliary ductal obstruction or gallstone pancreatitis need to have the common bile duct cleared. For many patients, the choledocholithiasis will clear spontaneously; this will be evident in improving symptoms and decreasing serum bilirubin.

(e) Patients with intermittent pain, who are afebrile, and can tolerate oral hydration can be scheduled for elective cholecystectomy as an outpatient.

5. Treatment:

(a) Patients with biliary colic can be considered for a trial of medication for alternate possible diagnoses, such as histamine 2 blockers, proton pump inhibitors, laxatives, and antispasmodics, but if the cause of the pain is biliary, these are unlikely to help.

(b) Asymptomatic gallstones in children can be observed, but should be checked for resolution with sonography in 6 months. Ursodeoxycholic acid can be administered. If there is a known underlying condition responsible for the gallstones, such as hemolytic disease, they are unlikely to resolve. Gallstones in children younger than eight without a known etiology are more likely to resolve. If at any point the gallstones become symptomatic, or if sequelae such as cholecystitis or obstruction occur, observation should be abandoned.
(c) Laparoscopic cholecystectomy is indicated for cholecystitis, symptomatic gallstones, and biliary dyskinesia. If there is any question of choledocholithiasis at the time of cholecystectomy, an intraoperative cholangiogram can be performed. Patients undergo cholecystectomy as an outpatient. Patients with sickle cell anemia should be admitted the day before surgery for hydration and transfusion when needed. After surgery they should be well hydrated to prevent a sickling crisis from the stress and fluid shifts of operation.
(d) Laparoscopic cholecystectomy can be converted to open cholecystectomy if needed for safe completion of the operation.

6. Outcomes:

(a) Possible complications of cholecystectomy: common bile duct injury, hemorrhage, bile leak, bileoma, diarrhea, gas.
(b) Nearly all patients with symptomatic gallstones have resolution of symptoms while 60–80 % of patients with acalculous biliary colic and biliary dyskinesia are improved with cholecystectomy.

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Varicocele

Jared M. Bieniek and Joel M. Sumfest

A varicocele represents dilated internal spermatic veins within the spermatic cord which can be palpated on physical exam.

1. Pathophysiology:
   (a) Incidence: 15 % of the adult male population has a varicocele which is often asymptomatic.
      (i) Varicoceles are typically seen around the onset of puberty and are noted with increasing incidence with increasing Tanner stage.
      (ii) Most varicoceles are left-sided or bilateral.
   (b) Pathophysiology thought to be related to multiple etiologies.
      (i) Angle of insertion of left gonadal vein into renal vein.
      (ii) Elevated hydrostatic pressure (longer left gonadal vein).
      (iii) Incompetence of venous valves.
   (c) Natural history may range from asymptomatic to symptomatic with testicular atrophy and subfertility.
      (i) Effects are thought to be secondary to elevated scrotal temperature with varicocele but exact reason remains unclear.

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2. Clinical presentation:
   (a) Often incidental finding on physical exam, especially sports physicals.
   (b) Testicular pain (uncommon).
   (c) Scrotal/testicular masses, swelling, or asymmetry.
3. Physical examination:
   (a) Scrotal examination:
      (i) Spermatic cord:
         1. Palpate each spermatic cord in upper scrotum for varicocele (may feel like a "bag of worms" on exam).
         2. Must be examined in both standing and supine positions for varicocele swelling and resolution.
            (a) Grade I: Palpable varicocele with Valsalva/standing only.
            (b) Grade II: Easily palpable varicocele but not visible.
            (c) Grade III: Varicocele visible before examination.
      (ii) Testicles:
         1. Examine each testis for tenderness, masses, and size.
4. Evaluation:
   (a) Laboratory:
      (i) In young adult males (typically over 21), may consider semen analysis to estimate fertility potential.
         1. Normal parameters not established for adolescents making interpretation difficult.
         2. Significant variability between specimens obtained through puberty and early post-puberty.
   (b) Imaging:
      (i) Scrotal ultrasonography (US) allows easy visualization of spermatic cord vessels and the testicles.
         1. In adult population, varicose spermatic veins often diagnosed on color Doppler US with diameter >3 mm with Valsalva.
         2. Each testicle volume should be calculated using Lambert formula and compared to contralateral side.
            (a) Significant testicular hypotrophy variously defined as 10–20 % difference in volume.
            (b) Testicular hypotrophy inversely correlated with semen parameters in some series [1].

(ii) If varicocele is right-sided only or does not decompress when supine and relaxed, must evaluate the abdomen/retroperitoneum with CT or US to rule out a mass compressing the gonadal vein.

5. Management:

(a) Observation:

(i) Significant testicular differential growth has been shown to occur during puberty; therefore testicular measurements should be trended over time [2].
(ii) Up to 82 % of adult men with varicoceles are able to father children suggesting that observation may be practical [3].

(b) Surgical:

(i) In cases of persistent >20 % left or bilateral testicular hypotrophy, pain, or abnormal semen analysis, surgery may be warranted.
(ii) Options for surgical repair.

1. Microscopic subinguinal/inguinal approach.

(a) Artery and lymphatic-sparing technique with ligation of several internal spermatic veins.
(b) Classic description includes delivery of the testicle with division of gubernacular veins though not all surgeons perform this step.
(c) Complications include low rate of hydrocele formation and testicular necrosis.

2. Open/laparoscopic suprainguinal approach.

(a) Testicular vessels are dissected out above the vas and ligated (usually single testicular vein at this level).
(b) Higher rates of post-operative hydrocele owing to ligation of lymphatics.
(c) Artery and lymphatic-sparing techniques are possible.

(iii) Testicular catch-up growth has been noted in a majority of patients following surgical repair [4].

(c) Endovascular:

(i) Sclerotherapy can be performed in retrograde or antegrade fashion.

1. Retrograde embolization performed by interventional radiologist with sclerosing agents or coils.
2. Antegrade sclerosis performed by direct injection into internal spermatic veins after isolation through subinguinal incision.

(ii) While lower success rates than surgical repair, sclerotherapy may be an option especially for recurrent varicoceles.

6. Follow-up:
   (a) Should follow patients managed with observation or surgical repair with serial scrotal US to trend testicular volumes.
   (b) If patient develops symptomatic post-operative hydrocele, may repair via normal approach.
   (c) If varicocele recurs or does not improve, consider more proximal ligation (suprainguinal approach) or sclerotherapy.
   (d) As patients get older, may evaluate semen analysis to better estimate fertility potential though paternity is the ultimate goal and only modestly predicted by semen parameters.

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Acute Scrotum

Jared M. Bieniek and Joel M. Sumfest

Acute pediatric scrotum, caused by a variety of etiologies, is the presentation of new onset scrotal pain, tenderness, or swelling in children less than 18 years-old.

1. Pathophysiology:
   (a) Testicular torsion: All acute scrotum presentations should be considered testicular torsion until proven otherwise.
   (b) Appendix testis/epididymis torsion.
   (c) Epididymitis/orchitis.
   (d) Cellulitis/Fournier's gangrene.
   (e) Inguinal hernia/hydrocele.
   (f) Varicocele.
   (g) Traumatic contusion, hematocele, or rupture.
   (h) Testicular/scrotal mass.
   (i) Referred pain (e.g. ureteral obstruction).
   (j) Other: idiopathic scrotal edema, insect bite, vasculitis.
2. Clinical presentation:
   (a) Age:
      (i) Infancy to puberty: Appendage torsion most common.
      (ii) Perinatal and pubertal periods: Epididymitis and testicular torsion common.

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(b) Onset, quality, duration.
(c) Precipitating/relieving factors: Relation to activity/trauma.
(d) Prior similar episodes.
(e) Associated symptoms such as fever, nausea/vomiting, hematuria, dysuria, discharge.
(f) Recent illnesses.
(g) Medical and surgical history.
(h) Sexual history.

3. Physical examination:

(a) Symptoms may be referred to the scrotum therefore evaluation should include complete abdominal and back/flank examination as well as genito-urinary examination.
(b) Penile examination for circumcision status or other meatal abnormalities which may predispose to urinary infections.
(c) Scrotal exam:

(i) Skin:

1. Examine for erythema, bruising, warmth, tenderness, swelling, crepitus, and discharge.
2. Local erythema, warmth, crepitus, or discharge raises concern for a scrotal skin/subcutaneous infection.
3. Cremasteric reflex: Normal reflexive retraction of the hemiscrotum with light stroke of the ipsilateral inner thigh in patients >2-years-old. Absence of this reflex raises concern for spermatic cord torsion.
4. Blue dot sign: Torsion of testicular appendage may produce blue discoloration seen through scrotal skin stretched over upper pole of the affected testicle.

(ii) Testicles:

1. Examine normal and affected side for tenderness, firmness, size, position, lie, and masses.

(a) Differentiate tenderness of testicle vs. epididymis or both.
(b) Firm testicle, horizontal lie, and elevated position in scrotum are all concerning signs for testicular torsion.
(c) Loss of testicular contour with history of trauma may indicate rupture of the tunica albuginea.
(d) Testicular masses rarely cause pain but this may represent initial symptom.

(iii) Spermatic cord:

1. Check for palpable mass coming down inguinal canal which may indicate inguinal hernia; check for reducibility.
2. If “bag of worms” appreciated within spermatic cord, evaluate patient in standing and lying positions for presence and resolution of varicocele.

4. Diagnosis:
   (a) Imaging:
      (i) Scrotal ultrasonography (US) is easily performed and reveals critical information about testicular size, homogeneity, blood flow, presence of fluid collections, and masses.
         1. Lack of blood flow within testicular parenchyma or weak flow only seen peripherally concerning for testicular torsion.
            (a) Sensitivity of color Doppler US for low/absent blood flow in torsion cases is only 63–90 % in studies [1].
            (b) May be difficult to perform and interpret in small children.
         2. Heterogeneous parenchyma throughout testicle also concerning for necrosis associated with testicular torsion.
         3. Enlarged testicular appendage with or without flow may or may not be seen in cases of appendix torsion.
         4. Increased size of/blood flow to epididymis suggestive of epididymitis.
         5. Scrotal/testicular abscess may appear as heterogeneous fluid collection with or without septations.
         6. Hematocele or testicular rupture can be seen following trauma.
      (ii) Abdominal x-ray or CT may reveal air in the scrotum if concerned for Fournier's gangrene.
      (iii) Consider renal US if history and physical suggestive of ureteral obstruction.
   (b) Laboratory data.
      (i) Urinalysis to examine for presence of bacteria, WBCs, leukocyte esterase, and/or nitrites as possible indicators of a urinary infection.
      (ii) Possible CBC to evaluate for leukocytosis.
      (iii) Consider sexually-transmitted disease testing in the sexually active male.
      (iv) Tumor markers (human chorionic gonadotropin, alpha fetoprotein, and lactate dehydrogenase) should be ordered in the patient with a testicular mass.
   (c) Further information regarding evaluation and management of inguinal hernias, hydroceles, and varicoceles can be found in their respective chapters.
5. Treatment:
   (a) If clinical concern is high for testicular torsion, patient should undergo emergent scrotal exploration though US is commonly performed initially due to its rapid availability.
      (i) A testicle that has undergone torsion should be untwisted and allowed several minutes for flow to return to determine viability; may use intraoperative Doppler to determine viability if color does not completely return to the testicle.

1. Viability related to duration of torsion with most testicles salvageable if duration <6–8 h [2].
2. If testicle viable, it should be fixated in multiple points to scrotal fascia.
3. If non-viable, simple scrotal orchiectomy should be performed.

(ii) While waiting to assess affected side, contralateral testicle should be exposed, examined, and fixated to prevent possible future torsion on that side.

(b) Alternatively for testicular torsion, bedside manual detorsion can be attempted though sedation may be required.

(i) Testicle should be twisted as if "opening a book". Rotate left side counter-clockwise and right side clockwise.
(ii) Series have demonstrated that testicular torsion may occur in non-traditional rotational direction in up to one-third of cases [3].

(c) In perinatal cases of testicular torsion, the rotation of the spermatic cord is typically extravaginal and present for extended period of time (likely occurred before birth).

(i) Very low chance of testicular salvage, therefore management deferred until patient stable and able to tolerate anesthetic for scrotal exploration.
(ii) Rare occurrences where exam was initially normal and acutely changed with findings of no blood flow should be explored emergently.

(d) With normal US findings during acute presentation but history suggestive of previous testicular torsion/detorsion episodes, may consider prophylactic bilateral scrotal orchidopexy.
(e) Appendix testis torsion may be a diagnosis of exclusion and can be treated conservatively with rest and anti-inflammatories.
(f) Cases of epididymitis/orchitis without abscess formation may also be treated conservatively.

(i) Antibiotics should only be used if clinical history or lab evaluation concerning for urinary infection (e.g. history of urinary tract infections, suspicious urinalysis).

1. Start with empiric antibiotics and adjust per urine culture sensitivities.

(ii) Empiric treatment for chlamydia/gonorrhea should be considered if the patient is sexually active.
(iii) Non-bacterial epididymitis (idiopathic or viral) treated conservatively with rest and anti-inflammatories.

(g) If concern for scrotal skin/subcutaneous infection, treatment should be directed at the severity and presence/absence of abscess.

(i) Cellulitis alone should be treated with antibiotics.

(ii) Scrotal wall abscess may be debrided at bedside or in the OR based on patient tolerability and abscess complexity.

(h) With a diagnosis of strangulated inguinal hernia, the patient should undergo immediate surgical exploration.

(i) Following testicular trauma, contusion or hematocele can be treated conservatively but testicular rupture requires surgical repair.

(i) On scrotal exploration, rupture appears as disrupted tunica albuginea with varying degrees of hemorrhage and extruded seminiferous tubules.

1. If salvageable, extruded tubules should be debrided and tunica closed.
2. If not salvageable, simple orchiectomy should be performed.

(j) Patients with a testicular mass should undergo semi-urgent inguinal exploration with partial vs radical orchiectomy as indicated by pathology findings.

6. Follow-up:

(a) After any surgical management of the acute scrotum, the affected testis (if salvaged) and contralateral testicle should be followed with ultrasound imaging.

(i) May offer testicular prosthesis after testicle loss if family of patient is interested.

(b) Patients treated for epididymitis/orchitis should be followed closely for resolution of symptoms.

(i) Consider abscess formation if patient has persistent infectious or scrotal symptoms.

(ii) If suspected viral orchitis does not resolve with conservative measures, consider infectious disease consult as atrophy can result if untreated.

(iii) Repeat scrotal US is recommended to ensure resolution of inflammatory findings and rule out an underlying testicular mass.

(iv) Further evaluation for genitourinary abnormalities which may predispose to urinary infection as clinically indicated and in recurrent cases.

1. Renal US, VCUG, cystoscopy as indicated.

(c) After treatment of scrotal skin/subcutaneous infection, patients' scrotal exam should be followed closely for improvement with further surgical debridement as needed.

(d) Follow-up of patients treated for a testicular mass will vary depending on their pathology.

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Pediatric Urolithiasis

Alison M. Rasper and Joel M. Sumfest

The incidence of urolithiasis in all age groups has been increasing in the USA. Generally males are affected more than females. Almost equal distribution of kidney and ureteral calculi; low incidence of bladder calculi in pediatric population unless in children with bladder augmentations. Children can develop urolithiasis in the first decade of life but peak age affected is 20–50-years-old. Signs and symptoms of stone disease include irritability, abdominal pain, vomiting, diarrhea, and hematuria; symptoms become the more classic unilateral flank colicky pain and hematuria as the child ages.

1. Pathophysiology:
   (a) Genetics: 20–40 % of patients have 1st or 2nd degree relative with history of stones.
      (i) Cystinuria: autosomal recessive defect in transport of cysteine, arginine, lysine, and ornithine in renal tubular and intestinal epithelium leading to recurrent stone disease from supersaturation; newborn screening checks for the 2 genes responsible (SLC3A1 and SLC7A9); accounts for 5–10 % pediatric stones
      (ii) Distal renal tubular acidosis (RTA type 1): urinary acidification defect; causes alkaline urine, hypokalemia, hypercalciuria, hypocitraturia leading to stone formation; can be autosomal dominant defect presenting in adulthood or autosomal recessive presenting in infancy with deafness, bone defects, and poor growth

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(iii) Other genetic disorders associated with urolithiasis: Lesch-Nyan syndrome, primary hyperoxaluria, and xanthine dehydrogenase deficiency.

(b) Environmental: western diet high in sodium, animal protein, and carbonated beverages with phosphoric acid increase risk of urolithiasis.
(c) Metabolic:

(i) Hypercalciuria: idiopathic, secondary to medications, hyperparathyroidism, RTA, hypermetabolic states from neoplasms.
(ii) Hyperoxaluria: rare genetic disorder in oxalate metabolism, altered diet, or altered gastrointestinal absorption.
(iii) Hypocitraturia.
(iv) Hyperuricosuria: dietary, metabolic disease, or malignancy.

(d) Anatomic: more common in younger stone formers; include ureteral anomalies, urinary diversion, bladder exstrophy, neurogenic bladder, obstruction and VUR.
(e) Most children with stones have no identifiable reason.

2. Diagnosis:

(a) History: evaluate for GI disease, anatomic malformations, metabolic derangements, and familial history.
(b) Diagnostic evaluation: UA (including urine pH), urine culture, serum chemistry studies
(c) Ultrasonography: good for nephroliths, less so for ureteroliths; should be initial assessment in children less than 10-years-old.
(d) KUB: detect radiopaque stones; diagnosis made best with combination of US and KUB.
(e) Non-contrast CT scan: only if diagnosis in question after US and KUB or need further anatomic assessment.

3. Treatment.

(a) Medical: pediatric patients are more likely to have metabolic derangements and suggested that all pediatric patients with urolithiasis undergo complete metabolic evaluation.

(i) Initial management includes increase fluid intake for all stone formers.

(ii) Hypercalciuria: risk of calcium oxalate and calcium phosphate stones.

1. Increased fluid intake.
2. Decrease sodium intake.
3. Maintain daily intake of diary of two to three servings to increase GI secretion of oxalate.
4. Moderate animal protein intake.
5. Increase citrate intake.
6. Thiazides: increase calcium reabsorption in distal convoluted tubules.

(iii) Hyperoxaluria: risk of calcium oxalate stones.

1. Increased fluid intake.
2. Low oxalate diet.
3. Citrate supplementation: potassium citrate, pyridoxine.

(iv) Hypocitraturia: Risk of calcium oxalate stones.

1. Increased fluid intake.
2. Increased dietary citrate.
3. Potassium citrate.

(v) Cystinuria: risk of cystine stones.

1. Increased fluid intake.
2. Potassium citrate, Thiola (tiopronin, Mission Pharmaceutical Co. San Antonio, TX).

(vi) Hyperuricosuria: risk of uric acid stones.

1. Increased fluid intake.
2. Moderate animal protein intake.
3. Potassium citrate to alkalinize urine, allopurinol.

(vii) History of UTIs:

1. Struvite stones.
2. Treat underlying causes of UTIs.

(b) Spontaneous passage (non-surgical management):

1. Pediatric patients: 55 % of stones smaller than 4 mm pass spontaneously.
2. Medical expulsive therapy with alpha blocker studied in adult literature. Pediatric literature suggests alpha blockade safe in children without episodes of asthenia or symptomatic hypotension.

(c) Surgical management:

(i) Shock wave lithotripsy: first line treatment for pediatric nephrolithiasis as well as ureteral calculus; predominately <2 cm calculi.

1. Requires general anesthesia.
2. Complications: petechial bruising of skin, tenderness at site of treatment; rare severe complications of pulmonary contusion, hemoptysis, perirenal hematoma.

(ii) Percutaneous nephrolithotomy: Stones larger than 1.5 cm, lower pole stones >1 cm, narrow UPJ, or struvite/cystine stones.

1. Access with either US or fluoroscopy.
2. Complications: infection, hemorrhage, pneumothorax.

(iii) Ureteroscopy: distal ureteral calculi with use of holmium: YAG laser.

1. More challenging in pediatric population due to anatomic size of ureter as compared to adults.
2. Occasionally need pre-operative placement of an indwelling JJ ureteral stent for 1–2 weeks to allow passive ureteral dilation.
3. Higher stone free rates and may be superior for symptomatic ureteral calculi management over SWL.
4. Complications: Ureteral stricture, infection.

