PEDIATRIC SURGERY MANUAL

Department of Surgery
Division of Pediatric Surgery
Milton S. Hershey Medical Center
Pennsylvania State University

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In dedication to the memory of Thomas Van Ness Ballantine whose love of surgery and children helped to establish the Pediatric Surgery program at the Milton S. Hershey Medical Center.

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Welcome to the Division of Pediatric Surgery. As a resident or medical student, we anticipate that you will have a worthwhile learning experience, as well as an opportunity to demonstrate your fund of knowledge and your enthusiasm for the care of children. We are all anxious to enhance your education, and are always available for consultations and discussions as well as the routines of daily rounds.

GOALS:

The primary mission of the Section of Pediatric Surgery is to provide the highest quality pediatric surgical and trauma care to our patients. It is expected that the students and residents will join the staff in working to accomplish this goal.

REFERENCES:

The Pediatric Surgery Library, in the Biomedical Research Building, C-4840 houses excellent sources of information on the care and surgery of children. Including the latest surgical texts, reprint handouts are also available on selected topics. Please feel free to utilize any book in the office. However, all books should remain in the office, unless special arrangements are made.

RESOURCES AVAILABLE

I) Nutritionist: Coleen P. Greecher, M.S., R.D., C.N.S.D., is an assistant professor in the Departments of Surgery and Pediatrics and is a pediatric dietitian. She is available for both formal and informal consultations regarding nutritional care for the neonatal and pediatric patients in the areas of enteral and parenteral nutrition, as well as didactic sessions on the principles of nutrition.

II) Clinical Nurse Specialists/Nurse Practitioners: Janet H. Shields, M.S.N., C.R.N.P., A.P.R.N., B.C., and Lynn Simmons, M.S.N., R.N., C.R.N.P., are members of both the Department of Surgery and the Department of Nursing. Their area of expertise is in pediatric nursing. They are helpful in preparing children and families for the transition from hospital to home, especially when the child will require technology in the home. They follow children pre-operatively, during the hospitalization, and in follow-up post discharge.

III) Pediatric Trauma Coordinator: Susan Rzucidlo, M.S.N., R.N. Sue works primarily with pediatric trauma patients and is helpful in coordinating the many services involved in their care. She will ensure that rehabilitative services are instituted early, and that there is a smooth transition to home. Sue has a special interest in injury prevention, and is actively involved in community projects on child safety and injury prevention.

IV) Pediatric Trauma Case Manager: Beverly Shirk, B.S.N., R.N., C.C.R.N. also follows the pediatric trauma patients, and coordinates the many services involved in their care. She will help you determine what rehabilitative services are required and will mobilize community resources for safe transitions to home.

V) All residents have access to the Pediatric Surgery office. Therein, you will find the best pediatric surgical library in the Medical Center, including journal articles and previous operative notes/patient charts. Please feel free to use the office as your own.

PEDIATRIC SURGERY RESIDENTS: As in any specialty area, it is our goal that the surgical residents should become aware of the surgical thinking that is used in selecting a given therapy or procedure for a child. In addition, the surgical resident should learn good pre- and postoperative care, as well as how to triage a patient to a tertiary medical facility. Finally, it is anticipated that all surgical residents will improve their technical skills in the operating room.

STUDENTS: During the rotation on this service, the clinical clerks are expected to function as the primary physician for our patients. It is incumbent upon the student to be aware of the patient's disease, the possible modalities of therapy, and the current status of the individual patient.

If you find errors in this manual, or would like to make suggestions for improvements or additions, please complete the form inside the back cover of this manual.

RESPONSIBILITIES

RESIDENTS

A. CHIEF RESIDENT

1. The PGY 4 is our Chief and in charge of the service. "The buck stops..."

2. Every weekday morning, the Chief Resident should contact or meet the attending on call to discuss the patients.

B. RESIDENTS

1. Education of the Medical Students
   a. Discuss the student's history and physical (H&P) to improve their clinical skills.
   b. Aid students in writing daily notes. Student notes should be countersigned, but a separate resident note documenting exam and assessment/plan is mandatory. It is not longer acceptable to write “seen and agree” at the end of a student note.
2. Perform MD history and physical for the record
   a. Document "case seen with..." or "case discussed with..."

3. Daily exam and progress notes on all patients and consults

4. Be prepared to present patients on rounds in the same format as the students are accustomed to using.

5. Residents should follow all consults as long as they are in the hospital.

6. Obtain operative permits and verify preoperative notes and orders, including anesthesia orders. **If you are unsure of the operative plan, discuss the consent with the attending before talking to the family.**

7. Assist in surgery. The resident who scrubs on a case is the primary surgeon. Before entering the OR, read about the procedure and plan what you wish to do.
   a. A resident must be with the patient at all times from the moment that patient enters the operating room. All patients MUST be seen in the preoperative holding area before their surgery for a final check of diagnosis before surgery and marking of the surgical site, if appropriate.

8. **PARENT CALLS:** It is very important to handle all calls and inquiries promptly, courteously, and compassionately. Quite often, the resident staff is the first line of communication between the family of a sick child and the attending staff. How you interact with the family will have a major impact on their perceptions of the hospital, care, etc. Remember, no question is a stupid question!

   It is important to elicit all information and to deal with concerns as appropriate. It is always wise to obtain a telephone number where the parent may be reached. The chief resident and/or attending may be consulted regarding the disposition of any problems. The clinical nurse specialist/nurse practitioner may be able to assist you in follow-up for outpatients. Documenting the telephone interaction is important. Use a Progress Note and turn it in to the Pediatric Surgery office for placement in the child's medical record, or dictate a telephone note for placement in the electronic chart.

9. **OUTSIDE PHYSICIAN CALLS:** Our surgical section is dependent on referrals from outside physicians, and accepts referrals from a wide geographic area. It is important that we maintain a cooperative and friendly relationship with the referring physicians. Most calls from outside physicians are diverted by the hospital operator to the attending physician. If you accept a call from a referring physician, always document the name and phone number of the referring physician so that prompt follow-up can be obtained. All calls regarding patients should be evaluated with the Chief Resident and/or Attending Physician on call. As a courtesy to referring physicians, please have copies of all dictated hospital summaries sent to them.

10. **TRANSFERS:** Ideally, all calls for transfer should be funneled through the **MD Network** (main hospital number – 531-0000). This connects the transferring physician with Pediatric Surgery Attending, along with admissions, so that the transfer (including transport) can be smoothly arranged. We will do everything possible to accept all transfers. Our motto: "You call, we haul, that's all!" In the event that a resident is arranging for the transfer (a rare event), these recommendations should be followed:
    a. Obtain referring physician's name and telephone number, patient's name and location, and probable diagnosis.
    b. Check with nurses in the appropriate unit for the availability of beds, and call the Admissions Office.

c. Before accepting or refusing the patient, contact the attending and the chief resident.
d. All post-operative patients are on the Pediatric Surgical Service unless special arrangements are made beforehand.

11. **OUTPATIENT OFFICE HOURS**

a. Residents and medical students initially see patients and begin the work-up. Patients are then presented to the attending and should include x-rays when appropriate.
b. Minor procedures such as suction rectal biopsies, suture removal, Broviac catheter removal, circumcision, etc. can be performed in the outpatient suites. If there is any doubt about performing such a procedure under local anesthesia, you are better off in the operating room.
c. Always ask the family who their primary care provider is, and who referred them to us, so that appropriate information can be relayed.

12. **DISCHARGES**

ALL DISCHARGE SUMMARIES ARE TO BE DICTATED BY THE HOUSE STAFF AT THE TIME OF DISCHARGE. Narrative summaries MUST be in the form of a narration with copies to all interested physicians, including referring physicians.

13. **MINOR PROCEDURES**

Procedures are performed with the attending physician. Always dictate a note and write a small notation in the chart. Remember to obtain consent from the family, unless an absolute emergency.

14. **SCHEDULING SURGERIES IN THE OUTPATIENT OFFICE**

Surgeries can be scheduled when patients are seen in Outpatient Department for their initial evaluation. Every effort should be made to complete the surgical work-up (H&P, signed consent, any lab work deemed necessary) and to have the child see the Anesthesiologist, if necessary, on that day so that families are not inconvenienced by the need to return on another day prior to the day of surgery. Many of our elective cases are now being performed at HOSC (Hershey Outpatient Surgery Center), which is separate from HMC. HOSC has different H&P forms and consent forms that must be used. The scheduling form is the same.

The Clinical Nurse Specialist and surgery scheduler can assist in scheduling cases and ensuring that the appropriate paperwork is completed prior to the child's leaving the clinic. The scheduler will need the following information to schedule the case:

Child's name
Date of Birth
Patient Number
Type of Admission (Regular, OPA, SDA, OPER)
Surgeon (if their is a preference)
Procedure and Diagnosis
Special Requests in the OR (fluoroscopy, equipment, etc.)
Postoperative Destination (PICU, 7 IMC, 7th Floor)
Position in the OR
How long patient will be in-house
How long will it take surgeon to do procedure

If a case needs to be scheduled on a day other than an Outpatient Office Day, please call the surgery scheduler at x8342 and ask to schedule the case.

Definitions:

Regular Admission: The child is admitted to the hospital at least one day preoperatively. There must be adequate documentation as to the necessity of this day in order for most insurance companies to assume this cost. Acceptable reasons are for bowel preparation in a very young child, I.V. hydration, neurology consults regarding anti-seizure medication, etc.

SDA (Same Day Admission): These patients are brought to the hospital on the morning of surgery, and admitted to the Same Day Unit. After surgery, they will be transferred to one of the inpatient units for their postoperative recuperation. An "SDA Packet" must be completed prior to the day of admission.

OPA (Outpatient Admission): These children are admitted to the Same Day Unit on the morning of surgery. Post-operatively, they recover in this unit, and are discharged to home once they are awake and able to tolerate fluids orally.

OPER (Outpatient Admission with Extended Recovery): These children are admitted to the Same Day Unit. Following surgery, they return to the Same Day Unit where they are observed for an extended period of time. Discharge must take place by 6:55 AM the following day. Reservations must be made for this option, and parents are requested to stay with the child postoperatively (as there is a nurse/patient ratio of 1:6). Apnea and bradycardia, as well as oxygen saturation monitoring, is available in this unit. Children who are likely candidates for this unit are infants who were premature and are at increased risk for apnea and bradycardia post anesthesia (hernia repairs, esophageal dilations, etc.).

Designated Donor Program: The families of children who may require that blood products be available for surgery should be made aware of the designated donor program available through the Blood Bank. There is a $100.00 fee which may or may not be covered by insurance companies, and families will need to acquire at least 2 donors for each unit of blood necessary. The Blood Bank personnel will be happy to explain the program to families, and make arrangements for the completion of the necessary forms as well as the actual donations. The patient must have an order for a type-and-cross, how many units of blood are needed, and the date of the surgery.

WEEKLY SCHEDULE

Monday	0715-	Operating Room
	1600-1700	Pediatric Radiology Conference
Tuesday	0715-	Operating Room
Wednesday	0715-800	Resident Core Lecture (Plastic Surgery Conf. Room)
	0800-	Outpatient Office Hours
	1700-1800	Surgery Morbidity and Mortality Conference
Thursday	0700-0800	Surgical Grand Rounds/Resident Education
	0830-	Operating Room
Friday	0700-	Trauma M&M
		Operating Room to follow
Saturday/Sunday		Rounds

ROUTINES

Admitting procedures:

When an unscheduled admission is called to admitting, have the information listed below ready:

Requested date
Name
Phone
Age
Diagnosis
Estimated length of stay
Brief plan of care
Attending M.D.
Referring M.D.
Referring Hospital
City
Bed request (location)
Insurance information

POSTOPERATIVE INSTRUCTIONS

1. In general, wounds should be kept dry for 5 days postoperatively. Sponge baths can be given during this time. After that period, baths and swimming are permitted. The child may shower then gently dry the wound, once the child is discharged. Dressings can be changed on a p.r.n. basis.

2. Most newborn and infant hernia incisions will be coated with collodion/Dermabond or covered with a Tegaderm dressing. Inform parents that this will harmlessly flake off in 3-5 days. The Tegaderm may be removed in 3-5 days. If there is gauze/Telfa underneath the Tegaderm, it should be removed in 3-5 days.

3. Alert parents to the signs and symptoms of wound infection - fever, redness, spreading away from the incision, pain, drainage, etc.

4. Families should leave the hospital with a prescription for pain medications and the appropriate dose of acetaminophen. In infants and toddlers, Tylenol with Codeine is the drug of choice. School age children and adolescents may prefer Percocet. Ibuprofen in an appropriate dose may be given every 6 hours as an adjunct to pain meds.

5. Some infants and children will experience persistent nausea and vomiting after discharge home. Tincture of time and simple home remedies such as ginger ale, jello, tea, etc. may be helpful. If the child cannot keep anything down after 6-8 hours post procedure, consideration should be given to admission and IV hydration overnight.

6. Post-op hernias, orchiopexy, and laparoscopic procedures have no restrictions on diet, activity, or bathing, so long as the Tegaderm remains in place at least 4 days.

7. The families of infants who have had Plastibell circumcisions should know that the Plastibell will fall off in 5-14 days. Vaseline should be applied to the penis for 48 hours with each diaper change. A convenient sticker with preprinted instruction is available to attach to a business card which can be given to the family. These stickers are available in the Pediatric Surgery office and the clinic.

8. Patients undergoing laparoscopy can return to full activity after two days or when their pain is adequately improved to allow full activity. This includes gym class and sports. This is in contrast to patients who have a laparotomy and, depending on age, may be kept with limited activities for 6 weeks (in older children; it is difficult to limit the activities of younger children).

MEDICATIONS

FORMULARY

All medications are given in pediatric doses, and these are based on the weight of the patient. There should be no guess-work or estimating weights and doses. Pediatric doses are available through the pharmacy intranet website, *http://infonet.hmc.psu.edu/pharmacy/*, then click on "The Formulary", or direct link to *http://www.crlonline.com/crlonline*. Additionally, you can page the pediatric pharmacist with questions. If there are questions regarding recent reports of side effects/toxicities of medications, the Harrell Gateway (http://www.hmc.psu.edu/library/eresources/index.htm) offers a link to Up To Date.

EMERGENCY MEDICATIONS

1. **Cardiac arrest:**

IV = intravenous
ET = endotracheal
IO = intraosseous
IC = intracardiac

Bicarbonate	1-2meq/kg (0.3 x kg x BE) IV,IC,IO (1-2cc/kg)
Epinephrine (1:10,000)	0.01mg/kg (0.1 ml/kg) IV,ET,IO,IC
Lidocaine	1mg/kg (0.05 ml/kg) IV,ET,IO,IC
Calcium Chloride 10%	0.2 ml/kg IV,IC
Atropine	0.02 ml/kg IV,ET,IO,IC,IM

2. **Cardiac Drips**:

Dopamine/Dobutamine	15 mg/kg in a 250 ml bag (1 ml/hr = 1 mcg/kg/min)
Epinephrine	1.5 mg/kg in a 250 ml bag
Isoproterenol	(1 ml/hr = 0.1 mcg/kg/min)
Norepinephrine	range: 0.1 - 1 mcg/kg/min
Lidocaine	30 mg/kg in a 250 ml bag (10 ml/hr = 20 mcg/kg/min) range: 20 - 50 mcg/kg/min

Nitroprusside	0.5 - 10 mcg/kg/min (max dose 70 mg/kg) (follow sodium thiocyanate levels)
Nitroglycerin	0.5 - 4.0 mcg/kg/min

CARDIOVERSION

Atrial arrhythmias (SVT)	0.5 - 1 joule/kg adult: 20 joules
Ventricular tach.	1 - 2 joules/kg
Ventricular fib	2 joules/kg (unsync)

PEDIATRIC DOSAGES (mg/kg)	
STEROID	
Airway Edema:	
Dexamethasone	0.5-2 mg/kg/day ÷ q 6h po/IV (Continue 4-6 doses after extubation) Croup: 0.6 mg/kg IM
Stress Dosing:	
Hydrocortisone (Solu-Cortef)	25-50 mg/m^2/day
Cerebral Edema:	
Dexamethasone	Loading dose: 1-2 mg/kg/dose Maintenance: 1-1.5 mg/kg/day ÷ q 6h
Immunosuppressive/ Anti-inflammatory:	
Cortisone	po 2.5-10 mg/kg/day ÷ q 8h
Hydrocortisone	po 2.5-10 mg/kg/day ÷ q 8h IV 1-5 mg/kg/day ÷ q 12h
Prednisone	0.5-2.0 mg/kg/day ÷ qd/bid

DRUG	GLUCOCORTICOID ANTIINFLAMMATORY	MINERALOCORTICOID
Cortisone	100 mg	100 mg
Hydrocortisone	80 mg	80 mg
Prednisone	20 mg	100 mg
Prednisolone	20 mg	100 mg
Methylprednisolone	16 mg	No effect
Triamcinolone	16 mg	No effect
9-" Fluorocortisol	5 mg	0.2 mg
Dexamethasone	2 mg	No effect
The above doses give approximately equivalent clinical effects		

MSHMC PEDIATRIC STANDARD DOSING PROGRAM

Pediatric standard dosing is a method in which approved standard doses are used in lieu of exact mg/kg/doses. Instead of administering 35mg of acetaminophen to a 3.5kg child, the standard dose would be 40mg. The standard dose option is available through Powerchart Order entry. This program was approved by the P and T Committee, and implemented in our Pharmacy in the hopes of providing increased medication safety for the patient, decreased opportunity for error due to fewer manipulations, and ease of use for nursing and pharmacy staff. Using this program the hospital has seen reduced product waste, as we do not discard partial amounts of drugs. Unadministered doses do not need to be destroyed, as they can be used for other patients, as the dose itself is no longer patient specific. No difference in treatment outcome should be seen when standard doses of wide therapeutic range medications are used. Standard dosing is used in many pediatric institutions around the country.

