Nemours. Children's Hospital

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Kathy Grasso, PA-C	(407) 225 - 5560

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Daily Schedule

Monday	7:30–8:00	Rounds -Ana
	8:00–4:30	OR
	8:00–12:00	Clinic
	1:00–4:30	Clinic
Tuesday	7:30–8:00	Rounds - Ana
	8:00-4:30	OR – Dr. Casas
	8:00–12:00	Clinic
	1:00– 4:30	Clinic
Wednesday	7:00–8:00	Surgery M&M, Grand Rounds
	8:00–8:30	Rounds – PA- time subject to change
	8:00–12:00	OR
	9:00–12:00	Clinic
	12:30-1:30	Tumor Board (Executive Conference Room)
Thursday	7:30–8:00	Rounds -Ana
	8:00–12:00	Clinic
	1:00 – 4:30	Clinic
	7:30 – 3:00	OR
Friday	7:30–8:00	Rounds- Ana
	8:00–5:00	OR
Saturday	Rounds*	
Sunday	Rounds*	

*timing of weekend rounds are determined by the service attending
Be prepare to do at least 2 weekends which includes rounds and surgical cases.

General Surgery Medical Education Program
Pediatric Surgery Rotation

1. GOALS AND OBJECTIVES/EXPECTATIONS

Patient Care

Goal: Practice skills in evaluation and management of hospitalized patients, ambulatory patients and patients requiring emergency care and demonstrate knowledge of basic technical procedures and operations related to pediatric surgery

Goals and Objectives Basic operative skills:
- Appendectomy
- Hernia repair (Inguinal, umbilical, epigastric)
- Soft tissue lesions

Objectives
A. Demonstrate proficiency at performing comprehensive history and physical evaluations including preoperative and postoperative evaluations

B. Formulate comprehensive plans of therapy for pediatric perioperative patients with attention to the unique physiology of children

C. Identify and initiate management of complications of procedures including those requiring urgent intervention

D. Accurately assess levels of pain in patients and institute therapy with appropriate dosing by weight

E. Apply knowledge of diagnostic testing including an assessment of risk benefit analysis

F. Learn the core components of procedures performed in children and begin to completely perform common basic surgical procedures such as herniorrhaphy, cirmcumcision and appendectomy

Medical Knowledge

Goal: Identify common acquired and congenital disease processes in pediatric patients and interpret diagnostic testing to narrow differential diagnoses

Objectives
A. Utilize diagnostic test results to identify disease processes associated with children

B. Apply knowledge of pertinent scientific information to the practice of pediatric surgery

C. Utilize basic knowledge of biomedical, clinical, epidemiological and social behavioral sciences in the management of pediatric patients, including an understanding of developmental milestones

D. Recognize disease processes and injuries due to congenital malformations including those of the genitourinary tract, acquired disease, trauma and infection in children

Professionalism

Goal: Practice carrying out professional responsibilities and adherence to ethical principles in the care of pediatric patients

Objectives
A. Provide care to patients and families in a sensitive and responsive manner with compassion, integrity, and respect for others

B. Apply knowledge of the diversity of gender, culture, race, religion, disability and sexual orientation in the pediatric population

C. Show respect for patient privacy and autonomy that is age appropriate and be aware of and comply with all HIPPA regulations.

D. Demonstrate proficiency in accountability of self to patients, society and the profession
Recognize the need to obtain informed consent from parents or guardians for procedures in patients who are minors

Interpersonal and Communication Skills

Goal: Apply interpersonal and communication skills that result in effective exchange of information and collaboration that is age appropriate with patients, their families, and health professionals

Objectives

A. Work effectively as an advanced member of a health care team

B. Communicate proficiently with patients, families, physicians, and other health care professionals

C. Efficiently communicate descriptions of disease processes

D. Accurately and comprehensively document care in medical records including surgical consultations

Practice Based Learning and Improvement

Goal: Demonstrate the ability to investigate and evaluate care of patients, appraise and assimilate scientific evidence and continuously improve patient care based on self-evaluation.

Objectives

A. Apply the strengths, deficiencies and limits in one's knowledge and expertise to the improved care of pediatric patients

B. Set learning goals for self and others and lead learning activities

C. Participate in activities related to patient safety within the pediatric population

D. Utilize formative feedback in daily practice

E. Present at morbidity and mortality conference

F. Demonstrate ability to utilize evidence-based medicine in the patient care of pediatric patients

Systems Based Practice

Goal: Apply principles of comprehensive patient care in the system of health care and demonstrate ability to call on other resources to maximize patient care in the pediatric setting.

Objectives

A. Monitor the discharge of patients from the hospital to ensure that all home care needs are arranged and educate parents and children about care

B. Master the ability to identify cost effective resource utilization in patient care with knowledge of the risk-benefit ratio for patients

C. Present patients at multidisciplinary patient care conferences

D. Practice patient advocacy and recognize the developmental variability related to age and health status in children

E. Work effectively in inter-professional teams

F. Participate in identifying system errors and identify possible system solutions

2. RECOMMENDED READING MATERIAL

- Ashcraft's Pediatric Surgery – Fifth Edition

- George W. Holcomb III and J. Patrick Murphy
- Fundamentals of Pediatric Surgery – Peter Mattei
- SCORE Curriculum
- NAT – available on Nemours website

3. CLINIC TIMES, CONFERENCE DAYS, ROUNDS SCHEDULE, OR DAYS, ETC.

Clinic Times are available almost all days of the week with different providers

We have clinic almost everyday 8-5 pm. The general surgery clinic is on the 2nd floor of NCH. Students will be expected to participate in clinic when they are not actively participating in patient care or are in the operating room.

Pediatric Surgery Education – 7A in the 6th floor VCR

*You will be assigned a weekly topic to present.

BROAD
- Umbilical hernia
- Inguinal hernia
- Malrotation
- Hypertrophic pyloric stenosis
- Intussusception
- Meckel's diverticulum
- Appendicitis

FOCUSED
- Gastroschisis
- Omphalocele
- Esophageal atresia
- Tracheoesophageal fistula
- Congenital diaphragmatic hernia
- Duodenal atresia/stenosis
- Intestinal atresia and meconium ileus
- Imperforate anus
- Necrotizing enterocolitis
- Congenital agangliosis (Hirschsprung's disease)
- Biliary atresia
- Choledochal cysts
- Cryptorchidism
- Wilms tumor
- Neuroblastoma
- Hepatoblastoma
- Other Tumors

Surgical Suture Education

Every other Tues afternoon at 4 pm in the Simulation Center. We will let you know the dates. You should be practicing knot tying as well as suturing on your own as well.

Rounds: Will start every morning at approximately 7:30 a.m. The team will round together until all patients have been seen and plans for care are made. The student will be expected to pre-round and present patients to the appropriate attending during team rounds.

4.   CONTACT INFORMATION

GME Program Coordinator – Maria Kierulf

Nemours Children's Hospital
(407) 567-3882
maria.kierulf@nemours.org

5.   EVALUATIONS

A computerized evaluation will be completed by the faculty at the end of each rotation. A mid-rotation evaluation will be done with Dr. Westmoreland. You are also required to write a case report. We are expectly a quality level for publication in a journal, such as the Journal of Pediatric Surgery Case Reports. https://www.journals.elsevier.com/journal-of-pediatric-surgery-case-reports

General Surgery Medical Education Program
Pediatric Surgery Rotation
Nemours Children's Hospital
PGY-4: 1 Month

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- Circumcision

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E. Apply knowledge of diagnostic testing including an assessment of risk benefit analysis

F. Learn the core components of procedures performed in children and begin to completely perform common basic surgical procedures such as herniorrhaphy, cirmcumcision and appendectomy. Student will also have exposure to pediatric urology with evaluation and management of common pediatric urology cases.

Medical Knowledge
Goal: Identify common acquired and congenital disease processes in pediatric patients and interpret diagnostic testing to narrow differential diagnoses. Evaluation and management of pediatric "index cases"

Objectives
A. Utilize diagnostic test results to identify disease processes associated with children

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Conferences – Pediatric Surgery Education: Fri 7 am in the 6th floor VCR

Surgical Suture Education: Every other Tues at 4 pm in the Simulation Suite

BROAD

- Umbilical hernia
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- Imperforate anus
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- Congenital agangliosis (Hirschsprung's disease)
- Biliary atresia
- Choledochal cysts
- Cryptorchidism
- Wilms tumor
- Neuroblastoma
- Hepatoblastoma
- Other Tumors

OR Schedule
In general the OR starts at 7:30 am daily, except Wed when it's at 8:30 am. You'll round and then go to the OR unless otherwise directed.

Rounds: Will start every morning at approximately 7:30 a.m. The team will round together until all patients have been seen and plans for care are made. The student will be expected to pre-round and present patients to the appropriate attending during team rounds.

4. CONTACT INFORMATION

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5. EVALUATIONS
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Structure of the Service

Rounds:

Rounds are done daily and involve the entire peds surgery team (attendings, PAs, nurse practitioner and students). Rounds during the week start promptly at approximately 0730. It is expected that the student will have pre-rounded on their assigned patients and have all of the patient data prior to morning rounds. After pre-rounding, the student should be prepared to present all pertinent data regarding the patient at morning rounds including: age and gender of the patient, diagnosis and procedure, postoperative or hospital day, overall condition, vital signs, I's & O's, physical findings, laboratory values, and a tentative plan for the day on that patient. New admissions or consults are discussed in more detail, typically with a complete, but concise presentation of the history and pertinent physical examination findings. Report total fluid intake in cc's per **kg** per **day** and urine output as cc's per **kg** per **hour**. A detailed "SOAP" note should be completed for every assigned patient, every day; the attending will co-sign the student's note or write a separate note. Cut and paste notes are unacceptable.

Clinic Hours:

There are multiple scheduled outpatient clinics every week, and frequent "unplanned" outpatient visits throughout the week. All students and medical students are expected to participate in clinic unless assigned to the operating room for a case. Clinic is for the most part observational; clinic is fast paced, though students should take advantage of all opportunities to do focused exams. All patients are to be seen by the attending physician or nurse practitioner.

Conferences:

Attendance at all conferences by all students and medical students is expected.

Operating Room:

Students are assigned to cases by the attending. Students are expected to be prepared for the case to which they are assigned, including reading about the procedure ahead of time. **Please be on time**. (You should be in the OR at the time the patient enters the OR, except when rounding). There is no reliable way for students to be notified when a case is being brought to the operating room; it therefore requires diligence and tenacity on the part of the individual student to find out when a particular case is actually going to be done. Operative notes are done by the attending surgeon. Also note that the usual rules of OR etiquette apply at all times. As a courtesy, always introduce yourself to the circulating RN and surgical technicians.