(iv) Laparoscopic surgery: only those with complex anatomy and very large stone burdens making other approaches challenging; however, percutaneous nephrolithotomy has reduced need for laparoscopic approach.

Part VII
Common Neoplasms in Children

Wilms' Tumor

Jagadeesh Ramdas

1. Pathophysiology.
   (a) Renal masses in children:
      (i) 5 % of cancer diagnosis below 20 years.
      (ii) 550 children/year diagnosed in USA.
      (iii) Wilms' tumor most common (95 %). Most commonly seen below 5-years-old.
      (iv) Rhabdoid tumors 1 %, primarily in infants and have bone metastasis.
      (v) Clear cell sarcoma of kidney (2 %), usually below 5-years-old and have brain metastasis.
      (vi) Renal cell carcinoma: Most common in adults, only 2 % occur in patients below 20-years-old.
   (b) Conditions associated with Wilms' tumor:
      (i) GU anomalies (6 %): Denys-Drash syndrome, intersex disorders, mesangial sclerosis, and Wilms tumor predisposition.
      (ii) Beckwith-Wiedemann syndrome (3 %): overgrowth anomalies.
      (iii) Hemihypertrophy.
      (iv) WAGR syndrome (5 %): Wilms' tumor, aniridia, genitourinary malformation, mental retardation.
   (c) Bilateral Wilms' tumor (5–10 %): stage V.

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(d) Wilms Tumor staging:

(i) Stage 1: tumor limited to the kidney and completely excised.
(ii) Stage II: tumor extends beyond the kidney but is completely excised.
(iii) Stage III: residual non hematogenous tumor confined to abdomen.
(iv) Stage IV: hematogenous metastasis: Lung, liver, bone, and brain.
(v) Stage V: bilateral renal involvement at diagnosis.

(e) Prognostic factors: Stage and histology.

(i) Histologic pattern of the tumor.
(ii) Presence of lymph node metastasis.
(iii) Degree of local invasion.
(iv) Presence of distant metastasis.

(f) Histology:

(i) Favorable histology: well differentiated tumors that resemble developing embryonic renal tissue. There are three histologic features: Tubules, blastema, and stroma.
(ii) Unfavorable histology: anaplastic Wilms' tumor.

2. Clinical presentation:

(a) Asymptomatic abdominal mass: most common.
(b) Pain and microscopic hematuria: 20–30 %.
(c) Hypertension (from distortion of renal vasculature).
(d) Anemia (from bleeding into tumor).

3. Diagnosis:

(a) History: Details regarding family history, congenital defects.
(b) Physical exam: blood pressure, presence of congenital anomalies, specifically hemihypertrophy, genitourinary anomalies, aniridia, etc.
(c) Labs: CBC, UA, BUN, creatinine, LFTs.
(d) Cytogenetic profile: deletion of short arm of chromosome 11 seen in many cases of aniridia and Wilms' tumor and familial Wilms' tumor.
(e) Radiological evaluation: CT chest, abdomen.

4. Therapy:

(a) Surgical management: total excision of the tumor, transabdominal incision, nodal sampling and inspection of opposite kidney and liver.
(b) Intravenous access for chemotherapy can be placed at the same operation.
(c) Radiation therapy: Given to stage III, stage IV, and stage II unfavorable histology.
(d) Chemotherapy: vincristine, actinomycin D, and adriamycin.

Neuroblastoma

Jagadeesh Ramdas

1. Pathophysiology:
   (a) Originates from neural crest tissue.
   (b) Second most common solid neoplasm in children, 10 % of all childhood cancers.
   (c) 700 children diagnosed each year. Six hundred and fifty are neuroblastoma others include ganglioneuroblastoma (more differentiated form) and pheochromocytoma.
   (d) Most commonly occur in the adrenal gland.
   (e) Most common cancer in infancy, incidence rate double that of leukemia in infants.
   (f) Incidence:
      (i) Median age at diagnosis is <2-years-old where 5 year survival is >83 %, as compared to 50–60 % for older children.
      (ii) 35 % cases occur below age 1-year-old.
      (iii) 70 % children present with metastasis.
   (g) INSS staging for Neuroblastoma:
      (i) Stage 1: localized tumor with complete gross excision.
      (ii) Stage 2A: localized tumor with incomplete gross excision.
      (iii) Stage 2B: localized tumor with or without complete gross excision, with ipsilateral nodes positive for tumor.
      (iv) Stage 3: unresectable unilateral tumor infiltrating across the midline.
      (v) Stage 4: any primary tumor with distant dissemination.
      (vi) Stage 4S: localized primary tumor with dissemination limited to skin, liver and/or bone marrow (limited to infants <1 year, marrow involvement in <10 %).

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(h) Neuroblastoma risk group.

(i) Low, intermediate, and high-risk group determined by:

1. Stage (INSS).
2. Age.
3. N-myc oncogene amplification.
4. Tumor cell ploidy.
5. Tumor histology.

2. Clinical presentation.

(a) Location of primary and degree of dissemination.
(b) Asymptomatic mass in the neck, thorax, abdomen and pelvis.
(c) Horner's syndrome, superior vena cava syndrome.
(d) Paraspinal tumors-spinal cord compression.
(e) Metastatic disease-bone and bone marrow.
(f) Bone pain, limp, refusal to walk, pallor.
(g) Orbital metastasis: proptosis, ecchymosis.
(h) Skin deposits.
(i) Massive hepatomegaly.
(j) VIP-oma –intractable diarrhea.
(k) Opsoclonus/myoclonus syndrome.

3. Diagnosis:

(a) Pathological examination of tumor tissue.
(b) Tumor cells in a bone marrow aspirate/biopsy.
(c) Elevated urinary catecholamine excretion.
(d) Staging workup:

(i) Primary: CT and/or MRI scan.
(ii) Metastatic sites:

1. Bone marrow: bilateral bone marrow aspirate/biopsy.
2. Bone: bone scan, metaiodobenzylguanidine (MIBG) or octreotide scan, plain x-rays of positive lesions.
3. Lymph nodes: clinical examination and CT scan.
4. Abdomen/liver: CT and/or MRI scan.
5. Chest: AP and lateral chest x-rays, CT scan.

4. Treatment:

   (a) Treatment schema by INSS stage:

INSS	Age	NMYC	Shima	DNA	Risk
1	0–21 years	Any	Any	Any	Low
2A/2B	<365 days	Any	Any	Any	Low
	>365–21	Non-A	Any	–	Low
	>365–21	Amp	Unfavorable	–	High
3	<365 days	Non-A	Any	Any	Intermediate
	>365–21	Non-A	Favorable	–	Intermediate
	>365–21	Amp	Any	–	High
4	<365 days	Amp	Any	Any	High
	>365–21	Any	Any		High
4S	<365	Non-A	Favorable	>1	Low
	<365	Non-A	Unfavorable	Any	Intermediate
	<365	Amp	Any	Any	High

   (b) Low-risk disease: surgical excision.
   (c) Intermediate-risk: moderate dose chemotherapy (cyclophosphamide, etoposide, doxorubicin, carboplatin), four to eight cycles.
   (d) High-risk: intensive chemotherapy, surgery, radiation, myeloablative therapy and stem cell rescue, immunotherapy.

Hepatoblastoma

Jagadeesh Ramdas

1. Pathophysiology:
   (a) Incidence: 1 % of all pediatric malignancies.
   (b) Age at diagnosis: 80 % between 6-months-old and 3-years-old. Neonatal and school aged children are also seen.
   (c) Etiology: Unknown.
      (i) Possible factors.
         1. Extreme prematurity and very low birth weight less than 1,000 g.
         2. Fetal alcohol syndrome.
         3. Hemi hyperplasia.
         4. Oral contraceptive use during pregnancy.
      (ii) Chromosomal Abnormalities:
         1. Beckwith-Weidman syndrome.
         2. Familial adenomatous polyposis.
   (d) Histologic subtypes:
      (i) Pure epithelial- Fetal.
      (ii) Embryonal/mixed.
      (iii) Macrotrabecular (MT).
      (iv) Small cell undifferentiated (SCU).
      (v) Mixed epithelial and mesenchymal (HB-MEM).

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(e) COG staging of hepatoblastoma:

(i) Stage 1: Complete gross resection at diagnosis with clear margins.
(ii) Stage II: Complete gross resection at diagnosis with microscopic residual disease at the margins of resection.
(iii) Stage III: Biopsy only at diagnosis, or gross total resection with nodal involvement or tumor spill or incomplete resection with gross intrahepatic disease.
(iv) Stage IV: Measurable metastatic at diagnosis to lungs or other organs.

2. Clinical presentation:

(a) Asymptomatic abdominal mass.
(b) Fatigue, fever, anorexia, weight loss, vomiting.
(c) Abdominal pain and hemorrhage after traumatic or spontaneous rupture of mass.

3. Diagnosis:

(a) Alpha fetoprotein (AFP) is extremely elevated in 90 % of cases (AFP levels at diagnosis <100 ng/ml are biologically more aggressive with a worse prognosis).
(b) Mild normochromic normocytic anemia may be seen.
(c) Thrombocytosis is common.
(d) CT scan shows a heterogeneous, large multinodular expansile mass, usually unifocal, but occasional multifocal mass with decreased enhancement and density compared to normal liver.
(e) MRI usually is more accurate in determining tumor extent and defining vascular involvement.

4. Treatment:

(a) Treatment of hepatoblastoma is multimodal.
(b) Complete surgical excision is the cornerstone of curative therapy.
(c) Chemotherapy: Neoadjuvant (preoperative) chemotherapy can help to shrink the tumor and consequently more likely to be completely resected, less prone for bleeding and more demarcated from the healthy liver parenchyma.

(i) Effective agents: Cisplatin, doxorubicin, 5 fluorouracil, vincristine.

(d) Liver transplant: treatment option for unresectable primary tumors; multifocal tumors invading all four sections of the liver and without demonstrable metastatic disease, unifocal large centrally located tumors involving main hilar structures or all hepatic veins.

5. Outcomes:

(a) Survival:

(i) Stage I: 3 year EFS 100 %.
(ii) Stage II: 3 year EFS- 96 %.
(iii) Stage III: 3 year EFS- 62 %.
(iv) Stage IV: 3 year EFS- 40 %.

Leukemia

Michal A. Miller

1. Epidemiology:
   (a) Acute leukemias are the most common pediatric malignancy.
   (b) Acute lymphoblastic leukemia (ALL) is the most common leukemia followed by acute myeloblastic leukemia (AML).
   (c) Annually, approximately 3,500 new cases of leukemia are diagnosed in the United States in children <20-years-old.
   (d) Peak incidence at 2–3-years-old for ALL and <2-years-old for followed by a decreasing incidence until 10-years-old when the incidence climbs with increasing age.
   (e) There is an increased incidence in children with congenital syndromes. The most common are: Downs syndrome, neurofibromatosis, Shwachman-Diamond syndrome, Bloom syndrome, ataxia telangiectasia.
2. Clinical presentations:
   (a) Children present with constitutional symptoms of fevers weight loss, night sweats, chills and mix of signs and symptoms related the bone marrow failure and tumor burden. Thrombocytopenia presents with petechiae, mucosal bleeding, easy bruising, and less commonly as a life threatening bleed. Anemia can lead to pallor, hypersomnia, fatigue, irritability and poor feeding in infants, tachycardia with varying degrees of cardiovascular compromise. Life threatening infections related to the neutropenia.
   (b) The leukemia cell infiltration resulting in adenopathy, hepatosplenomegaly, and bone pain.

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(c) Urgent complications encountered at diagnosis related to the tumor burden need to be anticipated.

(i) Tumor lysis syndrome: spontaneous or therapeutic lysis of tumor cells leads to the rapid release of intracellular cytokines, potassium, and phosphorus. Rapid cell turn over results in the buildup of uric acid which can precipitate in the renal tubules resulting in renal failure.

1. Treatment is anticipatory.
2. Hyperhydration to maintain urine output of >2 ml/kg/h. This can usually be achieved with D51/2NS or NS given at two times the patient's IVF maintenance rate.
3. No supplemental potassium.
4. Monitor intake and output for the development of oliguric/anuric acute renal failure.
5. Allopurinol to reduce uric acid production
6. Aggressive treatment of electrolyte disturbances: Hyperkalemia, and hyperphosphatemia and hypocalcemia.

(ii) Fever and neutropenia: for patients with neutropenia ANC <1,000 need urgent antibiotic coverage with empiric broad septum antibiotics. Cefepime is the front line therapy.

(iii) Symptomatic or prophylactic platelet transfusions for thrombocytopenia.

(iv) Leukostasis: most commonly encountered in AML patients with a WBC of >100,000 and ALL patients with >300,000. Can present with respiratory insufficiency and/or stroke-like signs. Requires urgent initiation of chemotherapy and/or leukophoresis.

(v) Mediastinal mass and airway compromise: most common in T cell leukemia or lymphoma.

(vi) Spinal cord compression from tumor mass effect.

(d) Urgent pediatric oncology consult should be obtained on any patient suspected of having leukemia or lymphoma. These patients frequently decline rapidly.

3. Diagnosis:

(a) The diagnosis is usually suspected or evident on CBC: Blast cells on the differential and/or varying degrees of pancytopenia.

(b) Bone marrow aspiration is needed to confirm and subtype the leukemia.

(c) CSF cytology is obtained to determine extent of the disease.

(d) Labs most helpful at diagnosis are BMP, LFTs, CBC with differential, uric acid, phosphorus, LDH, PT, PTT, fibrinogen and a type and screen.

4. Treatment:

   (a) The prognosis and treatment is dependent on the classification and the risk stratification.
   (b) Children with leukemia will usually require a tunneled intravenous line for chemotherapy, such as a Broviac (C. R. Bard, Murray Hill, NJ) or Port-a-cath (Smiths Medical, Dublin, OH).

      (i) A single lumen catheter is used for patients with ALL.
      (ii) A double lumen catheter is used for patients with AML.
      (iii) This decision should be discussed with the treating oncologist, who may also do a bone marrow biopsy at the time of catheter placement under general anesthesia.
      (iv) Parents of children receiving an intravenous line should be warned not to give their child non-steroidal anti-inflammatory medications such as ibuprofen for analgesia, because these medications will carry a risk of stress ulcer when chemotherapy causes thrombocytopenia.

   (c) The Children's Oncology Group (COG) has developed a complex stratification for ALL and AML.

      (i) Subtype by flow cytology: determines B cell ALL, T cell ALL and Myeloid phenotypes.
      (ii) The initial risk stratification for ALL starts with the National Cancer Institute criteria age >1-year-old and <10-years-old with a white blood cell count on presentation of <50,000/μL are considered low risk. Age outside of that range and/or a WBC >50,000 is high risk.
      (iii) Cytogenetics: for ALL, unfavorable cytogenetics include: Hypodiploidy (<45 chromosomes), Philadelphia chromosome (BCR-ABL1), and the translocations t(9;22), t(1;19), and t(4;11). Favorable cytogenetics for ALL include: Combined trisomies of chromosome 4 and 10 and ETV6/RUNX1 fusion. For AML, unfavorable cytogenetics include monosomy 5 or 7 FLT3, Favorable cytogenetics for AML include: Inversion of chromosome 16, and t(8;21), t(15;17), CEBPA and NPM1 mutations.
      (iv) CNS and/or testicular involvement.
      (v) Response to the therapy.

   (d) Chemotherapy for ALL consists of phases- induction, consolidation, interim maintenance, delayed intensification, and a prolonged maintenance. The first four phases consist of intensive chemotherapy that results in severe immunosuppression and pancytopenia. The maintenance phase last 2–3 years and is fairly mild. The patient's blood counts are generally fairly good and the child can undergo minor procedures safely. Therapy for AML

is much more intense and consists of six cycles of chemotherapy with prolonged hospitalizations for severe neutropenia. In the presence of unfavorable features and the availability of a marrow donor, many children will undergo bone marrow transplantation. The prognosis for children with ALL is good. The long-term survival rate for children with ALL, as a group, is 70–80 %. The cure rate for the low-risk subset of ALL with favorable features is a >90 %. The prognosis for children with AML lower than it is for ALL.

Suggested Reading

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Rhabdomyosarcoma

Jagadeesh Ramdas

1. Pathophysiology:
   (a) Sarcomas in children:
      (i) 6 % of childhood cancer.
      (ii) Rhabdomyosarcoma (RMS) most common soft tissue sarcoma.
      (iii) Approximately 350 new RMS cases/yr.
   (b) Age of incidence: 2–6-years-old.
   (c) Site of origin (In order of frequency):
      (i) Head and neck: 29 %.
      (ii) Extremities: 24 %.
      (iii) Genitourinary Tract: 18 %.
      (iv) Trunk: 8 %.
      (v) Orbit: 7 %.
      (vi) Retroperitoneum: 7 %.
      (vii) Other: 7 %.
   (d) Sites of metastasis: Lung, bone, bone marrow.
   (e) Patterns of spread:
      (i) Lymphatic: 40 % of paratesticular and 20 % of extremity tumors.
      (ii) Hematogenous: 10–20 % at diagnosis (lung, bone, bone marrow, liver).
      (iii) CNS extension: 50 % of parameningeal (cranial nerve palsies, erosion of cranial bone, direct intracranial growth).

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(f) Pathology:

(i) Embryonal RMS: 60–70 % of cases.

1. Simulates immature skeletal muscle, MyoD, myogenin expressed.
2. Embryonal RMS variants:

(a) Solid ("embryonal"): Favorable.
(b) Botryoid (polypoid grossly): Very favorable.
(c) Spindle cell (leiomyomatous with cross striations): Very favorable.

(ii) Alveolar RMS: 20–30 % of cases.

1. Growth pattern reminiscent of pulmonary alveoli with fibrovascular septa MyoD, myogenin expressed.
2. Associated with either a t(2;13)(q35;q14) or t(1;13)(p36;q14).
3. Extremity primary, lymph node involvement, and unfavorable prognosis.

(g) IRS Grouping System:

(i) Group I: Localized disease completely resected.
(ii) Group II: Gross total resection with evidence of regional spread.

1. (A) Gross resection of tumor with microscopic residual disease.
2. (B) Complete resection of primary tumor, regional nodes involved.
3. (C) Regional disease with involved nodes – microscopic residual disease and/or histologic involvement of the dissected regional node most distal to the primary tumor site.

(iii) Group III: Incomplete resection with gross residual disease.
(iv) Group IV: Distant metastatic disease.

(h) IRS staging system:

Stage	Sites	T	Size	N
1	Orbit, non-parameningeal head and neck, non-bladder/prostate GU, biliary tract	T_1 or T_2	a or b	N_0, N_1, or N_x
2	Parameningeal, bladder/prostate, extremity, other	T_1 or T_2	a	N_0 or N_x
3	Parameningeal, bladder/prostate, extremity, other	T_1 or T_2	a	N_1
			b	N_0, N_1, or N_x
4	All with distant metastases	T_1 or T_2	a or b	N_0 or N_1

Definitions: T = tumor invasiveness; T_1 = confined to anatomic site of origin; T_2 = extension and/or fixation to surrounding tissue; a = $\leq$5 cm; b = >5 cm; N = regional nodes; N_0 = regional nodes not clinically involved; N_1 = regional nodes clinically involved; N_x= clinical status of regional nodes unknown

(i) Prognosis:

(i) Stage/group/extent of disease; important predictor of outcome.
(ii) Primary site; affects time to diagnosis, likelihood of lymphatic spread, ease of excision, and potentially radiation delivery; represents an important prognostic factor (favorable sites; orbit, non-bladder prostate GU, non-parameningeal head and neck, biliary tract).
(iii) Histologic type; (most favorable) botryoid and spindle cell → solid embryonal → alveolar (least favorable).
(iv) Biologic factors; t (2; 13) unfavorable if stage 4.

2. Clinical presentation:

(a) Signs and symptoms:

(i) Relate to location of primary tumor.
(ii) Mass: Most common feature.

(b) Symptoms by site:

(i) Head and neck "parameningeal":

1. Nasopharyngeal/paranasal sinus- nasal obstruction, epistaxis, local pain or swelling.
2. Middle ear tumors: Signs and symptoms of otitis media (pain. ear discharge, facial palsy).
3. Orbital tumors: Rapidly developing, proptosis of eye with or without loss of EOM.

(ii) Urogenital tumors: Bladder or prostate- urinary tract obstruction or dysuria.
(iii) Perineal RMS: Mass in the pelvis and may develop constipation.
(iv) Extremity tumors: 25 % of all RMS: Firm mass and have a tendency to disseminate early.