The medications for the standard dose formulary were chosen because they are commonly used medications in pediatrics. Medications with a narrow therapeutic range (digoxin, phenytoin, etc.) were purposely left off of the list and will continue to be dispensed dosed on an mg/kg/basis. Chemotherapeutic agents and anti-retroviral agents were also excluded from the list.

The chart of pediatric standard doses can be found in our drug formulary. The formulary can be accessed from the Pharmacy web site, or by clicking on the Departments and Support Services tab on the home page of the Info net. The chart of the medications and standard doses can be found by initiating a search for "pediatric standard doses".

As a prescriber, you should write an order as usual, but round to a standard dose that is still within the dosing range. Continue to write the "mg/kg/dose" or "mg/kg/day" in parentheses following the order. If you write an order that is not a standard dose, either the nurse or pharmacist will contact you to convert to a standard dose.

Antiemetic Guidelines for Prevention of Postoperative Nausea and Vomiting

Ondansetron is indicated for patients at high risk, alternative agents (droperidol, metoclopramide, prochlorperazine, or promethazine) should be considered for other patients.

Adults: Ondansetron 4 mg IV
Pediatrics: Ondansetron 0.1 mg/kg IV

BOWEL PREP

Clear liquid diet for 24-48 hours prior to the surgical procedure, depending on age of patient. Infants may only need 12-24 hours of Pedialyte before their procedure.

Metronidazole (Flagyl) 7.5 mg/kg/dose po at 2pm, 10pm and 11pm the day before surgery.

GoLYTELY (Colyte) 25-40 ml/kg po, NG - discuss need with attending

- Discuss the need for enemas/irrigations with the attending physician.

Pediatric Radiology - Osmolarity of Contrast Agents

Blood approx 284 mOsm

Non-Ionic Water Soluble

Optiray	160	355mOsm	Omnipaque	180	408mOsm
	240	502		240	520
	300	651		350	844
	320	702			
	350	792			

Ionic Contrast

Hypaque	30 %	633
Hypaque	60%	1415
Conray	30%	600

Gastrografin (not ever for IV use) >1500
Gastrovue (not ever for IV use) >1500

1. Do not use any hypertonic solution in the upper GI tract of an infant or small child. Aspiration may prove fatal.

2. Do not use extremely hypertonic solutions in the lower GI tract of dehydrated infants or those infants or small children with potential Hirschsprung's disease (retention).

3. Optiray 160 is always safe and is the contrast of choice in the upper GI tract of an infant when barium cannot be used for fear of peritoneal or pleural spill.

ELECTROLYTE ABNORMALITIES

REPLACEMENT:

HYPOPHOSPHATEMIA K Phos, Na Phos PO/IV
Acute: 5-10 mg/kg/dose over 6h
Maintenance: IV: 15-45 mg/kg/day
PO: 30-90 mg/kg/day ÷ TID-QID

HYPOGLYCEMIA Children: 1-2 mL/kg D25 IV over 5 min
Neonates: 2ml/kg D10W bolus, then 6-8mg/kg/min drip

HYPERGLYCEMIA Regular insulin 0.1 unit/kg, then 0.1 unit/kg/hr drip

HYPOKALEMIA Oral: 1-4 mEq/kg/24hr ÷ BID-QID
IV: KCL 0.5-1 mEq/kg over 1-2 hr

HYPERKALEMIA Δ ECG: 10% Calcium gluconate 1 mL/kg over 3-5 min

D25W 2ml/kg over 30 min withInsulin 0.1U/kg

$NaHCO_3$ 1-2 mEq/kg IV over 5-10 min

No Δ ECG: Kayexalate 1-2 gm/kg PO/PR (with 20% sorbitol) Q6hr

HYPOMAGNESEMIA IV: $MgSO_4$ 25-50 mg/kg/dose Q6h (max single dose: 2 gm)
PO: $MgSO_4$ 100-200 mg/kg/dose QID
Maintenance: $MgSO_4$ 30-60 mg/kg/day IV
MgOxide 5-10 mg/kg/24hr ÷TID-QID PO

HYPOCALCEMIA CaGluconate 200-500 mg/kg/24hr IV or PO ÷Q6hr

NUTRITION AND METABOLISM

A Nutrition Assessment Consult should be part of all pediatric surgery admission orders, with the exception of simple single organ problems (i.e. uncomplicated appendicitis, hernias, etc.). This assessment will provide guidance on calorie, protein, and specific nutrient requirements, as well as optimal mode of administration (enteral, parenteral, or both).

ALL history and physical exams should include admission height and weight measurements, as well as percentile group for age (growth chart). Feeding regimens at home, including type of formula and schedule, must be documented.

Baseline Requirements

I) Fluid requirements

1-10 kg	100 ml/kg/d
10-20 kg	1000 ml + 50 ml/kg/d
> 20 kg	1500 ml + 20 ml/kg/d

II) Electrolyte requirements

Na	2-4 mEq/kg/d
K	2-3 mEq/kg/d
Cl	3-4 mEq/kg/d
Ca	1-3 mEq/kg/d
P	0.5-1 mMol/kg/d
Mg	0.5-1 mEq/kg/d

III) Caloric requirements

Based on an estimation of Basal Metabolic Rate (BMR)

Basal Metabolic Rate (weight in kg)

age range	females	males
0-3 yrs	(61 x wt) - 51	(60.9 x wt) - 54
3-10 yrs	(22.5 x wt) + 499	(22.7 x wt) + 495
10-18 yrs	(12.2 x wt) + 746	(17.5 x wt) + 651
18-30 yrs	(14.7 x wt) + 496	(15.3 x wt) + 679

Activity Factor

non-ambulatory = 10% BMR
ambulatory = 20% BMR

Growth Factor

1 - 3 months of age = 30% BMR

Stress Factor

Burns	70-120% BMR
Infection	30-50% BMR
Trauma	50-70% BMR
Cardiac failure	10-20% BMR
Post-op	0-5% BMR
Fever	12% BMR increase for each degree > 37^{o}C

Total Caloric Requirement = BMR + Activity + Growth + Stress

IV) Protein Requirements

age	gm/kg/day
premature + term infants	2.0 - 2.5
older infants	2.5 - 3.0
children	1.5 - 2.5
adolescents	1.0 - 1.5

PEDIATRIC INTRAVENOUS NUTRITION (PIN):

For patients 14 years and older, use the Adult TPN order form. For Pediatric surgery patients <13 years of age, use the pediatric order form.

The following guidelines are for filling out the Pediatric order form:

1. Calculate maintenance fluid requirements:

1-10 kg	100 ml/kg/d;
10-20 kg	1000 ml + 50 ml/kg/d
>20 kg	1500 ml + 20 ml/kg/d

This is typically a safe volume to start PIN.

This will provide minimal calories and will need to be advanced as the baby/child demonstrates tolerance to TPN.

2. Determine the volume of fat needed. Initial dose is 1 gm/kg/d. Multiply grams of fat desired x weight in kg x5. This will result in ml of fat to enter on the order sheet. Increase fat infusion using 1 gm/kg/d increments to a maximum of 3 grams/kg/day. 20% intralipid is the fat source of choice. Total fat should not exceed 40% of non-protein calories. Lipids should be infused over <24 hours to allow serum clearance of fat.

3. Subtract the ml of fat you ordered from the calculated fluid requirement. The adjusted volume is the fluid available for dextrose and amino acids. The rate of administration can be calculated by dividing the total volume by 24 hours.
The percentage of dextrose ordered is dependent on the route of administration and the % of dextrose previously tolerated. Dextrose is usually ordered as 5%, 10%, 12.5%, 15%, 17.5% or 20%. The maximum concentration recommended for a peripheral line is 12.5%. If central access is available, start the dextrose concentration at the % above the percentage last tolerated (i.e., if 12.5% was tolerated, start the PIN at 15%). The concentration of dextrose can be manipulated to increase or decrease calories. Concentration should be advanced after checking urine sugars and/or blood glucose.

4. Full term babies and older children can safely receive 1 gm/kg/day of protein on day one of TPN. Advance the daily amino acid dose to a maximum of 2 gm/kg/d. Please consult with the clinical dietitian for protein orders greater than 2 gm/kg/d. Use the equation gm/kg/d protein desired x weight (kg) x100 divided by total volume of dextrose fluid (ml) to figure out the percentage of amino acids to order. Protein is not included in calculation of calories.

5. Standard electrolytes should be ordered whenever possible. Remember that they are ordered via 100 ml volume. Therefore, if mEq/kg is desired, this amount must be calculated on the basis of total volume. The following electrolyte guidelines are recommended:

Na	2-4mEq/kg/d	K	2-3mEq/kg/d
Cl	3-4mEq/kg/d	Ca	1-3mEq/kg/d
P	0.5-1.5Mmol/kg/d	Mg	0.25-0.5mEq/kg/d

6. Multivitamins and trace elements will be added to the PIN solution automatically in the Pharmacy. The dose is calculated according to the weight of the baby/child. If a patient has cholestasis, it is recommended that “Special Neonatal Trace Elements” or SNTE be ordered.

This solution is made without copper and manganese, both of which are potentially toxic to the liver if bile flow is not optimal.

7. It is recommended that a minimum of 100 units of heparin be ordered if the PIN is to be infused through a central line.

8. If a patient receiving PIN is also NPO, consider the addition of Zantac or other H2 blocker to the PIN solution to control gastric hyperacidity (dose is 4-5 mg/kg/day).

9. How to calculate PIN values for patient list:

A. Weight in kilograms

B. Total fluid from PIN and IL in ml/kg $\frac{\text{Total Volume (in ml)}}{\text{Weight of Child (in kg)}}$

C. Non-protein calories/kg (see 1 and 2 below)

1) Calories from Dextrose = 3.4 kcal/gm

% Dextrose = gm Dextrose/100mL

Convert $\frac{\text{gm Dextrose}}{100 \text{ ml}}$ = x volume ml of PIN

x = total grams of Dextrose

x (3.4) = calories from dextrose

2) Lipid

Multiply mL of IL ordered x 2 = calories from IL

Add the calories from Dextrose + lipid (1 and 2) and divide by weight

D. Calories from carbohydrate = calculated in 1 above

E. Grams of fat/kg - $\frac{\text{mL of Intralipid}}{5}$ = gm of lipid $\frac{\text{grams of lipid}}{\text{wt in kg}}$

F. Grams of protein/kg

?

?gm aa / 100 ml x total volume of PIN = $\frac{\text{gm aa}}{\text{wt in kg}}$

ORAL/ENTERAL NUTRITION

Infants: Oral feeding is always the preferred method of feeding infants, using breast milk (full strength) or a standard lactose-based infant formula. Human milk and standard formulas contain 0.67 kcal/ml or 20 cal/oz. Formulas can be diluted or concentrated to fit the growth needs of individual patients. Concentrating formulas increases osmolality, and if done abruptly, can result in increased stool volume and frequency. A list of available formulas is contained in Table 1.

FEEDING TIPS:

Never advance volume and concentration simultaneously.

Use the GI tract whenever possible. Trophic feeds (non-nutritive low volume feeds) have been shown to stimulate normal GI function, play a role in preventing bacterial translocation from the gut to other sites in the body, and help prevent cholestasis in children maintained on lengthy courses of TPN.

Combination oral, enteral/PIN nutrition plans can be developed to maximize GI function. Nasogastric or transpyloric tubes can be used for short term enteral support. Pediatric tubes tend to be small in diameter, therefore, consistency and digestibility of the formula should be considered. The need for long term tube feedings usually requires the insertion of a gastrostomy tube.

Gastrostomy feeds can be administered after oral meals, as a fluid and calorie supplement, as a part-time continuous feeding (usually overnight for 8-12 hours at a slow rate) or as several bolus feeds throughout the day. It is most practical for home use to schedule no more than 4-5 bolus feedings per day for older children and to use a q 3 hour or q 4 hour feeding schedule for infants. A recommended feeding progression for primary gastrostomy tube feedings is as follows:

On the morning after G-tube placement, clamp the G-tube for 4 hours and check a residual. If the residual is less than 15ml, start feeds through G-tube (if patient takes PO and will only use the G-tube for supplemental feeding at night, start ad lib PO feeds):

Most patients will already be on pre-op tube feeds, and the goal will be to resume the pre-op regimen.

Infants: Pedialyte 30 ml bolus every 3 hours x1; if tolerated, advance to 30 ml regular formula or full strength breast milk every 3 hours x1; if tolerated, advance to 45 ml every 3 hours x2; then 60 ml every 3 hours. This system will provide maintenance fluids in the first 24 hours for a term baby of 3.5 kg. Not every baby will tolerate this advancement. If a baby develops vomiting, abdominal distension or diarrhea, drop back to the step last tolerated. Do not stop IV fluids until the baby is tolerating maintenance fluids via mouth or G-tube. When PO fluids are tolerated at maintenance rates, advance feeding to ad lib to meet calorie goals.

Toddlers: Pedialyte 50 ml bolus q 4 hours x1. Advance to 50 ml toddler formula (Nutren Jr. is often used but is not mandatory) bolus q 4 hours x2. Advance 30-50 ml per feed q 4 hours, depending on size and tolerance of the child, as well as on the child's ability to take PO liquids and foods.

Children: Initiate Pedialyte 75 ml bolus q 4 hours. Advance to Nutren Jr. or Pediasure at the same rate and sequence. Advance volume a minimum of 50 ml per feed, depending on size and tolerance of the child, as well as on the ability to take PO liquids and foods.

Adolescents: Initiate Pedialyte 100 ml bolus. Advance as children.

If continuous gastrostomy feeds are needed, initiate:

Infants: 10 ml/hr
Children 1-5 years: 20 ml/hr
Children >5 years: 30 ml/hr
Advance rate every 4 hours as tolerated to goal rate. May continue advancement after discharge to volume needed to support weight gain.

Table 1

Patient Group	Formula Description	Formulas
< 34 weeks GA	Premature infant formula	Premature Enfamil
Healthy term infant	60:40 whey/casein ratio	Similac Enfamil
Primary or secondary intolerance, casein sensitive	lactose lactose-free soy protein	ProSobee Isomil
Renal dysfunction	low electrolyte	PM 60/40
Steatorrhea, bile acid deficiency, ileal resection	MCT as primary fat	Pregestimil
Sensitivity to casein or soy, Hypoallergenic	casein hydrolysate	Nutramigen Pregestimil
Malabsorption	hydrolyzed casein with MCT (Pregestimil is sucrose free)	Pregestimil Alimentum

To increase calorie concentration of standard formulas using:

Polycose	to make 24 Kcal/oz - add 2 ml/oz formula to make 27 Kcal/oz - add 4 ml/oz formula
MCT	to make 24 Kcal/oz - add 0.5 ml/oz formula to make 27 Kcal/oz - add 1 ml/oz formula

NEWBORNS - PHYSIOLOGY

PHYSIOLOGY ***- GENERAL PRINCIPLES***

PHYSICAL EXAM

Normal findings in a full-term newborn:
Head circumference – 33-35 cm
if appreciably greater, should suspect increased intracranial pressure
anterior fontanelle - soft and < 5 cm

Liver - palpable at 3 cm

Spleen - tip palpable in 10% of infants

Kidneys - lower pole may be 1-2 cm above umbilicus (right lower than left)

Breasts - engorgement resolves in 2-4 weeks

Prepuce - not retractable until 4-6 months or later

Umbilical artery - single artery may signal presence of a major congenital anomaly

Anus - should be approximately half-way between coccyx and base of scrotum or vaginal orifice.