Admissions and Consults:

Newly admitted patients need to be evaluated as soon as possible after arrival to the floor; attendings will see every new admission and consult, though students may be involved in the admission process. Admission orders need to be placed as soon as possible and the plan needs to be discussed with the responsible attending.

Emergency Room consults should be seen and evaluated as soon as possible. All consults are to be evaluated by and discussed with the attending surgeon prior to disposition of the patient.

Clinical Responsibilities

Surgery Student:

The surgery student is an active member of the team, with an appropriate amount of clinical and patient-care responsibility. They should see consults and discuss them with the on call attending. They should participate fully in the OR, clinic hours and conferences. The surgery student will be assigned by the attendings to scrub for OR cases.

Pediatric Student:

Pediatric students may choose to do a peds surgery rotation as an elective. The pediatric student is an active member of the team, with an appropriate amount of clinical and patient-care responsibility. They should see consults and discuss them with the on call attending. They should participate fully in clinic hours and conferences. The pediatric student will be assigned by the attendings to observe and scrub in on certain cases for the learning experience.

Medical Students:

Medical students should participate in patient care aspects of the service as appropriate for their level, experience and abilities. These responsibilities are delegated by the attending and nurse practitioner. Students may follow individual patients from admission through discharge. They should also participate in clinic hours and observe/ assist in the operating room.

Nurse Practitioners and Nurses:

Our nurse practitioner and nurses are very knowledgeable and extremely valuable members of the surgical service. Our nurse practitioner participates fully in the care of both inpatients and outpatients; they are the cornerstone of continuity on our service and are experts in the care of our patients. The student and the nurse practitioner will work together to cover the patients on the floors. The nurses are mostly clinic-based. They are particularly valuable resources when it comes to teaching of daily patient care issues and problems unique to pediatric patients. Please feel free to collaborate with them in order to help us provide quality care to outpatients, but please do not abuse this privilege. During clinic, be sure to respect the nurse's work area and work space as to not interfere with the clinic workflow.

Daily Management

Ideally, a plan for each and every patient should be formulated in the morning and then instituted as soon as possible. This means that orders are written and clarified early, every member of the team is aware of any changes in the plan, and the floor nurses are aware of our plan for their patient that day. Remember that parents need to know the plan also.

- Evaluate <u>IV fluid</u> orders and <u>diet</u> orders every morning.

- Review all medications, especially <u>antibiotics</u> and <u>analgesics,</u> every day, and discontinue or taper those that are no longer needed.

- Remember to inquire about all medications that the child was on prior to admission (e.g. nebulizers, anti-seizure meds, etc.)

- Orders to discontinue NG tubes should be entered as soon as the decision is made.

- Remember to update activity orders.

- Pokes hurt! Avoid unnecessary blood draws; do not order blood tests if the result will not change your patient management. Minimize blood draws and try to group them together to minimize needle sticks. "Routine" blood draws are used only by TPN protocol.

- Discuss the plan with families ONLY if it is clear to you and you have been given permission by the attending. For complex patients this should be left to the attending. Absolutely avoid conjecture and expressing personal opinions.

- Every effort should be made to avoid parental confusion or dissatisfaction. If you feel that a parent is getting upset and the situation is likely to get out of hand, DO NOT try to defuse the situation yourself – consider this an emergency to be dealt with by the attending.

- Interpreters can be arranged by the social worker or case manager.

Fluids and Electrolytes

Fluid and electrolyte management in children requires careful attention to fluid intake, fluid losses, clinical state, and electrolyte needs. Fluid requirements need to be calculated accurately and individually for every patient, as the margin of error is often small.

A. Maintenance fluid requirements

As a general rule, maintenance fluid requirements can be calculated according to body weight:

<u>Infants and children:</u>

Weight	Per Hour	Per Day
2 – 10 kg	4 mL/kg/hr	100mL/kg/day
10 – 20 kg	40mL/hr + 2mL/kg /hr for wt > 10kg	1000mL/day + 50mL/kg/ day for wt > 10kg
20 – 50 kg	60mL/hr + 1mL/kg /hr for wt > 20kg	1500mL/day + 20mL/kg/ day for wt> 20kg
>50 kg	80-90 mL/hr	2000 mK/day

<u>Neonates and premies:</u>

Birth Weight	Day 1	Day 2	Day 3
Term newborn	80 mL/kg/day	100 mL/kg/day	120 mL/kg/day
1500-2000g	80 mL/kg/day	110 mL/kg/day	130 mL/kg/day
1250-1500g	90 mL/kg/day	120 mL/kg/day	130 mL/kg/day
1000-1250g	100 mL/kg/day	130 mL/kg/day	140 mL/kg/day
750-1000g	110 mL/kg/day	130 mL/kg/day	140 mL/kg/day
500-750g	120 mL/kg/day	140 mL/kg/day	150 mL/kg/day

The choice of which standard formula to write for is based on electrolyte requirements as discussed below. As a general rule, use D51/2 NS + 20 mEq KCI/L in infants and children, and the calculated electrolyte needs will be met.

These guidelines are approximations and should be modified for the clinical situation. In situations of high insensible losses, the fluid requirements will be greater, especially in infants. For example, patients with large open wounds, a silo for an abdominal wall defect, or high fevers may require up to 11/2 x maintenance. Also, infants receiving phototherapy for jaundice should have their fluids increased by 10-15%.

Fluid needs require frequent adjustment, often more than once or twice daily, based on the clinical assessment of the degree of hydration. This is especially true in infants. Signs of adequate hydration include good peripheral perfusion, moist mucous membranes, a level fontanel, and the presence of tears. <u>In most clinical situations, the best measure of hydration is urine output,</u> which should be

approximately 1 to 2 mL/kg/hour in the newborn-infant and 0.5 to 1 mL/kg/hour in children. Note that patients in the NICU will have their fluid requirements managed by the NICU team.

B. Electrolytes <u>Neonates:</u>

Electrolyte	Day 1	Day 2	Day 3
Na+ mEq/kg/day 0	1-2	2-3	
K+ mEg/kg/day 01	1-2		
Cl' mEg/kg/day 01*	1*		

*In newborns (exp. Premies) 1-2 mEq/kg/day of acetate is substituted for Cl' to prevent acidosis.

<u>Infants and Children:</u>

Sodium	2 – 4 mEg/kg/day
Potassium	2 – 3 mEg/kg/day
Chloride	2 – 4 mEg/kg/day
Calcium	0.25 – 1 mEg/kg/day
Phosphorus	0.5 – 1.5 mEg/kg/day
Magnesium	0.5 - 1 mEg/kg/day

C. Fluid losses and Replacements

Patients may experience loss of specific bodily fluids that may become significant not only from a fluid standpoint but from the loss of electrolytes or protons. Acutely hypovolemic children should receive fluid boluses of 10-20 mL/kg of NS (or LR). Blood is given in boluses of 5-15 mL/kg. (remember that total blood volume is approximately 80 mL/kg).

Fluid	Replacement	Volume
Nasogastric	0.45NS + 20mEg KCI/L	"cc per cc" or "0.5cc per 1 cc"
Stoma, stool, biliary, pancreatic	Lactated Ringer's Soln.	"cc per cc"
"third space"	NS or LR	Quantity sufficient to maintain hydration

These are usually written such that the total volume of fluid loss is measured every 4 hours and then replaced over the next 4 hours. For example, a typical order may read: "Replace NG drainage cc/cc with 0.45NS+20mEq KCI/L IV Q4 hr." Because fluid losses and their replacement cancel each other out and thus serve only to obscure the true fluid status, remember not to count fluid losses or fluid replacements when calculating I's or O's unless you are asked to do so by the person to whom you are presenting the data.

Nutrition

A. Total Parenteral Nutrition

In general, TPN orders are written by the NNP's and neonatologists for our patients in the NICU; our service may be responsible for TPN orders on surgical patients in the pediatric wards. Although a lengthy and detailed discussion of TPN is beyond the scope of this handbook, a summary of basic principles, especially as they apply to neonates, might be helpful. TPN orders for newborns typically involve a predetermined volume to which nutrients and electrolytes are added in incremental amounts until daily goals are being met.

a. <u>Glucose</u>

1. The initial concentration of glucose is usually 10%. This is then increased daily to a maximum of 25% for central TPN and 12.5% for PPN.

2. The minimum glucose infusion rate to prevent hypoglycemia in newborns is 4-6 mg/kg/min. The maximum is 15-16 mg/ kg/min.

3. The ideal glucose infusion rate is 10-12 mg/kg/min.
1 gram dextrose = 4 kcal.

b. <u>Protein</u>

1. Protein requirements vary with age. Newborns need 2 -3.5 gm/kg/day and have a higher essential amino acid requirement. Older children and adolescents require only about 1.5 – 2.0 gm/kg/day of protein.

2. Protein is given as an amino acid solution and is typically begun at a rate of 1.0 gm/kg/day and increased by about 0.5-1.0 gm/kg daily until the goal is reached.

3. Calories derived from protein are usually not counted when calculating total calorie requirements.

c. <u>Fats</u>

1. Calories derived from fat should total 25 – 40% of total non-protein calories and should not excess 55% of the total calories.

2. Fats are given as 10% or 20% Intralipid and started at 1.0 gm/kg/day and increased by 1.0 gm/kg daily to a maximum of 3.0 gm/kg/day.

3. 1 gram of fat = 9 kcal. 1 mL of 20% Intralipid = 1.8 kcal.

d. <u>Calorie</u>

Caloric requirements in a non-stressed patient can be estimated as follows:

Age/Weight	Calories
Newborn or Premie	120 kcal/kg/cal
<1 year, up to 10 kg	90 – 100 kcal/kg/day
10 – 20 kg	1000 kcal/kg/day for weight over 10 kg
>20 kg	1500 + 20 kg/kg/day for weight over 20 kg

e. <u>Cholestasis</u>

Newborns on long term TPN often develop cholestasis. This is manifested clinically by jaundice. The first laboratory value to become abnormal is serum bile acids (not a routine study), followed by direct hyperbilirubinemia, and later hepatic enzyme elevations. The best treatment is conversion to complete enteral nutrition. If this is not possible, alterations in the TPN should be made: fat is decreased and cycled, manganese is eliminated, and copper is decreased to 10 mcg/kg/day. Fat soluble vitamins are given also (A, D, E, and K). Omegaven is an Omega-3 based fat that is administered to patients who are allowed into the research study; strict criteria are used for admission into the study, including chronic hyperbilirubinemia and TPN/IL dependence.