3. Treatment: Multimodality approach.

(a) Surgery: Wide resection of primary tumor with margin of normal tissue.

(i) Local control: Excision of primary tumor upfront whenever possible without causing major functional or cosmetic deficits. Primary re-excision for residual tumor.
(ii) Special anatomic sites requiring surgical assessment of lymph nodes

1. Para testicular (ISRLND).
2. Extremity (node sampling).

(iii) Second look surgery during treatment sometimes done for residual tumor.

(b) Radiotherapy: Doses of 3,600–5,040 cGy generally used; dose depends on group (microscopic vs. gross disease), primary site, nodal involvement, histology, and whether second look surgery performed.
(c) Chemotherapy:

(i) For local and systemic tumor control.
(ii) Multi-agent/intensive/governed by risk-group.
(iii) Standard: vincristine, dactinomycin, and cyclophosphamide (VAC).
(iv) Other active agents used in recent trials; irinotecan, topotecan, doxorubicin, etoposide, and ifosfamide.

Part VIII
Surgical Procedures

Parenteral Access

Christopher P. Coppola

Surgical treatment of children usually requires parenteral access. Children's smaller size and severity of illness makes intravenous access difficult.

1. Percutaneous approach can be used for peripheral and central veins. For central veins, sonography before and during line placement reduces complications and increases rate of success. Check coagulation function before line attempt. Most children require anesthesia for safe line placement, and success rate will be higher in a patient who is still.

   (a) Peripheral: For most therapy, peripheral access will suffice. Use of peripheral IV catheters reduces the risk of line sepsis and thrombosis. Vein is located, tourniquet is applied, skin is prepped, and catheter is inserted at a low angle with skin, parallel with vein, attempting to thread the needle into the vein, then slide catheter off needle. Catheter is flushed, secured with an occlusive dressing, and for young patients the limb is secured with a padded immobilizer. In small children, the scalp veins can be used.

   (b) Jugular: Patient is positioned supine with 15–30° Trendelenburg to distend veins and avoid air embolus. Head is turned away from the side being attempted and a rolled towel under the shoulders helps extend the neck. Sonography is used to locate the jugular vein and carotid artery. Vein can be approached from a vector that starts posterior to the sternocleidomastoid muscle, between the clavicular and sternal heads of the muscle, or anteriorly, through the belly of the muscle itself. Skin is prepped and sterile technique is used. Vein is accessed with a needle and syringe using sonography and palpation to avoid the carotid artery. Seldinger technique is used to place a wire in the vein, advance a catheter over the wire, and then the wire is removed. It is verified that the catheter withdraws blood and flushes saline

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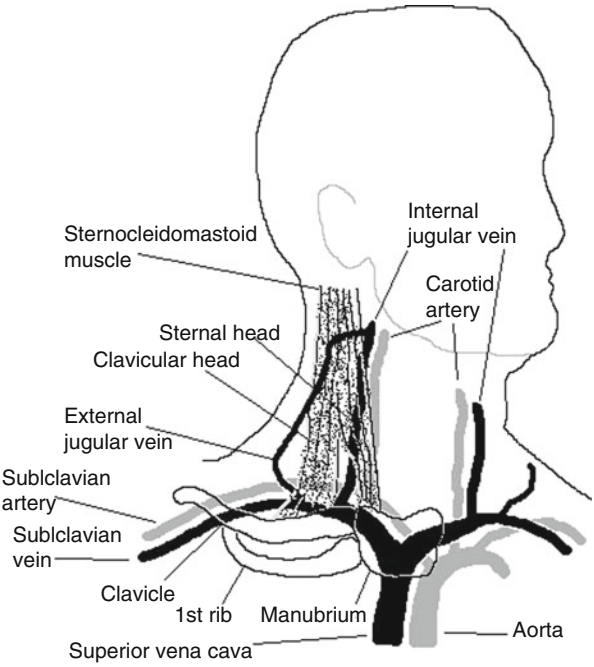

Fig. 1 The jugular vein, subclavian vein, and associated anatomic landmarks (*Source*: Christopher Coppola)

easily. Catheter is secured to skin with drain stitches and an occlusive dressing. X-ray is used to verify that the tip of the catheter is at the cavoatrial junction and there is no pneumothorax.

(c) Subclavian: Patient is positioned supine, with Trendelenburg, and a roll between the shoulder blades. Sonography is used to identify the subclavian vein. Shoulder and neck are prepped and alcohol and sterile technique used. Vein is accessed with a needle and syringe by entering skin lateral to the site where the clavicular head of the pectoralis muscle attaches to the curve in the clavicle. Needle is kept parallel to the floor, the tip is allowed to contact the clavicle, and then is redirected between the clavicle and the first rib until the subclavian vein is entered and venous blood can be withdrawn. Aim the tip of the needle at the manubrium or larynx. Avoid directing the needle toward the floor as this will increase the chance of pneumothorax. Using Seldinger technique, the catheter is placed, ensuring that the wire is completely removed. Fluoroscopy during procedure verifies the wire is directed to the superior vena cava and not to the opposite arm or up into the neck. An 11 blade scalpel and a dilator widen the entry so the catheter can enter. Catheter is secured and dressed, then chest x-ray is performed to verify placement in the superior vena cava and ensure there is no pneumothorax.

(d) Femoral: Patient is placed in supine position. Femoral vein is located with sonography and note is taken of the femoral artery, lateral to the vein. After sterile preparation and drape, vein is accessed with a needle and syringe, at a downward angle with needle parallel to the vein. When vein is entered, using

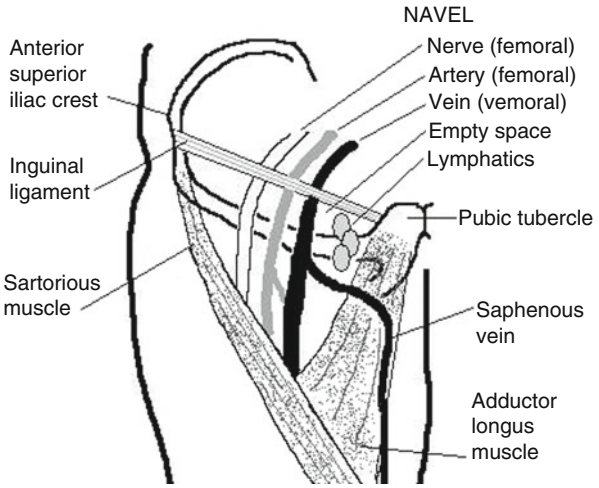

Fig. 2 Femoral artery, vein, and associated landmarks (*Source*: Christopher Coppola)

Seldinger technique a wire is inserted into vein and is used to advance a catheter into the femoral vein. The wire is removed. It is verified that the catheter can withdraw and flush, and it is secured with a drain stitch and occlusive dressing.

2. Cut-down:

   (a) Jugular:

      (i) Technique: Patient is positioned as for percutaneous access. The space between the sternal and clavicular heads of the sternocleidomastoid muscle is palpated. A transverse incision is made a finger breadth above the clavicle. The subcutaneous tissue and platysma are separated and wound is deepened until carotid sheath is encountered. The internal jugular vein is identified. It is carefully separated from the carotid artery and the vagus nerve. Proximal and distal control of the vein is obtained by circumferential dissection and encircling vein with a 4–0 absorbable ties above and below the planned point of vein entry. The distal vein is ligated. Venotomy is created. Catheter is inserted and advanced until the tip lies within the superior vena cava. The proximal tie is tied down ensuring that it does not crush the catheter. After verifying that the catheter withdraws blood and flushes saline, the wound is closed in layers of absorbable suture. The catheter is secured to skin and covered with an occlusive dressing.

      (ii) Alternatives:

         1. The internal jugular does not necessarily need to be ligated during catheter placement. A purse string of 4-0 polypropylene suture can be used to secure the edge of the venotomy, which is then tied down around the catheter, preserving jugular flow from the head.
         2. A branch of the internal jugular, the facial vein, can be used as the entry point for the catheter, and the internal jugular will not need to be ligated.

3. In larger children, the external jugular can be used for placement. After proximal and distal control is obtained, catheter is inserted via venotomy and must be guided into external jugular, which leads to the subclavian, and then enters the superior vena cava, via the innominate vein if on the left side.

(b) Femoral: Cut-down is usually performed parallel and just inferior to the inguinal ligament, but a vertical incision is an alternative. From lateral to medial, the orientation of structures in the femoral triangle are Nerve, Artery, Vein, Empty space (occupied by fat), and Lymphatics (NAVEL). The femoral vein is identified, dissected out circumferentially and secured with ties proximally and distally. Catheter is inserted via venotomy. Flow in the femoral vein is preserved.

(c) Saphenous vein runs from ankle to groin, on the medial surface of both legs. It is found easiest at the ankle, where a transverse incision is made a finger breadth superior and anterior to the medial malleolus. The vein is identified, isolated with proximal and distal control, distal vein is ligated, and catheter is inserted through venotomy, then proximal ligature is tied down to secure vein around catheter. Saphenous vein can also be found two finger breadths inferior and posterior to the femoral vein at the inguinal ligament.

3. Peripherally inserted central catheter (PICC): A PICC line offers the advantages of a central line, since the tip lies in the superior vena cava, but with fewer mechanical and infectious complications. The best vein for PICC insertion is the basilic vein above the antecubital fossa. The vein is identified with the help of sonography. Distance from entry site to the upper chest along path of venous return is measured to estimate length of catheter that will be required. Tourniquet is applied above entry site. Vein can be accessed by percutaneous technique or cut-down. Vein is accessed by a breakaway needle that splits in half, or a short catheter, large enough to pass the PICC line, depending on which type of kit is available. PICC is inserted through this device into vein. Tourniquet is removed. PICC line is advanced until the tip reaches the superior vena cava

4. Intraosseous (IO):

(a) Indications and use: An intraosseous line is a useful rescue when an intravenous line cannot be obtained. In an emergency, an intraosseous line should be placed when three attempts have been made or 90 s have passed without progress. Line can be used for IV fluid, medications, transfusion, and phlebotomy. It can be started easily on children up to age 6 years old, above that age, entry into bone can be difficult without specialized devices.

(b) Devices:

(i) Hand-driven device with detachable handle: Cook Intraosseous, threaded and smooth needles, (Cook Medical, Bloomington, IN); Jamshedi 15 g, (CareFusion, San Diego, CA)

(ii) Battery powered drill: EZ-IO, (Vidacare, San Antonio, TX)

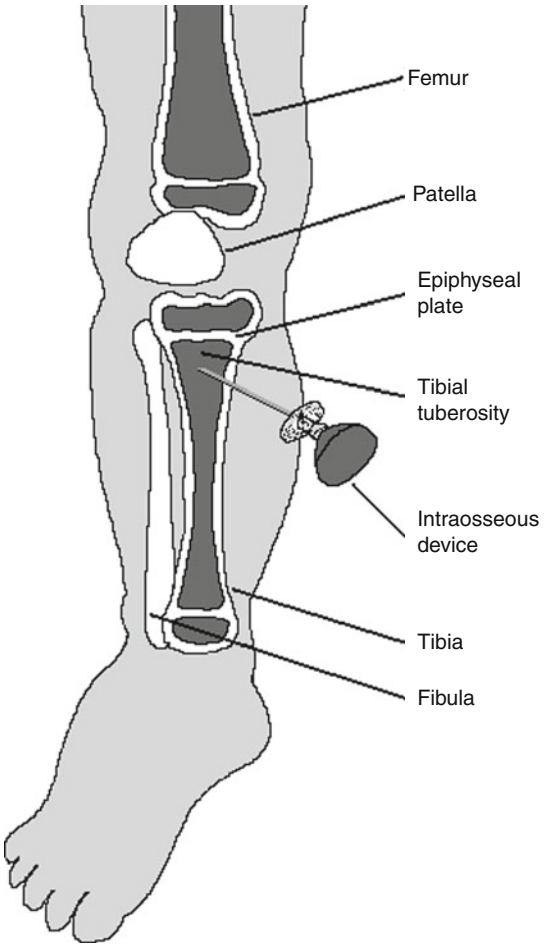

Fig. 3 Intraosseous line insertion into the proximal tibia (*Source*: Christopher Coppola)

(iii) Spring-loaded needle: "Bone Injection Gun" (BIG), (Waismed Ltd., New York, NY)
(iv) Sternal intraosseous insertion device (age 12-years-old and above): "First Access for Shock and Trauma 1" (Fast 1), (Pyng Medical Corp., Richmond, British Columbia, Canada)

(c) Entry site:

(i) Proximal tibia is the preferred insertion site and works in children and adults. Medial surface of the tibia is flat, and overlying tissue is thin. This allows for palpation of landmarks and minimum tissue to traverse before bone. From the tibial tuberosity, measure 1 cm distal and 1 cm medial (2 cm for children > 6 years), finding a broad flat area where skin is thin over the tibia.
(ii) Sternal placement of an intraosseous needle requires a commercially available device which can be used in patients 12 years old and older.

The device comes with a positioning patch which guides needle entry approximately 2 cm below the sternal notch. The catheter comes with a small extension set, protective dressing, and specialized removal tool, which is kept with the patient.

(iii) Alternate placement sites used when primary sites are unavailable due to trauma or previous attempt include the distal tibia on medial aspect, distal femur on anterior aspect, proximal humerus on antero-medial aspect, iliac crest, and others. In general, choose a site without overlying neurovascular structures to be injured, in the portion of bone with marrow cavity, and with minimum thickness of subcutaneous tissue.

(iv) Contraindications (most are site specific and an alternate site can be chosen):

1. Fracture of bone.
2. Previous attempt in same bone.
3. Vascular injury to limb.
4. Disease of target bone (tumor, osteoporosis, or osteogenosis imperfecta).
5. Infection or tissue deficit of soft tissue at target site.

(v) Complications: Intraosseous lines are used for expedient access when prompt intravenous access is unavailable. Although they may endure for up to 96 h, they should be replaced with intravenous access as soon as it is safely and accurately available.

1. Erroneous placement outside bone or completely through bone: This will cause extravasation.
2. Extravasation around needle: This increases the longer the needle is used.
3. Compartment syndrome from extravasation.
4. Infection.
5. Dislodgement of needle.
6. Occlusion of needle by bending, bone, or clot.
7. Hematoma.
8. Air or fat embolus.
9. Pneumothorax, hemothorax, or mediastinitis from through and through penetration of sternum.

5. Creating a tunnel under the skin for passage of the catheter before it enters the vein provides protection against infection. Also, the catheter can be secured to the subcutaneous tissue by cuffs of fibrous material into which the body grows and attaches. Tunneled catheters are safer for outpatients as there is less chance of catheter dislodgement and hemorrhage. Tunneled catheters are placed in the operating room or interventional radiology suite with anesthesia. Catheter tip is positioned at the junction of right atrium and superior vena cava, confirmed by x-ray. Catheter is accessed with aseptic technique to avoid contamination. Heparin 100 U/mL is left indwelling in the catheter to avoid thrombosis, unless contraindicated. In some scenarios, use of an indwelling ethanol lock can reduce infection.

(a) Externalized catheters enter the skin at one location, traverse the subcutaneous space, then enter a vein at a different location. Examples are Broviac and Hickman catheters. (C.R. Bard Inc., Murray Hill, NJ). Cuff is positioned under the skin for catheter fixation.
(b) Completely implanted catheters have a reservoir under the skin, and a catheter that traverses the subcutaneous space before entering the vein. For infusion or phlebotomy, the reservoir is accessed through the skin using a Huber needle, which is a non-coring needle that will not create leaking voids in the silicone surface of the port. Examples are the mediport and Port-a-cath (Smiths Medical, St. Paul, MN). The best location for the reservoir is on the surface of the pectoralis muscle fascia. The skin in the midline chest can be too thin and will erode with repeated use. The breast should be avoided because incision creates future scarring in breast, breast tissue thickness makes access of the port difficult, and when the patient stands, the weight of the breast can pull the port inferiorly and retract the catheter tip out of proper position. Care is taken to ensure that the catheter is securely fastened to the spout exiting the port to avoid leakage. The port should be secured to the pectoralis fascia in at least two locations to avoid port migration and flipping. Port placement will leave a larger scar than an externalized catheter and will require a second operation to remove. Ports give the advantage of being more difficult to break or dislodge. Patients will be able to swim and shower when the port is not accessed.

6. Dialysis:

(a) Hemodialysis: Various catheter options exist included tunneled vs. percutaneous, and a range in sizes of catheter diameter and length to cover pediatric patients across changes in age and size. The catheters have two lumens, corresponding to the need to simultaneously withdraw blood from the patient and return blood after it has passed through the dialyzer. The tips of the two lumens are staggered to reduce the recirculation of dialyzed blood directly back into the catheter. Catheter is positioned so that the tips of both lumens are clear with good withdrawal of blood and easy flushing of saline. Heparinized saline is left indwelling in both lumens of the catheter. The concentration of heparin used can vary between institutions; one example is 1,000 U/mL. Before flushing the catheter, the indwelling heparin must be withdrawn and discarded to avoid infusing the heparin and causing bleeding.

(i) Percutaneous hemodialysis catheters are used when the need is urgent and the catheter is intended to be used temporarily, while the child is an inpatient, and removed when the child recovers renal function, or a more durable access for dialysis is provided. The best access for percutaneous hemodialysis is the left internal jugular vein because it provides a straight, short path to the right atrium without flexing of the catheter which can reduce flow rate.
(ii) A tunneled hemodialysis catheter is placed when a patient is intended to have the catheter for a prolonged period of time and have the catheter while they are an outpatient. The routes most commonly used are the

right internal jugular vein and the right subclavian vein because the catheter tip will descend the superior vena cava in a straight superior to inferior orientation, avoiding occlusion caused when the catheter is pressed up against the inside surface of the superior vena cava.

(b) Peritoneal dialysis is preferred by most families because therapy can be provided in the home. It also avoids the fluctuations in blood pressure that occur during hemodialysis.

(i) Specialized hemodialysis catheters called Tenkhoff catheters are used. There are different sizes available for different ages. Catheters have multiple holes to allow for efficient flow of dialysate and peritoneal fluid, and to reduce the incidence of catheter occlusion from adhesions. There is a portion of the catheter intended to lie in a subcutaneous tract, and one or multiple fibrous cuffs to encourage tissue ingrowth for fixation. The catheters are pre-curved to assist with creation of a subcutaneous tract and creation of a region of the peritoneum that serves as a reservoir for dialysate. Examples are the Quinton Curl Cath and Argyle Swan Neck Cath (Covidien, Mansfield, MA).

(ii) Catheter is inserted sterilely in the operating room. A supraumbilical incision is made to gain entry to the peritoneal cavity. The catheter tip is directed downward, to lie in the anterior pelvis. The peritoneum and midline fascia are closed with a purse-string suture, attempting to create a water-tight seal. The external portion of the catheter is tunneled out to an incision in the right abdomen, where it is secured with a drain stitch. The fibrous cuff or cuffs of the catheter must be under the skin.

(iii) When the need is urgent, the peritoneal dialysis catheter can be used immediately, but there will often be leakage of dialysis from the two incisions. When possible, peritoneal dialysis should be delayed at least 2 weeks to allow the body to create a tight seal around the catheter.

7. Arterial access is often needed in critically ill patients and during operations with anticipated fluctuations in blood pressure. They can be used to continuously monitor blood pressure and to withdraw blood specimens.

(a) Placement: Arterial site is selected. Skin is prepped. An arterial catheter is selected: Size should be 18–24 gauge depending on age of patient. Either Intravenous catheters can be used, or specialized kits which include a wire to guide advancement of catheter into artery. While palpating artery, catheter is inserted into skin at a 45° angle until a flash of blood is seen. Needle angle is lowered, and needle is slightly advanced in an attempt to thread into the lumen of artery. If a wire is present, it can be advanced into artery at this time. Catheter is advanced off of needle into the artery and the needle and wire are removed. There should be a pulsatile flow of blood from the catheter and an arterial wave form when connected to a monitor. Catheter is secured with a suture and an occlusive dressing is applied. When percutaneous

placement attempt is unsuccessful, open cut-down to visualize artery during catheter placement can be performed.