Hemodynamics:

Respiratory rate: 40-60

Newborns breathe with their diaphragms and are obligate NASAL BREATHERS. Preemies may have apneic episodes up to 10 seconds, which is called periodic breathing. This is normal, but the infant should be watched closely or placed on an apnea monitor. Periodic breathing is uncommon during the first 24 hours of life and may be pathologic.

Heart rate: 120-140 beats/min

Preemies rate: 130-150
Blood Pressure: full-term infants 60-80 systolic
45-60 diastolic

premature infants 40-60 systolic

Temperature Regulation:

COLD STRESS: Use warming lamps for all neonates when out of isolette for examinations, procedures, etc. Newborn responds to cold stress by increasing metabolic rate. If protracted, this leads to anaerobic breakdown of glycogen, depletion of energy stores, production of lactic acid, and progressive metabolic acidosis. Thermogenesis also impaired by anoxia when PaO_2 drops to 40 mm Hg or less.

Skin temperature should be at least 97.5^{o} F. Rectal temperature may be misleading and can be normal in presence of cold stress. Rectal temperature becomes abnormal after increased metabolic activity can no longer maintain normal core temperature.

REMEMBER, the major source of heat loss is from the skin, and a full-term infant has approximately 5% of an adult's body weight but 15% of an adult's surface area.

Gestational Age and Risk Factors:

Premature infant: 37 weeks GA or less
-most less than 2500 gms weight

Dangers: hypoglycemia, respiratory distress (RDS), apnea, hypothermia, hyper-bilirubinemia, hypocalcemia, intracranial hemorrhage, sepsis, retrolental fibroplasia

Full-term infant: 38 weeks to 41 weeks GA

Postmature infant: 42 weeks GA or later

Dangers: Increased perinatal mortality due to marginal or depressed intra-uterine fetal oxygenation

Small for gestational age (SGA): < 10th percentile weight (most born at or close to term)

Dangers: hypoglycemia, perinatal asphyxia, respiratory distress (meconium aspiration, pneumothorax, pulmonary hemorrhage), hypothermia, polycythemia cerebral edema, intrauterine infections, hyperbilirubinemia, hypocalcemia

Large for gestational age (LGA): >90th percentile

Low Birth weight Infants: < 2500 gms

High birth weight Infants: ≥4000 gms
- high incidence of diabetic mothers

Dangers: birth injury, congenital anomalies, hypoglycemia, hypocalcemia

Respiration:

Normal Values	Respiratory Failure
pH 7.35-7.45	< 7.25-7.20
PaO_2 60-90 (room air)	< 50 (on 40% O_2)
$PaCO_2$ 35-40	> 45 or increasing

Newborn respiratory status is satisfactory IF:
PaO_2 > 60 mm Hg on room air

No respiratory assistance needed IF:
PaO_2 > 50 mm Hg on 40-50% oxygen without an increasing $PaCO_2$

Respiratory assistance needed IF:
pH < 7.25
PaO_2 < 50 torr despite supplemental O_2
$PaCO_2$ increasing into 65 torr range

Management principles:
Isolette: O_2 conc < 30%, O_2 mist
Nasal Prongs: O_2 requirements 25-30%
Head Box: O_2 requirements 25-35%
CPAP: O_2 requirements > 40%
Mechanical ventilation

Mechanical Ventilation
All infants undergo time cycled-pressure limited ventilation because of the small tidal volumes involved.

Standard ventilator settings:

Peak inspiratory pressure:	16-20
End expiratory pressure:	0-4
PEEP:	3-15
Flow rate:	6-10 L/min
Rate:	20-40
FiO_2:	0.23-1.0

Weaning from Respirator:
- Reduce O_2 concentration first unless pressures are excessively high
- Consider reducing rate into 20-40 range before reducing pressures

INFECTION

Symptoms are often vague and non-specific, and may be opposite the expected adult response: **Apnea** or tachypnea, **bradycardia** or tachycardia, hypotension, lethargy, poor suck, refusal or intolerance of feedings, **hypothermia or temperature instability**, gastric retention, abdominal distention, jaundice, cyanosis. Antibiotics are usually started on the suspicion of an infection instead of waiting for results of cultures.

Lab findings:

WBC (normal) 15,000-20,000 at birth with neuts.
sepsis birth to 1 week < 4,000 or > 25,000
1 week < 4,000 or > 15,000
2 months < 4,000 or > 10,000

CSF

> 10 total WBC's or > 3 polys in meningitis
glucose 1/2 blood level
increased protein (normal 15-40)

Meningitis occurs in about 30% of babies with sepsis

Hypoglycemia

Hemolysis

Hyperbilirubinemia

COMMON ORGANISMS

B-hemolytic strep, E. coli, Klebsiella, enterococcus, pseudomonas, proteus, staph aureus

Management:

Pan culture: blood, urine, CSF, trachea
Broad spectrum antibiotics - usually Amp and Gent or ceftriaxone
(unless risk of line sepsis or MRSA infection - Vanc and Gent)

HEMATOLOGY

HCT and Hgb on cord blood normally less than capillary blood
Anemia: Hgb < 13.5; HCT < 45%
Polycythemia: Hgb > 23; HCT > 70%
consider phlebotomy with plasma volume replacement

Physiologic anemia: Hgb 10 gms at 10-12 weeks of age
(7-8 gms at 8 weeks for preemie)

Normal Blood Volume Neonate:

85-95 ml/KG (8-10% of body weight)

Shock from blood loss in a neonate will often be manifested by BRADYCARDIA rather than tachycardia (decreased blood volume leads to hypoxia which leads to bradycardia)

Colloid Administration:

Na poor albumin	1-2 gm/kg/24 hrs
FFP	10-20 ml/kg
Whole blood	10-20 ml/kg
Packed cells	10-20 ml/kg

Coagulation:

DIC: decreased platelets, fibrinogen, factors II, V, VIII
PT and APTT increased

HEMOPHILIA: factor VIII deficiency
APTT increased

CHRISTMAS: factor IX deficiency; accounts for 15% of patients with hemophilia
(IX and VIII deficiencies account for 90% of congenital coagulation disorders)

VON WILLEBRAND'S: platelet count normal but defect in platelet adhesiveness;
factor VIII deficiency varies; APTT variable; bleeding time elevated
Although the majority of infants with congenital deficiencies of the coagulation factors do not bleed during the first weeks of life, all of the congenital defects may manifest themselves with hemorrhage in the newborn period.

Tests:

APTT: abnormal when any factor other than VII is < 30% normal
PT: assays V, VII, X, also II (prothrombin) and fibrinogen
Neither PT nor APTT will detect XIII deficiency

Normal values:
APTT: < 45 secs
Term AGA infant: 55 +/- 10
Preterm: up to 80

PT: 12-14
Preterm: up to 17
May be prolonged in AGA term if baby has not received Vitamin K

Transfusions:

Packed Red Blood Cells

Decisions for transfusion of PRBC are made based on many factors, including hematocrit, vital signs and patient disease. Transfusions should not be ordered without a discussion with the Attending.
Heme-Onc patients and premature infants will often require irradiated and leukocyte-reduced PRBC for transfusion, which must be stated in the order.

- usual volume of transfusion is 10-20 mL/kg.

Fresh frozen plasma (FFP) and Cryoprecipitate

FFP will replace clotting factors II, V, VII, X, XI, and XIII. Cryoprecipitate is prepared from FFP and serves as a source of concentrated Factor VIII, Fibrinogen, and Factor XIII. The most common use for cryoprecipitate is for fibrinogen replacement. Each unit is approximately 10-15 mL in volume.
As with PRBC, transfusion should occur only after discussion with the Attending.
Since FFP and cryoprecipitate are considered "acellular" blood components, irradiation is usually not required. Fresh plasma (i.e., not previously frozen) should be irradiated, if irradiation is indicated.

- usual volume of transfusion is 10-20 mL/kg (for FFP).
- There are formulas for calculating the dosage of cryoprecipitate. Please consult the Blood Bank.

Factor VIII and IX concentrates

Patients with hemophilia A and B are treated with concentrated factor VIII or IX. Serum levels must be monitored for adequacy of treatment. Hematology consultation is mandatory and their recommendations are followed.
Other factor concentrates also require a Hematology consultation in conjunction with a Blood Bank consultation.

Platelet Components

Platelets: A unit of platelets is a concentrate of platelets separated from a single unit of whole blood and suspended in 40-70 ml of plasma , and is stored at room temperature (20-24C). One unit of platelets contains no fewer than 5.5×10^{10} platelets. The dosage in children is 1 unit per 10 kg, or approximately 5 mL/kg (e.g., transfused into an infant with a 50 mL/kg plasma volume, and correcting for 30% retention in the spleen, a "standard" dose of 5 mL/kg of platelets should raise the platelet count by approximately 100,000/uL under ideal circumstances). Because of the plasma present, platelets must be given ABO compatible with the recipients RBC's. One bag of platelets would be expected to raise the count of an 18 kg child by 20,000/µl

Pooled platelets: Six units of platelets are combined (from six different donors), then the plasma is adjusted to a volume of 200-350 ml. Once platelets are mixed, they must be infused within four hours. Once ordered, it takes 1/2 - 1 hour to prepare..

Concentrated platelets: The volume of plasma in a unit of platelets may be reduced a minimum of approximately 15±5 ml/unit. Therefore, a dose of pooled platelets may be reduced to a volume of approximately 60-80 ml.

Platelets, Pheresis: These are platelets that are collected from one individual, at one sitting using a cell separator. The amount of platelets in one unit of pheresed platelets is equal to the amount of six to eight units of platelet concentrates. These must be given ABO compatible with the recipients RBC's. This component is especially useful if HLA matched for patients who are refractory to platelets from unmatched donors, or for limiting donor exposure.

Irradiated Blood Components

Platelets, red blood cells, and granulocytes are irradiated to kill lymphocytes that can potentially cause Transfusion-Associated Graft-versus-Host-Disease (TA-GVHD). Indications are: bone marrow transplant recipient or candidate, congenital cellular immune deficiency, pediatric hem-onc patients (BMT recipients/candidates, Hodgkin's Lymphoma, Fludarabine treated [or other nucleoside analog therapy], intrauterine transfusion and exchange transfusion, transfusion from a blood (biologic) relative. Other possible at-risk patients include: other hematologic malignancies (e.g., acute leukemia, non-Hodgkin's Lymphoma), immunosupressed patients with solid tumors (e.g., neuroblastoma, rhabdomyosarcoma, glioblastoma, immunoblastic sarcoma), immunosuppressed solid organ transplant recipients, and premature infants (<1200 g at birth). Immunocompetent, full-term infants do not routinely require irradiated blood compoents.

Leukocyte-reduced Blood Components

Red blood cells and platelets are leukocyte-reduced by a filter to remove WBC's. This is done primarily to remove CMV virus that may be in WBC's, to prevent HLA sensitization and alloimmunization, and to prevent febrile, non-hemolytic transfusion reactions. TA-GVHD is NOT prevented by leukocyte-reduction alone. Indications are; transplant candidate or recipient (solid organ or BMT), neonate, intrauterine transfusion, exchange transfusion, congenital cellular immune deficiency, pediatric hem-onc patients, HIV patients.

Transfusion Reactions

All transfusions should be closely monitored for possible adverse effects. Fever, rash, respiratory distress, and hypotension can be signs of an adverse reaction. The transfusion should be immediately stopped, and the Blood Bank notified whenever a transfusion reaction is suspected.

Informed Consent

All transfusions require that proper informed consent be obtained from the patient or guardian, and documented in the medical record.

HYPERBILIRUBINEMIA

Physiologic jaundice

Well baby, full-term. Peaks day 3-4 and is gone by day 7 (appears after 24 hrs).

Pathologic

- appearance within first 24 hours of life
- greater than 8 mg% within 48 hrs of age
- greater than 12 mg% at any time
- persists longer than 10 days

Causes of Indirect Hyperbilirubinemia

Breast milk jaundice, hemolytic disease, hypothyroidism, pyloric stenosis, Crigler-Najjar syndrome, Gilbert syndrome.

Causes of Direct Hyperbilirubinemia

Hepatitis of infectious origin (viral origin, rubella, echovirus, coxsackie, herpes, toxoplasmosis), biliary obstruction (atresia, bile plug, cyst), hyperalimentation, erythroblastosis fetalis, metabolic diseases (Trisomy E, Zellweger's syndrome, Wilson's disease, Alpha-1-antitrypsin, galactosemia, tyrosinemia, Niemann-Pick disease, Gaucher's disease, paucity of intrahepatic biliary ducts).

Evaluation:

Routine blood work:

total and direct bilirubin
type and Coombs
CBC with reticulocytes
TORCH titers
Metabolic screen, alpha-1 levels

Imaging studies: KUB, ultrasound, HIDA scan

Guidelines for the Management of Hyperbilirubinemia

RECOMMENDED MAXIMAL TOTAL SERUM BILIRUBIN CONCENTRATIONS (MG/DL)		
BIRTH WEIGHT CATEGORY (GM)	**UNCOMPLICATED COURSE**	**COMPLICATED COURSE**
Less than 1250	13	10
1250 - 1499	15	13
1500 - 1999	17	15
2000 - 2499	18	17
2500 and up	20	18

Direct-reacting bilirubin concentrations are not subtracted unless they amount to more than 50% of the total serum bilirubin concentration. This table is applicable during the first 28 days of life.

Equivalent gestational age categories may be used in lieu of birth weight for small for gestational age (SGA) infants.

Complications include perinatal asphyxia and acidosis, postnatal hypoxia and acidosis, significant and persistent hypothermia, hypoalbuminemia, meningitis, and other significant infections, hemolysis, hypoglycemia, and signs of clinical or CNS deterioration.

NEONATAL FLUIDS AND ELECTROLYTES

General calculations:

In general, fluid requirements for premature and full term infants are calculated in the following manner:

DAY #1: 80 ml/kg/day of D10W

DAY #2: 100 ml/kg/day
at 24 hours of life, check electrolytes and Ca and add electrolytes to IV fluids.

Na 3 mEq/kg/day
K 1-2 mEq/kg/day
Ca 2 mEq/kg/day

DAY #3: 120 ml/kg/day
Additional guidelines:
Check lytes and Ca at 12 hours of life in unstable premature infants, asphyxiated infants, septic full-term infants and infants of diabetic mothers. Calcium may need to be added to IV at this time.

In babies who are not expected to take PO for some time (e.g. gastroschisis), TPN may be started on the first day of life.

In very low birth weight infants (< 750 gms), initial fluid requirements tend to exceed the usual 80 ml/kg/day on day #1 due to the increased insensible losses via the skin. These infants require 120-150 ml/kg/day initially with careful assessment of hydration status - urine output, specific gravity, serum and urine osmoles, fontanelle size.

Increase fluids 20% when infants are under phototherapy.

Infants with abdominal surgery or with NEC have third space losses, fluids should be increased to 150 ml/kg/day.

Magnesium sulfate is often given during labor; check infant's Mg level. Feedings should not be started until Mg level is less than 3.0 and/or infant has passed meconium.

Electrolyte Abnormalities:

Hyponatremia (salt loss)

(conc. desired (140) - actual conc.) x 0.6 x kg = mEq Na required
(use 0.7 for infants and children < 3 years) Replace over 48-72 hours.

Metabolic acidosis

For pH < 7.25 or less due to metabolic acidosis:
bicarb dose = base deficit x 0.3 x kg

Hypoglycemia

Must treat whether symptomatic or not
SGA, diabetic mother, low birth weight, polycythemia, toxemic mother, asphyxia, stress
25% glucose: 2-4 ml/kg IV
then: D 15W infusion with gradual reduction to D 10 W over 48 - 72 hours

Hypocalcemia

< 7 mg % (normal 8-10 in full-term)
10% calcium gluconate 1 ml/kg over 5 mins
d/c if HR < 100
avoid extravasation

NEWBORN SURGICAL CONDITIONS

GENERAL PRINCIPLES:

There are four cardinal signs that should alert one to a possible surgical emergency in a newborn:

1) BILE STAINED VOMITING
2) DISTENSION
3) BLOODY STOOL
4) RESPIRATORY DISTRESS

Bile Stained Vomiting

Usually means intestinal obstruction (mechanical or functional) until proven otherwise. It is usually associated with distension. Such distension may be absent with such problems as duodenal atresia, stenosis, annular pancreas, malrotation with midgut volvulus. Bilious vomiting in a newborn must be evaluated for a surgical cause. Usually, this will involve a contrast study.