B. Enteral Nutrition a. <u>Newborns</u>

1. Caloric requirements are approximately 120 kcal/kg/day for most newborns and premies.

2. With adequate nutrition, newborns should gain 20-30 grams per day.

3. Breast milk and standard infant formulas have 20 kcal per ounce or 0.67 kcal/ mL.

4. For premies and babies with short or damaged gut, a hydrolyzed (partially pre-digested) formula such as Pregestimil or Elecare is used. The caloric density of any formula can be increased up to 30 kcal/oz if necessary, though remember that increasing the density also increases the osmolarity.

5. Feedings are typically given as boluses (e.g. Q3hr) but can be given continuously via an NG or G-tube, which is often required in children with short gut. (Feedings given via NJ-tube or GJ-tube must be continuous, not bolus.)

6. Feeding intolerance should be suspected if:
   - Stool output is >45 mL/kg/day
   - Gastric aspirates are >10 mL/kg.
   - Abdominal girth is significantly increased.

Caloric requirements can be calculated using complex formulas that actually calculate metabolic rates but cam be approximated depending on age and weight. Non-newborn formulas are generally 30 kcal/ounce.

Age/Weight	Rate
<20 kg. < 1 yr	90kcal/kg/day*
>20 kg. >1 yr	80kcal/kg/day*
7 -- 10 yr	70kcal/kg/day*
11 – 14 yr	60kcal/kg/day*
15 – 18 yr	50kcal/kg/day*

*Most non-infant formulas are 30/kcal/ounce (1kcal+1mL).

Always request a nutrition consult before writing any enteral feeding orders. They will calculate the patient's goal for calories, recommend the specific formula that is best for the child, and often suggest an advancement regimen as well.

Enteral Feeding devices

Calls about enteral feeding devices are very frequent and also very important. Time is of the essence in order to avoid spontaneous closure of the fistula and a return to the OR to replace the tube. As a rule, any questionable tube replacement needs to be confirmed by a radiologist using contrast and fluoroscopy.

A. Gastrostomy tubes ("G-tubes")

Gastrostomy tubes come in a variety of shapes and sizes. A gastrostomy tube that "falls out" should be replaced as soon as possible and can be ordered through One Chart. Tubes are also available in clinic during regular clinic hours, though the correct paperwork needs to be completed when a tube is utilized.

Replacement of G-tubes requires skill and patience. Use plenty of water-soluble lubricant and never force the tube in. If the tract is very small, put in whatever tube will fit to prevent it from closing up and notify the attending or CNP. After the tube is placed, fill up the balloon with the appropriate amount of water and attempt to aspirate gastric contents through the tube. If you are unsure about the position of the tube or if it has been less than 6 weeks since surgical placement of the tube, a water soluble contrast study needs to be done immediately, before anything is placed through the tube.

Types of G-tubes

AMT: These are low profile, clear, silastic G-tubes that are very short and do not project very far from the skin. They have 4mL balloons and require extensions to allow venting or feeding; these come with the kit. They are as easy to replace and are sized by length and diameter. A special sizing device may be used to

determine the proper length of the tube. This should be used to identify the appropriate tube for each patient (e.g. 14F 1.2cm,etc.)

<u>MicKey bolster G-tube:</u> These are clear silastic tubes that have a balloon at the tip and a silastic wafer that prevents the tube from being drawn into the GI tract.

<u>Foley catheter:</u> Should be used rarely, and only when a more appropriate tube is not available. Specific problems include obstruction due to the catheter being drawn into the GI tract, poor tolerance of the tube to gastric secretions, and the requirement for another hospital visit for it to be replaced with a "real" G-tube.

B. Jejunostomy tubes

Gastro-jejunostomy (G-J tubes): These are tubes that are placed through a gastrostomy but are designed so that the tip lies in the jejunum. They have a balloon that resides in the stomach and two "ports": One in the stomach and one in the jejunum. These can only be replaced by an interventional radiologist. In general, it is best to replace it with standard G-tube until a radiologist is available to replace it.

Central Lines

There are several different types of central lines that you will be asked to evaluate or remove while rotating on Pediatric General Surgery:

- "Percutaneous" central lines (short term)
- Indwelling tunneled central lines ("Hickman" or "Broviac"), and
- Subcutaneous injection ports ("Bard Power Port" or "Bard Slim Port"or "Port-a-Cath")
- Peripherally inserted central catheter (PICC line)

Consults for placement of central access are rarely emergent. Always do a full consult with attention to the age and size of the patient, indication for the line, whether the patient has had multiple central lines in the past, the type of line requested, whether the patient is septic, and the platelet count (at least >50,000). Patients who have positive blood cultures should be treated with antibiotics for at least 2 to 3 days before placement of a central line. Patients who have a history of having multiple central lines require a duplex ultrasound study of the central vessels prior to an attempt at placement of another line. Leave a consult and notify the attending to discuss appropriateness and timing of the line placement.

Essentially all central lines in children are placed in the operating room under general anesthesia. Exceptions are central lines placed as part of an emergency resuscitation, neonates in the NICU, or PICC lines which may be placed at the bedside or in Peds outpatients. Percutaneous central lines are those that enter the skin and the vein in the same puncture; they are not "tunneled" or indwelling. They are designed to be temporary; percutaneous lines can be removed at the bedside or in the office in most patients without sedation or local anesthetic by laying the patient supine, cutting any sutures that may be present, gently removing the line, and holding pressure for a least two minutes. Be careful when removing sutures that the entire suture is removed – do not cut the suture in such a way as to leave suture material buried under the skin.

Indwelling central lines are usually "tunneled" meaning that the entry site in the skin is several centimeters away from the entry site in the vein. Tunneled central lines which have been in place for more than 2-3 weeks are usually removed in the OR under general anesthesia. An exception is central lines in NICU babies. These can be removed at the bedside with local anesthesia. Lines in place for less than 2-3 weeks will often not have much scar tissue holding the cuff in place. These warrant a brief attempt a removal at the bedside with gentle, continuous traction (NO tugging). If the cuff is within 2 to 3 cm from the skin, gentle spreading at the skin entry site usually allows one to snip the scar tissue that holds the cuff. Be VERY careful not to cut the catheter or allow the catheter to snap apart. The intravascular portion can then become an embolus that requires surgery or interventional radiology to retrieve. With careful technique, this should never happen. If any resistance is encountered – STOP – and notify the attending. No line should be removed without prior discussion with the surgical attending.

You will sometimes be asked to repair a tunneled catheter that has developed a leak. There are repair kits available. Ask for help from the nurse practitioner if you have never used one before, or if you think the catheter is not repairable. Leakage around the catheter can be due to a leak in the subcutaneous portion of the catheter. This usually requires replacement of the catheter. Catheters whose cuff has extruded from the skin also require replacement. Catheter infection is typically treated with antibiotics first but may also require removal of the catheter. Pertcutaneous (i.e. non-tunneled) catheters can be changed over a guide wire and the tip sent for culture while the patient is treated with antibiotics. Catheters obstructed by clot can be cleared in most cases with tPA. Other causes of catheter obstruction include kinks, overzealous knot-tying on stay sutures, pinching between the clavicle and first rib, and organized clots. Work-up includes: direct inspection, aspirating and flushing the catheter during changes in patient position, CXR, and contrast injection under fluoroscopy.

Subcutaneous injection ports are entirely covered with skin and require percutaneous access with a Huber needle after application of a topical anesthetic cream (never use a standard needle). They have several advantages over tunneled lines in that they are more cosmetically acceptable, have fewer infectious complications, and require less daily care. These always require general anesthesia for placement or removal. You will sometimes be asked to evaluate these ports for infection. The signs of infection vary depending on whether there is an abscess (infected fluid around the port or infection in the tissues around the port). The consult is usually prompted by redness and or pain at the site. Always evaluate the ports with the dressings removed. Gently palpate the port for evidence of fluctuance. Fluid, redness, and/or tenderness does not always mean infection of the port site as extravasation of intravenous medications or a hematoma can give the same appearance. Generally, these are treated expectantly, often with empiric antibiotics, unless unmistakable evidence of infection develops (positive blood cultures, aspirated pus from the site, lack of resolution of local signs for 2 or 3 days). Infection of a port almost always requires removal of the port as these infections do not respond to antibiotics alone.

Bowel Preparation

Patients will require a bowel prep before certain abdominal or rectal procedures. Outpatients are often admitted the day before a scheduled operation specifically for completion of a bowl prep. The type of bowel prep will vary depending on who the operating surgeon is. ALWAYS ask for specific instructions from the attending or CNP before starting a bowel prep. Bowel preps should be started as soon as possible on the day prior to surgery. There is a standard order set in Epic.

Appendicitis

Clinical Presentation:

Although the first clue that a child may have appendicitis is typically abdominal pain, there is a great deal of variability in the presentation. Making the diagnosis requires a high index of suspicion, attention to the details of the history and physical examination, and good judgment based on personal and collective experience. Never second guess a request for surgical evaluation of a child with abdominal pain, regardless of how unlikely the story is over the phone. Always evaluate the patient personally and discuss the case with more experienced members of the surgical team. Similarly, do not order imaging studies or discuss a detailed plan with the family until it has been discussed with the attending surgeon.

History:

The classic history of appendicitis is an initially vague, crampy pain located in the periumbilical area or epigastrium, followed by a change to a more intense, constant pain located in the right lower

quadrant ("McBurney's point"). This typically progresses over a 12 to 24 hour period. Unfortunately, few patients present with this classic history. They may never localize the pain or may localize from the earliest onset of pain. The pain may localize in the suprapubic region or anywhere else in the abdomen. Any child with localized abdominal pain should be assumed to have appendicitis until proven otherwise.

Most patients will present with other symptoms such as nausea and/ or vomiting, fever, and anorexia. Anorexia is not a reliable sign of appendicitis in children; similarly, the absence of anorexia does not reliably rule out appendicitis. Suprapubic discomfort, especially with micturition, may indicate an appendicitis localized to the pelvis. Flank pain may suggest a retrocecal appendix. A history of pain elicited by bumps in the road during the car ride to the hospital are also notoriously unreliable.

The duration of symptoms is important: appendicitis rarely presents in less than 6 to 12 hours. Similarly, pain of more than 72 hours duration suggests either another diagnosis or ruptured appendicitis. Most cases of perforation occur after 36 to 48 hours.

The bottom line: there are no absolutes when it comes to appendicitis.