(b) Upper extremity sites:

(i) Radial artery: Before placing a radial arterial line, the Allen's test is performed. Radial and ulnar arteries are compressed, hand is squeezed to blanch skin, ulnar artery pressure is released, and it is verified that color returns to the hand within 5 s. If test is abnormal, consider another site. Hand and wrist are stabilized on an arm board with dorsiflexion of the wrist for placement. During radial artery line use, the hand and fingers are observed for signs of thrombosis.

(ii) Axillary artery: This site is a secondary site when radial arteries are not available or palpable. It can also be used for catheter based interventions. When axillary hair is present, it should be trimmed to allow for more effective skin preparation and application of an occlusive dressing. There is more motion possible in the axilla than the wrist so patients should be stabilized and sedated when necessary while an axillary artery catheter is in place.

(c) Lower extremity:

(i) Femoral artery: Artery crosses the inguinal ligament half way between the anterior superior iliac spine and the pubic tubercle. A long enough catheter should be selected to traverse the subcutaneous fat and so that tip does not become dislodge from the artery with patient movement. Length varies with age; 5 and 8 cm lengths are available. After femoral artery catheter placement, the leg needs to be monitored for signs of thrombosis and ischemia.

(ii) Dorsalis pedis artery: This artery runs over the dorsum of the foot, toward the medial aspect. It can usually be located at the crease on the anterior ankle. Foot is dorsiflexed and secured. Catheter should enter artery below ankle crease so it is not occluded with foot flexion.

(iii) Posterior tibial artery: This artery runs posterior to the medial malleolus of the ankle. It is usually encountered deeper than expected by palpation of the pulse.

Enteral Access

Filip Moshkovsky

Infants and children who are otherwise in good health can tolerate 5–7 days without significant caloric intake. Significant illness or injury in children requires earlier nutrition. If possible the caloric intake should be through enteral nutrition. Oral intake is the preferred method for enteral nutrition but if not possible several delivery modalities are available.

1. Nasogastric or orogastric tube: In an intubated patient with short term feeding requirements an orogastric tube is an excellent method of delivering nutrients to the gastrointestinal tract.
   (a) Orogastric tube: Placed into the esophagus through the mouth and advanced into the stomach
   (b) Nasogastric tube: Placed into the esophagus through the nasal passage and advanced into the stomach.
      (i) Check abdominal x-ray to confirm the tube is below the gastroesophageal junction.
2. Percutaneous gastrostomy tube (PEG) placement is a common option to pursue if there is no history of abdominal surgery, hepatomegaly or malrotation.
   (a) Gastroscope placed after sedation initiated. The stomach is fully inflated with air which displaces the colon inferiorly and creates a surface area between stomach and abdominal wall. The lighted gastroscope end is located though the skin and pressed firmly upward at this point. This is usually halfway between the costal margin and the umbilicus.
   (b) Local anesthesia is injected and a stab incision is made. A needle with catheter is passed into the stomach under direct visualization and a wire is intro-

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duced through the catheter into the stomach. The wire is grasped endoscopically and withdrawn through the patient's mouth with the scope.

(c) A PEG catheter is secured to the end of the wire at the mouth and pulled through the esophagus and into position by pulling the wire back out of the abdominal incision, with the catheter following.

(d) The endoscope is reintroduced and the position of the PEG is verified. An external bumper is applied, and secured with non-absorbable suture.

(e) PEG tube can be used for feeds the same day as the procedure.

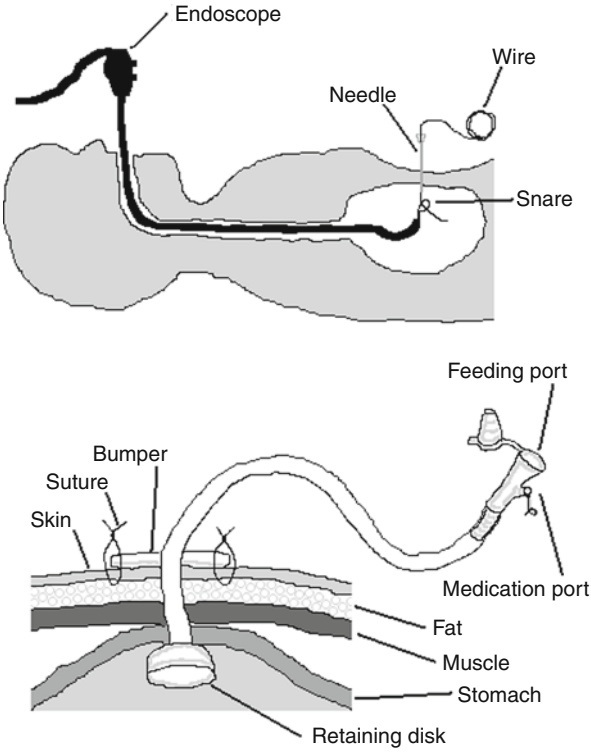

Fig. 1 Endoscopy for percutaneous endoscopic gastrostomy tube placement (*Source*: Bryan Walters and Christopher Coppola)

3. Open Gastrostomy should be considered if there are contraindications for a PEG or if abdomen is open for another procedure.

   (a) An abdominal incision adequate to identify the stomach and suture it to the abdominal wall is made. Create the incision far enough from the gastrostomy tube to allow adequate healing.

   (b) Incision for the tube is planned to provide a comfortable tube position without tension or twisting of the stomach, typically between the left costal margin and the umbilicus.

(c) The mid anterior gastric wall is grasped with a Babcock forceps, and positioning is tested. Two concentric purse-string sutures are placed in the mid anterior wall of the stomach. A gastrotomy is created in the center of the purse string sutures.
(d) A gastrostomy tube is introduced into the stomach and balloon is inflated. Caution is taken to not overinflate balloon and obstruct distal stomach.
(e) The gastric wall is anchored to the inside of the abdominal wall to approximate gastric incision with the abdominal wall. Caution must be taken to not puncture the balloon while placing anchoring sutures.
(f) The abdominal incision is closed and the gastrostomy tube is anchored to the skin, taking care to not puncture the balloon.

4. Other modalities:

(a) Gastro-jejunostomy tube: Similar to the gastrostomy tube except the catheter extends beyond the balloon as far as the jejunum. This is an option for children with risk of reflux and aspiration.
(b) Gastric bypass and gastrostomy with roux-en-Y esophago-jejunostomy.
(c) Roux-en-Y feeding jejunostomy.

5. Common tube site problems:

(a) Granulation tissue surrounding tube.

(i) Try application of silver nitrate.

(b) Clogged tube:

(i) Flush with water or saline. Can also instill carbonated beverage into tube and flush after 15–30 min.

(c) Mechanical failure:

(i) Fluoroscopic gastrostomy tube study and replacement of tube.

(d) Leak:

(i) Get a tube study. Make sure the external bumper is flush with skin.
(ii) Keep tube perpendicular to the skin.

(e) Skin irritation:

(i) Keep area dry.
(ii) Keep tube perpendicular to the skin.
(iii) Skin surrounding tube may become infected – cellulitis. Clinical examination important before initiating antibiotics.

(f) Obstruction of pylorus:

(i) If using tube with an intragastric balloon then partially deflate the balloon enough to relieve the obstruction.

(g) Tube dislodgement or removal:

(i) After 2 months it is safe to replace the tube in an office setting. If there are signs of obstruction or leak, a fluoroscopic gastrostomy tube study should be performed to confirm the balloon has not migrated causing obstructed at the pylorus.

(ii) If tube is removed and stomach has fallen away from the abdominal wall, then this must be repaired immediately via laparotomy – treat like a perforated viscus.

Chest Tube

Luiz G. Foernges

Tube thoracostomy is indicated for drainage of air or fluid accumulated between visceral and parietal pleura.

1. Indications:
   (a) Emergent:
      (i) Tension pneumothorax.
   (b) Urgent:
      (i) Pneumothorax in patients on mechanical ventilation.
      (ii) Tension pneumothorax after needle decompression.
      (iii) Pleural effusions affecting respiratory function.
      (iv) Traumatic hemopneumothorax.
   (c) Elective:
      (i) Empyema or complicated parapneumonic pleural effusion.
      (ii) Malignant pleural effusion.
      (iii) Transudates from cardiac, renal and hepatic diseases.
      (iv) Postoperative after cardiac and thoracic procedures.
2. Contraindications:
   (a) Coagulopathy.
   (b) Pulmonary bullae.
   (c) Pulmonary collapse "whiteout" from mucus plug.
   (d) Dense pleural adhesions.
   (e) Drainage of a post pneumonectomy space.

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3. Diagnosis:

   (a) Clinical exam: primary diagnosis for tension pneumothorax. Absent breath sounds, deviation of the trachea away from the side with the tension pneumothorax, hyper-expansion of the chest side with tension pneumothorax, increased venous pressure with dilated neck veins, and cardiorespiratory collapse.
   (b) Chest x-ray: initial diagnostic test, cheap, fast, and readily available. Will require lateral and/or decubitus views for small effusions. Bilateral effusions more commonly associated with transudates. Can also suggest diagnosis cause for the effusions (consolidation, tumors, and cardiomegaly).
   (c) Ultrasonography: available in most emergency departments and intensive care units. Operator dependent. Able to detect small amounts of pleural fluid. Useful tool for multiloculated pleural effusions and identifying landmarks before chest tube placement, especially in small children.
   (d) Computed tomography: more sensitive to differentiate pleural thickening and focal pleural masses. Main disadvantage is radiation exposure.
   (e) Magnetic resonance imaging: limited role on diagnosis of pleural disease.
   (f) Pleural fluid analysis: pleural fluid can be obtained prior to chest tube placement to help diagnosis and guide treatment. Pleural fluid sample can be sent for protein, lactate dehydrogenase, glucose, pH, amylase, hematocrit, cell count and differential, triglycerides, cholesterol, chylomicrons, culture, Gram stain and cytology analysis.

4. Equipment:

   (a) Mask, head cover, sterile gloves and gown.
   (b) Skin antiseptic solution.
   (c) Sterile drapes.
   (d) Gauze.
   (e) Syringes and needles.
   (f) Scalpel and blade.
   (g) Suture:

      (i) Preterm and term infants #3-0 non-absorbable suture.
      (ii) 1–6 years #2-0 non-absorbable suture.
      (iii) Above 6 years #0 non-absorbable sutures.

   (h) Instruments for blunt dissection, curve hemostatic and Kelly clamps.
   (i) Needle driver.
   (j) Mayo scissors.
   (k) Chest tube:

      (i) Preterm infants: 8Fr.
      (ii) Term infants: 10Fr.
      (iii) 1–3 years of age: 12Fr.
      (iv) 3–10 years of age: 16-18Fr.
      (v) 10–15 years of age: 20-28Fr.
      (vi) Above 15 years of age: 28Fr.

   (l) Connecting tube.

(m) Closed drainage system.
(n) Adhesive tape.

5. Procedure:

(a) Review imaging. Check and confirm the side of the procedure.
(b) Administer intravenous analgesia and sedation. Prophylactic antibiotics should be given in trauma cases.
(c) Position patient supine, with the arm on the side of the procedure abducted 90° with the palm of the hand resting under or near the head, secure the wrist with adhesive tape or soft restraint, being sure that the hand is perfused.
(d) Identify the landmarks, "safe triangle":

(i) Inferior: level of nipple (4th intercostal space), placement of chest tube below the 4th intercostal space will increase the chance of intra-abdominal chest tube placement.
(ii) Anterior: lateral border of pectoralis major muscle, placement of chest tube anterior to this line will increase the chance of injury to the pectoralis muscle and breast tissue.
(iii) Posterior: anterior border of the latissimus dorsi, placement posterior to this line will increase the chance of injury to the long thoracic nerve.

(e) Ultrasonography can guide insertion site. Select a place within the safe triangle, close to the midaxillary line.
(f) Use 1 % lidocaine (maximum dose 3.3–4.4 mg/kg); infiltrate a wheal for the skin. Redirect the needle to the expected course of the chest tube towards the superior border of the rib injecting subcutaneous tissue, intercostal muscle and pleura.
(g) After entering the pleura with the needle, aspirate air, blood or pleural effusion to confirm position, if necessary aspirate fluid for diagnosis after anesthesia.
(h) Make a small incision following the curvature of the rib (approximately the diameter of the chest tube) in the skin overlying the rib that is below the intercostal space to be used for placement of the chest tube.
(i) Bluntly dissect the subcutaneous tissue with a hemostat clamp, spreading the tissue. Direct the tip of the hemostat towards the superior border of the rib. Palpate the tract with a finger to be sure of the correct intercostal space, and superior border of the rib during dissection. Small bore chest tubes can also be inserted by the use of Seldinger technique over guidewire and dilators.
(j) Gently advance the hemostat over the superior border of the rib, opening the hemostat in the same direction of the ribs to dissect the intercostal muscles.
(k) With the hemostat closed, carefully pierce the pleura and spread in a controlled manner to not injure intrathoracic organs. A rush of air or fluid may occur when the pleura is entered.
(l) For older children, palpate the tract with a finger, passing the finger through the intercostal space into the pleural cavity to evaluate the presence of adhesions.

(m) Measure the length between the insertion site and the apex of the lung to estimate the length of the chest tube, place a clamp in the chest tube to mark the estimated length measured.
(n) Grasp the fenestrated portion of the chest tube with a Kelly clamp and introduce it in the pleural space, release the Kelly clamp and gently advance the chest tube posteriorly and superiorly. Be sure that all the side holes in the chest tube are within the pleural cavity.
(o) Holding the tube in place, connect the chest tube to the drainage device. Release the clamp from the chest tube. Connect the closed drainage system to suction if indicated.
(p) Secure the chest tube to the skin with non-absorbable sutures. Approximate the skin around the chest tube with sutures. Place sterile dressing around the chest tube site. Strap the chest tube to the flank with adhesive tape to prevent kinking and tension. Tape chest tube connections to prevent dislodgement.
(q) Obtain chest radiography to ensure correct placement.

6. Complications:

(a) Severe:

(i) Puncture of the heart, lung, liver and spleen.
(ii) Damage to intercostal vessels, anterior thoracic artery and trachea.
(iii) Wrong side.

(b) Moderate:

(i) Infection.
(ii) Nerve damage.
(iii) Chest tube disconnection or displacement.

(c) Mild:

(i) Subcutaneous emphysema.
(ii) Poor drain management.
(iii) Underwater seal not maintained.

7. Chest tube removal:

(a) Remove dressings.
(b) Remove sutures around chest tube, holding the chest tube in place.
(c) Drain should be removed during Valsalva maneuver or expiration.
(d) Pull the tube out quickly while covering the insertion site with Vaseline gauze.
(e) Place folded 4×4 gauze over the petrolatum gauze and cover with adhesive tape.
(f) Order a follow up chest x-ray.

Scoliosis Exposure

Christopher P. Coppola

Several procedures, such as repair of scoliosis or resection of tumors, require operative exposure of the thoracic or lumbar spine.

1. Exposure of thoracic spine only:

   (a) Operation is similar to a postero-lateral thoracotomy.
   (b) Patient is positioned in lateral decubitus with padding and deflatable beanbag for stability.
   (c) Flex table or raise kidney rest if anatomy and target site require it.
   (d) Incision is made over the intercostal space corresponding to the midpoint of the lesion measured from top to bottom.
   (e) After dividing latissimus and serratus muscles, the intercostal muscle is divided over the top of the lower rib, avoiding the intercostal neurovascular bundle below the rib above.
   (f) Pleura can be dissected off of the parietal surface of the ribcage and retracted anteriorly, or if necessary for broader exposure, the pleura can be entered and the lung is retracted anteriorly.
   (g) If more space is needed, one or both of the ribs can be divided posteriorly or detached from the vertebra, then spread wider, taking care not to injure the intercostal bundle. If the intercostal artery is lacerated, it is suture ligated.
   (h) If still present, pleura is reflected off of the desired vertebral levels, attempting to preserve pleura to close over vertebra at end of case.
   (i) Carefully suture-ligate segmental vertebral veins that cross the body of vertebrae.
   (j) After spinal procedure is complete, obtain hemostasis, use fibrin glue if indicated.

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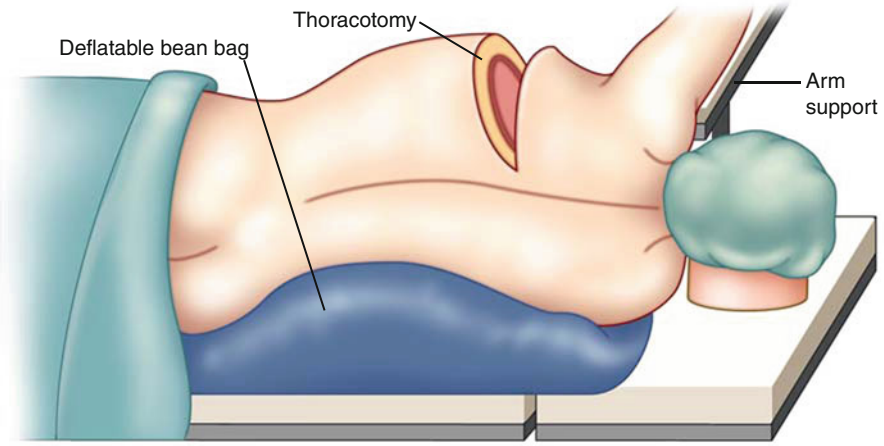

Fig. 1 Exposure of Thoracic spine only (*Source*: Christopher Coppola)

(k) Cover spine with pleura.
(l) Place chest tube to reinflate lung and position postero-laterally.
(m) Place intercostal sutures and close as for thoracotomy.

2. Exposure of lumbar spine only:

(a) Patient is positioned lateral decubitus with proper padding and a deflatable beanbag for stability.
(b) Incision follows the curve of the superior iliac crest from anterior to posterior choosing the distance between the bone and incision such that the incision is centered over the level of vertebra desired.
(c) Divide the external oblique muscle and aponeurosis in direction of fibers from the edge of the rectus posteriorly to the front edge of the latissimus dorsi.
(d) Divide across the internal oblique muscle fibers.
(e) Carefully divide the transversus abdominus muscle, preserving the peritoneum, pushing it anteriorly and bluntly dissecting it off of the underside of the flank body wall. This is easier in the posterior portion of the transversus abdominus where it is not so densely adherent to the peritoneum. Repair any holes created in the peritoneum with absorbable suture.
(f) Continue mobilizing the peritoneum and viscera contained within to the front of the body, working around the posterior edge until the spine comes into view. Often there is a layer of fat in this plane that facilitates dissection.
(g) Retroperitoneal structures, namely ureter, aorta, and vena cava branches, are carefully identified and retracted anterior, suture ligating the segmental vertebral venous and arterial branches as necessary.
(h) After spinal procedure is completed, obtain hemostasis, using fibrin glue if needed.
(i) Close layers of the abdominal wall, taking care not to injure ureter or viscera.

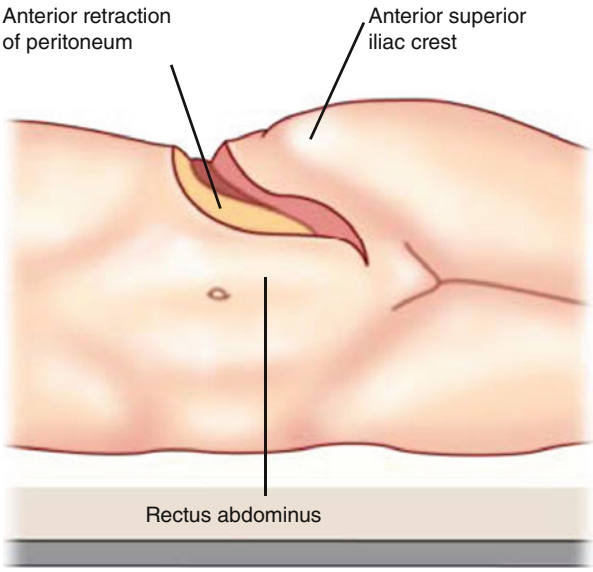

Fig. 2 Exposure of lumbar spine only (*Source*: Christopher Coppola)

3. Exposure of thoracic and lumbar spine simultaneously:
   (a) Patient is positioned partially over into lateral decubitus position with proper padding and deflatable beanbag for stability.
   (b) A thoracoabdominal incision is made that follows the course of the tenth rib posteriorly and descends from the costal margin anteriorly, following the lateral edge of the rectus muscle inferiorly to the level of the umbilicus.
   (c) The intercostal muscles above the tenth rib are divided, entering the thoracic cavity, taking care not to injure the lung or the diaphragm which should be under the incision and may be pressed against the inside of the ribcage.
   (d) Divide the abdominal muscles under the incision and enter the peritoneal cavity avoiding injury to the viscera.
   (e) The diaphragm is detached from the spine posteriorly, and around the lateral edge of body wall, leaving a margin of diaphragm muscle on the body wall, allowing for reconstruction of the diaphragm to the body wall at end of the case.
   (f) After the diaphragm is divided, the thoracic and abdominal cavities are connected. The lungs and the abdominal viscera are protected and retracted anteriorly. Both a rib spreader and a Bookwalter self-retaining retractor are useful for this retraction.
   (g) The retroperitoneal tissue is carefully dissected off of the abdominal portion of spine, suture ligating the segmental vertebral arterial and venous branches where necessary, until sufficient length of spine is exposed for spinal procedure.
   (h) When spinal procedure is complete, obtain hemostasis, using fibrin glue if needed.