Distension

May be caused by a number of problems:

- Intestinal obstruction
- Pneumoperitoneum (perforated viscus, severe RDS)
- Pseudocyst (meconium, bowel perforation)
- Ascites (hydrops)(urine, blood, chyle, bile)
- Mass effect
- Most common newborn intra-abdominal mass is due to multicystic kidney or UPJ obstruction
- Duplication, mesenteric cyst
- Hydro- or hematocolpos
- Intestinal inflation (vent support, CPAP)

Bloody Stool

Implies a break in the mucosal lining of the bowel. Suspicious for necrotizing enterocolitis or volvulus.

Respiratory Distress

May indicate:

- Diaphragmatic hernia or eventration
- Pneumothorax
- Pulmonary cyst
- Congenital lobar overinflation (emphysema)
- Congenital cystic adenomatoid malformation (CCAM)
- Congenital heart disease
- Esophageal atresia/TEF
- Laryngo-tracheal abnormality

There are only a few STAT EMERGENCIES where a baby must be operated upon as soon as possible. They are:

- suspected malrotation with volvulus
- pneumoperitoneum (due to perforated viscus)
- gastroschisis or ruptured omphalocele

ESOPHAGEAL ATRESIA

Etiology:

Originates 3-6 weeks fetal development when trachea and lungs develop and then separate from the foregut.

5 types, the main ones are:

Type C - proximal pouch/distal fistula	85%
Type A - pure esophageal atresia	7-10%
Type E - H - type fistula	5%

Associated anomalies:

Up to 30% may have other "midline" anomalies. Cardiac, GI (duodenal atresia, imperforate anus), GU, skeletal (vertebral and peripheral) VACTERL association and its variants.

Symptoms:

Esophageal atresia present: excessive salivation, feeding difficulty from birth with gagging/coughing and regurgitation of feeds
No esophageal atresia (H-type): recurrent aspiration or pneumonias

X-rays:

Replogle tube coiled in proximal pouch
+/- air in distal GI tract
vertebral/spinal abnormalities

Differential Diagnosis:

Laryngotracheal cleft, tracheal agenesis (with esophageal lung)

Management:

Pre-op:

1) replogle tube in upper pouch with constant suction
2) head up or prone
3) IV hydration
4) antibiotics
5) CXR with downward pressure on replogle tube
6) cardiac ECHO (determine aortic arch side and anomalies)
7) renal ultrasound (associated anomalies)
8) spinal cord ultrasound (evaluate for tethered cord, can be done post-op)

Surgery:

If infant stable

1) gastrostomy tube (12, 14, or 16 Fr Malecot) if necessary (rare)
2) R thoracotomy (or side opposite arch)
3) retropleural dissection
4) chest tube drainage of retropleural space

If infant unstable or any of the following:

- extreme prematurity
- extreme low birth weight
- associated life threatening anomalies
- sepsis, meconium, RDS

1) gastrostomy (under local if necessary) – consider needle decompression of stomach if acute respiratory compromise due to distension
2) consider fistula ligation without repair of esophageal atresia
3) treat underlying illness

Post-op:

1) extubate when weanable (usually 24-48 hours) or in the OR
2) chest PT and ET suction q 2 h
3) TPN if no trans-anastamotic feeding tube
4) NPO until Day 7 (if trans-anastamotic feeding tube ok to start feeds POD1)
5) Esophagram at day 7-10
6) if NO LEAK, remove retropleural tube, start oral feeds - 5 ml q feed (breast milk or 1/2 strength formula), advance 3-5 ml's every other feeding
   - if LEAK, continue NPO, antibiotics, and drainage until POD #14, repeat study
7) clamp GT for each feeding, release to "burp" after each feeding x 1 month
8) Discharge:
   - clinic appointment 2 weeks post discharge, then every month until 6 months
   - if no problems feeding, clamp GT continuously p 2 months
   - postoperative esophageal study if symptomatic
   - consider removing GT at 6 months if no problems with feeding or study

REMEMBER: 80-100% of infants with esophageal atresia have dysmotility problems and gastroesophageal reflux

20-50% will require an antireflux procedure

50-80% will require esophageal dilatation for anastomotic stricture

LONG GAP ESOPHAGEAL ATRESIA

Children with long gap esophageal atresia (usually Type A or pure esophageal atresia without fistula) may be allowed to grow for several weeks prior to surgical repair. The upper esophageal pouch can be decompressed with a Replogle tube. A surgical gastrostomy will allow for enteral findings until surgery can be performed.

Periodic radiologic studies can be performed to identify the proximity of the esophageal pouches. These “gap-o-grams” are done in the fluoroscopy suite. The resident will need the esophageal bougie used for upper pouch stretching and the Bake’s Dilators from the OR When the esophageal pouches are only 1-2 vertebral bodies apart, surgical repair is considered.

For patients with a long-gap that does not shorten adequately, options include a repair with the Foker technique (sutures through each end of esophagus with 1-2 weeks of progressive stretching of esophagus before repair), or cervical esophagostomy and later esophageal replacement.

GASTROINTESTINAL CONDITIONS

COMMON CAUSES OF NEWBORN BOWEL OBSTRUCTION:

Mechanical:

- Intestinal atresia or stenosis
  - pyloric, duodenal, ileal, jejunal, colon
- Malrotation with midgut volvulus
- Meconium ileus (cystic fibrosis)
- Meconium plug
- Small left colon syndrome (diabetic mothers)
- Segmental volvulus (without malrotation)
- Meconium peritonitis (with perforation, adhesions, and pseudocyst formation)
- Incarcerated hernia (inguinal, diaphragmatic, internal)
- Necrotizing enterocolitis
- Duplication
- Anorectal malformation

Functional:

- Hirschsprung's disease (aganglionosis)(behaves like a mechanical obstruction)
- Paralytic ileus (usually secondary to sepsis)
- Shock
- Peritonitis
- Maternal drugs - magnesium, Valium, heroin
- Electrolyte - hypokalemia, hypermagnesemia, uremia

RADIOGRAPHS IN NEWBORNS

General:
When evaluating a newborn for a possible intestinal problem, all infants should have a flat plate of the full abdomen including chest (i.e. a "babygram"), as well as a left lateral decubitus film (liver side up)to evaluate for free air. In the newborn, it is impossible to determine the position of the colon without a contrast study.

Radiographs to determine etiology of obstruction:

- Plain x-rays
  - High obstruction - "double bubble" sign
    - duodenal atresia vs. malrotation/volvulus
  - Low obstruction - multiple dilated loops
    - jejunal, ileal, or colonic lesions
- UGI - malrotation
- Barium enema (with a bowel obstruction), and limited UGI
  - High cecum: malrotation, volvulus
  - Microcolon: small bowel atresia, meconium ileus
    - long segment Hirschsprung's
  - Normal colon: sepsis, small bowel stenosis
    - NEC, segmental volvulus
  - Dilated colon: Hirschsprung's disease, meconium
    - plug, meconium ileus

Other possible findings:

- Pneumoperitoneum
- Ascites
- Mass effect
- Pneumatosis

INITIAL MANAGEMENT OF NEWBORN BOWEL OBSTRUCTION:

1) Resuscitate infant
   - correct shock, dehydration, electrolyte and acid base abnormalities
   - intubation and respiratory support if needed
   - keep infant warm
2) Secure IV access
3) Nasogastric tube
4) Vit K
5) Baseline blood studies - lytes, CBC, type and cross bilirubin, glucose, calcium
6) Antibiotics
7) Preoperative x-rays: CXR, AXR
   special studies: Ba enema, abdominal US, UGI as needed
8) Talk to family/op permit

DUODENAL ATRESIA - 50%

Etiology:

Probably due to failure of recanalization (mucosal proliferation with complete obstruction at 5-6 weeks).

Associated anomalies:

Low birth weight - (50%)
Down's syndrome - (30%)
Malrotation - (50%)

Symptoms:

Bilious vomiting, maternal polyhydramnios

X-rays:

"Double bubble" sign, gasless abdomen (rarely may have distal gas due to biliary tract abnormalities

Differential Dx:

Annular pancreas, windsock, malrotation

Management:

Pre-op:
1) NG decompression
2) IV hydration
3) AXR, CXR
   UGI if diagnosis in doubt
4) Cardiac ECHO

Surgery: transverse supraumbilical incision, duodenoduodenostomy, duodenojejunostomy, possible gastrostomy, duodenal plication.

Post-op:
1) NG decompression, replace output
2) NPO until NG output minimal
3) TPN via Broviac catheter if prolonged ileus
4) Start po feeds - 3-5 ml q 3h
   Enfamil, Pregestimil, Similac
5) Advance feedings slowly
6) Karyotype

SMALL BOWEL ATRESIAS:

Etiology:

Vascular occlusion with aseptic necrosis and resorption. Probably due to defect in mesenteric arcade or intrauterine volvulus.

Associated Anomalies:

Low incidence of other anomalies
Low birth weight
Meconium ileus
Multiple atresias

Symptoms:

Maternal polyhydramnios, bilious vomiting, abdominal distension

X-rays:

Multiple dilated loops, air-fluid levels
Ba enema: microcolon

Differential Dx:

See Newborn Obstruction

Management:

Pre-op:
1) IV hydration (fluid bolus)
2) NG decompression
3) Labs
4) Antibiotics
5) CXR, BaE, UGI for suspected stenosis

Surgery:
Transverse supraumbilical incision
Resection with reanastomosis
Tapering enteroplasty
Protective stoma for multiple atresias

Post-op:
1) NG decompression
2) IV hydration - 120-150 ml/kg/day x 24 hrs
3) Antibiotics for 48 hours
4) Consider TPN for prolonged support (will need PICC/Broviac)
5) Begin feedings when bowel function returns. Pedialyte, then breast milk or formula
6) Distal limb study/BaE prior to closure of stomas
7) Close stomas at 6-8 weeks
8) Outpatient visit 2-4 weeks post discharge

MALROTATION AND MIDGUT VOLVULUS

Etiology:

Failure to achieve retroperitoneal fixation and proper orientation of intestines (mid and hind guts) when intestines become intra-coelomic again during week 6 in utero. Cecum may normally reside in the RUQ with proper rotation. Fixation of colon to retroperitoneum and descent of cecum into RLQ takes place postnatally. With failure of rotation, the mesenteric root is narrow with fixation only around superior mesenteric vessels. The duodenum and small intestine lie to the right of the spine, while the cecum and colon usually resides in the LUQ. Ladd's bands are peritoneal attachments extending from the RUQ to the cecum, crossing the duodenum; this results in the characteristic duodenal "kinking" or "coiling" and may result in partial or complete obstruction of the duodenum.

Associated anomalies:

Few anomalies:

- duodenal atresia
- omphalocele/gastroschisis
- diaphragmatic hernia

Differential Dx:

Duodenal atresia, annular pancreas

Symptoms:

Acute: bilious vomiting often with a scaphoid unimpressive abdomen
75% onset symptoms in first week of life (usually first three days), volvulus present in 85%

Chronic: (due to Ladd's bands or intermittent volvulus) cyclic abdominal pain, bilious vomiting, malabsorption

Radiographs:

ABXR: duodenal obstruction - dilated stomach, gasless abdomen or paucity of gas beyond duodenum

UGI (gold standard for diagnosis): absent ligament of Treitz (normal LOT position is left of spine, level or superior to pylorus and posterior to pylorus), coiled spring or reverse "3" sign

BaE - cecum in LUQ (helps confirm, but not diagnostic)

Management:

Pre-op:

1) IV hydration; 120-150 ml/kg per day
2) NG decompression
3) Labs
4) Antibiotics
5) CXR, UGI

Surgery:

Emergency laparotomy when diagnosed or unable to rule out volvulus
RUQ transverse incision
Ladd Procedure

1) Reduction of volvulus/derotation of bowel
2) Division of Ladd's bands
3) Broaden mesentery (divide other adhesions, replace small bowel on right side and colon on left)
4) Appendectomy

Resect gangrenous bowel if present, with ostomy vs. primary anastamosis
Consider second look operation for extensive intestinal ischemia (24-48 hours)

Post-op:

1) IV hydration (150 ml/kg/day x 24 hrs)
2) NG decompression
3) Antibiotics 3 days
4) Begin oral intake when bowel function returns

NECROTIZING ENTEROCOLITIS (NEC)

Etiology:

A highly lethal disease predominantly in premature infants characterized by ischemic necrosis of the GI tract leading to perforation. High incidence of associated neonatal problems: RDS, sepsis, cyanosis, hypothermia, premature rupture of membranes, exchange transfusions, apnea, prior cardiac arrest, PDA. Usually occurs in premature infants after initiation of feeds, and may occur in multiple patients at the same time. Spontaneous intestinal perforation (isolated ileal perforation) is a mild variant of NEC.

There is a high association with indomethacin treatment for PDA. The perforation is in the distal ileum on the antimesenteric border with the remainder of the bowel appearing healthy; there is usually minimal associated peritonitis and the baby is usually not overtly septic.

Associated anomalies:

Prematurity (see above).

Signs/Symptoms:

Bile stained aspirates, blood in stool, abdominal distension, poor feeding, apnea, increasing jaundice, lethargy, temperature instability, bradycardia, glucose intolerance, worsening respiratory status.

Labs:

WBC may be elevated or depressed; thrombocytopenia; acidosis.

X-rays:

Bowel distension, thickened bowel wall, pneumatosis intestinalis, portal vein gas, persistently dilated loops, free air.

Management:

Unless there is evidence of perforation, initial therapy is non-operative

Non-operative:
1) NG decompression
2) IV hydration/parenteral nutrition
3) Broad spectrum antibiotics (amp/gent) for 7-14 days
4) Serial AXR (esp. left lat decub)
5) Serial examinations
6) Begin oral feeds after completing antibiotic therapy

Surgery:

Indications
1) Perforation
2) Persistent obstruction
3) Fixed abdominal mass
4) Fixed loop on serial x-rays
5) Abdominal wall erythema
6) Clinical deterioration

Technique

Peritoneal drainage
1) VLBW infants (usually < 1000g) with evidence of perforated NEC or too unstable for laparotomy
2) done at bedside with local anesthetic through lower quadrant incisions (careful of liver on right side)
3) may be only treatment required, or may develop strictures or fistulas that require surgical repair when patient is larger and stable
4) proceed to laparotomy if patient fails to improve or deteriorates

Laparotomy
1) transverse incision (in ELBW, VLBW infants use infraumbilical incision to avoid liver)
2) limited resection with stoma (resect only clearly necrotic bowel), may require multiple resections with multiple stomas
3) with severe ischemia of majority of bowel, consider second look procedure 24-48 hours later rather than initial resection
4) primary anastamosis possible with minimal peritoneal contamination (rare to do this, and usually only in spontaneous intestinal perforation)

Post-op:
1) NG decompression
2) Antibiotics x 2 weeks, include anaerobic coverage
3) Start small feedings - expressed breast milk (EBM), Pregestimil
4) Consider closing stomas when infant's weight reaches 2-3 kg
5) Pre-op GI contrast studies (distal limb and BaE)

Intestinal strictures develop in 10-30% and are most likely in the colon (though most common after NEC treated non-operatively).

Peritoneal Tap: rarely performed currently (place drains instead), but may use if
1) Portal vein gas with one of the following:

- metabolic acidosis
- persistent thrombocytopenia
- clinical deterioration
- persistent sepsis

2) Clinical deterioration without overt radiographic signs of perforation positive tap brown fluid, bacteria on gram stain

Modified Bell's Staging Criteria for NEC

Stage IA	suspected NEC	ileus, stasis, apnea
Stage IB	suspected NEC	gross hematochezia
Stage IIA	definite NEC	pneumatosis
Stage IIB	definite NEC	metabolic acidosis
Stage IIIA	advanced NEC	DIC, peritonitis
Stage IIIB	advanced NEC	bowel perforation

HIRSCHSPRUNG'S DISEASE

Pathology:

Developmental arrest in migration of ganglion cells from neural crest tissue resulting in an absence of ganglion cells in the intermuscular Auerbach's plexus and in the submucosal Meissner's plexus. Spastic contraction of denervated smooth muscle with proximal obstruction:

rectosigmoid	75%
splenic flex	15%
otal colon	10%

Associated anomalies:

Rare in premature infants

associated anomalies	5-20%
Down's syndrome	5%

Symptoms:

80-90% of babies will show symptoms in the first days of life.
95% normal babies pass meconium within 24 hours and remainder pass meconium within 48 hours of birth.