Physical Examination:

Localized tenderness is the most common finding suggesting appendicitis. Typically there are localized peritoneal signs evident by examination, but localized tenderness alone should raise the suspicion of appendicitis. "Peritoneal signs" can be subtle or obvious; tenderness with sudden movement, voluntary guarding, involuntary guarding, percussion tenderness, muscular rigidity. In children, "peritonitis" refers more to a severe degree of tenderness, rather than any rigidity. In children, "peritonitis" refers more to a severe degree of tenderness, rather than any particular physiologic reflex. This is especially true in children under 5 years of age whose only signs of peritonitis may be grunting, distension, and mild diffuse tenderness. "Rebound" tenderness is an unreliable sign of peritonitis (over-sensitive) and, moreover, can be extremely painful: do not test for rebound in the right lower quadrant. Rebound in the left lower quadrant that causes pain in the right lower quadrant ("Rovsing's sign") is somewhat more reliable but is not 100% accurate either.

Always check for flank tenderness, which can suggest an alternative diagnosis such as pyelonephritis or renal calculi, or a retrocecal appendicitis. A rectal examination is rarely useful and unnecessarily traumatic for the young patient. Use judgment when deciding in which patient it is likely to provide any useful information. Likewise, pelvic examinations should only be performed in sexually active teens in whom you suspect PID or an adnexal mass.

Laboratory Studies:

A patient with a clinical diagnosis of acute appendicitis based on history and physical examination needs no confirmatory studies before operation. Most patients should have a urinalysis and all girls beyond menarche or older than 12 years <u>must</u> have a urine pregnancy test, <u>regardless</u> of the sexual history. "Borderline" patients should have a urinalysis and CBC. An elevated WBC is suggestive of but not diagnostic of acute appendicitis, and a normal WBC does not rule out acute appendicitis (Sensitivity-85%, Specifically-50%). A left-shifted differential is slightly more accurate. Remember that an inflamed appendix adjacent to the bladder or ureter can result in a few WBC's and/or RBC's on the urinalysis. Most other laboratory studies are unnecessary and expensive but should be considered if clinically indicated: amylase/lipase, LPT's basic metabolic panel, sed rate, etc.

Abdominal radiographs are rarely indicated except when bowel obstruction, abdominal mass, or free air is suggested by history or physical examination. A chest radiograph may be done in children under 6 years of age with a complaint of abdominal pain, regardless of findings on adominal exam, to rule pneumonia.

Special Studies:

Studies such as ultrasound or CT scan are useful in confirming or ruling out the diagnosis of acute appendicitis. They may be helpful in certain "borderline" situations. Ultrasound is most useful in girls when there is a concern about ovarian pathology. Abdominal CT scan is more accurate than ultrasound but is much more labor intensive: it requires IV contrast, and exposes the patient to radiation. It is helpful under very specific circumstances, for example, when a patient is unable to provide a reliable physical exam or for patients whose risk of surgery is very high such as Crohn's disease. Always check with the attending before ordering any diagnostic study as they are expensive and often unnecessary.

Diagnostic laparoscopy is useful in a few clinical situations, for example, in the female patient in whom an ovarian process cannot be ruled out or distinguished from appendicitis, or in a morbidly obese patient. The procedure needs to be thought of a true operative procedure with the same level of risk that accompanies a laparotomy. Because of the common public misconceptions about laparoscopic surgery, do not casually mention diagnostic laparoscopy to a family of a patient with abdominal pain unless directed to do so by the attending, as it is often then very difficult to explain why the procedure is not indicated or not necessary.

Admission for observation: "Rule out appendicitis"

In borderline cases, the safest and most accurate approach is admission to the hospital for serial examinations. The patients are admitted to the surgery service and made NPO. IV fluids are given at the maintenance rate. DO NOT give antibiotics; Medicate appropriate to treat pain. Patients need to have a careful abdominal examination (preferably by the same examiner) every 4 to 6 hours, with any worsening in the exam reported to the attending surgeon. In most cases the appropriate treatment (operation or discharge) will become clear in 8 to 12 hours. Further studies should be discussed with the attending surgeon before being ordered.

Operation:

Once the decision is made to take a patient to surgery for an appendectomy, they should be given IV antibiotics, Zosyn

(or alternative for PCN allergy), and narcotic analgesics (morphine 0.1 mg/kg IV). The patient remains NPO, IV hydration is started, and informed consent is obtained. Most appendectomies can be safely delayed by up to several hours if an OR is not available immediately.

Appendectomies for acute appendicitis are performed using a standard or single-site laparoscopic approach. Post-operative management depends on whether the appendix was non-perforated or perforated. Non-perforated patients may advance to regular diet as tolerated. Most patients should be ready for discharge in 24 to 36 hours. Passage of flatus or a bowel movement is not a prerequisite for discharge. Perforated patients receive antibiotics and, depending on the anticipated degree of post-op ileus, may need to remain NPO for days. A typical length of stay varies between 5 and 10 days, generally until the patient has normal gastrointestinal function, is afebrile, and has a normal WBC. Always discuss the post-operative plan with the surgical attending and collaborate with the CNP.

Post-operative abscess:

The diagnosis of abscess should be considered in any patient who by POD 5 or 6 has: persistent ileus, poor PO intake, fever, or elevated WBC. Diarrhea can be a sign of an abscess also. An abdominal/pelvic CT scan should be performed on such patient no sooner than POD 5 or 6. Most abscesses can be treated non-operatively, multiple small ones are treated with continued antibiotics, large ones can usually be drained percutaneously or trans-rectally. Discuss the plan with the attending.

Umbilical hernia
Umbilical hernia results from failure of closure of the fascial defect through which the structures of the umbilical cord pass and therefore presents in infancy. It is more common in premature infants and African-American children, but occurs equally in boys and girls.
The majority of umbilical hernias will resolve spontaneously before the age of 4 or 5. They are rarely symptomatic and diagnosis is made on physical examination. The defect itself should be palpated after reduction of the hernia and its size should be documented. The amount of protruded bowel or redundant skin, although sometimes of great concern to the family, is less important clinically than the size of the defect itself. The exception is the occasional child with a huge projection of bowel and skin, which can resemble a proboscis or phallus and can influence the timing of operation. There is no indication for binders or trusses (e.g. taping a quarter to the defect) as these are not effective and can cause discomfort to the child and skin breakdown.
Complications are uncommon. Incarceration and strangulation of umbilical hernias is quite rare but can occur. As with all incarcerated hernias, an attempt should be made to reduce the hernia. This is best done with the child in the supine position and occasionally requires mild sedation.
Because most will resolve spontaneously and the risk of incarceration is so low, the mere presence of an umbilical hernia is not an indication for surgery. **Indications for surgery** include; failure of spontaneous resolution by the age of 4 or 5, a history of incarceration, and hernias that are symptomatic. Defects that are smaller than 1 cm in diameter can probably be watched beyond age 4, although even small defects pose a risk for pregnant women, suggesting that our threshold to recommend surgery should be lower in girls. Children younger than 3 sometimes require repair if the hernias are very large, especially if there is a huge projection of skin.
The operation is typically performed electively and in an outpatient setting. Most surgeons employ an infra-umbilical curvilinear incision, which provides good exposure and heals with minimal scarring. The sac is usually adherent to the skin of the umbilicus and is dissected circumferentially. The sac can then be excised or left intact to be inverted into the defect. The defect is closed transversely with interrupted absorbable or nonabsorbable sutures. The skin of the umbilicus is tacked to the fascial repair with one or two sutures and the skin is closed in the usual fashion. A light pressure dressing is sometimes used to prevent a postoperative hematoma. Resection of excess skin or formal umbilicoplasty should be avoided whenever possible due to the often unsatisfactory cosmetic appearance of the umbilicus. Only when there is a truly excessive amount of redundant skin should be considered.
Complications include wound hematoma, wound infection, (both uncommon) and recurrence (very rare). The results are generally very good and most patients and their families are satisfied with the ultimate cosmetic result even if the umbilicus is not typical in appearance.
Inguinal hernia and Hydrocele
Inguinal hernias and hydroceles are considered together because they share a common etiology (patent processus vaginalis), they can be difficult to distinguish clinically, and the operation to repair them is essentially the same.
Inguinal hernias occur in 3-5% of full-term infants and in 10% or more of premature infants.
Herniorrhaphy is one of the most common operations performed in children. Some present in infancy though they can present at any age. Nearly all inguinal hernias that present in children are "indirect" hernias (i.e. congenital patency of the processus vaginalis) rather than "direct" hernias (due to fascial incompetence of the floor of the inguinal canal). The repair therefore requires only high ligation of the hernia sac and almost never requires a formal repair of the musculature or the use of mesh.
Most hernias **present** with an asymptomatic bulge in the inguinal region, usually noted by the parents or pediatrician. Diagnosis can only be confirmed by physical examination and can sometimes be difficult. An impulse noted when the patient coughs or strains is <u>not</u> enough to make the diagnosis of inguinal hernia in a child. There must be a visible or palpable bulge that can reduce spontaneously but more often requires manual reduction. One can sometimes appreciate a "silk glove sign" (a palpable membrane

within the spermatic cord) but is also insufficient evidence of a hernia to justify operation. It is best to make the child valsalva using a painless and safe maneuver such as standing, performing an abdominal examination, asking the child to inflate a balloon or examination glove, or in an older child simply requesting that they strain. Sometimes the diagnosis cannot be confirmed in the surgeon's office and then a judgment needs to be made regarding the reliability of the history. In most cases, it is best to recommend observation until the hernia can be confirmed on examination by the surgeon or rarely if the historical evidence is incontrovertible surgery may be recommended. There is <u>no</u> role for imaging studies. The **history** should focus on overall health and behavior of the child, feeding issues, and bowel habits. **Physical examination** should also include a thorough abdominal and scrotal examination. Masses should be noted, and testicular descent should be documented. The overall appearance and disposition of the child should be noted, as a restless and irritable child raises concerns for incarceration.

Inguinal lymphadenopathy can sometimes be confused with a hernia but is usually easily distinguished on physical examination by an experienced examiner. Lymph nodes are usually more laterally positioned, are firm, immobile, and occasionally tender, and can usually be separated from a normal spermatic cord. Although ultrasound can be useful in rare cases, in general, diagnostic uncertainty can only be resolved adequately with inguinal exploration.

Easily reducible hernias are repaired on an elective outpatient basis. All inguinal hernias should be repaired surgically because: they never resolve spontaneously, they often enlarge and can become symptomatic, and they are at risk of incarceration and strangulation. Parents are instructed regarding signs and symptoms of incarcerated hernia and an OR date is scheduled by the office. Hernias that are initially incarcerated but that are eventually reduced can be repaired semi-electively (next day or next week) according to the judgment of the attending surgeon. For hernias that cannot be reduced or in cases of diagnostic uncertainty, operation is performed emergently after rapid fluid resuscitation. In girls, it is often the ovary that is incarcerated. If it is non-tender and otherwise showing no signs of torsion or ischemia, these can be repaired on the semi-elective basis.