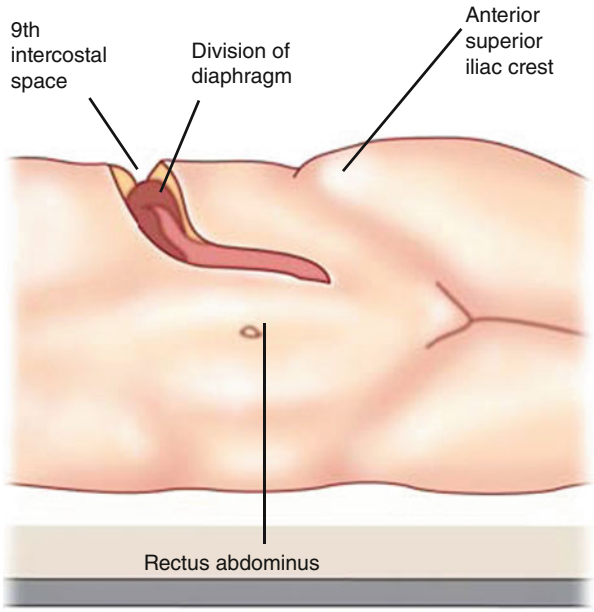

Fig. 3 Exposure of Thoracic and lumbar spine simultaneously (*Source*: Christopher Coppola)

(i) The diaphragm is reconstructed with permanent suture, taking care not to leave gaps which would become sites of diaphragmatic hernia.
(j) Chest tube is placed posterolaterally to reinflate lung and is secured.
(k) Intercostal sutures are placed and chest wall is reconstructed.
(l) Abdominal wall is reconstructed avoiding injury to the viscera.

Hydrocephalus: Ventricular Shunts and Endoscopy

Amir Kershenovich

Hydrocephalus and ventricular shunts: Hydrocephalus is a pathological accumulation of CSF in the ventricles which can result from multiple conditions such as injury, hemorrhage, tumor, infection or congenital malformations. In some children, operative shunting of the fluid is required to control increase intracranial pressure and symptoms.

1. Pathophysiology:

   (a) Ventricular anatomy: two lateral C-shaped ventricles separated by a midline septum pellucidum; each has a frontal foramen of Monroe which connects into the third ventricle; the third ventricle has an anterior pre-mammillary membrane anterior to the hypothalamic mammillary bodies and posterior to the chiasmatic and infundibular recesses; posteriorly in connects with the fourth ventricle through the Aqueduct of Sylvius; the fourth ventricle sits between the brain stem anteriorly and the cerebellum posteriorly and drains cerebrospinal fluid (CSF) through the central foramen of Magendie and two lateral foramina of Luschka to be spread around the brain and spinal cord.

   (b) Cerebrospinal fluid (CSF) physiology: clear colorless fluid produced by choroid plexus within the ventricles (80 %), interstitial space, ventricular ependymal and nerve root sleeves dura mater; resorbed by arachnoid villi of the dural venous sinuses, choroid plexus and cervical lymphatics back into the venous system driven by hydrostatic gradient. Daily CSF production and volumes: Newborns: 25 ml produced per day; total volume of 5 ml; adults: 0.3–0.35 ml/min (450–750 ml/day) produced; total volume 150 ml. The 150 ml volume is reached by age 5 years old. Production is pressure independent.

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(c) Hydrocephalus (HCP): abnormal accumulation of CSF, resulting in raised intracranial pressure (ICP). Prevalence 1–1.5 %; incidence of congenital HCP 0.9–1.8/1,000 births.

(i) Types:

1. Obstructive (O, non-communicating): internal or external obstruction at any of the communicating paths of CSF flow within the ventricles.
2. Non-obstructive (NO, communicating): CSF circulation blocked at the arachnoid granulations.

(ii) Etiologies: congenital: myelomeningocele/Chiari II malformation (O/ NO), Chiari I malformation (O), aqueductal stenosis (O), intrauterine infection (NO), germinal matrix hemorrhage (NO), Dandy Walker malformation (O), arachnoid cyst (O), syndromic craniosynostosis (NO), X-linked inherited disorder (NO); acquired: Infectious (NO), post hemorrhagic (germinal matric or traumatic, NO), masses (O), postoperative (O/NO), dural sinus thrombosis (NO), unknown origin.

(d) Hydrocephalic syndromes:

(i) Acute hydrocephalus: can occur within hours from the onset of obstruction; characterized by a rapid and progressive rise in intracranial pressure if the cranial sutures are closed; the patient becomes progressively obtunded and exhibits signs of central trans-tentorial herniation with increasing intracranial pressure; death usually occurs rapidly unless treatment is instituted promptly.

(ii) Chronic hydrocephalus: the ventricular obstruction is incomplete; compensatory changes like expansion of the skull if the cranial sutures are open, contraction of the cerebral vascular volume or brain atrophy prevent a rapid death.

(iii) Arrested hydrocephalus: chronic hydrocephalus with ventriculomegally or progressive smaller ventricles where there is neither increased intracranial pressure nor new symptoms.

2. Signs and symptoms:

(a) Infants: macrocephaly, increased head circumference, bulging fontanelle, splaying of cranial sutures, irritability, vomiting, engorgement of scalp veins, upward gaze palsy, 6th cranial nerve palsy bradycardia, apneas, failure to reach milestones.

(b) Older children: headache, nausea, vomiting, papilledema, up-gaze and or 6th cranial nerve palsy, gait changes.

3. Diagnosis: ultrasound, CT, MRI.
4. Treatment options: CSF shunting, endoscopic third ventriculostomy (ETV), endoscopic choroid plexus coagulation (CPC), fetal surgery.

(a) ETV: for specific cases where an obstruction occurs at or caudal to the aqueduct of Sylvius and for some cases of NO HCP, an ostomy (hole) is created in the pre-mammillary membrane of the third ventricle. This creates an outflow of CSF from the lateral and third ventricles into the CSF cisterns anterior to the brain stem which in turn circulate outside the cranium to the spinal subarachnoid spaces.
(b) CPC: has been utilized by few surgeons in recent years together with ETV, improving outcomes in some infants younger than 1 year-old.
(c) CSF shunts: most common and worldwide available treatment for hydrocephalus. Became available almost 50 years ago; before, hydrocephalus used to be a lethal or a severely disabling condition. Notwithstanding, they have high complication rates from infections (1–15 %), occlusions, disconnections, shunt fractures, improper placement within or outside the ventricle and shunt migrations, requiring multiple surgical interventions.

    (i) Types: common: ventriculoperitoneal (VP), ventriculopleural (VPl), ventriculoatrial (VA), Ventriculosubgaleal (VS), lumboperitoneal (LP), external ventricular drainage (EVD). Rare: Ventriculogallbladder, ventriculoureter, ventriculovenous.
    (ii) Shunt components: different companies manufacture different shunts. Catheters can be or not impregnated with antibiotic or a chemical coating such as silver to decrease infections. Shunts may be unitized or have some or all components separated requiring to be connected during surgery; this is usually done through in-site male connectors and non-absorbable ties (usually silk). Components from cephalic to caudal are:

        1. Ventricular (proximal) catheter.
        2. Reservoir: depressible/pumpable (or not) chamber that can be tapped or depressed percutaneously; a valve may be included/unitized with the reservoir; may be unidirectional or bidirectional.
        3. Valve: unidirectional, pressure (most) or flow related; may open and allow outflow only after a certain fixed intracranial pressure, or may be adjustable (aka programmable) percutaneously; of the later, the newest are compatible with MRIs, not requiring to be re-programmed afterwards.
        4. Distal catheter: same as proximal catheter, but longer.

    (iii) Other devices: shunt assistant device (anti-syphon/anti-gravity overdrainage system), connectors.

(d) Treatment strategies: are based on trying to avoid a shunt insertion; ETV when possible; removing or draining obstructing masses like tumors, cysts or hematomas; temporary EVD and challenging its need prior to converting it to a shunt.

(e) Surgical considerations when inserting a shunt:

(i) Ventricular access: anatomical land mark based vs. guidance based (image, platform); ideal place is in the lateral ventricle away from the choroid plexus; entry site can be frontal or parieto-occipital.

(ii) Peritoneal access: open: midline upper abdomen in newborns through the abdominal raphe, para-umbilical, trans-umbilical. Minimal invasive: trans-trocar; laparoscopic assisted. Away from gastrointestinal tubes or ostomies. Enough time after abdominal surgery, intestinal anastomosis, recovery of necrotizing enterocolitis, etcetera, so that the peritoneum is able to absorb CSF and for the shunt not to get infected from enteric flora.

(iii) Subcutaneous tunneling: trajectory needs to avoid closeness to tracheostomies, central lines, ostomies, tubes, etc.

(iv) Shunt selection: surgeon's preference. Regional and economic factors influence the decision. Antibiotic impregnated catheter, programmable valves and shunt assistant devices are more expensive.

(v) When the peritoneum is not an option: (From multiple surgeries and adhesions preventing CSF absorption, pseudo-cyst, peritonitis, NEC, etc.), the pleura (if older than 4 years old) or the atrium are considered. Ventriculoatrial shunt: the distal catheter is inserted through the external jugular vein, internal jugular vein or subclavian vein; the tip is positioned in the right atrium. Ventriculopleural shunt: Any part of the pleura can be used, typically below the 4–5th rib at the anterior axillary line.

(f) Surgery in brief: duration varies but commonly lasting between 30 and 60 min on a new insertion. Scalp incision is recommended to be curved so no part of the shunt is underneath it. Once the fontanels is small or closed, a burr hole is required when the entry is frontal. The dura mater is opened and the catheter inserted to the ventricle confirmed by obtaining CSF. The peritoneum can be opened before or after opening the dura mater and tunneling the system. In the case of the atrium or pleura, preferentially the ventricular catheter insertion and tunneling have been done. Tunneling is done with a special shunt tunneler. Prior to inserting the distal catheter, CSF flow is confirmed along the shunt system. Closure of the peritoneum around the shunt entry is not always required if the entry site is small.

(g) Postoperative care and follow up: observation in a floor room is adequate. Most patients can be discharged 24–48 h after surgery. Postoperative imaging is optional. Fast or short MRIs are preferred over CT to prevent radiation as these children will require many brain images in their lives. Analgesia is usually adequate with acetaminophen supplements if needed with NSAIDS or opiates. Close outpatient follow up the first year at any of 1, 3 and/or 6 months after surgery and then annually, bi-annually or per need. Annual images are not necessary.

Suggested Further Reading

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Circumcision

Jacob A. Baber, Alysia A. Agnoni, and Joel M. Sumfest

Circumcision has been performed for centuries dating back to 2100 BC for a variety of reasons including religion, rite of passage, and social norms. Routine circumcision is a widely debated topic. Although it is frequently performed soon after birth in the United States, it is currently not routinely indicated. Local anesthesia is recommended for all newborn circumcisions. Possible benefits of circumcision include reduction in urinary tract infections, sexually transmitted diseases, and penile cancer. Circumcision can be categorized into neonatal circumcisions, performed a few days after birth, and non-neonatal circumcisions, performed at months or years of age.

1. Physiology: The majority of males are born with fusion of the underside of the prepuce to the glans; the foreskin often hides the meatus. This fusion prevents the foreskin from being retractable. The natural process of separation of the prepuce and the glans occurs over months but may take several years. By puberty, the foreskin should be completely retractable.
2. Neonatal circumcision:

   (a) Indications:

      (i) Elective.
      (ii) Religious practice.

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3. Contraindications to neonatal circumcision:

   (a) Penile curvature.
   (b) Hypospadias or epispadias.
   (c) Ambiguous genitalia.
   (d) Dorsal hood deformity.
   (e) Buried and webbed penis.
   (f) Blood dyscrasias (relative contraindication).

4. Procedure:

   (a) Circumcision devices: These are primarily used in newborns. Common devices include: the Gomco clamp, Plastibell, or Mogen clamp.

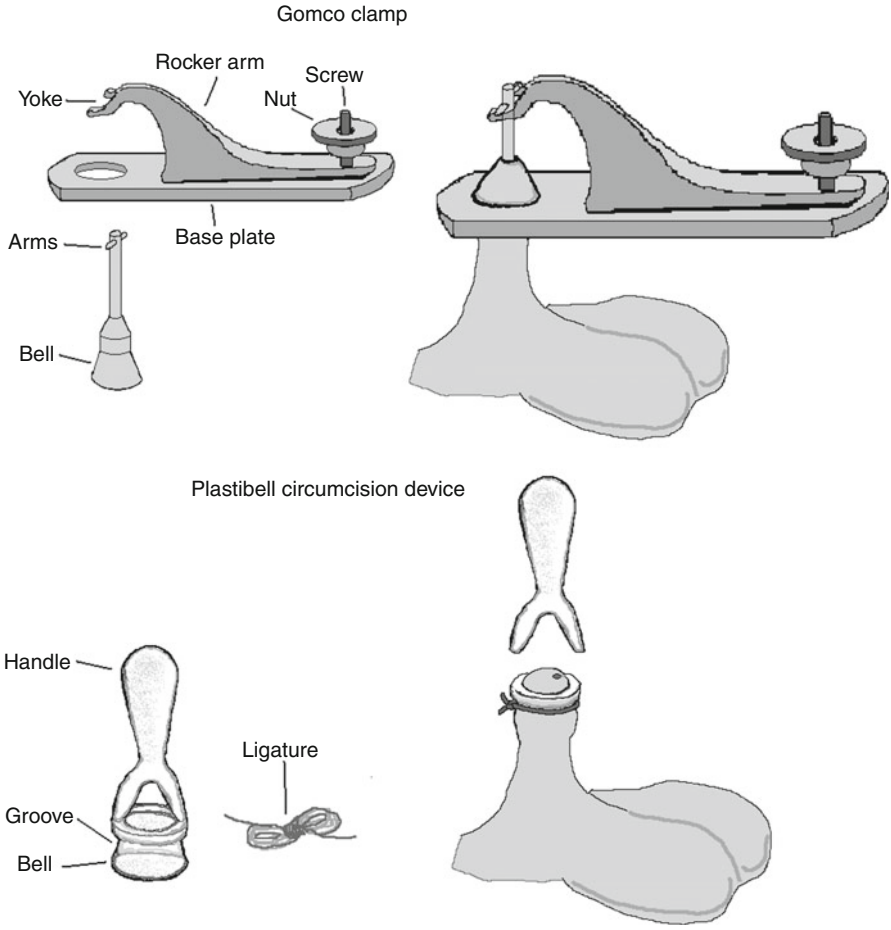

Fig. 1 Devices used to perform circumcision. The three most common devices used to perform circumcision are the Gomco clamp, the Plastibell circumcision device, and the Mogen clamp (*Source*: Christopher Coppola)

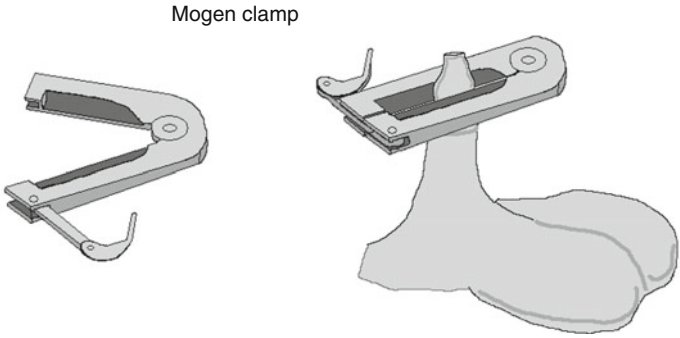

Fig. 1 (continued)

(i) General procedure guidelines:

1. Adequate anesthesia should be obtained: Tylenol, local anesthetic, etc.
2. Antisepsis: Sterile technique.
3. Adhesions between the glans and the underside of the foreskin are bluntly separated.

(ii) Gomco clamp: Most common device:

1. Named after Goldstein Medical Company.
2. A dorsal slit is made to allow for the bell device to fit over the glans and under the foreskin.
3. Bell is placed over the glans.
4. Foreskin is pulled over the bell.
5. The bell and overlying foreskin are brought through the clamp and the yoke is tightened, clamping the vessels and skin of the foreskin.
6. The device is removed after about 3–5 min.
7. The excess prepuce is then excised using a scalpel.
8. Antibiotic ointment is applied.

(iii) Plastibell: Uses strangulation to remove foreskin.

1. Dorsal slit is made to allow for Plastibell device to fit over glans and within the foreskin.
2. A sterile string is tied around the foreskin at a groove in the Plastibell near the glans to cut off the innervation and blood supply.
3. The excess skin is trimmed and the handle of the Plastibell snapped off the bell.
4. The remaining Plastibell and foreskin will slough off in the next 1–2 weeks.

(iv) Mogen clamp: Currently not performed due to significant complication risk (glanular amputation).

5. Non-neonatal circumcision:
   (a) Indications:
      (i) Phimosis.
      (ii) Infection of the foreskin or glans.
      (iii) Recurrent urinary tract infections.
      (iv) Balanitis xerotica obliterans.
   (b) Technique: Sleeve resection or "free-hand": Performed on older patients in the operating room.
      (i) A general anesthetic is used, the patient is prepped in the typical sterile manner, and the adhesions between the glans and foreskin are bluntly taken down.
      (ii) The prepuce is placed in its usual position and marked at the level of the coronal sulcus.
      (iii) Usually dorsal and ventral slits are made.
      (iv) The prepuce is then retracted and an incision is made circumferentially along the base of the glans. Dissection is carried down to Buck's fascia.
      (v) Hemostasis is achieved using low-amperage electrocautery.
      (vi) A second incision is made along the marked foreskin and this cuff of skin is excised.
      (vii) Care should be taken not to injure the urethra or ventral frenulum.
      (viii) The remaining cuff of skin is then approximated with interrupted absorbable sutures to the skin just below the glans at the coronal sinus.
6. Post-op care:
   (a) Some edema, bleeding, and crusting should be expected in the first few days.
   (b) Antibiotic ointment or a petroleum based ointment can be applied to the area.
   (c) Penis should be gently cleansed with soap and water as needed.
7. Complications:
   (a) Occur in 0.2–5 % overall.
   (b) Minor complications:
      (i) Bleeding.
         1. Most common (0.1 %).
         2. Seen more in older children/non-neonatal circumcision.
         3. Treatment:
            (a) Before operation, the provider must obtain detailed family history regarding bleeding disorders and treat underlying coagulopathy.
            (b) Apply direct pressure.

(c) Assess for a location of blood vessel (frenulum versus penile shaft), ligate if present.
(d) Apply Surgicel (cellulose polymer, Johnson & Johnson, Piscataway, NJ).
(e) Laboratory tests: complete blood count, protime, activated partial thromboplastin time, and specific coagulation factors as indicated by history.

(ii) Wound dehiscence:

1. More common after newborn circumcision.
2. Results from too much foreskin being pulled into the clamp and amputated.
3. Treatment:

(a) Local wound care.
(b) Allow to heal by secondary intention.

(iii) Wound infection:

1. Rare.
2. Treatment:

(a) Apply bacitracin to the circumcision site post-operatively.
(b) IV antibiotics if severe.

(iv) Penile scarring:

1. Can result in torsion, lateral deviation, or chordee.
2. Treatment:

(a) Scar can be softened with betamethasone and manual retraction.
(b) Formal repair may be necessary.