1) failure to pass meconium
2) abdominal distention with bilious vomiting in newborn period

Exam:

Abdomen often distended. May get “explosion” of stool with rectal exam.

X-rays:

Dilated loops of bowel on AXR

Bariumenema:

Contracted rectum or rectosigmoid with dilated colon proximal to transition zone.
Retention of contrast for > 24 hours

Differential Dx:

Meconium plug, small left colon syndrome, intestinal stenosis or atresia

Management:

All infants with the suspicion of Hirschsprung's Disease will undergo a barium enema and a suction rectal biopsy. If the patient is stable, the work-up is done before irrigations begin. If the patient is septic or ill-appearing, rectal irrigations and IV antibiotics should start immediately with diagnostic work-up performed after stabilization.

Suction Rectal Biopsy: (“gun” in Pediatric Surgery office, procedure performed with attending present)

1) 2 biopsy sites – 2 and 4cm
2) Make sure each biopsy specimen is adequate in size
3) Hold infant in crossed leg position
4) Place cutting capsule facing posterior midline

5) Notify and hand deliver specimens in separate containers on saline soaked Telfa to Pathology Department, as these are fresh specimens (x8246, transfer to the Gross Room)
6) Results should be available in 12-24 hours
7) If this procedure is performed after hours or on the weekend, the gun should be disassembled and soap/water cleaned, taking care not to lose any small parts. The equipment must be sterilized before re-use. Return the instrument to the office and staff will clean and sterilize it.

Saline Rectal Irrigation: 10-15 mL/kg (use 15 if tolerated) q 2 h until clear return
Administer with 12F red rubber catheter.
Massage abdomen when enema is administered.
Catheter should be left in place to assist with drainage of the enema.
Passing fifth finger into rectum may also assist in drainage of the enema.
This should result in significant decompression of the abdomen.
If this does not occur, it is important to notify the Attending Surgeon, since the child may then need a decompressing colostomy.

Intravenous antibiotics are administered to all newborns with lower GI obstruction.
Ampicillin and Gentamicin.

Surgery:
Transanal pull-through as newborn if transition zone in rectosigmoid and patient responds to irrigations.
Decompressing colostomy with second-stage pull-through if longer segment lesion, poor response to irrigations, or perforation. Second-stage pull-through performed once baby is gaining weight and other medical issues resolved (usually 2-6 mo).

Post-op:
1) NG decompression until bowel function returns
2) IV hydration 1.25-1.5x maintainence x 24h then maintenance rate
3) Antibiotics 24-48 hours
4) Sign on bed: **No rectal exams or temperatures**
5) TPN (will need PICC or Broviac)
6) Start feeding (breast milk, formula) when bowel function returns
7) Start anal dilations > 2 weeks post-op
8) Follow-up in clinic 2 weeks post discharge; frequency of appointments depends on dilations

COMPLICATIONS:

Enterocolitis
Can occur in newborn or older child before or after colostomy or definitive procedure. Can be rapidly fatal due to dehydration, shock, and sepsis.

Treatment:
1) Rapid IV hydration
2) Broad spectrum antibiotics
3) Rectal decompression irrigations

Stricture: treated with anal dilations, rarely requires revision of anastamosis

MECONIUM ILEUS

Etiology:

Occurs in 10-15% of patients with cystic fibrosis. Very rarely occurs in the absence of cystic fibrosis pancreatic enzyme deficiency.

Associated anomalies:

50% simple or "uncomplicated"
50% are complicated and associated with volvulus, atresia, meconium peritonitis, meconium pseudocyst.

Symptoms:

Abdominal distension with bilious aspirates, doughy feeling abdomen with palpable loops (especially in RLQ), no meconium in rectum.
Distal ileum filled with thick, tarry meconium down to a terminal ileum filled with clay-like pellets of inspected meconium.

X-rays:

Multiple dilated loops with great variation and coarse granular appearance (“ground glass”), no air fluid levels, calcifications

BarumEnema:

Microcolon

Diagnosis:

Positive sweat test:
Na > 60 mEq (Results dependent on amount of sweat
Cl > 60 mEq accumulated, study may need to be repeated one or more times.)
Genetic testing

Differential Dx:

- See intestinal obstruction
- Hirschsprung's disease, atresias, etc.

Management:

Simple Meconium Ileus

40-60% can be handled non-operatively with enema (Gastrografin) therapy
1) IV hydration (fluid bolus prior to enema)
2) antibiotics
3) NG decompression
4) Gastrografin enema (hyperosmolar)
- monitor urine output and hemodynamics since the hyperosmolar enema can lead to severe hypovolemia.
- must reflux back into dilated loops to be successful repeat in 12-24 hours if necessary

5) N-acetyl-cysteine enema if gastrograffin unsuccessful in providing full evacuation
6) serial AXR's
7) begin oral feedings (Pregestimil) when bowel functions and is decompressed

If N-acetyl-cysteine unsuccessful, consider total colon Hirschsprung's Disease as a possibility.

Surgery: if enema fails, proceed to surgery
1) supraumbilical transverse incision
2) enterotomy with evacuation of meconium and pellets
3) irrigate intestine with 1% N-acetyl cysteine
4) primary closure of enterotomy if bowel healthy or consider ostomy if health of bowel questionable.

Post-op:
1) IV hydration (150 mL/kg/day x 24 hrs)
2) NG decompression
3) broad spectrum antibiotics 3-5 days
4) BID intestinal irrigations with 1% N-acetyl cysteine (given via stoma, enema or NGT)
5) if ostomy placed, plan closure after 6 weeks if patient gaining weight and healthy

Complicated Meconium Ileus

Proceed to surgery as above
- repair volvulus or atresias as indicated
- consider end ileostomy and mucous fistula if size discrepancy marked

MECONIUM ILEUS EQUIVALENT

Usually seen in older child with CF. Secondary to impaction of stool in terminal ileum and right colon. May mimic appendicitis. Usually can palpate doughy loops of intestine.

Treat with N-acetyl cysteine enema, done by radiology. Start MOM or mineral oil or GoLYTELY PO.

MECONIUM PLUG SYNDROME

Etiology:

Unknown, maternal diabetes common; also reported with maternal magnesium administration.

Symptoms:

Abdominal distension, bilious aspirates, failure to pass meconium

Differential Dx:

Long segment Hirschsprung's Disease

X-rays:

Multiple dilated loops of bowel

BariumEnema:

small left colon to splenic flexure with proximal dilatation. Rectal width usually equals splenic flexure width.

Management:

BaE is often curative with passage of large meconium plug.
Consider suction rectal biopsy to prove diagnosis.

ANORECTAL MALFORMATION

Etiology:

Occurs in fifth to seventh week in utero when cloaca forms the urorectal septum and lower urinary tract.

usually full-term infants (males 3:2)
1 in 5000 births

Associated Anomalies:

High incidence of major associated anomalies: 30-60%; vertebral, urologic, cardiac, esophageal atresia, bowel atresias/malrotation.
Urologic anomalies:
70% in high male and female
5-10% in low female
Fistulas: (not a true fistula since it is really the ectopic ending of the bowel)

Symptoms:

Bowel obstruction, failure to pass meconium
meconium in urine

Exam:

Inspect for anal opening or fistula. Fistula may not become apparent for 24 hours, and may be to perineum, vestibule (females) or scrotum (males). Evaluate shape of buttocks

("rocker-bottom" buttocks associated with high lesions). Catch urine to inspect for meconium.

Radiology:

1) AXR - include sacrum, CXR
2) Echocardiogram
3) Renal US
4) Perineal US – if no obvious fistula may determine location of rectal stump
5) Spinal cord US

Management:

Pre-op:

1) NG decompression
2) TPN
3) Pre-op radiographic evaluation (renal and spinal ultrasounds can be post-op)
4) Broad spectrum antibiotics
5) If no opening or obvious fistula, re-examine after 24h to assess if fistula present.

Remember: R/O VACTERL Association

Vertebral anomalies
Anal anomalies
Cardiac anomalies
Tracheoesophageal fistula
Esophageal atresia
Renal anomalies
Limb anomalies (absent radii)

Surgery:

Low lesion: anal proctoplasty (cutback) procedure, minimal PSARP, or progressive dilatations (daily).

High lesion: diverting colostomy followed by delayed Posterior Sagittal Anorectoplasty (PSARP) reconstruction, or primary PSARP as a newborn.

Post-op:

1) NPO until bowel function returns
2) EBM or formula feeds as tolerated
3) antibiotic coverage 3-5 days

Cutback procedure:

- saline wash with diaper change
- **"NO RECTAL TEMPS OR TREATMENTS"** sign at bedside
- size rectum with Hegar dilators at 2 weeks post-op - rectal dilatations start daily at 2 weeks

Colostomy:

- clinic visit at 2 weeks and 6 weeks postoperatively
- schedule distal loop colostogram study ± VCUG to locate fistula
- schedule PSARP at 3-4 months

PSARP:

- **"NO RECTAL TEMPS OR TREATMENTS"** sign at bedside
- antibiotic coverage 24-48 hours
- Foley catheter in for 3-5 days
- clinic visit at 2 weeks
  calibrate anus with dilator that fits snugly. Instruct parents to dilate twice daily
- follow-up outpatient visits every 1-2 weeks until dilatations easily performed at desired size
- close colostomy when dilatations are easy (usually at 8 weeks)

AGE	Hegar Size
1-4 months	12
4-8 months	13
8-12 months	14
1-3 years	15
3-12 years	16
more than 12 years	17

Once the dilator passes become easy and without pain (twice a day), the parents may start tapering the frequency of the dilatations.

- once a day for a month
- every other day for a month
- every third day for a month
- twice a week for a month
- once a week for a month
- once a month for three months

If at any time the process of dilatations becomes difficult, painful, or bloody, it is a specific indication to dilate twice a day and restart the process.

Some children will require a bowel management program to accomplish a daily evaluation of the colon. Refer to the section on constipation.

At the Hershey Medical Center, a parent-to-parent support group exists to link the families of children born with anorectal malformations for the purpose of mutual support and education. The support group, the A.R.M.S. Network (AnoRectal MalformationS) publishes a newsletter.

ABDOMINAL WALL DEFECTS

OMPHALOCELE

Etiology:

Failure of fusion at umbilicus of cephalic, caudal and lateral mesenchymal folds at tenth week of development. Results in three types of defects. Classic omphalocele - midline defect with sac (amnion) covering peritoneum and abdominal contents. Umbilical cord comes off inferiorly.

Associated anomalies:

High incidence of midline defects

Malrotation	100%
Cardiac	30%
Renal	30%

Symptoms:

Amnion covering peritoneum and abdominal contents. "Giant omphalocele" contains most abdominal organs and has an underdeveloped peritoneal cavity (usually involves liver).

Differential Dx:

Umbilical cord hernia, gastroschisis

Management:

Pre-op:
1) IV hydration
2) NG decompression/NPO
3) Vit K
4) KEEP INFANT WARM
5) CXR
   Cardiac Echo
   Renal USG
6) Wrap with Xeroform and coban as a circumferential dressing

Surgery:
1) Small defect (< 5cm) Primary closure
2) Large defect
   - continue Xeroform and coban silo wrap until maximal reduction has taken place (3-6 weeks) then consider closure or delayed abdominal wall reconstruction after epithelialization of defect
   - TPN (will require PICC or Broviac)
3) Ruptured omphalocele – surgical emergency
   - cover intestines with warm, saline sponges.
   - antibiotics
   - primary closure
     or
   - silo construction with Gore-Tex/reinforced silastic, compress silo over 7-10 days until fully reduced, then proceed to OR for closure.

Post-op: 1) NG decompression
2) Consider TPN and Broviac if prolonged ileus
3) Consider possible complications
Respiratory distress due to too tight an abdominal closure with pressure on diaphragm
IVC compression due to tight abdominal wall closure with poor perfusion

GASTROSCHISIS

Etiology:
Believed to result from an *in utero* rupture of a cord hernia late in gestation.

Associated anomalies:
GI (malrotation) 100%
Bowel atresia 16%
Low incidence of other anomalies

Symptoms: abdominal wall defect to the right of the umbilical ring. Ring is intact. Loops of bowel foreshortened and matted together with chronic peritonitis.

Management:
Pre-op: 1) IV hydration - 20ml/kg bolus of NS or LR when IV placed
2) NPO
3) NG decompression
4) Vit K
5) Cover intestines with warm saline soaked pads, place the lower half of the baby in a “bowel bag"
6) KEEP INFANT WARM
7) CXR
8) Antibiotics

Surgery:
1) Silo placement at bedside – preformed silo with expandable ring (requires some narcotic/sedation, may require intubation)
2) Primary closure is less commonly performed now, but possible
3) If atresia is clearly defined and easily mobilized for repair, proceed with anastamosis or stoma (do not perform in thickened, matted bowel). If atresia is noted but not easily accessible, leave it alone and decompress the GI tract. Second look operation in 3-6 weeks to repair the atresia can then be performed.
4) Consider gastrostomy (uncommon)
5) Daily or twice daily reduction of silo until ready for closure

Post-op: 1) NG decompression
2) TPN (will need PICC/Broviac) since ileus can last up to > 1 month. Bowel may then act like a "short gut" with rapid transit time and feeding intolerance

3) Respiratory distress due to too tight an abdominal closure with compression of diaphragm
4) IVC compression due to too tight a closure
5) Antibiotic therapy for 3-7 days.

*If surgery is delayed for several hours, the attending physician will wrap the bowel in kerlex and suspend it over the baby to reduce edema and facilitate reduction.

CONGENITAL DIAPHRAGMATIC HERNIA

Etiology:

Diaphragm forms eighth-tenth week in fetus. Midgut returns to abdomen at this time
males > females
posterolateral (Bochdalek) defect is the most common
Anteromedial (Morgagni) very rare;
left side: 4:1

Associated anomalies:

- Intestinal malrotation - 100%
- Cardiac anomalies - 5-25%
- Pulmonary hypoplasia
- Pulmonary hypertension, severe right to left shunting

Symptoms:

- Severe respiratory distress
- Scaphoid abdomen, maternal polyhydramnios

X-Rays:

Bowel loops in chest, mediastinal shift NG in chest

Differential Diagnosis:

- Cystic adenomatoid malformation
- Pneumothorax, mediastinal tumor (teratoma)

Management:

Pre-op:

1) immediate intubation if respiratory distress (DO NOT AMBU)
2) IV hydration
3) prevent hypothermia
4) pre- and post- ductal O_2 saturation monitors
5) maximize pre-ductal O_2 saturation
6) NG decompression
7) minimize manipulation of baby (pulmonary hypertension worsens with agitation)
8) CXR
9) Cardiac ECHO – assess cardiac anomalies and pulmonary hypertension
10) vasodilator therapy (Nitric Oxide, Viagra)

11) aggressive ventilatory support, including jet ventilator and oscillator
12) ECMO if inadequate oxygenation despite maximal support
13) follow pulmonary compliance

ONCE MEDICALLY STABILIZED AND THE PULMONARY HYPERTENSION HAS RESOLVED, THE INFANT THEN UNDERGOES ELECTIVE OPERATIVE REPAIR OF THE DEFECT (THIS MAY TAKE DAYS TO WEEKS).

Surgery:

1) transabdominal approach through subcostal incision
2) some right side lesions require thoracotomy
3) repair with native tissues if possible, use PTFE if inadequate diaphragmatic rim

Post-op:

1) maximize pre-ductal PO_2. Wean ventilator only to keep pO_2 > 100 torr pre-ductal
2) follow post-op pulmonary compliance

ECMO:

If infant is placed on ECMO prior to medical stabilization, surgical repair of the diaphragmatic defect may be considered while on bypass or after bypass is completed, though it is usually performed after bypass at this institution. The operation is performed in the NICU with all equipment moved up from the OR. If performed during ECMO, a chest tube is placed due to higher risk of post-op bleeding. There should be a low threshold for reoperative exploration if bleeding is a problem unresponsive to heparin and platelet adjustments or Amicar administration.

PEDIATRIC SURGICAL CONDITIONS

HERNIAS AND HYDROCELES

INGUINAL HERNIA

One of the commonest surgical conditions of infants and children. Almost all are indirect and due to persistent patent processus vaginalis.