Incarcerated hernias merit an attempt at reduction by a surgeon in most cases. Steady pressure should be applied to the end of the bulge as well as the sides of the hernia sac while keeping the hernia lined up properly to avoid folding it over on itself. Successful reduction more commonly occurs with experience rather than the use of specific technical maneuver. Young children will sometimes require sedation (IV morphine and/or midazolam). Notify the attending surgeon prior to sedating a child with an incarcerated hernia.

A **hydrocele** is a collection of fluid within a portion of the tunica vaginalis and can occur in the scrotum (most common), anywhere along the spermatic cord ("hydrocele of the cord") or, rarely, in girls ("hydrocele of the canal of Nuck"). Daily fluctuation in size suggests the presence of a communication between the sac and the abdomen and is referred to as a communicating hydrocele. Those that are more stable in size are thought to lack a continuously patent processus and are referred to as noncommunicating hydroceles. Communicating hydroceles are essentially hernias in which the patent processus is only large enough to allow fluid to pass but not bowel. Nevertheless, the risk of complications is low enough that a more conservative approach is sometimes appropriate for hydroceles. For example, a period of observation or waiting until a child is 1 year old before recommending operation is considered appropriate. Communicating hydroceles are unlikely to resolve spontaneously and should probably be treated like hernias.

The **diagnosis** of hydrocele is made on physical examination. Classically, there is an asymptomatic, nonreducible, fluctuant mass in the scrotum that may have a bluish tinge and transillumiates when a light is placed behind it in a dark room. The key to distinguishing a hydrocele from an incarcerated hernia on physical examination is that one should be able to palpate a normal spermatic cord above the hydrocele. Unfortunately, some hydroceles extend up into the inguinal canal and can be indistinguishable from a hernia. Because bowel can transilluminate in infants and ultrasound is not able to distinguish a hernia from a hydrocele with certainty, these babies should undergo inguinal exploration without delay. Except for this rare situation, most patients with hydrocele can be discharged from the ER with follow-up or scheduled for elective hydrocelectomy.

Surgical approach

These are as many modifications of herniorrhaphy as there are pediatric surgeons. Nevertheless, certain principles remain relatively constant for open inguinal hernia repair. The basic steps include:

1. General anesthesia + regional block;
2. Small inguinal incision, usually in the skin crease;
3. Expose the external inguinal ring and incise the external oblique aponeurosis to open the ring (not all surgeons perform this step);
4. Split the cremasteric muscle fibers longitudinally;
5. Deliver the cord structures into the wound;
6. Separate the sac (patent processus) from the spermatic vessels and vas deferens;
7. Dissect the sac to the level of the internal ring.
8. Make certain that the sac is empty (open it, if necessary, to be sure);
9. Twist and ligate the sac at the level of the internal ring;
10. Excise the excess sac distal to the ligature;
11. If a hydrocele is present, open it widely;
12. Return the testicle to its anatomic position;
13. Close the wound in layers with absorbable suture.

In girls, the operation is somewhat more straightforward. The sac is usually very long and redundant. There is no spermatic cord of course, but the round ligament should be divided just distal to the sac. The sac must be opened and the absence of a sliding hernia needs to be confirmed, as failure to do so can result in injury of the fallopian tube, ovary, bladder, or bowel.

The incidence of contralateral hernia when only one is evident on examination is unknown but might be no more that 10%. Traditionally, boys under the age of 1 or 2 and girls under the age of 5 were thought to be at higher risk and were therefore recommended to undergo contralateral groin exploration at the time of hernia repair. Risks of spermatic cord injury are high enough that contralateral exploration is no longer advocated in all cases. It appears that premature infants, boys with left sided hernias, and those with a strong family history of inguinal hernia may be at higher risk and should be considered for contralateral exploration. Laparoscopy may be used to "explore" the contralateral side by placing the lens through the hernia sac or through a small umbilical incision. Hernias may be repaired via a laparoscopic approach, which is the primary repair by our surgeons.

Complications

Complications after inguinal herniorrhaphy are rare. Infection, bleeding, and injury to adjacent structures are discussed preoperatively but are extremely rare occurrences. All parents should be warned that recurrence can occur, although the incidence is probably about 1%. Post-operative hydroceles are relatively common, but typically resolve within several weeks of surgery. Those that don't may respond to serial trans-scrotal needle aspiration, or rarely, reoperation. A recurrence can be due to a direct inguinal hernia, which requires formal reconstruction, e.g. Cooper ligament repair.

<u>The most common variants of inguinal hernias and hydroceles</u>

    A.   Normal (closed) processus vaginalis

    B.   Noncommunicating hydrocele

    C.   Communicating hydrocele

    D.   Hydrocele of the cord

    E.   Inguinal hernia

    F.   Scrotal hernia

Pyloric Stenosis

History:
Pyloric stenosis typically occurs in newborns from 2 to 12 weeks of age. The classic presentation is progressive, projectile, non-bilious emesis. If there is any bile (green or bright yellow), pyloric stenosis is unlikely and malrotation must be ruled out immediately with a STAT UGI. Males are more frequently affected than females and a positive family history strongly supports the diagnosis.

Physical Examination:
With experience and patience, an "olive" may be palpable in the epigastrium. If an olive is palpable by an experience examiner, no further diagnostic studies are required. The diagnostic work-up should be discussed with the attending surgeon.

Laboratory:
All patients require a STAT basic metabolic panel. The classic derangement is hypokalemic, hypochloremic, metabolic alkalosis. A urinalysis may reveal paradoxic aciduria and an elevated specific gravity. No other labs are necessary although occasionally the anesthesiologist will request that a CBC be sent.

Fluids:
Most patients will require a bolus of 0.9% Normal Saline
(20 ml/kg over 1 hour). D51/2 NS +/- 20 mEq KC/L is given at 1.5 maintenance until electrolytes are corrected (HCO3 < 28 CI > 100), then given at maintenance. This may take 8 to 12 hours.

Work Up:
An ultrasound should be ordered. As always, diagnostic work-up should be discussed with the attending surgeon.

Operation:
The standard treatment is Ramstedt pyloromyotomy performed through a peri-umbilical incision or laparoscopy (our preferred method).

Post-operative:
A feeding schedule is started post-operatively. Patients are initially NPO for hours, and then started on breast milk or formula. Limit to < 3 oz. initially, then advance full (ad lib) feedings, if tolerated. It is common for babies to spit up after the surgery and is not necessarily a sign of a complication.

Intussusception

History:
Classic ileocolic intussusception typically occurs in patients from 3 months to 3 years. The classic presentation is of episodic abdominal pain, manifested by crying and drawing of the legs toward the body, with quiet periods in between. The pain recurs every 15 to 20 minutes and the child may appear entirely

well between episodes. All children in the proper age group with this history have intussusceptions until proven otherwise. Another classic but less common presentation is that of lethargy which can be easily confused with sepsis or meningitis.

When consulted for "R/O intussusception" our role is to assess and examine the child to rule out peritonitis or other contraindication to contrast enema. Intussusception cannot be ruled out by history or physical examination, although another diagnosis may be suspected.

Physical Findings:
- palpable "sausage-shaped" mass in right upper quadrant (rarely appreciated)
- "currant-jelly" stool (occurs late)
- abdominal distention due to SBO (occurs late)
- peritonitis due to ischemic bowel (occurs very late)

Diagnosis:

An AXR may suggest the diagnosis but can neither confirm nor rule-out the diagnosis regardless of the findings. Suggestive findings include: a soft-tissue mass in the right upper quadrant, absence of gas in the cecum, or a picture suggestive of a bowel obstruction. An AXR should always be done first, but should never dissuade you (or the radiologist!) from proceeding with a more definitive study unless, of course, free air is seen. Ultrasound is the test of choice, but is user dependent and may not be helpful in the middle of the night.

The definitive diagnostic study is a contrast enema with air or water soluble contrast. This can confirm the diagnosis and treat the condition. The patient must be examined by a surgeon to rule out evidence of peritonitis (an indication for immediate surgery, not an enema). It is preferred for patients to have an IV in place, and some surgeons prefer that antibiotics are given prior to the contrast enema. The surgical attending should be made aware of the study before it happens but does not need to be present or even in the hospital when it occurs. The student should be present during the study for their education as well as for efficient patient care.

Treatment:

A contrast enema is successful in reducing the intussusception in up to 85% of cases. After successful reduction, the patient is admitted to the peds surgery service and diet should be advanced. The patient should be observed for 24 hours. There is a 5-7% recurrence rate; most occur in the first 24 hours. If symptoms recur, the enema should be repeated. Failure of enema reduction is usually an indication for urgent surgery.

In the event of a perforation during the enema, don't panic. Notify the surgery attending immediately, accompany the patient back to the floor, and make preparations for an urgent operation: ensure good IV access, give a bolus of crystalloid (20mL/kg over 1 hour), give a dose of Zosyn or similar antibiotic and try to reassure the parents.

Gastroesophageal Reflux

Gastroesophageal reflux (GER) is relatively common in infants and children. Those with symptomatic GER that have failed medical management, especially those with significant neurologic impairment, are candidates for an anti-reflux operation. During the course of your rotation you will be asked to consult on patients being considered for gastrostomy placement and Nissen fundoplication. You will also be participating in the operations as well as the pre- and post-operative care of these patients.

Work-up:

There are several standard diagnostic procedures that are useful for patients with GER. Most patients will require one or more studies before being considered for surgery.

1. Upper GI: Must be done for all patients before an operation is performed. Although not a sensitive study for GER, it provides critical anatomic and functional information. The three important questions are: (a) Is there evidence of malrotation or other anatomic abnormality?, (b) Is there evidence of esophageal stricture, esophageal dysmotility, or hiatal hernia?, (c) Does the stomach appear to empty normally?

2. <u>pH Probe Impedence study</u>: The most sensitive and specific test for the diagnosis of GERD. This important test is coordinated by GI and requires an overnight hospital stay. This should be done in most patients who are considered for surgery unless there is clear clinical evidence of severe reflux.
3. <u>Gastric Emptying Scan:</u> Sensitive for reflux but most useful in assessing gastric emptying. A nuclear medicine study that uses 99m-Technetium sulfur colloid, it is no longer a required study before surgery. It is now felt that poor gastric emptying may improve after fundoplication alone, thus avoiding the risks associated with pyloroplasty (i.e. dumping syndrome) which was routinely done in these patients in the past.
4. <u>Esophagoscopy:</u> Useful for evaluating strictures, esophagitis and webs. Not a required study but useful in some patients. This is performed by the peds gastroenterologists in most cases.