(v) Glanular adhesions and skin bridges:

1. Attachments between circumcision line and shaft or glans; most patients will have a suprapubic fat pad that may have contributed to the issue.
2. Treatment:

(a) Usually resolves on its own without treatment as suprapubic fat pad recedes and erections become more frequent and firmer.
(b) Persistent adhesions can be lysed in the office with local anesthetic.
(c) Skin bridges must be sharply divided after compression with a hemostat.
(d) Petroleum jelly can be used to prevent recurrences.

(vi) Concealed penis:

1. Results from too much removal of shaft skin with a concurrent prominent suprapubic fat pad ultimately causing healing within the fat pad.
2. Treatment:
   (a) Pressing on fat pad initially to determine how much skin to remove.
   (b) Compress fat pad regularly after the procedure to allow penis to protrude.
   (c) Operation may be warranted if inadequate protrusion of the penis.

(vii) Meatal stenosis:

1. Most occur secondary to inflammation or irritation; possibly related to ligation of frenular artery.
2. Symptoms include deviation of urinary stream, high velocity stream, or pain with voiding.
3. Treatment:
   (a) Meatotomy.
   (b) Meatoplasty.

(c) Major complications:

(i) Urethral injury/urethrocutaneous fistula:

1. Can occur with newborn circumcision due to compression injury or free-hand with ventral dissection and inadvertently injuring the urethra.
2. Treatment: Delayed flap repair.

(ii) Glanular necrosis:

1. Can be as a result of cautery injury during the use of clamp device.
2. Treatment:
   (a) Allow skin to slough and treat with local wound care.
   (b) May need formal repair with suprapubic diversion and delayed urethroplasty.

(iii) Glanular amputation:

1. Most commonly seen with the use of Mogen clamp circumcision.
2. Treatment: Primary repair has been successful with this injury.

Patent Ductus Arteriosus Ligation

Anastasios C. Polimenakos

Patent ductus arteriosus (PDA) is an abnormal communication between the aorta and the pulmonary artery. It occurs in 1 in 2,000 term births but it can be present in over 70 % of infants born at or prior to 32 weeks. There is a female predominance.

1. Pathophysiology:
   (a) Embryology [1]:
      (i) The PDA originates from the left sixth aortic arch. It is rarely absent or dual.
      (ii) The circulation in-utero differs markedly from postnatal circulation. Gas exchange occurs in the placenta, which receives blood from the umbilical arteries. Oxygenated blood then returns through the ductus venosus, which joins the inferior vena cava at the level of the hepatic veins. This blood is mixed with the venous return from the superior vena cava in the right atrium. Blood in the right atrium may be shunted across the foramen ovale to the left atrium, and through the left ventricle into the systemic circulation. Blood in the right atrium may also follow the right ventricle path and be pumped into the pulmonary artery. Patency of PDA in fetal life is maintained by regional and circulating prostaglandin. Pulmonary vascular resistance is very high in utero, and much of the blood pumped into the pulmonary artery returns through the PDA into the descending thoracic aorta. Because the PDA is large and communicates with the aorta, pressure in the pulmonary artery is the same as that in the aorta (systemic).

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(iii) After birth the umbilical cord is cut, thereby decreasing ductus venosus return to zero and causing the ductus venosus to constrict and obliterate. Pulmonary vascular resistance drops with expansion of the lungs, which causes pulmonary blood flow to increase. Because return to the left atrium is increased, left atrial pressure causes the foramen ovale to close. Increased oxygen partial pressure, decreased level of circulating prostaglandin and the interplay between circulating bradykinin, histamine, and catecholamines contribute on ductal constriction and, ultimately, its obliteration usually within 72 h from birth in terms neonates. Closure occurs with smooth muscle contraction of circular and longitudinal fibers within the media. Dense fibrous proliferation of the subintima permanently seals the lumen over 2–3 week period. The pulmonary and systemic circulations at this point become separate. In premature (less than 32 weeks) infants 88 % will close within the first 8 weeks.

(b) Anatomy and associated anomalies:

(i) PDA usually arises from the upper descending thoracic aorta and attaches to the main pulmonary artery at the origin of the left pulmonary artery. However, its size, shape and location might be variable. Different locations include, but not limited, the undersurface of the right aortic arch to the right pulmonary artery, the Kommerell's diverticulum in a vascular ring or the innominate/subclavian artery.
(ii) The PDA is in close proximity with left recurrent laryngeal nerve.
(iii) PDA may be associated with almost any congenital cardiac anomaly. One of the exceptions is tetralogy of Fallot with absent pulmonary valve where embryologically the PDA is absent in surviving fetuses.

2. Clinical features [2]:

(a) Patency of ductus arteriosus in premature babies usually involves a structurally normal ductus that fails to close because of the decreased response to rising oxygen tension and the presence of circulating mediators (e.g., prostaglandins) that maintain patency of the ductus. In a term baby, patency is caused by a structurally abnormal ductal wall.
(b) If a PDA is large, there is a large left-to-right shunt, with the pulmonary artery being exposed to systemic pressure. Initially, the additional volume work is handled by the left ventricle. Early diastolic systemic pressure reduction can compromise coronary flow and, especially in preterm infants, due to reversal flow from the abdominal circulation might lead to transient or persistent visceral malperfusion.
(c) With longstanding flow via a large PDA these children experience the same course of congestive heart failure followed by increased pulmonary vascular resistance as do children with a large ventricular septal defect. Eisenmenger's

syndrome may, then, develop in children with a large PDA with irreversible fixed elevated pulmonary vascular resistance and cyanosis.

(d) The usual clinical features of PDA are the signs and symptoms of left sided heart failure. Tachypnea at rest might be present early. The child might be prone to recurrent respiratory infections and failure to thrive is, commonly, present. Oxygen saturation remains normal and it might be affected only after evidence of elevated pulmonary vascular resistance. Patients, especially premature infants, might develop respiratory distress syndrome (RDS) secondary to left to right shunt. Endocarditis and Eisenmenger's syndrome are late complications. Eisenmenger's syndrome will be the end point of irreversible elevated pulmonary vascular resistance.

3. Because of the runoff from the aorta into the low-resistance pulmonary artery during diastole, diastolic pressure in children with a PDA tends to be low, with widened pulse pressure both by palpation, as well as, blood pressure measurements. Auscultation at left parasternal border or midscapular region will reveal a holosystolic murmur that often extends into diastole to give the typical machinery continuous murmur.

4. Diagnosis [1, 2]:

   (a) Chest x-ray: Plethoric congested lung fields with left atrial and ventricular enlargement.
   (b) Twelve-lead EKG: Might demonstrate increased left sided forces or/and signs of right ventricular hypertrophy later with persistent PDA.
   (c) Transthoracic echocardiogram: Gold standard for diagnosis and confirmation of shunt flow across the PDA.
   (d) Cardiac catheterization: Indicated only in older children with evidence of persistent PDA flow in order to evaluation pulmonary vascular resistance. If resistance is >75 % of systemic or unresponsive to nitric oxide and oxygen the patient should be considered inoperable without prior documentation of reversal of above findings.
   (e) MRI: Might be a useful adjunct to determine if PDA is associated with other congenital aortic arch anomalies where early surgical intervention will be advisable together with PDA closure.

5. Treatment [1, 2]:

   (a) Maintaining ductal patency:

      (i) Prostaglandins (prostacyclin [PGE1]) are useful for maintaining ductal patency in newborns. The indications may be for either right- or left-sided obstructive lesions.
      (ii) If a patient has a right-sided obstructive lesion (e.g., pulmonary atresia), pulmonary blood flow may be duct dependent. Prostaglandins may be used to keep the ductus patent until a surgically created systemic pulmonary artery shunt or definitive repair can be accomplished.

(iii) With left-sided lesions (e.g. interrupted aortic arch), blood flow to parts of the systemic circulation may be duct dependent; that is, before repair can be undertaken, the ductus is the only means by which part of the systemic circulation can receive blood flow. Closure of the ductus is lethal in the setting of a severe left sided obstruction, such as an interrupted aortic arch. In this situation, blood through the ductus flows from the pulmonary artery to the aorta.

(b) Medical treatment and therapeutic interventions: Three main strategies are available to ICU care providers for the treatment of PDA:

(i) Watchful waiting and fluid restriction: This strategy can be useful in extremely low birth weight premature infants (≤1,000 g; estimated gestational age 26 ± 3 weeks) who exhibit early signs of heart failure considering that over 30 % will demonstrate spontaneous closure within 4–5 days postnatally. Furosemide is not recommended as it is associated with an increase in treatment failure. Major advantage of this strategy is limiting infant's exposure to pharmacologic or surgical means of treatment and their risk. The major drawback is that, if this strategy fails, has a potential (>40 %) for diminishing efficacy of COX inhibitors.

(ii) Pharmacologic closure: PDA in premature babies usually involves a structurally normal ductus that fails to close because of the low oxygen tension and the effect of circulating mediators (e.g., increased prostaglandins: especially PGE_2) that maintain patency of the ductus. By changing the hormonal environment with non-selective COX inhibitors, such as intravenous indomethacin or ibuprofen lysine, which block prostaglandin production, a PDA in a premature infant may be closed. PDA closure can be achieved up to 95 %. Responsiveness to PGE_2 decreases with increased age. In a term baby ductal patency caused by a structurally abnormal ductal wall. COX inhibitors, therefore, have little effect on term infants and after significant delay in introducing therapy.

(iii) Surgical closure: Is indicated in premature infants, who had failed pharmacologic treatment, have hemodynamic compromise with contraindications to COX inhibitors, significant CHF, or in older patients with significant shunt across the PDA. Few reports associate surgical closure of PDA with development of chronic lung disease when compared to pharmacologic alternatives.

(c) When surgical closure of PDA is deemed necessary, then this can be accomplished by means of open or minimally invasive approach. In general, ducti less than 1.5 mm in diameter by echocardiographic measurements have limited short and long-term hemodynamic implications and do not warrant surgical closure after failure of pharmacologic treatment (even in preterm infants). Eisenmenger's syndrome is a contraindication for PDA closure.

(d) In open approach, small left posterolateral thoracotomy via 3rd or 4th intercostal space is performed. The serratus muscle is usually spared. The lung parenchymal is gently retracted and high frequency/low tidal volume ventilation is introduced to optimize exposure. The parietal pleura is entered and gentle dissection is performed superiorly and inferiorly to the ductus avoiding any manipulation of the PDA. Injury or rupture of the friable ductal wall can result in a catastrophic hemorrhage and patient's demise. Care must be practiced to identify and spare the left recurrent laryngeal nerve. Test occlusion of PDA to confirm anatomy (place pulse oximeter/blood pressure cuff on right arm and leg) is advised. Probably the commonest error is to misinterpret the left pulmonary artery as the ductus especially when PDA is frequently larger than the aortic arch isthmus and the left subclavian artery might be perceived as the aortic arch. Simple clip ligation is sufficient in neonates, ligation and division to prevent recanalization recommended in older children. In minimally invasive approach, three small (3 mm) ports are inserted via the 4th or 5th intercostal space between anterior and posterior axillary lines. The same principles followed in open approach should be honored for VATS surgical closure. The use of VATS approach in infants smaller than 1.0 kg might not be feasible. This approach is associated with superior cosmetic results and minimal discomfort.

(e) Complications, though rare, include: hemorrhage, inadvertent ligation of pulmonary artery or aortic isthmus instead of PDA, chylothorax, recurrent laryngeal nerve injury or ductal recanalization.

(f) Coil occlusion of the PDA might be entertained in relatively small and of adequate length (not an option in premature infants) ducti and usually for patients older than 2.0 kg.

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Tracheostomy

Evan B. Young

1. Indications:
   (a) Upper airway obstruction that cannot be controlled medically or with alternative means.
      (i) Congenital (craniofacial anomalies such as Pierre-Robin sequence, etc.)
      (ii) Acquired (subglottic stenosis, iatrogenic bilateral vocal cord paralysis, etc.)
   (b) Need for prolonged ventilation (neurologic, cardiopulmonary dysfunction, etc.)
2. Medical, alternative, preventative and temporizing strategies, depending on etiology.
   (a) Medical:
      (i) Supplemental O_2, anti-reflux medications, antibiotics, racemic epinephrine.
   (b) Alternative surgical:
      (i) Microlaryngeal surgery – balloon dilation, debridement, supraglottoplasty.
   (c) Preventative:
      (i) Anti-reflux medications, avoiding high cuff pressures on endotracheal tube.

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(d) Temporizing:

(i) Endotracheal intubation, nasopharyngeal airway, heliox.

3. Assessment/preoperative evaluation:

(a) Multidisciplinary discussion with parents, intensivists, other consultants, and social workers.
(b) Preoperative counseling:

(i) Requires frequent attention and care, usually by the primary caregiver.

1. May not be permanent and can be removed, leaving a scar.

(ii) Potential complications:

1. Major: Tube dislodgement, false passage, mucous plugging, tracheal injury, tracheoinnominate fistula, pneumothorax, death.
2. Minor: Stoma breakdown, scarring, suprastomal granulations, persistent tracheocutaneous fistula, tracheal granulations or ulcerations.

(c) Evaluation:

(i) Unless urgently needed, the child should be medically optimized for surgery with stable/low ventilator settings, normal coagulation parameters, removal of obstructing medical devices.
(ii) Palpate laryngotracheal landmarks, noting previous scars and cervicofacial anatomy.

4. Surgical procedure:

(a) Anatomic considerations:

(i) Younger patients have smaller tracheas, a higher larynx, soft tracheal arches and difficult to palpable landmarks.

(b) Positioning:

(i) Supine, with a shoulder roll and neck in extension (tape submental area if necessary).
(ii) Mark cricoid and sternal notch.

(c) Skin incision below the cricoid, horizontal or vertical depending on surgeon's preference.
(d) Remove a core of subcutaneous fat for exposure.
(e) Frequent palpation of trachea is critical as midline dissection prevents surgical misadventure.
(f) Strap muscles are swept laterally and thyroid is divided with electrocautery or swept cephalad.

(g) Two non-absorbable stay sutures placed lateral to planned vertical incision (2 tracheal arches, ideally between arches 2–4).

  (i) Caution (airway fire): When using high oxygen concentration, electrocautery should be avoided after the airway has been entered.

(h) Using the stay sutures for gentle lateral retraction, anesthesia slowly withdraws the existing orotracheal tube under direct visualization and stopping when just above tracheostomy.
(i) Place appropriately sized tracheotomy tube: Once correct placement is confirmed it is secured with a snug tie around the neck (1 finger insertion) and four flange sutures (optional).
(j) Remove orotracheal tube (1 tube → 2 tubes → 1 tube).

5. Postoperative care:

(a) Immediate chest x-ray (looking for PTX).
(b) Spare tracheostomy tube, smaller tracheostomy tube, suction, suture removal kit; lubrication should ALWAYS be at the bedside. Cuff should be down when pressure support is not needed.
(c) Using stay sutures for retraction, the tube is changed around post-operative day 5 and the stay sutures are removed.

Part IX
Resources

Note Templates

Christopher P. Coppola

1. Admission orders
   (a) Admit
   (b) Admitting service/physician/location
   (c) Covering resident(s) and contact information
   (d) Diagnosis
   (e) Condition (stable, fair, poor, critical, moribund, etc.…)
   (f) Vital sign frequency (also neurologic checks, pulse checks, weights, body part diameters, pulse oximetry)
   (g) Continuous monitoring/telemetry needed
   (h) Activity (bed rest, out of bed to chair, ambulate, assistance needed, fall risk, seizure prophylaxis)
   (i) Allergies
   (j) Nursing procedures
      (i) Wound and dressing care/changes
      (ii) Parameters for which to call surgeon (temperature, pulse, blood pressure, respiratory rate, urine output, change in examination, function, or mental status)
      (iii) Tubes and drains (both placement and monitoring of output)
      (iv) Tubes/orifices which cannot be manipulated, any signs to post over bed
      (v) Compression hose and/or sequential compression devices
   (k) Diet or NPO orders
   (l) In/Out monitoring and frequency
   (m) Intravenous access and fluids

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(n) Medication

(i) Scheduled medication
(ii) Continuous infusions
(iii) Symptomatic medication
(iv) Deep venous thrombosis prophylaxis
(v) Stress ulcer prophylaxis
(vi) Home medications to stop/continue
(vii) Sliding scale for insulin

(o) Laboratory specimens to collect/process
(p) Blood bank orders for type and cross or transfuse
(q) Diagnostic imaging to obtain
(r) Electrocardiogram
(s) Services to consult
(t) Respiratory therapy orders
(u) Physical/occupational therapy orders

2. Preoperative note and checklist

(a) Diagnosis
(b) Planned Procedure(s)
(c) Surgeon(s)
(d) Consent form status
(e) Anesthesia evaluation requested/accomplished
(f) Allergies
(g) Vital signs
(h) Relevant laboratory values

(i) Serum studies
(ii) Evaluation of coagulation
(iii) Urine
(iv) Blood bank type and cross or type and hold

(i) Relevant diagnostic imaging
(j) Electrocardiogram and other cardiac studies
(k) Evaluation of pulmonary function: pulmonary function test, arterial blood gas
(l) NPO order status and timing
(m) Intravenous fluids
(n) Preoperative antibiotics
(o) Bowel preparation
(p) Medications to continue/hold for operation

(i) Anticoagulation
(ii) Cardiac medication
(iii) Insulin

(q) Skin preparation/bathing

3. Brief operative note
    (a) Patient name/record number
    (b) Date
    (c) Location
    (d) Preoperative diagnosis
    (e) Operation
    (f) Postoperative diagnosis
    (g) Surgeon
    (h) Assistant(s)
    (i) Anesthesia type
    (j) Findings
    (k) Fluids
        (i) Intravenous fluid/blood volume administered
        (ii) Estimated blood loss
        (iii) Urine output
    (l) Specimens for pathology and laboratory, intraoperative frozen section pathology report
    (m) Drains and locations
    (n) Diagram if necessary
    (o) Condition
    (p) Disposition location
    (q) Physician responsible for operative report
4. Operative report
    (a) Patient name/record number
    (b) Date of operation and operative report
    (c) Dictating physician
    (d) Location
    (e) Preoperative diagnosis
    (f) Operation
    (g) Postoperative diagnosis
    (h) Attending surgeon and service
    (i) Assistant(s)
    (j) Findings
    (k) Intravenous fluid/blood volume administered
    (l) Estimated blood loss
    (m) Urine output
    (n) Specimens for pathology and laboratory, pathology results if available
    (o) Intraoperative consultation
    (p) Drains and tubes
    (q) History and Indications
    (r) Consent obtained and Documented
    (s) Anesthetic used

(t) Antibiotics given
(u) Wound class
(v) Complications
(w) Deep venous thrombosis prophylaxis
(x) Positioning and padding
(y) Skin preparation
(z) Documentation of final time out

(aa) Narrative description of operation
(ab) Condition
(ac) Disposition location
(ad) Attestation of attending surgeon presence and participation
(ae) Documentation of modifiers to coding

5. Postoperative note

(a) Patient identification
(b) Time and location
(c) Diagnosis
(d) Operation performed
(e) Elapsed Time since operation (4–6 h is appropriate)
(f) Narration of events since operation
(g) Subjective account of patient's symptoms
(h) Control of pain
(i) Vital signs
(j) Intake of diet, tube feedings, and intravenous fluid
(k) Output of urine, tube drainage, vomiting, stool/stoma output
(l) Examination of wound and/or dressing, and drains
(m) Evaluation of distal neurovascular perfusion and function
(n) Postoperative lab results
(o) Overall assessment and plan

6. Progress note

(a) Patient identification
(b) Location
(c) Hospital day/postoperative day
(d) Diagnosis
(e) Operation
(f) Events of past 24 h
(g) Subjective account of patient's symptoms
(h) Vital signs
(i) Intake and output
(j) Diet
(k) Medications

(i) Continuous infusions
(ii) Antibiotic date, duration, indication for treatment
(iii) Symptomatic medication usage

(l) Stress ulcer prophylaxis
(m) Deep venous thrombosis prophylaxis
(n) Decubitus prophylaxis
(o) Head to toe examination
(p) Operative wound or dressing evaluation
(q) Evaluation of distal neurovascular perfusion and function
(r) Lab results
(s) Diagnostic imaging results
(t) Consultation reports
(u) Injury survey, if trauma
(v) Assessment, system-based if critical care

(i) Neurologic
(ii) Respiratory/ventilator
(iii) Cardiac/circulation
(iv) Renal
(v) Fluids/Electrolytes/Nutrition
(vi) Gastrointestinal
(vii) Hematology
(viii) Immune/infectious disease
(ix) Endocrine
(x) Psychological/social
(xi) Extremities

(w) Plan

(i) Goals
(ii) Diet
(iii) Pain control
(iv) Antibiotics
(v) Consultations

(x) Discharge planning and estimated date of discharge

7. Discharge summary

(a) Patient identification
(b) Physician completing summary
(c) Location
(d) Type of admission
(e) Admission date
(f) Discharge date
(g) Diagnosis, primary and secondary
(h) Discharging service
(i) Discharging physician
(j) Disposition destination
(k) Condition on discharge
(l) Diet
(m) Activity

(n) Medications on discharge
(o) Future appointments
(p) Pending test results
(q) Future testing
(r) Brief admission history and physical

(i) Complaint
(ii) History of present illness
(iii) Past medical/surgical history
(iv) Mediations/allergies
(v) Social history and family history
(vi) Birth history and vaccinations for infants

(s) Hospital course
(t) Procedures performed
(u) Complications
(v) Laboratory test results
(w) Diagnostic imaging results
(x) Discharge plan
(y) Providers who should receive discharge summary

Common Medications

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- Acetaminophen (Tylenol): Non-narcotic analgesic, antipyretic.
  - <12 years old:
    - Oral: 10–15 mg/kg/dose every 4–6 h as needed; do not exceed 5 doses in 24 h.
    - Rectal: 10–20 mg/kg/dose every 4–6 h as needed; do not exceed 5 doses in 24 h.
  - 12 years and older:
    - Oral: 325–650 mg every 4–6 h or 1 g 3–4 times/day (max dose 4 g/day).
- Albuterol (AccuNeb, ProAir HFA, Proventil HFA, Ventolin HFA, VoSpire ER): Beta2-Adrenergic agonist agent, bronchodilator.
  - Acute exacerbation, asthma (children):
    - Inhalation: 90 mcg/spray, 4–8 puffs every 20 min for 3 doses then every 1–4 h.
    - Nebulization: 0.15 mg/kg (minimum dose 2.5 mg) every 20 min for 3 doses then 0.15–0.3 mg/kg (not to exceed 10 mg) every 1–4 h as needed or 0.5 mg/kg/h by continuous nebulization.