Incidence: 1% of all children

- 8% present as incarcerations, with 50% of these occurring in infants < 6 months of age.
- 60% right-sided, 25% left, 15% bilateral

Management:
Surgical repair scheduled within reasonable amount of time for family and OR schedule - UNLESS the hernia is troublesome (i.e. always "out" with an irritable infant) or very difficult to manually reduce.

Children with a unilateral inguinal hernia are at higher risk of having a contralateral processus vaginalis still patent, and therefore at increased risk of developing a contralateral inguinal hernia. The risk is most associated with prematurity and the age at which the initial hernia is diagnosed (younger patient = greater risk). There are three options for management of these patients: 1) observation of contralateral side, 2) exploration of contralateral side during initial surgery with repair of hernia if found, and 3) diagnostic laparoscopy (through hernia sac of known hernia) and repair of hernia if patent processes vaginalis identified. Given the pros and cons of surgery/no surgery, we have decided on the following general guidelines:

Children < 6mo: contralateral exploration + hernia repair if present
Children 6mo-10yo: diagnostic laparoscopy + hernia repair if present
Children > 10yo: observation of contralateral side

In girls we tend to explore the contralateral side at older ages since the risks of surgery are less (no vas deferens or testicular vessels to injure)

Incarcerated hernia:

1) Manually reduce (+/- sedation)
2) Admission
3) Allow PO formula/Pedialyte
4) Surgical repair - 24-48 hours
5) If unable to reduce, emergent surgery indicated

complication rate - manual reduction: 1.7%
complication rate - surgery (incarc): 20%

In girls, a firm, incarcerated mass is often an ovary and does not require emergent surgery (if asymptomatic), though should be repaired within 1-2 weeks due to risk of torsion of ovary.

POINTS TO REMEMBER:

1) Always know where the testicle is on the side of the hernia. Some hernia repairs (particularly premature infants) will require some form of orchiopexy. The family needs to know this fact.

2) 0.1% of females with inguinal hernias will have androgen insensitivity syndromes. Highly suspicious if BOTH gonads are palpable in the inguinal canal. Buccal smear preoperatively or identify normal tubes at operation.

HYDROCELES

In infants and children, almost all due to patent processus vaginalis that has sealed off proximally. Most hydroceles present at birth will resolve by 1 year of age (85%). Repair rarely recommended before 1 year of age, unless there is an associated hernia.

UMBILICAL HERNIAS

In general, delay repair until 2 years of age, or longer if defect is becoming smaller The size of the fascial defect is important, not the size of the sac/protrusion.

Fascial defect: < 0.5 cm :spontaneous closure by 2 years
" ": 0.5 - 1.5 cm: spont closure usually by 4 years
" ": >1.5 cm : spont closure rare

Incarceration is rare, but does occur.

<u>Management:</u> (hernias, hydroceles, umbos)

1) schedule OPA
   OPER (extended recovery) if premature infant < 50 weeks gestational age

<u>Post-op:</u>

1) sponge baths until POD #4
2) Tylenol and ibuprofen p.r.n. for pain (can alternate doses Q3hr)
3) follow-up outpatient appointment 2-3 weeks, though if patient is doing well parents may call and cancel follow-up

GASTROINTESTINAL CONDITIONS

GASTROESOPHAGEAL REFLUX DISEASE (GERD)

Pathology:

Common pathophysiologic problem in neurologically impaired children develops due to incompetence of the anti-reflux barrier.

Anatomy

Esophageal diaphragmatic hiatus

- Formed by R crural fibers
- Insertion of phrenoesophageal ligament at angle of HIS (sharp angle of the lower esophagus and gastric fundus)

Lower esophageal sphincter - 1.5-2.5 cm in length

6-8 cm H_2O intra-abdominal pressure

Resting pressure:

2 week infant	4-6 mm Hg
3 months	12-15 mm Hg

Symptoms:

1) Vomiting, feeding difficulties, FTT
2) Respiratory: aspiration, pneumonias, reactive airway disease, asthma, apnea and bradycardia
3) Esophagitis, bleeding, strictures
4) Life threatening event

Work-up may include:

1) Upper GI study (important to exclude gastric outlet obstruction, e.g. malrotation)
2) Gastric emptying scan
3) pH probe (with pneumogram)
4) Esophagoscopy with biopsy
5) Bronchoscopy (lipid laden macrophages)

Differential Diagnosis:

Pyloric stenosis, achalasia, esophageal stricture, malrotation, gastric web, gastric duplication, peptic ulcer disease.

Management:

Almost all patients are treated medically with an anticipated success rate of 90%.

- prokinetic agent (Reglan)
- H_2 blockers (ranitidine) or Omeprazole (Prilosec)
- thickened feedings 1 tsp. rice cereal to each oz. of formula
- positioned feedings (prone with head elevation >30 degrees)

Delayed gastric emptying may contribute to symptoms. Diagnosis is made by history and UGI at this institution, with less use of pH probe studies.

Surgical procedures:

1) Nissen fundoplication (360^{o} wrap), laparoscopic or open
2) Gastrostomy, laparoscopic or open (done in all infants < 1yo)
3) Pyloroplasty, pyloromyectomy, or pyloromyotomy
4) Thal fundoplication (270^{o} wrap)

Belsey Mark IV and Hill posterior gastropexy are rarely done in children.

Surgery is tailored to the child's symptoms and needs and may include any of the following procedures: antireflux procedure, pylorus procedure, and gastrostomy or feeding jejunostomy.

Postop Feeds:

Post-op feeding regimens will be tailored for individual patients, but the overall goal is that gastrostomy tube patients will be discharged on POD 1 and Nissen + G tube patients will be discharged POD 1 or 2. To do this feeds must be started early on POD 1.

Coleen Greecher should be consulted on the day of surgery to plan out a post-op feeding schedule for each patient. The orders for the feeding should be written on the day of surgery so that there is adequate time for the formula to be available on the floor the next day.

G tubes will be placed to gravity drainage immediately post-op. If drainage overnight is <15ml for infants or <30ml for older children, feeding should begin by 8am on POD 1, according to the schedule arranged with Coleen Greecher. In general, residuals are checked before each feed, or every 4 hours for those on continuous feeds, and the feeds are held if >15ml for infants or 30ml for older children.

For patients on a regular diet pre-op who only underwent placement of a G tube, they can start a clear liquid diet with the G tube clamped by 8am POD1 if the above drainage requirements are met, and be advanced to regular diet by noon if clears are tolerated. For patients on a regular diet pre-op who underwent a Nissen, they are advanced only to a soft diet, which they stay on for 1-2 weeks.

Complications:

1) Gas Bloat syndrome
2) Delayed gastric emptying (high residual volumes)
3) Dumping syndrome (following pyloroplasty)

All medications must be known pre-operatively, as well as appropriate serum levels. A plan should be made for the postoperative administration of anti-seizure medications until the child is taking PO or GT feedings.

All gastrostomy feedings must be accompanied by some oral stimulation. Even in the most oropharyngeally compromised child with chronic aspiration, oral stimulation is important for future feeding behavior. Consider an occupational therapy or speech therapy consult.

GASTROSTOMY TUBE PLACEMENT AND PROBLEMS

Consults for gastrostomy tube placement constitutes one of the most common consults for the Pediatric Surgery service. We have moved to placing Mic-Key buttons (skin-level tubes) in the OR, rather than mushroom or Malecot catheters, though the latter are still used occasionally. The first G-tube change is performed 4 weeks after surgery. Replacement of a tube before this must be performed carefully and usually requires a contrast study to confirm placement and absence of leak before use of the new tube; contact the Attending Surgeon for tubes dislodged in this time period. Percutaneous endoscopic gastrostomy (PEG) tubes should not be removed for a minimum of three months post insertion. At that time, a skin level device can safely be inserted.

Gastrostomy Tube Problem Solving:

Leakage At The Site:

The most common cause of leakage at the site is that the balloon or mushroom of the tube is not resting up against the stoma on the inside of the stomach, and the tube moves freely in the tract. The tube should be pulled gently back and secured to the skin. Use of an anti-migration device such as a nipple or other clamping device may help in keeping the tube secure, decreasing the amount of leakage.

The nipple or anti-migration device will also aid in keeping the gastrostomy tube in good position, maintaining the tube perpendicular to the skin, allowing the skin to heal around the tube, and prevent undue pressure on the stoma. If there is no anti-migration device, the tube will move freely in and out of the tract, causing irritation and leakage.

When replacing a gastrostomy tube, always pull the nipple (anti-migration device) over the tip of the catheter prior to inserting the tube. It is easier to do this, than it is to get the nipple on after insertion. Do not cut a hole in the end of the nipple, as this will defeat the purpose of having the nipple “clamp” onto the tube. Instead, using a hemostat, push through the tip of the nipple and gently stretch the hole to accommodate the tube by opening the hemostat. Then pull the tube through the nipple.

Skin Excoriation:

The most common cause of skin breakdown at the site is from the leakage of gastric contents onto the skin. It is easier to prevent this than it is to heal. Skin protection includes adequate daily cleansing and the use of techniques to protect the skin. This can include the use of dry gauze dressings, protective ointments and creams (Balmex, vitamin A and D ointment, zinc oxide ointment, Critic-Aid), or skin barriers (Coloplast, Duoderm, No Sting Skin Barrier). Some like to paint the skin with an antacid like Maalox or Mylanta, or to make a paste from Carafate to buffer the gastric acid that leaks from around the tube, in the hopes that this will prevent skin excoriation.

Once skin breakdown has occurred, it is important to identify if there is bacterial or fungal infection present, or if the excoriation is purely from irritation. If there is a bacterial infection, the skin will be red, warm and have the appearance of a cellulitis. Antibiotics will be necessary. If monilia or fungal infection is present, the characteristic satellite lesions will be noted. These are small red, macular-papular lesions spreading away from

the area of erythema. An anti-fungal ointment or powder should be ordered and applied three to four times per day (nystatin ointment, Mycolog ointment, Mycostatin powder). Stopping the leakage will allow an environment that will promote healing.

Tube Migration:

The gastrostomy tube will readily migrate into the small intestine and cause symptoms of obstruction unless some sort of system is used to prevent this. Parents are taught to use an anti-migration (nipple) device and secure the tube to the skin. They are also instructed to measure the length of the tube from the abdominal wall to the tip daily. If it is normally 12 to 14 inches from the abdominal wall, and today they measure only 9 inches, you know that the tube has migrated.

Other symptoms may include bilious vomiting, yellow bilious drainage from the tube, feeding intolerance with gagging, diarrhea, and abdominal discomfort with feedings.

If the tube in place is a balloon catheter, deflate the balloon and remove the tube. Be sure to put the nipple on the new tube before replacing the tube in the stomach. Check for gastric contents, then secure the tube to the skin. If the tube in place is a mushroom catheter, the tube will need to be removed and replaced with a Foley catheter. Usually 3 to 5 ml of water is instilled into the balloon. Remember, the balloon will take up part of the volume of the stomach, so the volume used will depend on the size of the child. Rarely is greater than 5 ml of water indicated.

Granulation Tissue:

The formation of this tissue is from chronic irritation by the tube. It is very prolific and often seems to blossom overnight. The tissue is moist and bleeds easily. We note that this tissue seems to be more problematic when silicone tubes are used instead of the silastic tubes.

The tissue is treated with the application of silver nitrate on a daily or every other day basis. It often takes several applications before the tissue dries up and flakes away. Parents can be taught to do this at home. Be sure to remind them that the silver nitrate is a chemical and can cause a chemical burn if they get it on their fingers and then touch the eye. Good hand washing is essential.

Tube Blockage:

Medications and nutritional formulas should be flushed through the gastrostomy tube with an ample amount of water following each feed or med instillation. This will cleanse the tube and insure that all of the drug or feed is flushed through the end of the catheter. Blockage occurs when adequate flushing is not accomplished. Precipitates can form, effectively occluding the tube. It is easier to prevent a blockage with adequate and appropriate flushing, than it is to unblock a tube.

The easiest way to handle the problem is to change the tube. If this is not an option, one can attempt to unclog the tube through instillation of a variety of different substances. Flushing with water is the first step. If no progress is made, one can try cranberry or pineapple juice, as these juices are very acidic and may aid in breaking up the clog.

Some recommend the use of soda, as the bubbles are helpful in clog busting. We also recommend making a slurry of meat tenderizer and water and instilling that into the tube.

Using a 1cc syringe, gently instill the solution of choice into the catheter. This will deliver the solution far down into the catheter, where it will hopefully be able to come in contact with the clog, and help to dislodge it. If this fails, a product called “clog zapper” is available in Central Stores (order #12647). Mix the solution per the package insert. It will be amber-colored and frothy. Use the syringe and introducer and instill into the catheter. Allow to remain in the catheter for several hours then flush through. The company states that if just a small amount of solution gets to the clog, it will break it up. If all else fails, the tube can be wired in an attempt to break up the clog. This is done skillfully and carefully, recognizing that gastric or bowel perforation is a possible complication. Use the guide wire from a central line insertion kit or that from a nasojejunal tube. Carefully measure the distance from the abdominal wall to the tip of the gastrostomy tube. Being careful not to insert the tube any farther than this will decrease the possibility of gastric perforation. Gently insert the wire until the clog is engaged. Remove the wire and flush with water or one of the above solutions.

Gastrostomy Skin Level Devices:

Skin level devices provide a more cosmetic appearance and can decrease some of the more common problems associated with gastrostomy tubes. Most families wait anxiously for the 6 to 12 weeks of gastrostomy tract healing until a skin level device can be placed. There are many products on the market these days. They fall into two categories: tubes that are placed over an obturator, and balloon devices. The decision to place one type over the other is often determined by size. The balloon catheters come as small as a 14 or 16 Fr, while the tubes placed over an obturator come only as small as an 18 Fr. No matter what type of tube is to be used, a good measurement of the length of the gastrostomy tract is essential; that is, the distance from the inside of the stomach to the skin. Each device comes in a variety of sizes based on the tube diameter (French) and the shaft length (measured in centimeters). A variety of sizing mechanisms and tools are available to insure that an appropriate sized skin level device is placed. These devices are designed to last for 6 to 12 months. We rarely replace anything that isn’t broken, as you can cause some damage in replacing the buttons. They require replacing when the anti-reflux valve becomes defective and leakage becomes problematic, or the fit is no longer appropriate.

While there are now several different companies manufacturing these devices, the hospital stocks only one of each of the types. The **Bard Button**, a tube that is placed over an obturator, is stocked in Central Stores. We carry sizes 18 Fr, 24 Fr, and 28 Fr. These can be ordered through the Stores catalog. The most common sizes are also stocked in the Pediatric Surgery clinic. The Pediatric Surgery Clinic also stocks the **MIC-KEY**, a skin level device developed by the Medical Innovations Corporation. We keep many different shaft lengths in the size 14 and 18 Fr diameters. Each patient unit has a box of extra feeding tubes to accommodate inpatients who come to the hospital without their feeding tubes. If a child needs a different company’s device, special orders can be placed through the Pediatric Surgery Clinic.

Each resident new to the service should spend some time with the clinical nurse specialist to learn how to place these devices. Inevitably you will be called to the Emergency Department late at night or on a weekend to evaluate problems associated

with the device. If you are comfortable, please feel free to replace the device. If not, the correct answer is to always remove the device and replace with a Foley catheter, then give the family a clinic appointment for replacement of the skin level device.

Skin Level Device Problem Solving

The most common problems associated with these devices are related to the fit of the device. If there is not a good fit, and the device is too long, leakage will be problematic. If the device is too short, pressure from the wings on the skin and resulting skin excoriation will be problematic. So, careful measurement is essential when placing a skin level device.

Leakage Around the Shaft:

This usually is an indication that the shaft of the device is too long, and that the device slides up and down in the tract. The mushroom or balloon of the device does not fit snugly against the stomach wall, and leakage occurs around the shaft. Placing a shorter device (if possible) or using a washer device may make a better fit and stop the leakage. The resulting skin excoriation will need to be treated.

If the tract appears to have stretched, removing the device and placing one of a larger diameter is an option. We tend to avoid this, as it tends to make the hole larger and larger. We find it better to attempt to get the tissue to heal down around the shaft of the device with methods which promote tissue healing. It may be necessary to remove the skin level device and place a small Foley catheter in the stoma until the tissue heals down around the tube. A Foley will allow you to inflate the balloon more fully to decrease the amount of leakage around the device while the tissue heals.

Leakage Through the Shaft:

This usually means that the anti-reflux valve in the device is either worn out, or something is stuck on the valve preventing it from closing completely. If the valve is defective, it will require changing of the device. Buttons and other skin level gastrostomy devices are designed to last for 6 to 12 months. Some last far longer than that, and others may require changing sooner than that.