Medical Management:

Standard management concepts include: small, frequent feedings, upright positioning; thickened feedings; H2-blockers or PPI, pro-motility agents such as metaclopramide (Reglan). Alternative feeding regimens may be useful in some patients: continuous NG feeds, or jejunal feeds via an NJ- or GJ-tube.

Indications for Surgery:

1. Vomiting – More than just a social concern, vomiting can interfere with activities or physical therapy and it can
contribute to poor total caloric intake. Vomiting can also lead to oral aversion.

2. Failure to thrive (FTT) – Other causes need to be ruled out, but vomiting due to reflux can be a cause.

3. Recurrent aspiration – Manifested clinically by coughing and choking, but can also result in recurrent pneumonia. More subtle manifestations include asthma, laryngospasm, or sinusitis.

4. Esophagitis + stricture – Documented by UGI or esophagoscopy. Persistence or recurrence of esophagitis despite maximal medical therapy is an indication for surgical intervention.

5. Apnea/Near SIDS – An indication for surgical intervention if the reflux work-up is positive and other causes have been ruled out.

6. Feeding gastrostomy in a neurologically impaired child – A relative indication for anti-reflux surgery but still somewhat controversial. Generally indicated if reflux is present, but rarely done as a prophylactic maneuver.

7. Asthma/Sinusitis – Some patients with asthma or sinusitis can be shown to have reflux and chronic aspiration. This is somewhat controversial and difficult to document.

Pre-operative evaluation:

Occasionally, however an inpatient consult will be called in to the surgery service. There needs to be documented evidence of significant reflux for a child to be considered for surgery. This is preferably in the form of a positive pH probe study, but a gastric emptying scan positive for reflux, esophagoscopy positive for esophagitis, strong clinical evidence, or some combination of these may be sufficient to justify an operation. A relatively recent UGI is also required to demonstrate upper GI anatomy. Reflux seen on an UGI is not sufficient evidence by itself of significant reflux, but must be correlated with the clinical findings. Severe esophagitis or active pneumonia should be treated prior to undertaking an operation.

Operation:

Most patients undergo a Nissen fundoplication (360° wrap). In many patients, especially those who are neurologically impaired, a gastrostomy is placed at the same operation. The fundoplication is performed laparoscopically.

Post-operative Care:

Protocols vary with the preferences of the surgeon and the needs of the patient. General guidelines are: NPO 24 hours, prophylactic antibiotics for up to 24 hours, nubain or morphine sulfate for pain (up to 0.1 mg/kg IV q2h PRN pain), attention to pulmonary toilet (incentive spirometry or respiratory therapy) and early mobilization. Gastrostomy tubes may be used for medications and venting. Resumption of feeds is usually the factor that determines when a patient is discharged after a fundoplication. Typically feeds are started PO or by gastrostomy on POD I at 1/2 volume and then gradually increased to full feeds over 2 to 3 days. Discuss the feeding advancement schedule with the attending before initiating it. The peds nutrition service may also be consulted at this time to suggest a feeding regimen for optimal nutrition. If the patient has a new gastrostomy tube, the parents must be instructed in its management and care; the CNP will coordinate the education plan.

As with all complex patients, discharge planning needs to be anticipated ahead of time. The CNP will work with the case managers to coordinate parental instruction, home therapy support, and supplies.

Complications:

In addition to the more common post-operative complications that can occur after any abdominal operation, there are a few that are unique to the fundoplication:

Esophageal perforation: Rare, but a dreaded complication. Usually presents within first 12 hours post-op with high fever, tachycardia, tachypnea, dysphagia, and chest pain. If suspected, a CXR should be obtained and may show a pneumothorax or pleural effusion. A (water solube) contrast esophagram should be obtained next to confirm or rule out the diagnosis.

Aspiration pneumonia: Fever, tachypnea, abnormal breath sounds on auscultation. Confirmed by CXR, treated with antibiotics.

Wound infection: Presents with fever, wound drainage 5 to 7 days post-op. Wound often needs to be opened, but discuss with attending first.

Subphrenic abscess: Very uncommon, typically presents 7 to 14 days post-op with fever, ileus, abdominal pain, elevated WBC. Confirmed by abdominal CT or ultrasound.

Gas Bloat/Poor gastric emptying: Often occur together and present with abdominal discomfort, retching/gagging. Usually relieved by venting the gastrostomy or placing an NG tube. Usually resolves on its own but may take up to several weeks.

Dumping: A characteristic syndrome of tachycardia, diaphoresis, crampy abdominal pain, and often diarrhea that is thought to be caused by rapid emptying of the gastric contents into the duodenum. It is more common after pyloroplasty. It is often temporary but can persist for a long time. Treatment includes smaller more frequent feedings, continuous feedings, or a change in formula (less hyperosmolar).

Dysphagia: Patients will often complain of difficulty swallowing for 4 to 6 weeks after fundoplication. Non-verbal children will demonstrate poor PO intake or increased secretions (drooling). A soft or liquid diet sometimes helps: the problem usually resolves spontaneously.

Loss of G-tube: A new gastrostomy is usually replaced no sooner than 6 weeks post-operatively. If a new gastrostomy is dislodged prior to its first post-operative replacement, it should be replaced by experienced personnel and then confirmed by radiology. If it cannot be replaced, a radiologist will sometimes be asked to attempt it under fluoroscopy. Dislodgement of a fresh gastrostomy tube may require an operation for replacement.

NICU Consults

Few patients are as intimidating as a newborn baby, especially when premature or small for gestational age. Remember that the care of the infants in the NICU is under the direct supervision of the neonatologists and that our role is that of consultants, albeit very involved consultants. When asked to evaluate an infant for a surgical problem, the same rules apply as for all other consults: obtain a detailed history, perform a complete physical examination, gather all pertinent laboratory values and results of imaging studies, and call the attending to discuss the plan. The following are some of the more common surgical problems.

Neonatal Bowel Obstruction:

A relatively common reason for a surgical consultation in the NICU, there are a handful of common diagnoses as well as a longer list of some uncommon ones to sort through. The diagnostic protocol is usually straightforward but subtleties in the presentation may warrant a slightly different approach; therefore, always discuss the plan with the attending and NICU before initiating a work-up.

Presentation:

Typically, a full-term newborn who develops feeding intolerance, abdominal distension, bilious emesis, and/or failure to pass meconium in the first day or two of life. But the presentation may vary. Important historical information includes gestational age and weight, history of oligohydramnios or polyhydramnious, results of antenatal ultrasound, and group-B strep status.

Work-up:

As always, start with a complete physical examination: overall appearance, vital signs, and abdominal exam are most important. Check for presence and location of the anus, but DO NOT perform a rectal examination (it may alter the results of subsequent studies). Always insist on placement of a Replogle or equivalent decompression NG tube and obtain an abdominal x-ray.

The common causes of neonatal bowel obstruction include:

- duodenal atresia
- bowel atresia (jejuna, ileal, or colonic)
- imperforate anus
- Hischsprung's disease
- meconium plug syndrome, meconium ileus,
  (sometimes with antenatal perforation and chemical peritonitis or a meconium "pseudocyst")
- malrotation w/ or w/o volvulus
- segmental volvulus Less common causes include:
- small left colon syndrome
- duplication cyst
- lactobezoar
- incarcerated hernia

Causes of ileus that can present as obstruction:

- generalized sepsis (most common)
- NEC (rare)
- hypokalemia (rare)
- prematurity (uncommon)
- hypothyroidism (very rare)

- hyerpmagnesemia (especially in pre-eclamptic moms)

The diagnosis is rarely made by history or physical examination alone, with the exception of *imperforate anus*, and *incarcerated hernia*. The diagnosis is often suggested by the abdominal x-ray: A "double-bubble sign" is diagnostic of duodenal atresia, no further studies are necessary: a very distended loop of bowel filled with stool likely represents meconium ileus; and a very dilated loop of bowel with air/fluid levels a bowel atresia. Most of the other diagnoses show a non-specific pattern of dilated loops of bowel (it is <u>impossible</u> to distinguish small bowel from colon on a plain film of a newborn) consistent with a bowel obstruction or sometimes a normal bowel gas pattern (which should not dissuade you from pursuing further studies, given a clinical picture of bowel obstruction.)

Some diagnoses are made with a high degree of certainty by this point and require no further studies (e.g. imperforate anus, duodenal atresia, bowel atresia {although some bowel atresias require contrast enema}). Similarly, a child who is gravely ill with evidence of ischemic bowel or a perforated viscus should undergo urgent laparotomy rather than a diagnostic study. Most other cases require a work-up. It is usually best to start with a water soluble <u>contrast enema,</u> if meconium ileus is suspected. This study is diagnostic and often therapeutic for *meconium plug syndrome* and *meconium ileus*. It may also suggest the diagnosis of *Hirschprung's disease* or *small left colon syndrome*, thus avoiding an urgent laparotomy. It does not accurately rule out malrotation, even if the cecum is in a normal location.

If the enema is non-diagnostic, a decision is then made to either prepare the patient for laparotomy or proceed with an upper GI contrast study specifically to rule out malrotation. (A small bowel follow through is not usually requested.) Most newborns with a bowel obstruction of unclear etiology will require a laparotomy at some point. A child with malrotation, regardless of how well he or she looks clinically, requires an emergent laparotomy. Many other patients with obstruction who are clinically well may benefit from NG decompression or further resuscitation, so an UGI can help determine the timing of surgical intervention. Patients with a small bowel atresia should not have an upper GI study performed. Remember to let the radiologist know ahead of time that you may want an UGI after the contrast enema; this may affect their choice of contrast material.

Ileus presenting as obstruction can be perplexing and in rare cases can even prompt a laparotomy if a true obstruction cannot be ruled out by other means. The diagnosis is usually apparent given other clinical factors and the diagnostic work-up described above is not always necessary. Always discuss the plan with the attending so as to avoid a potentially life-threatening diagnostic procedure in an infant who is already seriously ill.

Most causes of obstruction do not require surgery emergently, with two exceptions: If there is a strong suspicion of midgut volvulus (bilious emesis, little or no gas beyond the duodenum on x-ray, a tender abdomen with a mass, blood per rectum) or antenatal perforation (distended, erythematous abdomen with intraabdominal calcification on x-ray), the work-up, resuscitation, and operation must be performed expeditiously.