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- Amoxicillin and clavulanate potassium (Augmentin): Antibiotic, Beta-lactam and Beta-lactamase combination, Penicillin.
  - General dosing:
    - Infants 1–3 months: 30 mg amoxicillin/kg/day orally in divided doses every 12 h using the 125 mg/5 mL suspension.
    - Infants 3 months and older, children, and adolescents: 25–45 mg amoxicillin/kg/day in divided doses twice daily. Max single dose 875 mg amoxicillin.
- Ampicillin: penicillin antibiotic.
  - Pediatric:
    - IV/IM: 100–400 mg/kg/day divided q6 h (max 8 g/day).
    - PO: 50–100 mg/kg/day divided q6 h.
  - Adult:
    - IV/IM: 1–2 g q4-6 h.
    - PO: 250–500 mg q6 h.
- Ampicillin Sulbactam (Unasyn): Beta-lactam and Beta-lactamase combination, antibiotic, penicillin.
  - Pediatric general dosing:
    - <40 kg: 100–200 mg ampicillin/kg/day IM/IV divided every 6 h (max 1,000 mg/dose).
    - >40 kg: 1.5–3 g IM/IV every 6 h (max 12 g/day).
  - Adult general dosing:
    - 1.5–3 g IM/IV every 6 h (max 12 g/day).
- Aspirin: Analgesic, antipyretic, Nonsteroidal Anti-Inflammatory Drug, Salicylate.
  - Typically aspirin is avoided in children less than 12 years who actively exhibit flu symptoms or are recovering from chicken pox due to association with Reye's syndrome.
  - Analgesia:
    - Infants, Children, Adolescents < 50 kg: 10–15 mg/kg/dose every 4–6 h (max daily dose is lesser value of either 120 mg/kg/day or 4,000 mg/day).
    - Children 12 years and older > 50 kg: 325–650 mg every 4–6 h (max daily dose 4,000 mg/day).
- Bisacodyl (Dulcolax): Laxative, stimulant.
  - Pediatric:
    - Tablet: 3–12 years: 5–10 mg once daily.
    - Suppository:
      - <2-years-old: 5 mg once daily.
      - 2–11-years-old: 5–10 mg once daily.

  - Adults:
    - Tablet: 12-years-old and older: 5–15 mg once daily.
    - Suppository: 12-years-old and older: 10 mg once daily.
- Cefazolin (Ancef): First generation cephalosporin antibiotic.
  - Pediatric:
    - IV/IM: 25–150 mg/kg/day divided q8 h (max 6 g/day).
  - Adult:
    - IV/IM: 250–1,000 mg q6-8 h.
- Cefepime (Maxipime): Fourth generation cephalosporin antibiotic.
  - Pediatric:
    - IV/IM: 2 months-16 years <40 kg: 50 mg/kg q12 h (max 2 g/dose).
  - Adult:
    - IV: 2 g IV q12 h.
- Cefoxitin (Mefoxin): Second generation cephalosporin antibiotic.
  - Pediatric:
    - IV/IM: >3 months: 80–160 mg/kg/day divided q4-8 h (max dose 12 g/24 h).
  - Adult:
    - IV/IM: 1–2 g q6-8 h (max 6–8 g/day).
- Ceftriaxone (Rocephin): Third generation cephalosporin antibiotic.
  - Pediatric:
    - Mild to moderate infection: 50–75 mg/kg/dose IV/IM once daily (max single dose 1,000 mg).
    - Severe infection: 100 mg/kg/day IV/IM divided every 12–24 h.
- Cephalexin (Keflex): First generation cephalosporin antibiotic.
  - Pediatric:
    - PO: 25–100 mg/kg/day divided q6 h (max 4 g/day).
  - Adult:
    - 250–500 mg q6 h.
- Cholestyramine (Prevalite, Questran): Antilipemic agent, bile acid sequestrant.
  - PO: 4 g powder BID mix with water/food.
  - Topical: mix with ILEX paste.

- Ciprofloxacin (Cipro): Quinolone antibiotic.
  - Pediatric:
    - IV: 6–10 mg/kg q8 h (max 400 mg/dose).
    - PO: 10–20 mg/kg BID (max 750 mg/dose).
  - Adult:
    - IV: 400 mg BID.
    - PO: 500 mg BID.
- Clindamycin (Cleocin): Antibiotic, anaerobic.
  - Pediatric:
    - PO: 10–40 mg/kg/day divided TID (max 1,800 mg/day).
    - IV/IM: 20–40 mg/kg/day divided TID-QID.
  - Adult:
    - PO: 150–450 mg q6 h.
    - IV/IM: 1.2–2.7 mg divided BID-TID.
- Dexmedetomidine (Precedex): Alpha-adrenergic agonist agent, sedative.
  - Pediatric:
    - Loading dose: 0.5–1 mcg/kg IV.
    - Maintenance: 0.2–0.7 mcg/kg/h (children < 1-years-old may require higher infusion rates).
  - Adult:
    - Loading dose: 1 mcg/kg.
    - Maintenance: 0.2–0.7 mcg/kg/h.
- Diphenhydramine (Benadryl): antihistamine, sedative.
  - Pediatric:
    - PO/IV/IM:
      - 2–5-years-old: 6.25 mg q4-6 h (max 37.5 mg/day).
      - 6–11-years-old: 12.5–25 mg q4-6 h (max 150 mg/day).
      - 12-Adult: 25–50 mg q4-6 h (max 300 mg/day).
  - Adult:
    - PO: 25–50 mg every 6–8 h.
    - IV/IM: 10/50 mg per dose; single doses up to 100 mg (max 400 mg/day).

- Docusate (Colace): laxative, surfactant, stool softener.
  - Pediatric:
    - Liquid:
      - <3-years-old: 10–40 mg daily.
      - 3–6-years-old: 20–60 mg daily.
      - 6–12-years-old: 40–150 mg daily.
    - Tablet:
      - <2-years-old: 25 mg daily.
      - 2–12-years-old: 50–150 mg daily.
  - Adult:
    - Tablet: 50–500 mg daily-QID.
- Ertapenem (Invanz): Carbapenem antibiotic.
  - Pediatric:
    - 3 months- 12-years-old: 15 mg/kg/dose q12 h (max 1 g/day) IV or IM.
  - Adult:
    - IV/IM: 1 g daily.
- Erythromycin ethylsuccinate (E.E.S, Eryped): Macrolide antibiotic, prokinetic agent.
  - GI motility agent dosing:
    - Pediatric:
      - PO: 3 mg/kg/dose q6 h (max 10 mg/kg/dose).
    - Adult:
      - PO: 400–800 mg q6-12 h.
- Fentanyl: Narcotic analgesic.
  - Pediatric:
    - Analgesia/sedation: IV: 0.5–4 mcg/kg/dose IV q1-2 h.
  - Adult:
    - Analgesia/sedation: IV/IM: 1–2 mcg/kg q30-60 min.
    - Severe pain: IV/IM: 50–100 mcg/dose q1-2 h.

- Fluconazole (Diflucan): Antifungal agent.
  - Pediatric:
    - Systemic: PO/IV: 6–12 mg/kg/day.
    - Oropharyngeal: PO/IV: 6 mg on day 1 then 3 mg/kg/day.
  - Adult:
    - PO/IV: 200 mg on day 1 then 100 mg/day.
- Flumazenil (Romazicon): benzodiazepine antidote.
  - Benzodiazepine reversal for sedation/anesthesia in children:
    - 0.01 mg/kg (maximum dose: 0.2 mg) given over 15 s; may repeat 0.01 mg/kg (maximum dose: 0.2 mg) after 45 s, and then every minute to a maximum total cumulative dose of 0.05 mg/kg or 1 mg, whichever is lower; usual total dose: 0.08–1 mg.
  - Benzodiazepine overdose in children:
    - Initial dose 0.01 mg/kg (max 0.2 mg) with repeat doses of 0.01 mg/kg (max 0.2 mg) given every minute to a maximum total cumulative dose of 1 mg.
- Furosemide (Lasix): Loop diuretic.
  - Pediatric:
    - Oral: 2 mg/kg once daily. May increase by 1–2 mg/kg/dose every 6–8 h (max 6 mg/kg/dose).
    - IV/IM: 1–2 mg/kg/dose every 6–12 h.
    - Continuous infusion: 0.05 mg/kg/h.
- Gentamicin: Aminoglycoside antibiotic.
  - Pediatric:
    - IV/IM:
      - <5-years-old: 2.5 mg/kg/dose q8 h.
      - 5–12-years-old: 2–2.5 mg/kg/dose q8 h.
  - Adult:
    - IV/IM: 1–2.5 mg/kg/dose q8-12 h.
- Heparin-Saline for line flushing.
  - 2–5 mL/flush depending on catheter size.
  - Dose of heparin flush used should not approach therapeutic unit per kg dose.
  - Heparin flush should be given stagnant blood is observed in catheter, after used for drug or blood administration, or after blood withdrawal from catheter.

  - Concentrations typically used to assure patient safety based on size.
    - <10 kg: 10 units/mL concentration.
    - >10 kg: 100 units/mL concentration.

- Hydromorphone (Dilaudid): Narcotic analgesic, antitussive.
  - Pediatric:
    - PO: 0.03–0.08 mg/kg q4-6 h.
    - IV/IM/PR/SC: 0.015 mg/kg q4-6 h.
  - Adult:
    - PO: 2–4 mg q4-6 h.
    - SC/IM: 1–2 mg q4-6 h.
    - IV: 0.2–0.6 mg q2-3 h.
  - Pediatric PCA:
    - Basal rate (mg/kg/h): 0.003–0.005.
    - Bolus dose (mg/kg): 0.003–0.005.
    - Lockout period (minutes): 6–10.
    - Boluses/h: 4–6.
    - Max dose per hour (mg/kg): 0.015–0.020.
- Ibuprofen (Motrin, Advil): Nonnarcotic analgesic, Nonsteroidal Anti-Inflammatory Drug.
  - Pediatric:
    - PO: 5–10 mg/kg/dose q6-8 h (max 40 mg/kg/day).
  - Adult:
    - PO: 300–800 mg q6 h.
- Ketamine (Ketalar): General anesthetic, noncompetitive NMDA receptor antagonist.
  - Oral: 6–10 mg/kg for 1 dose given 30 min prior to procedure.
  - IM: 3–7 mg/kg.
  - IV: 0.5–2 mg/kg or for minor procedures use 0.5–1 mg/kg. Usual induction dose 1–2 mg/kg.
  - Sedation: 5–20 mcg/kg/min.
- Ketoconazole: Antifungal agent.
  - Pediatric:
    - Infants and children: 3.3–6.6 mg/kg/day once daily (max 800 mg/day).
  - Adults:
    - 200–400 mg/day once daily (max 800 mg/day).

- Lactulose: Ammonium detoxicant, laxative.
  - Pediatric:
    - PO: 0.7–2 g/kg/day (max dose 40 g/day).
  - Adults:
    - PO: 10–20 g daily.
- Lansoprazole (Prevacid): Gastric acid secretion inhibitor, proton pump inhibitor.
  - Pediatric (oral):
    - <10 weeks: 0.2–0.3 mg/kg/dose once daily.
    - >10 weeks: 1–2 mg/kg/dose once daily.
    - 1–11 years: <30 kg: 15 mg daily for up to 12 weeks. >30 kg: 30 mg daily for up to 12 weeks.
    - Children > 12 years: 15–30 mg once daily for up to 8 weeks.
  - Adult (oral):
    - 15–30 mg once daily for up to 8 weeks.
- Lorazepam (Ativan): Benzodiazepine.
  - Anxiety:
    - Oral: 0.05 mg/kg/dose (max 2 mg/dose) every 4–8 h.
  - Sedation:
    - IV: 0.01–0.03 mg/kg and repeat every 20 min, titrate to effect.
  - Status epilepticus:
    - Infants and Children: 0.05–0.1 mg/kg (max 4 mg/dose) slow IV over 2–5 min (max rate 2 mg/min). May repeat every 10–15 min.
    - Adolescents: 0.07 mg/kg (max 4 mg/dose) slow IV over 2–5 min (max rate 2 mg/min). May repeat in 10–15 min. usual total dose 8 mg.
- Meropenem (Merrem): Carbapenem antibiotic.
  - Pediatric:
    - IV: 10–20 mg/kg q8 h. (Max 3 g per day).
  - Adult:
    - IV: 0.5–1 g q8 h.

- Metronidazole (Flagyl): Amebicide, antiprotozoal, anaerobic antibiotic.
  - Pediatric general dosing:
    - Oral: 30–50 mg/kg/day divided three times daily (max 2,250 mg/day).
    - IV: 22.5–40 mg/kg/day divided three times daily (max 1,500 mg/day).
  - Adult general dosing:
    - Oral/IV: 30 mg/kg/day divided every 6 h (max 4,000 mg/day).
- Midazolam (Versed): Benzodiazepine.
  - Pediatric sedation/amnesia:
    - Oral:
      - >6 months, children, and adolescents: 0.25–0.5 mg/kg once (max dose 20 mg).
    - Rectal:
      - >6 months and children: 0.25–0.5 mg/kg once (max dose 20 mg).
    - IV:
      - 6 months to 5 years old: Initial 0.05–0.1 mg/kg carefully titrated. Usual total dose 6 mg.
      - 6–12 years: Initial 0.025–0.05 mg/kg carefully titrated. Usual total dose 10 mg.
      - 12–16 years: Dose as adults, usual total max dose is 10 mg.
- Morphine: Opioid analgesic.
  - Pediatric:
    - Oral: 0.08 mg/kg/dose every 4–6 h.
    - IM, IV, SC: 0.05–0.1 mg/kg/dose every 4–6 h (max 0.1 mg/kg/dose).
    - Continuous IV: 0.01 mg/kg/h (max 0.03 mg/kg/h).
    - PCA:
      - Basal rate (mg/kg/h): 0.01–0.03.
      - Bolus dose (mg/kg): 0.01–0.03.
      - Lockout period (minutes): 6–10.
      - Boluses/h: 4–6.
      - Max dose per hour (mg/kg): 0.1–0.15.
- Nitrofurantoin (Macrobid, Macrodantin, Furadantin): antibiotic.
  - UTI:
    - Infants > 1 month and children: 5–7 mg/kg/day divided every 6 h (max 400 mg/day).

  - UTI prophylaxis:
    - Infants > 1 month and children: 1–2 mg/kg qHS or 2 divided doses.
- Omeprazole (Prilosec): Gastric acid secretion inhibitor, proton pump inhibitor.
  - Infants: 0.7 mg/kg/dose orally once daily.
  - Infants and children (oral):
    - 5–10 kg: 5 mg once daily.
    - 10–20 kg: 10 mg once daily.
    - >20 kg: 20 mg once daily.
  - Adults: 20–40 mg orally once daily.
- Phenobarbital: Barbiturate, anticonvulsant.
  - Pediatric loading dose:
    - Infants and children: 15–20 mg/kg (max 1,000 mg/dose). May repeat dose after 15 min as needed.
  - Pediatric maintenance dose:
    - Infants: 5–6 mg/kg/day in 1–2 divided doses.
    - 1–5 years: 6–8 mg/kg/day in 1–2 divided doses.
    - 5–12 years: 4–6 mg/kg/day in 1–2 divided doses.
- Phenytoin (Dilantin): Anticonvulsant.
  - Initial pediatric seizure prophylaxis for traumatic brain injury:
    - Variable guidelines regarding dosing. In general, use the standard listed below.
    - 18 mg/kg IV over 20 min followed by 6 mg/kg/day divided every 8 h for 48 h.
  - Maintenance prophylaxis:
    - 6 months – 3 years: 8–10 mg/kg/day.
    - 4–6 years: 7.5–9 mg/kg/day.
    - 7–9 years: 7–8 mg/kg/day.
    - 10–16 years: 6–7 mg/kg/day.
- Polyethylene glycol (Miralax, Glycolax): Laxative, Osmotic.
  - Children > 6 months:
    - Maintenance dosing: 0.5–1.5 g/kg daily.
    - Bowel preparation: Children > 2-years-old: 1.5 g/kg/day (max daily dose 100 g).

  - Adults:
    - Maintenance: 17 g daily.
    - Bowel preparation: 17 g in 8 oz. liquid and administer every 10 min until 2 l are consumed. Consider also giving Bisacodyl<6 h prior to bowel preparation.
- Ranitidine (Zantac): Histamine H2 Antagonist.
  - Pediatric:
    - 1 month to 16 years: Oral 4–10 mg/kg/day divided twice daily (max 300 mg/day).
  - Adult:
    - 16-years-old and older: 150 mg/dose twice daily or 300 mg at bedtime.
- Trimethoprim-sulfamethoxazole (Bactrim): Sulfonamide antibiotic.
  - Pediatric:
    - PO: 8–12 mg/kg/day divided BID (max 160 mg/dose).
  - Adult:
    - Bactrim DS: 1 tab PO daily.
- Vancomycin: Antibiotic.
  - Pediatric:
    - Mild to moderate infection: 40–45 mg/kg/day divided every 6–8 h (usual max daily dose 2,000 mg/day).
    - Severe infection: 45–60 mg/kg/day divided every 6–8 h (usual max daily dose 4,000 mg/day).
- Electrolyte *Repletion* (not to be used for maintenance):
  - Calcium:
    - Notes:
      - Hypomagnesemia is a common cause of hypocalcemia. If serum magnesium concentration is low, this should be replaced prior to calcium repletion in most cases.
      - If IV calcium in required, calcium gluconate is typically preferred over calcium chloride because it is less likely to cause tissue necrosis if extravasated.
      - Milder cases of hypocalcemia may be treated with oral elemental calcium given as calcium carbonate or calcium citrate.

- Calcium gluconate:
  - IV: 200–500 mg/kg/day as a continuous infusion or in 4 divided doses (max 2–3 g/dose).
- Calcium carbonate (Tums):
  - Oral: 45–65 mg/kg/day (elemental calcium) in 4 divided doses.
- Calcium citrate (Calcitrate OTC):
  - Oral: 45–65 mg/kg/day (elemental calcium) in 4 divided doses.