One can attempt to clear the valve by filling a syringe with 10 ml of water and forcefully instilling it directly into the device (do not attach the feeding tube). This may be successful in cleansing the valve of formula or dislodging a medication precipitate. After instilling the water from the syringe, attempt to withdraw the water by pulling back on the plunger of the syringe. By aspirating with the syringe, you may successfully pull the valve back into a correct position.

We never recommend changing the skin level device if it is not defective. Some parents worry that leaving a device in place for longer than 12 months will lead to infection. This is not true, and there is danger of causing damage to the tract in removing the old device and replacing it. Careful assessment of the cause of the problems will help in determining the best solutions.

Skin Excoriation:

Skin breakdown is handled the same way it is with gastrostomy tubes. Determine the cause of the breakdown (leakage either through or around the shaft of the device) and treat the underlying problem. Ointments may protect the skin. If this is unsuccessful, a skin barrier may be helpful. Be sure that the most appropriate sized device is in place.

HYPERTROPHIC PYLORIC STENOSIS

Etiology:

-Hypertrophy and hyperplasia of pyloric musculature
-Cause is unknown - may be a deficiency in nitric oxide synthesis

Males 4:1, typically first-born male

Peak incidence: 2-6 weeks of age (average 3 weeks)

Symptoms:

-Progressive non-bilious vomiting of undigested formula
-Occasionally with coffee grounds

Signs:

-"Olive" may be palpable in epigastrium or RUQ (best felt with small NG tube and sham feedings)
-Dehydration, oliguria, sunken fontanelle

Labs:

Hypochloremic, hypokalemic metabolic alkalosis; hyperbilirubinemia
Serum potassium may be elevated, but patients are whole-body depleted

Radiology: - no tests needed if “olive” palpated

1) flat plate shows dilated stomach
2) US – standard for diagnosis, usually only test needed
   Pyloric wall thickness >4mm
   Pyloric length > 17 mm
3) UGI - string sign or double track sign with shoulder

Differential Dx:

Gastroesophageal reflux, duodenal web, duodenal stenosis, duplication

Management:

Pre-op:

1) IV hydration
   - bolus 10-20 ml/kg NS
   - start 1 1/2 maint D5NS
   - switch to D5 1/2NS + 20 mEq KCl when lytes are known
2) potassium – in dehydrated patients potassium is only added to IVF after urine passed; usually does not require extra K^+ beyond IVF replacements
3) NG tube not needed unless persistent emesis; strict NPO
4) repeat lytes 12-24 hours, depending on level of abnormalities, follow urine output - before surgery prefer chloride >100, potassium > 3.5, and HCO_3 < 30

Surgery:

1) infant ready for surgery when electrolytes and fluid status corrected. This is not a strict emergency, and repair is usually scheduled within 24 hours after admission.
2) be present at induction to ensure that stomach is aspirated before beginning anesthesia.
3) pyloromyotomy (Ramstedt) – laparoscopic or open

Post-op:

1) ad lib feeds when awake from anesthesia
   - many infants vomit with early feeding attempts – holding the next feed after emesis is routine, but do not completely stop feeds because of vomiting
2) consider Reglan if persistent emesis
3) discharge usually POD #1
   - follow-up visit in outpatient at 2-3 weeks (parents may call and cancel if patient well at home)

INTESTINAL OBSTRUCTION

Infants to 24 months:

- pyloric stenosis
- incarcerated inguinal hernia
- intussusception
- aganglionosis (Hirschsprung's Disease)
- intestinal stenosis
- congenital bands, duplications, cysts, omphalomesenteric duct remnants
- internal hernia
- malrotation with midgut volvulus
- trauma

Toddler on Up:

- incarcerated inguinal hernia
- appendicitis
- adhesions
- duplications, cysts
- malrotation (anomalies of fixation)
- trauma
- granulomatous disease
- tumors

ACUTE APPENDICITIS

Most common condition in children requiring intra-abdominal surgery. Pediatric peak in mid-teens. Not rare under 3 years, just less common. Runs a more rapid and deadlier course in children (perforation can be within 6-12 hours of onset in very young).

- poor localization and walling-off
- systemic toxicity occurs early with high fever, vomiting, diarrhea, and dehydration
- 60-70% perforation rate in children < 4 years

Symptoms:

Pain, Vomiting, Fever

Pain almost always the first symptom - often poorly localized.

Vomiting may be first presenting symptom in young child because pain may not be complained about.

Fever usually 100-101, but may be up to 106 with diffuse perforation. Fever of 104 or greater without signs of peritonitis and especially if early in course makes one hesitate to diagnose acute AP.

WBC almost always > 10,000.

Radiology:

An x-ray will often show non-specific bowel gas pattern, localized ileus in RLQ look for mass effect in RLQ, FECALITH

CT scan or US should be considered for indeterminate history/exam, but with classic history and exam there is no need for radiographic work-up.

Differential Dx:

GI: gastroenteritis, regional enteritis, acute ileitis, constipation, mesenteric adenitis, neutropenic colitis, hemolytic uremic syndrome, Henoch-Schönlein purpura, pneumonia
GYN: ruptured or twisted ovarian cyst, pregnancy, mittelschmerz, PID
GU: ureteral stone, renal abscess, testicular torsion, epididymitis

Management:

Pre-op:

1) IV hydration - 20 mL/kg bolus over 30 minutes
   2X maintenance
2) antibiotic coverage
   Broad spectrum coverage
3) Foley and NG tube if infant seriously sick, otherwise wait until OR/or may not be needed at all
4) OR consent for subclavian line if perforation suspected or if anticipate prolonged ileus (alternative is wait until post-op and request a PICC line)

Post-op:

1) IV hydration 1.5-2 X maintenance initially postoperative
2) Start PO diet same day if not perforated
3) Consider TPN or peripheral support if perforation found and expect prolonged ileus
4) if perforated, IV antibiotics until afebrile, normal WBC and tolerating regular diet; convert to PO antibiotics for total 7-14 days of treatment

5) if perforated, consider CT scan about 5-7 days postoperative if persistent fever, worsening abdominal pain or ileus (looking for abscess).

Fecaliths:

Acute abdominal pain in a child associated with a fecalith on x-ray is almost certainly due to acute appendicitis. Asymptomatic fecaliths should be scheduled for elective appendectomy as soon as possible.

Perforated appendicitis with abscess:

Patients who present after 4-5 days with localized symptoms may have a contained abscess. We will typically obtain a CT scan if the symptoms have lasted this long. Surgery in this situation has higher complication rates (including inability to find the appendix and stump leak), and because of this other treatment options are attempted. Stable patients with an abscess are started on IV broad spectrum antibiotics and undergo percutaneous drainage of the abscess, if possible, by Interventional Radiology. If the symptoms (pain, fever, ileus) resolve, they remain on antibiotics for approximately 2 weeks (converting from IV antibiotics to PO when afebrile and ready for discharge from the hospital) and the drain is removed when it has minimal output. These patients will then return for an elective interval appendectomy approximately 6 weeks later. If the symptoms do not resolve with drainage, the patient will need the appendectomy performed early.

Additionally, patients with prolonged symptoms and a phlegmon on CT scan (no distinct abscess but severe inflammatory changes) may also be treated with IV antibiotics and interval appendectomy, as long as their symptoms resolve with antibiotic therapy.

INTUSSUSCEPTION

80% under 1 year; peak 5-9 months.
males 2:1

80% ileocolic; 15% ileo-ileocolic; 5% ileoileal

Etiology:

Unknown 90-95% (probably enlarged Peyer's patches)
Rarely, Meckel's diverticulum, polyp, duplication, purpura (in HSP – often ileoileal)

In infants over 2 years of age, small bowel tumor (lymphoma, soft tissue sarcoma)

Symptoms:

Healthy child who develops sudden episodes of severe abdominal pain, vomiting, and "currant jelly" stool; in between pain episodes the patient may appear fine. May have had a recent URI.

May also present insidiously with lethargy, vomiting, diarrhea.

Abdomen may be soft initially with slightly tender epigastric mass and progress to distension, signs of peritonitis.

Mass effect in RUQ with empty RLQ

Radiology:

Flat plate may show small bowel distension consistent with obstruction. Paucity of gas in colon - particularly in RLQ. However, plain films cannot rule out the possibility of an intussusception!

US - halo or bull's eye effect on colon.

Contrast enema (barium, air, water) – diagnostic and therapeutic

Management:

Pre-op:

Barium/air enema reduction may be attempted if child is not "sick", symptoms < 24 hours, and no peritoneal signs.

Prior to starting the enema:

1) IV hydration - fluid bolus
2) antibiotic coverage - cefoxitin
3) check labs
4) NG tube may be necessary

If reduction is successful:

1) admit child for 23 hr observation
2) IV hydration - 1 1/2 x maint.
3) NPO until passing gas, minimal NG output, abdomen soft

Recurrence rate after enema is 5-7%

Any episode of abdominal pain, repeat AXR and enema. One can repeat enema as often as necessary, as long as each reduction proceeds easily.

If reduction is unsuccessful, proceed to OR

Surgery:

1) transverse RLQ incision vs. laparoscopy
2) manual reduction of intussusception (push intussusceptum out, do not try to pull it out), appendectomy
3) resection for gangrene, serosal disruption
   end-to-end anastamosis usually possible

Recurrence rate after surgery is < 2%

Post-op:

1) continue antibiotics 72 hours (unless bowel resected)
2) NPO until bowel function returns

LOWER GI BLEEDING

Infants:

Necrotizing enterocolitis, infectious enterocolitis, anal fissure, sigmoid mucosal prolapse, hemorrhagic disease of the newborn (Vit K deficiency)

Toddlers: (to 4 years):

Anal fissure, infectious enterocolitis, intussusception, Meckel's diverticulum, intestinal duplication, rectal prolapse, juvenile colonic polyps (unusual before 2 years)

Puberty: (4 years and up)

-Juvenile colonic polyps (other polyps are rare: Peutz-Jeghers, adenomatous)
-Meckel's diverticulum, duplication proctitis, ulcerative and granulomatous colitis, anal fissure

Neoplasms, hemangiomas and other vascular malformations may be seen in all age groups

Always remember the possibility of a duodenal/gastric ulcer in all age groups

Basic work-up:

CBC, ESR, coag. studies, barium enema, proctoscopy or colonoscopy, Meckel's scan, bleeding scan, UGI, endoscopy

MECKEL'S DIVERTICULUM

Usually presents as sudden, painless bleeding with a large amount of dark red or burgundy blood. Usually stops spontaneously. Male predominance 3:1. Ulceration occurs in the ileal mucosa adjacent to the ectopic gastric mucosa. The diverticulum is usually within 100 cm of the ileocecal valve and can cause other problems such as: intussusception, volvulus around a persistent fibrous cord to the umbilicus, diverticulitis ± perforation. Diagnosis by Meckel's scan.

POLYPOID CONDITIONS

JUVENILE POLYPS

Incidence:

May be as high as 3%

Pathology:

Inflammatory NOT NEOPLASTIC and NO MALIGNANT POTENTIAL. Usually pedunculated mass of granulomatous tissue with edema and "lakes" of mucus

- 70% in rectum and sigmoid with another 15% in left colon
- 70% solitary; 30% multiple

Rare reports of co-existent juvenile and adenomatous polyps

Symptoms:

Painless rectal bleeding

Treatment:

-Polyp removal via proctoscope or sigmoidoscope or colonoscope
-Surgery should be avoided

PEUTZ-JEGHERS

Incidence:

-Mendelian, autosomal dominant gene of high penetrance: 50% positive family history
-No gender predominance

Pathology:

-Hamartomatous: some adenomatous (especially in duodenum). Majority in small bowel and colon

Symptoms:

-Rectal bleeding or melena; recurrent episodes of abdominal pain due to intussusception. Melanin spots on lips buccal mucous membranes

Treatment:

-Surgical intervention for complications (intussusception, obstruction etc). Risk of malignant degeneration is unknown

FAMILIAL ADENOMATOUS POLYPOSIS (FAP)

Incidence:

-1 in 6,000-12,000 births
-Autosomal dominant inheritance, high penetrance; no gender predominance
-10% spontaneous mutation
-Mutation is in APC gene on chromosome 5

Symptoms:

-Occasionally asymptomatic, usually rectal bleeding and frequent stools
-Risk of colon cancer: rare before 10yo; 7% by 20yo, 15% by 25yo; probably 100% by 50yo
-~3% develop adenocarcinoma of duodenum and therefore all are screened

Treatment:

-Usually between ages 10-20 years. Total colectomy and rectal mucosectomy with J-pouch endorectal pull-through. This can be done open or laparoscopically, and usually is performed with a diverting ileostomy. Surgery is usually performed after 18 years of age, but may be performed earlier in presence of symptoms (e.g. frequent bleeding) or development of dysplasia seen on biopsy.

LYMPHOID HYPERPLASIA:

-Commonly seen in pediatric age group as incidental finding on BE or at time of surgical exploration
-No treatment necessary

ILEO-ANAL PULL-THROUGH WITH J-POUCH FORMATION

Children require total colectomy with this pull-through procedure for a variety of reasons including ulcerative colitis and familial polyposis. The procedure is performed in two stages with a protecting ileostomy. The first stage (total colectomy, ileo-anal J-pouch pull through and diverting ileostomy) is usually performed laparoscopically. The children usually spend about four days in the hospital for the first stage, and three to four days following closure of the ileostomy. Following the colectomy and pull-through procedure, the children return to the outpatient office in two to three weeks for the first post-op visit. Rectal examination at that time determines healing and the need for anal dilations. Daily dilation with an appropriate sized Hegar dilator may be necessary. Most children are ready for closure of the ileostomy in six to eight weeks. Many choose to wait until school is out so they can use the summer months for bowel retraining. Most find that a combination of careful dietary management and the "triple threat" medications will allow them to achieve a manageable stooling pattern of four to five times per day without soiling.

<u>Post-Op Medications after total colectomy and ileo-anal pull-through and stoma closure</u>

LOPERAMIDE (Imodium)
- it is a narcotic-like antimotility agent, overdose is treated with naloxone
- no evidence of abuse, dependence, or tolerance to the antidiarrheal effect has been observed
- Imodium A/D 5 ml = 1 mg, 2 mg capsules
- DOSE: 0.5-1.5 mg/kg/24h bid-qid, dose up to 3.8 mg/kg/24h has been given

DIPHENOXYLATE WITH ATROPINE (Lomotil) this is a prescription drug
- a meperidine congener with constipating effects
- typical opioid activity at very high doses, not soluble (not injectable), atropine prevents abuse also
- 1 tablet = 5 ml liquid = 2.5 mg diphenoxylate + 0.025 mg atropine
- DOSE: 0.3 -0.4 mg/kg/24h bid-qid, adults up to 20 mg/24h, <2y do not use

KAOLIN WITH PECTIN (Kaopectate Liquid)
- Kaolin is hydrated aluminum silicate, pectin is obtained from citrus fruit rinds or from apple pomace
- Kaolin is an adsorbent. The mechanism of Pectin is unknown.
- DOSE: children 3-6y 15-30 ml/dose
  children 6-12y 30-60 ml/dose q4h

ATTAPULGITE (Diasorb, Rheaban, Kaopectate tabs/caps)
- Rheaban liquid contains 4.2 g activated attapulgite/30 ml, tablets contain 600 mg
- not narcotic, not habit forming
- Attapulgite, thermally activated is an absorptive substance which absorbs nutrients, digestive enzymes, noxious gases, irritants, toxins and some bacteria and viruses
- DOSE: children 3-6y 1/2 tbsp
  children 6-12y 1/2 to 1 tbsp
  adults 1 to 2 tbsp

METHYLCELLULOSE, CARBOXYMETHYLCELLULOSE

hydrophilic, semisynthetic cellulose derivatives, absorb water and provide an emollient intestinal mass in the treatment of diarrhea and in modifying ileostomy output
marketed as laxatives
available as powders, granules, 500 mg capsules or tablets
DOSE: children 500 mg/dose
adults 1 to 6 g/d in divided doses

PSYLLIUM (PLANTAGO) (Metamucil)

obtained from various species of plantain, contains natural mucilage and forms a gelatinous mass on contact with water, outer seed coats are a mechanical irritant
DOSE: 4 to 10 grams, one to three times per day

After closure of the diverting ileostomy in patients who undergo ileo-anal pull-through procedures, a combination of agents is used to decrease stool frequency and increase stool consistency. This includes:

1) an antimotility agent such as Imodium or Lomotil or sometimes even Codeine
2) an absorbant such as kaolin/pectin (pectin is available over the counter as Certo. Start 1/4 teaspoon 2 times per day with po liquids) or attapulgite
3) a bulking agent such as Metamucil or Fibercon

Diphenoxylate is a prescription medication. Others are available over the counter. PECTIN - available over the counter as Certo. Start 1/4 tsp. 2 times per day with oral fluids.