Esophageal atresia with or without tracheo-esophageal fistula:

Esophageal atresia (EA) with or without trachea-esophageal fistula (TEF) is a complex congenital anomaly with multiple variations and a number of significant associated anomalies. The conventional classification system with approximate frequency is as follows:

Type A	Isolated esophageal atresia	8%
Type B	EA w/proximal fistula	2%
Type C	EA w/distal fistula	85%
Type D	EA w/prox. And dist. Fistulas	<1%
Type E	Isolated TEF ("H type")	5%

Patients usually present with excessive drooling and choking with feeds shortly after firth. The diagnosis is confirmed with placement of an NG tube, which typically cannot be advanced more than 10-12cm. A plain x-ray ("babygram") will demonstrate the tip of the NG in the upper mediastinum. By injecting air into the tube and taking the film right away, the proximal esophageal pouch can often be visualized. Contrast should not be used. The presence of intestinal gas is diagnostic of a TEF to the distal esophagus. A gasless abdomen usually signifies a pure EA without fistula. "H-type fistulas usually present weeks after birth with a history of coughing or choking with feeds.

Associated anomalies:

VACTERL: Vertebral anomalies, Anorectal malformations, Cardiac defects, Tracheal and Esophageal anomalies, Renal abnormalities, and Limb anomalies. Cardiac defects are sometimes lethal. All patients with EA +/-TEF should be screened for additional anomalies.
This includes: careful physical examination (esp. cardiac and anus), plain x-rays (vertebral anomalies), cardiac echocardiography, spinal
ultrasound and renal ultrasound. The other important question for the echocardiographer is the location of the aortic arch, as this may determine the surgical approach (right vs. left thoracotomy).

Initial Management:

The definitive treatment for esophageal atresia is surgical: through right (usually) thoracoscopy, or right thoracotomy the fistula is ligated and the two ends of esophagus are joined end-to-end. The timing of the operation is semi-elective (next day). While awaiting surgery, the infant is kept NPO with a Replogle(depocmpression) tube left in the proximal pouch and left to constant suction and IV fluids are started. Anti-reflux precautions are instituted and the screening studies (ECHO, renal ultrasound, abdominal x-ray) are performed. In preparation for surgery, the infant is given antibiotics (Amp/Gent) and an H2-blocker).

Post-operative care:

A feeding tube is sometimes placed during surgery to help stent the two ends of the esophagus; the corpak may also be used for enteral feedings during the postoperative period. Patients are typically left on the ventilator overnight. A chest tube is left as a drain of the anastomosis and usually remains until after POD 7, when a water soluble contrast study is done to rule out a leak. Antibiotics are continued post-op at the discretion of the surgical attending.

Abdominal Wall Defects

Infants with gastroschisis or omphalocele are usually known about well in advance due to the prevalence of antenatal ultrasound screening. The distinction between these two types of abdominal wall defects is important because of differences in associated anomalies, timing of repair, and the anticipated clinical course after surgical repair:

	Gastroschisis	Omphalocele
Location	To the right of umbilicus	midline at the umbilicus
Membrane	absent	present (may be ruptured)
Umbilicus	intact	attached to membrane
Associated anomalies	Rare	common
Post-op ileus	prolonged	varies

The size of the defect in gastroschisis is almost always 2 to 4 cm, whereas the size of the defect in omphalocele is very variable, from a tiny "hernia of the cord" to a so-called "giant" omphlocele (larger than 10 cm, often with liver with it). Patients with gastroschisis rarely have other anomalies although intestinal atresias occur with slightly higher frequency due to in utero volvulus of the exposed bowel. Omphalocele is associated with sometimes severe and life-threatening anomalies, such as Beckwith-Wiedemann syndrome (macroglossia, hypoglycemia), trisomy 13, 18, or 21, and cardiac defects. Patients with omphalocele should therefore undergo STAT glucose monitoring, cardiac echo, and renal ultrasound.

Initial management:

Gastroschisis is repaired urgently. In preparation for surgery, the infant is placed within a clear vinyl bag up to the axillae into which a small amount of warm saline is placed. IV access is obtained and IV antibiotics (Amp/Gent) are given. A Replogle NG tube is inserted and placed on constant suction. IV fluid and (sodium) requirements are often extremely high.

It is extremely important to inspect the exteriorized bowel, as it may become ischemic as the baby swallows air and the intestine becomes more distended, or if the bowel hangs dependently over the side of the abdomen. The viscera should either be kept on top of the abdomen, or the baby placed on its side. If the bowel remains cyanotic, one must relieve the compression by urgently enlarging the abdominal wall opening, which can be readily performed at the bedside using a right angle clamp and a concept cautery, cutting the skin for a centimeter in the right lateral direction.

Patients with omphlocele with an intact membrane can be treated semi-electively, IV fluids, IV antibiotics, and an NG tube are instituted. Renal ultrasound, cardiac echo, and blood glucose are obtained.

Surgical repair:

Depending on the size of the defect and the ability to place the viscera within the abdominal space, the defect is repaired in one of three ways: primary closure (bowel reduced, fascia closed), or creation of a silo with staged primary closure (bowel unable to be reduced). The factor that determines safety of closure is peak inspiratory pressure (PIP). A fascial closure that is under a great deal of tension will not allow proper ventilation. The PIP should be less than 30 cm H2O at the completion of the closure. Silos are made of silastic and are partially reduced daily until the bowel is completed reduced, at which time the infant is returned to the operating room for definitive closure (typically 5 to 7 days).

Post-operative care:

Patients with gastroschisis require TPN for up to 2 weeks or more due to prolonged ileus. Patients with omphalocele generally resume enteral feeds much sooner. The sign that signals resumption of bowel activity is typically a transition in the color of NG drainage from green to clear and a decrease in NG volume. More pressing initial concerns include: respiratory function and timing of extubation, prevention of sepsis, and attention to IV fluid status with avoidance of dehydration as well as over-hydration.

Necrotizing Enterocolitis

Necrotizing enterocolitis (NEC) is an invasive and sometimes severe intestinal infection of unclear etiology that affects newborn (usually premature) infants. It can occur in epidemics, sometimes closing down whole nurseries for periods of time. No single infectious agent has been identified as a causative agent. Although the disease can affect any newborn up to 3 months of age, there are risk factors that place certain newborns at much high risk for NEC: <31 weeks gestation, weight <1500 grams, presence of pulmonary disease, hyperosmolar feedings, sepsis, jaundice, and umbilical artery catheters. Feedings seem to play a role in that children who are breast fed have slightly lower rates of developing NEC and those who are not fed enterally at all may be at even lower risk.

Clinical presentation:

NEC typically presents after 7 days of age with feeding intolerance, abdominal distension, vomiting, bloody stools, temperature instability and/or abdominal tenderness.

Diagnosis/Work-up:

Clinical signs may be enough to prompt empiric treatment for NEC, although the diagnosis can usually be confirmed by a combination of laboratory data and imaging studies. Associated laboratory abnormalities include leukopenia (WBC<3,000), thrombocytopenia (plt ct < 150,000, or a decline of >100,000), anemia, and metabolic acidosis. An abdominal x-ray should be

obtained on every patient suspected as having NEC. In the appropriate clinical setting, the finding of pneumatosis intestinalis is diagnostic of NEC. Other potential findings include dilated loops of bowel, portal vein air, ascites, and free air. Occasionally the x-ray findings are within normal limits.

Treatment:

The therapy of NEC is primarily medical: NPO, NG decompression of GI tract, IV fluids, IV antibiotics. Umbilical artery catheters should be removed and plans for TPN should be made. Patients are usually transfused with blood to keep the Het < 30-35%. Treatment is continued for 7-10 days depending on the severity of the illness. Meanwhile, careful surveillance for complications is ongoing. Surgical intervention is reserved for life-threatening complications of the disease such as bowel perforation or dead bowel.

Indications for surgical intervention:

- pneumoperitoneum
- cellulitis of the abdominal wall (erythema and/or edema)
- clinical deterioration despite medical therapy
- "fixed loop" on radiograph or physician examination
- paracentesis positive for meconium or dead bowel (rusty brown color)
- a significant fall in platelets, or persistent metabolic acidosis

Although in some cases the need for surgical intervention is obvious (e.g. free air on x-ray), others may involve a great deal of judgment and careful decision making. An important strategy is that of frequent serial assessments: examination, x-ray, CBC, ABG every 4 to 6 hours. In this way, changes are detected early and trends are followed closely.

Surgical intervention in babies with NEC generally means a formal laparotomy. The parents are prepared for the likelihood of bowel resection and enterostomy. At operation, all frankly necrotic bowel is resected and usually no attempt is made to perform a repair or anastomosis. If a large amount of intestine is involved, small segments of frankly necrotic bowel are excised, a stoma is brought out, and plans are made for a "second-look" laparotomy in 24 to 48 hours.

Some infants who are very premature or very small (<1000g) may be considered too ill for a formal laparotomy. These infants are candidates for drainage for of the peritoneal cavity. This is performed at the bedside with the patient under sedation and using aseptic technique. One or more small incisions are made on the abdomen and penrose drains are placed into the peritoneal cavity. Decisions regarding further surgery are made based on the patient's clinical progress after drainage.

Stomas are usually taken down after at least 6 weeks or when the patient has achieved some weight milestone. (e.g. 2.5kg). However, the timing of surgery is highly dependent on surgeon's preference as well as patient factors. Prior to take down of the stoma, all infants with a history of NEC should undergo a contrast enema to rule out the presence of a distal stricture. An infant with medically-treated NEC may also develop a stricture that usually presents as bowel obstruction. The diagnosis is confirmed by contrast enema and/or UGI and the treatment is surgical resection and primary anastomosis.

Need for Central IV Access
NICU babies often require central IV access for TPN, antibiotics, or blood draws. PICC lines are most commonly used. The neonatal nurse practitioners place the PICC lines in the NICU and will consult the peds surgery CNP for those PICC placements that benefit from her expertise. When this fails we may be asked to place a central line. Central line placement may require a cutdown

technique; this is usually performed in the NICU with the infant under sedation. With rare exception, the baby <u>must</u> be intubated prior to central line placement. The potential access sites are external jugular vein, facial vein, internal jugular vein, and saphenous vein at the thigh. Informed consent is obtained as for any surgical procedure. The patient is made NPO for at least 4 hours and anesthesia is coordinated by the neonatology staff. Bacteremia is an indication to wait 48 hours prior to placing the line. Platelet count should be at least 50,000/ mm3. The pediatric central line cart is stored in the OR and is available to be transported for use in the NICU.

Imperforate Anus

Anorectal malformations occur with a incidence of approximately 1 in 5000 live births, and is slightly more common in males. It typically occurs sporadically and is an isolated anomaly, however it may be associated with duodenal atresia, Down syndrome, tethered cord, and any of the VATERL-associated anomalies (vertebral
anomalies, trachea-esophageal anomalies, renal, radial limb, and/or cardiac anomalies.)