– Hypertonic saline:
- Patients with serum sodium less than 125 mEq/L:
  - Signs and symptoms: CNS symptoms, lethargy, seizures.
  - Rx: hypertonic saline (3 % sodium chloride) using equation based on sodium deficit: (desired serum sodium – current serum sodium concentration) × 0.6 × (weight in kg).
  - Typically given over a few hours with serum sodium checks throughout.
  - Do NOT correct sodium faster than 12 mEq/L within 24 h.

– Magnesium sulfate:
- Notes:
  - Up to 50 % of infused magnesium will be excreted in the urine due to inhibition of magnesium reabsorption in the loop of Henle caused by the infusion.
  - If given IV, beware of respiratory depression, hypotension, heart block, or hypermagnesemia. Calcium gluconate via IV may be used as antidote.
  - Normomagnesemic magnesium depletion (cellular magnesium depletion) should be considered as a possible cause of refractory hypokalemia or unexplained hypocalcemia.
- Children with severe symptoms:
  - IV: 25–50 mg/kg (0.2–0.4 mEq/kg) every 4–6 h for 3–4 doses, with max single dose of 2 g (16 mEq).
- Children with minimal or no symptoms (hypomagnesemia or hypocalcemia):
  - Oral: 100–200 mg/kg/dose four times per day.

– Potassium chloride:
- Notes:

  - IV potassium should be reserved for severe depletion, and continuous ECG monitoring should be utilized. Monitor serum potassium closely.
  - Severe depletion or ongoing losses may require > 200 % of normal daily limit needs.
  - Consider alternative solutions for atypical presentations: potassium bicarbonate for hypokalemia with metabolic acidosis or potassium phosphate for hypokalemia with hypophosphatemia.

- Treatment of Hypokalemia (infants and children):

  - Oral: 2–5 mEq/kg/day in divided doses (max single dose 2 mEq/kg).
  - Intermittent IV infusion (diluted): 0.5–1 mEq/kg/dose (max dose: 40 mEq) to infuse at 0.3–0.5 mEq/kg/h (max dose/rate: 1 mEq/kg/h).

– Phosphate:

- Notes:

  - Asymptomatic patients with serum phosphate less than 2 mg/dL may be treated with oral phosphate.
  - Serum phosphate levels below 1 mg/dL are typically treated with IV phosphate, switching to oral therapy when serum phosphate exceeds 1.5 mg/dL.
  - Consider halting repletion when serum phosphate is greater than or equal to 2 mg/dL.
  - Contact Children's pharmacy for available/appropriate IV dilution.

- IV: 15–45 mg/kg over 24 h.
- Oral: 30–90 mg/kg/24 h divided TID-QID.

Vaccination Schedule

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1. Vaccination schedule and supporting information
2. Source: ACIP Childhood/Adolescent Immunization Work Group: Akinsanya-Beysolow I, Jenkins R, Meissner HC. Advisory Committee on Immunization Practices (ACIP) Recommended Immunization Schedule for Persons Aged 0 Through 18 Years—United States, 2013, Morbidity and Mortality Weekly Report (MMWR), 62:2–8, 2013, Centers for Disease Control and Prevention.
3. The most recent vaccination information can be found at : http://www.cdc.gov/vaccines
4. **Fig. 1** Recommended immunization schedule for persons aged 0 through 18 years—2013
5. **Fig. 2** Catch-up immunization schedule for persons aged 4 months through 18 years who start late or who are more than 1 month behind—United States, 2013
6. Full footnotes available at: http://www.cdc.gov/mmwr/preview/mmwrhtml/su6201a2.htm
7. Vaccines, minimum age of administration, and recommended schedule
   (a) Hepatitis B (HepB)
      (i) Minimum age: birth
      (ii) Birth/1 month/6 months
   (b) Rotavirus (RV)
      (i) Minimum age: 6 weeks
      (ii) 2 months/4 months/6 months (if RV-5 used)

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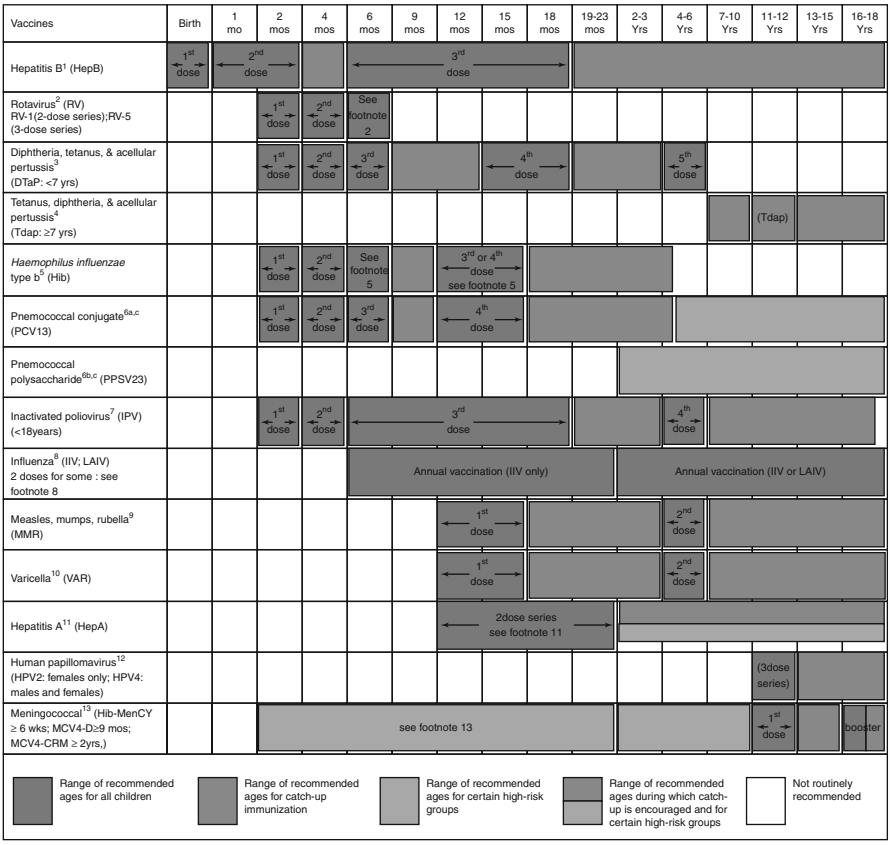

Fig. 1 Vaccination schedule for children 0–18 years old (From Centers for Disease Control, Atlanta, GA. http://www.cdc.gov/mmwr/preview/mmwrhtml/su6201a2.htm. Downloaded 31 Dec 2013)

(c) Diphtheria/tetanus toxoids/acellular pertussis (DtaP)

(i) Minimum age: 6 weeks
(ii) 2 months/4 months/6 months/15 months/4 years

(d) Tetanus and diphtheria toxoids and acellular pertussis (Tdap)

(i) Minimum age: 10 years
(ii) 11 years

(e) Haemophilus influenza type b (Hib)

(i) Minimum age: 6 weeks
(ii) 2 months/4 months/6 months/1 year

Persons aged 4 months through 6 years					
Vaccine	Minimum Age for Dose 1	Minimum interval Between Doses			
		Dose 1 to dose 2	Dose 2 to dose 3	Dose 3 to dose 4	Dose 4 to dose 5
Hepatitis B'	Birth	4 weeks	8 weeks and at least 16 weeks after first dose; minimum age for the final dose is 24 weeks		
Rotavirus[2]	6 weeks	4 weeks	4 weeks[2]		
Diphtheria, tetanus, pertussis[3]	6 weeks	4 weeks	4 weeks	6 months	6 months[3]
Haemophilus influenzone type b[5]	6 weeks	4 weeks if first administered at younger than age 12 months 8 weeks (as final dose) if first dose administered at age 12–14 months No further doses needed if first dose administered at age 15 months or older	4 weeks[5] if current age is younger than 12 months 8 weeks (as final dose)[5] If current age is 12 months or older and first dose asministered at younger than age 12 months and second dose administered at younger than 15 months No further doses needed if previous dose administered at age 15 months or older	8 weeks (as final dose) This dose only necessary for children aged 12 through 59 months who received 3 doses before age 12 months	
Pneumococcal[6]	6 weeks	4 weeks if first administered at younger than age 12 months 8 weeks as final dose for healthy children) if first dose administered at age 12 months or older or current age 24 through 59 months No further doses needed for healthy children if first dose administered at age 24 months or older	4 weeks if current age is younger than 12 months 8 weeks (as final dose for healthy children) If current age is 12 months or older No further doses needed for healthy children if previoud dose administered at age 24 months or older	8 weeks (as final dose) This dose only necessary for children aged 12 through 59 months who received 3 doses before age 12 months or for children at high risk who received 3 doses at any age	
Inactivated poliovirus[7]	6 weeks	4 weeks	4 weeks	6 months[7] minimum age 4 years for final dose	
Meningococcal[13]	6 weeks	8 weeks[13]	See footnote 13	See footnote 13	
Measles, mumps, rubella[9]	12 months	4 weeks			
Varicella[10]	12 months	3 months			
Hepatitis A[11]	12 months	6 months			
Persons aged 7 through 18 years					
Tetanus,diphtheria;tetanus, diphtheria, pertussia[4]	7 Years[4]	4 weeks	4 weeks if first dose administered at younger than age 12 months 6 months if first dose administered at 12 months or order	6 months if first dose administered at younger than age 12 months	
Human papillomavirus[12]	9 years	Routine dosing intervals are recommended[12]			
Hepatitis A[11]	12 months	6 months			
Hepatitis B[1]	Birth	4 weeks	8 weeks (and at least 16 weeks after dose)		
Inactivated poliovirus[7]	6 weeks	4 weeks	4 weeks[7]	6 months[7]	
Meningococcal[13]	6 weeks	8 weeks[13]			
Measles, mumps,rubella[9]	12 months	4 weeks			
Varicella[10]	12 months	3 months if person is younger than age 13 years 4 weeks if person is aged 13 years or older			

Fig. 2 Catch up vaccination schedule (From Centers for Disease Control, Atlanta, GA. http://www.cdc.gov/mmwr/preview/mmwrhtml/su6201a2.htm. Downloaded 31 Dec 2013)

(f) Pneumococcus

(i) Pneumococcal conjugate (PCV)

1. Minimum age: 6 weeks
2. 2 months/4 months/6 months/1 year

(ii) Pneumococcal polysaccharide (PPSV23)

1. Minimum age: 2 years
2. After age 2 years for certain high risk groups (asplenia, sickle cell disease, immunocompromised, transplant recipient, chronic heart disease, chronic lung disease, diabetes, cerebrospinal fluid leak, cochlear implant)

(g) Inactivated poliovirus (IPV)

(i) Minimum age: 6 weeks
(ii) 2 months/4 months/6 months/4 years

(h) Influenza (IIV)

(i) Minimum age: 2 years
(ii) 6 months (annual)

(i) Measles/mumps/rubella (MMR)

(i) Minimum age: 1 year
(ii) 1 year/4 years

(j) Varicella (VAR)

(i) Minimum age: 1 year
(ii) 1 year/4 years

(k) Hepatitis A (HepA)

(i) Minimum age: 1 year
(ii) 1 year (2 dose series)

(l) Human papillomavirus (HPV)

(i) Minimum age: 9 years
(ii) 11 years (3 dose series)

(m) Meningococcal conjugate (MCV)

(i) Minimum age: 6 weeks
(ii) 11 years

Developmental Milestones

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1. Source: Centers for Disease Control and Prevention, http://www.cdc.gov/NCBDDD/ACTEARLY/milestones/, as adapted from Shelov S, Altmann TR. *Caring for Your Baby and Young Child: Birth to Age 5*, 5th ed., American Academy of Pediatrics and Hagan Jr J, Shaw JS, Duncan PM. *Bright Futures: Guidelines for Health Supervision of Infants, Children, and Adolescents*, 3rd ed. American Academy of Pediatrics, Elk Grove Village, IL, 2008.
2. Up to date information on developmental milestones is available at www.cdc.gov/actearly or via 1-(800) CDC-INFO.
3. There is great variation between individual children. Progressive gain in milestones is important. Loss of previous milestones is particularly worrisome. Regular visits with a primary care provider are important to track normal development of children.
4. Age: 2 months
   (a) Motor: holds head up, smoother limb motion
   (b) Speech: coos, gurgles, turns to sound
   (c) Cognitive: attends faces, follows with eyes, gets bored with repetitive activity
   (d) Social: smiles, can briefly calm self, tries to look at parent
5. Age: 4 months
   (a) Motor: Holds head steady, pushes down with feet, rolls from prone to supine, shakes a toy, hands to mouth, when supine can prop on elbows
   (b) Speech: babbles, copies sounds, had different cries

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(c) Cognitive: expresses happy or sad, responds to affection, reaches out with one hand, directs hand to seen object, follows with eyes side to side, watches faces closely, recognizes people at distance
(d) Social: smiles at people, likes play, copies expressions

6. Age: 6 months

(a) Motor: can roll supine to prone and back, sits unsupported, bounces on legs while standing, rocks back and forth on knees
(b) Speech: responds to sound with sound, strings together vowels, responds to name, has happy and sad sounds, beginning consonants
(c) Cognitive: looks around a things, brings things to mouth, is curious about objects, pass object from hand to hand
(d) Social: can recognize a stranger as such, enjoys play, responds to emotions, gazes in mirror

7. Age: 9 months

(a) Motor: stands, can sit up, can pull self to standing, crawls
(b) Speech: understands "no", "mama", "baba" noises, copies sounds and gestures, points
(c) Cognitive: watches falling objects, looks for hidden objects, peek-a-boo, puts things in mouth, smooth passing from hand to hand, pincer grasp
(d) Social: fear of strangers, clings to know persons, has favorite toys

8. Age: 1 year

(a) Motor: can sit up unaided, cruises on furniture, isolated unsupported steps, stands alone
(b) Speech: responds to questions, can shake head no, can wave goodbye, can change tone of sounds, says "mama", "dada", tries to copy words
(c) Cognitive: explores objects, finds hidden objects, knows names of pictures, uses simple objects as intended, bangs objects together, moves objects in and out of a container, releases objects, pokes, follows commands
(d) Social: shy with strangers, cries when parent gone, has favorites, fearful, hands book to adult for story, repeats sounds for attention, assists with putting on clothing, plays "patty-cake"

9. Age: 18 months

(a) Motor: Walks alone, climbs steps, pulls toys, helps remove clothing, drinks from cup, uses spoon
(b) Speech: says single words, says "no" and shakes head, points at desired object
(c) Cognitive: knows purpose of everyday items, points for attention, pretends to feed doll, points to body parts, scribbles, follow verbal commands without gestures
(d) Social: enjoys handing off objects, has temper tantrum, plays pretend, clings to parent in new situation, points out objects of interest, explores alone near parent.

10. Age: 2 years
    (a) Motor: stands on tiptoe, kicks ball, runs, climbs furniture, walks stairs, throws ball, draws lines and circles
    (b) Speech: knows names, short sentences, follows two step instructions, repeats conversation, points in a book
    (c) Cognitive: sorts shapes and colors, completes sentences and rhymes, plays make believe, builds towers, prefers one hand, names pictures
    (d) Social: copies others, excited with other children, shows independence, disobeys, plays next to other children
11. Age: 3 years
    (a) Motor: climbs well, runs easily, rides tricycle, walks stairs with one foot per step
    (b) Speech: follows three step instructions, names familiar things, understands prepositions, can name self, age, and gender, names a friend, uses pronouns and plurals, can be understood by stranger, can converse
    (c) Cognitive: works moving parts, completes four piece puzzle, understands "two", copies circle, turns page, builds six block tower, opens jar and door
    (d) Social: affection for friends, takes turns, has concern for crying friend, understands possession, has range of emotions, can separate from parent, upset with change in routine, can dress/undress
12. Age: 4 years
    (a) Motor: hops on one foot, catches bounced ball, pours, cuts, and mashes food
    (b) Speech: basic grammar and gender, sings song, recites poem, tells story, states first and last name
    (c) Cognitive: names colors and numbers, understands counting, understands time, remembers story, understands same/different, uses scissors, copies letters, plays board or card games, anticipates events in story
    (d) Social: enjoys new things, plays "mom" or "dad", creative make-believe, prefers playing with others, cooperates with playmates, confuses real and make-believe, discusses likes and interests
13. Age: 5 years
    (a) Motor: Stands on one foot 10 s, hops and skips, can do a somersault, uses fork and spoon, uses toilet, swings
    (b) Speech: speaks clearly, tells story with sentences, uses future tense, states name and address
    (c) Cognitive: counts to ten, draws person with six body parts, prints letters and numbers, copies triangle, understands money and food
    (d) Social: wants to be like and please friends, agrees with rules, sings, dances, acts, aware of gender, differentiates reality and make-believe, increased independence, can be demanding and cooperative at different times.

Phone List

Resource	Phone	Contact	Fax
Admissions			
Adolescent Medicine			
Anesthesiology			
Blood Bank			
Cafeteria			
Cardiology			
Catheterization Lab			
Chemistry			
Clinic			
CT			
Cytology			
Dermatology			
ENT			
ER			
ER			
Frozen Section			
Gastroenterology			
Genetics			
Hematology			
Hematology			
Hemodialysis			
Infectious Disease			
IR			
Lab			
Medical Records			
Microbiology			
MRI			
Neonatology			
Nephrology			
Neurology			
Neurosurgery			

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Resource	Phone	Contact	Fax
NICU			
Nuclear Medicine			
Nutrition			
OB/GYN			
Occupational Therapy			
Oncology			
Operator			
Ophthalmology			
OR			
Orthopaedics			
Paging Operator			
Pain Service			
Pathology			
Pediatric Surgery			
Pediatrics			
Perioperative Care			
Pharmacy			
Physical Therapy			
PICU			
Plastic Surgery			
Police			
Psychiatry			
Pulmonary Lab			
Pulmonology			
Radiation Therapy			
Radiology			
Reading Room			
Rehabilitation			
Respiratory Therapy			
Scheduling			
Security			
Social Work			
Speech Pathology			
Surgical Pathology			
Thoracic Surgery			
Transplantation			
Ultrasound			
Urology			
Vascular Lab			
X-ray File Room			

Index Case List

Pediatric surgery index cases:

1. Source: Accreditation Council for Graduate Medical Education, Chicago, IL
2. Minimum number of cases for pediatric surgical (2 year fellowship) experience
   (a) Total major operations: 800
   (b) Major trauma non-operative: 90
   (c) Tumor: 25
   (d) Index cases: 55
   (e) Neonatal: 75
3. Most current figures available at www.acgme.org
4. Qualifying cases
   (a) Tumor
      (i) Major excision soft tissue tumor
      (ii) Resection chest wall tumor
      (iii) Cystic hygroma/lymphangioma
      (iv) Major head and neck tumor
      (v) Excision mediastinal tumor
      (vi) Excision neuroblastoma/adrenal/other retroperitoneal tumor
      (vii) Major hepatic resection/repair for tumor
      (viii) Nephrectomy, total or partial
      (ix) Oophorectomy, total or partial
   (b) Index cases
      (i) Repair chest wall deformity
      (ii) Excision mediastinal cyst
      (iii) Pulmonary resection for tumor or congenital malformation
      (iv) Esophageal resection or replacement
      (v) Perineal procedure for imperforate anus
      (vi) Pull through for imperforate anus

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(vii) Operation for Hirschsprung's disease, open
(viii) Operation for Hirschsprung's disease, laparoscopic
(ix) Orchidopexy, open
(x) Orchidopexy, laparoscopic
(xi) Vaginal reconstruction for disorders of sexual differentiation

(c) Neonatal

(i) Repair esophageal atresia or tracheoesophageal fistula
(ii) Repair diaphragmatic hernia
(iii) Operation for malrotation
(iv) Repair intestinal atresia, stenosis, or web
(v) Resection, repair, or enterostomy for necrotizing enterocolitis
(vi) Ostomy for anorectal malformation
(vii) Ostomy for Hirschsprung's disease
(viii) Repair of omphalocele
(ix) Repair of Gastroschisis
(x) Excision sacrococcygeal teratoma

5. Most common pediatric surgery operations

(a) Inguinal hernia repair
(b) Appendectomy
(c) Placement of long term IV access (Broviac or Mediport)
(d) Cholecystectomy
(e) Pyloromyotomy
(f) Nissen fundoplication
(g) Gastrostomy
(h) Closure of stoma
(i) Excision of subcutaneous cyst or mass
(j) Umbilical hernia repair
(k) Splenectomy

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