Numerous classification schemes have been suggested over the years, however for practical purposes, anorectal anomalies can be divided into *low* lesions (perineal fistula, suitable for early primary repair) and *high* lesions (rectourethral fistula, treated with early colostomy and delayed repair). Most anorectal anomalies in girls present with a perineal fistula, although many have a fistula located at the posterior aspect of the vaginal orifice (*fourchette* or *vestibular fistula*) which are considered by some to be *intermediate* lesions.* Many boys with imperforate anus present with a perineal fistula covered by a thin membrane and may therefore initially appear to have high lesions. The classic appearance is that of a perineal dimple within the medial raphe' with a thickened strand of skin crossing it in the midline that is sometimes referred to as a *bucket-handle* deformity. These infants nearly always have a low lesion.
The **initial management** of an infant with imperforate anus focuses on three things. The first issue is to prevent intestinal distension and sepsis. Babies should be made NPO and an OG or NG tube should be placed. Intravenous fluids are started and IV antibiotics are given. The fistula can be gently dilated with a small Hagar dilator or a hemostat to encourage passage of meconium. The second issue is to rule out associated anomalies. All babies with imperforate anus should have a thorough <u>physical examination</u> to rule out a cardiac murmur, radial limb anomalies (thumb and forearm), stigmata of Down syndrome, evidence of esophageal atresia (excessive secretions, inability to pass an NG tube into the stomach), and GU anomalies (hypospadias, undescended testes). A whole body plain <u>radiograph</u> ("babygram") should be done to rule out vertebral anomalies and excessive bowel distension. Ultrasound examination should be performed of the kidneys and spinal cord and an <u>echocardiogram</u> with cardiology consultation are obtained.

The third issue is an assessment of the type of lesion and plans for operative intervention. Most lesions can be classified accurately on the basis of physical examination. In general, most low lesions will present with some form of a perineal fistula while high lesions rarely do. Fourchette fistulas (and some scrotal fistulas) can be associated with high lesions. Careful examination can usually determine the proximity of the rectum to the perineal skin: a blunt instrument such as a hemostat is placed in the fistula and directed posteriorly; if a rectal pouch is present and close to the perineal skin, the lesion can be treated as low lesion. In the absence of a fistula, a determination needs to made whether the rectum is low or high. The operation (repair or colostomy) is not urgent and some cases it is best to delay the operation for up to several days until classification of the lesion can be made with certainty.

Medical imaging techniques can be used to determine the level of the lesion. An <u>invertogram</u> (Wagensteen-Rice view) is a cross-table lateral x-ray of the pelvis with the baby in a knee-chest (i.e. bottom-up) position. The air-filled rectum can usually be visualized and if it is proximal to a line drawn between the pubis and the coccyx ("PC line") it is considered to be a high lesion. If it is distal to a line drawn at the ischial tubersoity and parallel to the PC line, than it is considered low. Anything in between the two lines is considered intermediate. There are important limitations to this study. It can take 12 to 24 hours for swallowed air to reach the rectal pouch

and even then, if the pouch is not visualized, it can be due to a lack of air, not necessarily the presence of a high lesion. <u>Ultrasound</u> can sometimes be helpful to visualize the rectal pouch and determine its distance from the perineal skin. Most would consider a distance less than 2 cm to be amenable to primary repair in the newborn period. Rarely, a constrast study by injecting the fistula can be helpful. After diverting colostomy, prior to planning definitive repair, a <u>distal colostogram</u> is done through the mucous fistula to determine the distal extend of the rectal pouch.

*The distinction between high and intermediate lesions is somewhat arbitrary in that the initial treatment, diverting colostomy, is the same regardless. There may be an implication for the extent of reconstruction necessary at the time of definitive repair but the question is: "colostomy or primary repair?"

Definitive repair of imperforate anus involves some form of anoplasty. For lesions that present with a fistula that very close to the normal location of the anus, especially in girls, a cutback anoplasty may be all that is necessary. Although performed at the bedside in the past, the current standard is general anesthesia. The procedure involves incising the perineum posterior to the fistula and performing what is essentially a V-Y plasty, so that the majority of the anal opening is encircled by sphincter muscle. Two to three weeks after surgery, daily dilatations are begun in the office and continued at home for 3 to 6 months, depending on the nature of the subsequent scar.

Laparoscopic assisted anorectal pull through is currently the most common approach to all high anorectal malformations. To avoid injury to the urethra, a foley catheter must be placed in all boys, so that the urethra is more easily identified intra-operatively. The patient is place in a supine position with a large roll under the abdomen and all pressure-points padded. A specialized electrical stimulator is used to identify the anal sphincter. The rectum is then mobilized circumferentially, while the fistula is identified. The anterior aspect of the rectum is invariable attached very closely to the vagina in girls or the urethra in boys, and this is therefore the most difficult and delicate part of the procedure. When enough rectum has been mobilized, and the fistula is divided, the location of the sphincter is confirmed with the stimulator. The anus is then pulled through and sutured to the skin. The rectum is sutured circumferentially to the anal skin, using the site of the fistula as the lead point. The new anus is gently sized with a Hagar dilator (should accommodate appx. #10) and antibiotic ointment is applied to the skin.

Postoperatively, the patients are allowed to eat when they stool consistently, usually the next day. Antibiotics are continued for 48 to 72 hours post-op to prevent the rare but dreaded complication of wound sepsis. Complications are rare and include would infection, skin breakdown, and dehiscence with retraction of the rectum. Patients are usually discharged within 3 to 5 days. Follow-up should be in appx. 2 weeks, at which time the parents are taught to dilate twice daily. The size of the dilator is determined by the largest dilator that is accommodated comfortably. Every week, the size of the dilator is increased one size until a #10 dilator is able to be passed easily. Dilatations continue until the circular scar is felt to be pliable and stable.

The results are generally good from the anatomic standpoint. Functionally, nearly all children with low imperforate anus are constipated, despite successful anoplasty. This is presumed to be due to an associated defect in the musculature and/or neurologic control of the sphincter. Those with high imperforate anus have a higher incidence of fecal incontinence, and may also be constipated. Patients are therefore followed long term and are maintained on a non-constipating diet and sometimes a medical regiment to prevent constipation.

Common On-Call Problems

There is no way to anticipate every potential problem to be encountered when one is on-call at night. The following represent some of the more common issues that can arise. For all calls, it is best to follow some simple rules: always be professional and courteous to the caller, always go and assess the patient yourself, always document the encounter in the chart, and never be afraid to call the on-call attending even in the middle of the night – we would much rather address the problem right away than hear about it from an angry nurse or parent in the morning!

1. **"Your patient is 6 hours post-op and hasn't voided yet."** Urinary retention can occur after any general anesthetic and is more likely to occur when the patient is given narcotic analgesics and when they have not yet ambulated. Contrary to the situation in adults, children rarely require catheterization: it is usually unnecessary, it is emotionally traumatic, and is associated with urethral strictures, especially in boys. As always, assess the child yourself. If they are able and permitted to do so, have them stand up and try to void. Consider asking the bedside RN to bladder scan the patient's bladder. If the bladder is not palpably distended, consider a NS fluid bolus (10-20 mL/kg over 1 hour) and try again in an hour or two. Consider catheterization only if the bladder is very distended and a reasonable effort to make the patient void has been made, or if 10 or more hours have passed since surgery. Of course, use good judgment to modify the rules depending on the clinical situation.

2. **"Your patient is post-op and in a lot of pain – can I give him more morphine?"** When analgesics orders are written appropriately, this should be a rare occurrence. Always consider whether the patient is in pain for some other reason, such as a complication. Assess the patient yourself and check the medication administration record – were the analgesics given appropriately? If the pain can be attributed to the incision and the pain meds as ordered are clearly insufficient, then increase the dose. A "full" dose of morphine sulfate is 0.1 mg per kg and should be given every 2 hours on a PRN basis. If the patient seems to need more, do so only cautiously. Consider notifying the attending before doing so. Children under 6 months of age should not receive narcotic analgesics except under rare circumstances and only when fully monitored. Infants should instead be given acetaminophen (up to 15 mg/kg PO/PR/IV every 4 hours PRN) or ibuprofen (up to 10mg/kg PO every 6 hours).

3. **"The IV is out and we can't get another on back in."** These calls can be particularly frustrating and time-consuming. In some patients, the IV can be left out, for example, if they are able to take liquids and are not likely to require any critical meds before morning. More often, the IV needs to be replaced.

4. **"Your patient got a bowel prep but he is still passing stool."** For some patients, it is critical that their GI tract is extremely well-prepared, while for others it is less important. The "cleanest" we can expect a patient's stools to be is slightly bile-stained with flecks of mucus – it will never be "like water." If the stool is still brown or significantly solid and it is before midnight, consider giving more of the cathartic used (e.g. GoLytely 20mL/kg over 2 hours). It is best to consult with the attending on-call first.

5. **"Your post-op patient is vomiting."** Nausea and vomiting are very common after exposure to a general anesthetic. If it is the night of surgery and the patient has a non-GI procedure, it is usually safe to give ondansetron (Zofran 0.1 mg/kg IV q6h PRN, max of 4 mg). Metaclopramide (Reglan) is usually not effective. Phenergan is not indicated for children. Also, don't forget to ask the color of the vomitus. Also consider an alternative to morphine, which can cause nausea. For patients who have had a GI procedure, especially if it has been more than 24 hours post-op, an ileus or obstruction might be the cause and anti-emetics might be contraindicated. A nasogastric tube should be considered but it is best to delay placement until it is
clearly indicated.

6. **"Your post-op patient has a fever."** The response to this issue depends on the clinical situation and the post-operative day. Fevers are common in the first three days after a major operation. Often attributed to "atelectasis" they are likely part of a physiologic response to the stress of surgery. These patients need to be examined to rule out unusual causes of post-op fevers, such as phlebitis due to an IV or a necrotizing would

infection. In general, blood work and x-rays are not required. Encourage ambulation and deep breathing and reassure the patient and family. High fevers and those that occur after the third post-op day need to be assessed more carefully to rule out serious infections. Use a careful, thoughtful diagnostic approach, rather than a "shotgun" approach. Examine the patient first

(don't forget vital signs and I/O's), then decide whether further studies are necessary. Consider ordering urinalysis, CBC, blood culture, and/or CXR. Consider wound infection, abscess, central line infection, and deep venous thrombosis (extremely rare in children).

7. **"Mom has some questions and she's very upset."** <u>Always</u> respond in person and try to answer all questions in a calm and reassuring manner. Never be defensive, condescending, or rude, even if you feel the family is being unreasonable. Of course, don't be afraid to say "I don't know" either. An angry parent should always be dealt with urgently and <u>always</u> notify the attending as soon as possible.