CONSTIPATION

Infants and children referred to us for constipation usually have a significant problem, since most parents seek help from their family doctor or pediatrician before they are referred to us. Often times, the child's constipation is a dominant issue in the household.

The first step is to reassure the family that we understand what an important problem it is, and that in all likelihood, it can be resolved. Ninety-nine percent of the constipation we see is related to three principal diagnoses:

1. *Anorectal abnormalities:* variants of imperforate anus, such as an anterior displaced anus. These can usually be discerned by the physical examination (unusual).
2. *Hirschsprung's Disease:* suggested by lifelong history of constipation. Diagnosis made by suction rectal biopsy or open rectal biopsy (unusual).
3. *Functional Constipation*: may be severe and debilitating (common). A better term to describe the problem is Fecal Retention, since children with this problem may have constant "smearing" (overflow incontinence), and parents may not understand this as constipation.

Other causes of constipation are very rare (unrecognized myelomeningocele, intestinal pseudo-obstruction, neuronal intestinal dysplasia [maybe less rare than is currently appreciated], hypothyroidism, cystic fibrosis, lead poisoning, constipating drugs).

Functional constipation/fecal retention are usually not present from early infancy. Onset is often related to a change in diet that results in increased hardness of the stool, such as weaning and

instituting table food. Onset may also be related to a specific event, such as a viral illness, with resulting dehydration and hard stools. A rectal fissure may cause pain with defecation and start the pattern of fecal retention. The child may have a BM as infrequently as once every 7 to 10 days. The BM is usually painful. Progressive abdominal distention often precedes the evacuation, as well as decreased appetite. Fecal soiling is the most common complaint. Examination is remarkable for normal sphincter tone and a vacuous, capacious rectum FOS. Barium enemas are often unhelpful (dilated rectum), and biopsies need not be done immediately as the suspicion of HSD is small.

The cornerstone of treatment is a BOWEL PROGRAM that results in a complete evacuation of the rectum each day, i.e., a program that mimics normal bowel function. A bowel program is tailored to meet an individual child's needs and includes some of the following:

- Good hydration, encourage oral fluid intake
- Fresh fruits and vegetables, roughage
- Stool-softening agents
  - Mineral oil-colorless, odorless, tasteless, cheap - should never be used in infants, secondary to the risk of aspirating it into the lungs. Instead, lubrication can be provided from below with a glycerin suppository.
  - Colace syrup
- Cathartics like Magnesium citrate, Milk of Magnesia (best used for short-term only)
- Lubricant (glycerin) suppository (acts as a good stimulant to defecation in infants)
- Dulcolax suppository (avoid in the neurologically normal child, used most often in spinal cord injury bowel programs, spina bifida, and the neurologically impaired)
- Stimulant enema (pediatric Fleets enema or equivalent) - phosphate solutions (should not be used on a daily basis without assessing serum electrolytes)
- Evacuating enema (10-20 ml/kg) of saline (2 tsp. salt per quart of water)

Enema programs are highly effective at producing a complete daily evacuation. They should be given at a specific time each day. Most families administer the enema after the evening meal. The child retains the enema for as long as possible, preferably 15 minutes. The child then sits on the toilet and evacuates as completely as possible. This pattern is rigidly adhered to for 1-2 months. The program is then withdrawn to q. 2 d., q. 3 d., and spontaneous evacuation usually resumes. There is a great deal of "behavior modification" involved in this approach. Failure to withdraw from the enema program is reason to pursue further work-up.

Most families are very satisfied with these bowel programs. Their child becomes immediately better (usually in the first week of the program). It is a little difficult to "sell" it to some families, but the results are nearly uniformly gratifying. The family is no longer focused on when their child is going to have his/her next bowel movement. The social embarrassment stops. The dirty underwear ends. The most common cause of failure is not getting started. It must be understood with the child that the enema is not a negotiable treatment - that it is being given for his/her good. Once this is understood by the child and the first enemas have been given, the treatment is accepted. Older children can participate in the administration of the enema.

For functional constipation, the desired goal of treatment is to return to normal bowel function. Many children with underlying abnormalities of the anorectum (such as imperforate anus) may benefit from similar treatments on a long-term permanent basis.

Children with presumed functional constipation who fail to wean from a bowel program should begin an evaluation with barium enema, anorectal manometry, and transanal myectomy (to diagnose an ultra short segment of Hirschsprung's Disease). Transanal myectomy is also the surgical treatment for an ultrashort segment of Hirschsprung's Disease and other forms of "anal achalasia."

TESTICULAR PROBLEMS

UNDESCENDED TESTICLE

5% absent, 10% abdominal, 3% ectopic, 82% inguinal
90% associated with patent processus or hernia
Bilateral 15%, right 45%, left 40%

Recommended age of repair: 6 months-1 year (earlier if associated with hernia)

- Relationship of histologic changes and fertility not completely clear. Histologic changes are noticeable by <2 years of age.

- Increased incidence of torsion in undescended testicle.

- Increased incidence of tumor in undescended testicle (10x) in the second or third decade of life (orchiopexy does not decrease the tumor risk, but does make it easier to examine the testicle and diagnose a mass earlier).

If you cannot palpate a testicle, may consider laparoscopy to identify an intra-abdominal testicle.

Main differential is retractile testicle, which is caused by a hyperactive cremasteric response. Retractile testicles will often descend into scrotum during a warm bath (important to ask the parents if they have ever noticed the testicle in the scrotum), and can be easily milked to the lower scrotum during examination. You can also overcome the cremasteric response by holding the testicle down in the scrotum for 45-60 seconds; if the testicle briefly stays in the scrotum when you release it, it is retractile; if it snaps immediately back up to inguinal canal it is undescended. No surgery is indicated for a retractile testicle.

TESTICULAR TORSION

Cause: unknown, may occur in sleep R = L

Symptoms:

Significant lack of symptoms in the newborn. Reddish or purple hard, non-tender scrotal mass. Sudden attack severe pain. Usually no fever or urinary symptoms. Testicle frequently elevated. Elevating testicle does not relieve the pain (as it would for epididymitis).

Differential Dx:

1) epididymitis - usually associated with fever, pyuria, leukocytosis and tender prostate
2) orchitis
   Both rare before puberty
3) torsion of appendix testis

Evaluation:

1) testicular scan - no perfusion to twisted side
2) Doppler spermatic vessels in canal
3) operative exploration if high suspicion, prolonged symptoms or delay in obtaining studies

Management:

Prompt surgery indicated - viability can usually be preserved if within 6 hours from onset of symptoms.

- Orchiopexy of contralateral testicle recommended
- Trans-scrotal incision (transverse)
- Orchiectomy if severe gangrene

Neonatal Testicular Torsion

Concern for newborn testicular torsion requires immediate evaluation and discussion with an Attending, as this is potentially a surgical emergency.

NECK MASSES

Thyroglossal Duct Cyst:

Midline cystic structure lying close to the hyoid cartilage (usually 1 cm below it). Remnant of thyroglossal duct extending from foramen cecum as a midline diverticulum from pharyngeal floor. Duct passes through hyoid bone.

Cyst should be excised when diagnosed. Cyst may be solid and consist of functioning thyroid tissue in 5% of cases, and rarely contains the patient's only thyroid tissue. Pre-op thyroid scan should be done if normal thyroid gland cannot be clearly felt.

Branchial Cleft Cysts and Sinuses:

1) Cervical cutaneous cartilaginous remnants - never associated with fistula.
2) 1st cleft: located in submandibular area and goes to external auditory canal lateral (but occasionally medial) to facial nerve.
3) 2nd cleft: located along anterior margin SCM muscle and passes between internal and external carotid arteries, over hypoglossal nerve to tonsillar fossa. Approximately 30% end blindly. This is most common lesion.
4) 3rd cleft: located along lower third of the sternocleidomastoid, enters at the pyriform sinus.

Lymph Nodes:

Common in children. Usually chronic lymphadenitis.

Any solitary, discrete, firm, non-tender node deserves consideration for excision. If not smaller over a period of observation, excision should be recommended. Diagnosis includes Hodgkin's lymphoma, thyroid neoplasm, atypical mycobacteria, cat scratch fever. In some patients a course of PO antibiotics should be considered, and biopsy performed if there is no resolution of adenopathy after treatment.

Torticollis:

In the newborn infant, a neck mass situated in the sternocleidomastoid muscle can appear between 1-3 weeks of age. This represents hemorrhage and fibrosis of the muscle body. There is usually a history of a difficult birth (breech presentation or forceps delivery). Differential diagnosis includes a cervical teratoma (based off of the thyroid) or a soft tissue tumor such as a rhabdomyosarcoma (very unusual). Diagnosis can be confirmed by cervical USG. Treatment is with physical therapy and stretching exercises.

Hemangiomas:

Vast majority require no treatment. Frequently noted at 2-3 weeks after birth with gradual growth to about one year of age. Undergo spontaneous regression and disappearance over next few years. RT may be indicated for rapidly growing facial lesions and for large lesions with platelet trapping. Steroid efficacy is debated.

Cystic Hygroma:

Congenital malformation of lymphatics with large cystic spaces containing lymph fluid. Most occur in cervical area, but are seen in almost all areas of the body. Excision at 6-12 months of age (unless involving mediastinum or compressing trachea - these should be removed as soon as possible). Important to attempt entire removal, but not at expense of sacrificing nerves and other important structures.

BREAST LESIONS

Neonatal:

Physiologic hypertrophy due to circulating maternal hormones: birth - 1 month if abscess, I & D ASAP.

Pre-pubertal:

(2-8 yrs) unilateral or bilateral slightly tender, non-fixed subareolar discoid mass. Physiologic hypertrophy, cause unknown. Boys and girls. Usually disappears at 6-12 months. DO NOT EXCISE OR BIOPSY IN GIRLS!

Adolescents:

Fibroadenoma most common tumor: should be excised. Cystic disease and carcinoma extremely rare under 18 years of age.

CHEST WALL DEFORMITIES

PECTUS EXCAVATUM

Etiology:

Unknown, though some incomplete inheritance pattern possible. Affects 2% of the population, 4x more common in males. Caused by costal cartilages curving inward and pulling lower sternum in.

Symptoms:

Deformity – usually present by 3 yo and may worsen in puberty
Decreased exercise tolerance – poor evidence of physiologic problem
Pain

Radiology:

CT – Haller index (width/depth)
PFT – usually normal, though exercise PFT's may be abnormal
Echo – if suspect Marfan

Management:

Observation for mild cases
Surgery for moderate/severe

Surgery:

Ravitch procedure – open operation with removal of affected cartilage

Nuss procedure – minimally invasive approach with placement of Lorenz bar to force the sternum to normal position. Chest remodels over time and then the bar can be removed. This is the most common procedure currently.

Post-Op:

Pain is major issue, and most patients have a thoracic epidural for 2-4 days. When tolerating diet oral narcotics are started. Toradol is also used for up to 3 days.
Diet starts night of surgery and advanced as tolerated.

All patients have a post-op CXR. Small pneumothoraces are common and need no intervention if the patient is breathing well.
Follow-up ~ 2 weeks after discharge, limited activity for 2 months.
Bar removal approximately 2 years later as outpatient.

PECTUS CARINATUM

Incidence/Etiology:

Unknown etiology, but same problem with costal cartilage as excavatum, except in carinatum the sternum is pushed outward. 5x less common than excavatum, but same male:female ratio.

Symptoms:

Develops later, with 50% > 11 yo, and worsens with puberty.
Deformity is main complaint
Pain – uncommon
NO exercise tolerance effect

Management:

Observation
Compression brace – requires at least 2 years of use, > 12 hours/day
Surgery – Ravitch procedure (may not be covered by insurance)

CAUSTIC INGESTION

Etiology:

Caustic injury due to ingestion of strong alkali (soap powders, Drano-type liquids, lye, etc.) or acid (rare). Most injuries occur only to the lips and oral pharynx, but esophageal damage must always be ruled out. Infants tend to drink liquids in bolus "gulps" and thus, material can traverse the oral pharynx and damage the esophagus. Acid burns tend to affect gastric antrum and pylorus.

Most commercial soaps (powders and liquids) do not contain enough alkali (hypochlorite) to do any damage. However, inhalation of the concentrated soap powders can cause significant oral and tracheal burns.

Symptoms:

- Drooling, refusal to swallow, pain
- Respiratory distress due to laryngeal swelling always a major concern in the first 12 hours
- Tachycardia, fever, abdominal pain - early signs of possible full thickness necrosis (thorax or abdomen).

X-rays:

1) CXR - aspiration, pleural effusion, mediastinal air
2) AXR
3) Esophageal swallow - contrast study usually at 7 days in cases of severe burns as a baseline study. Obtain water contrast study earlier if full thickness necrosis and possible perforation suspected.

Management:

Admit all suspected injuries - 7 IMC or PICU

1) Children with burns of lips or oral pharynx
   - Admit for observation
   - NPO, IV hydration
   - If pharynx is free of burns and the infant is swallowing normally with no drooling after 12 hours, consider starting oral fluids. Esophagoscopy not mandatory.

   If there is any question as to the severity of injury, schedule flexible esophagoscopy in 12-24 hours.

2) Children with pharyngeal burns and/or severe burns
   - Admit to PICU
   - Consider elective intubation if respiratory symptoms develop
   - NPO IV hydration
   - Broad spectrum antibiotics
   - Flexible esophagoscopy within 24 hours
   - If extensive or severe burns are found, insert subclavian line for TPN

Post-op:

- Begin oral feedings as tolerated
- Esophageal swallow at 7-10 days for baseline
- Repeat endoscopy at 3 weeks after injury
- Start dilatations if necessary

If severe stricture formation, consider gastrostomy tube and string placement

- Discharge home when tolerating oral or gastrostomy feedings
- Schedule repeat dilatations every 1-2 weeks as OPA until minimal stricture

All esophageal dilatations should be done under fluoroscopic control in the OR (make sure C-arm fluoro unit, flexible esophagoscope, and video adapter and cart are scheduled as special requests).

- Repeat esophageal swallow 8-12 weeks post injury and prn

FOREIGN BODIES

AIRWAY FOREIGN BODY

Etiology:
- Usually 6 months to 5 years of age
- Most aspirated objects are radio-opaque

Symptoms:
- Witnessed coughing episode post ingestion
- Intermittent episodes of cyanosis
- Wheezing, decreased breath sounds

X-rays:
- CXR-pa and lat
- Left and right decubitus films

Early:
Hyperinflated lung field
Mediastinal shift

Late:
Lobar collapse
Infiltrate

Management:

Pre-op:
1) NPO IV hydration
2) CXR
3) Obtain surgical consent:
   Bronchoscopy and removal of foreign body, possible tracheostomy

Surgery:
- Ventilating bronchoscope
- Alligator forceps
- Fogarty balloon catheters

Post-op:
1) PIMC/PICU observation if indicated
2) Albuterol treatments if necessary
3) Chest PT
4) Most discharges 24 hours post-op
   - no F/U necessary

GI FOREIGN BODY

Esophageal foreign bodies are removed endoscopically on presentation (usually the next day if the patient is stable).

Intragastric foreign bodies usually pass. If the parents do not note that it has passed, an X-ray should be done in 2-4 weeks, to be certain that it has passed from the stomach. If not, removal can be attempted endoscopically. Even batteries may be observed under certain circumstances.

COMMON TUMORS IN CHILDREN

THORACIC MASSES IN INFANTS AND CHILDREN

The tables listed below are meant to be an overview of the differential diagnosis of various masses that can appear in the thoracic cavity of infants and children. A number of lesions represent derivatives of Bronchopulmonary and Neuroenteric Foregut anomalies, while others represent acquired lesions. Anatomical location - particularly with the mediastinal lesions - has an important bearing on the potential diagnosis.

Mediastinal masses

- Neural tumors
- Neuroenteric cyst
- Bronchogenic cyst
- Enterogenous cyst
- Teratoma
- Thymoma
- Cystic hygroma
- Intrathoracic meningocele
- Intrathoracic stomach (hiatal hernia)
- Pericardial cyst
- Lymphoma
- Neoplasm
  - Neurogenic tumors
  - Soft tissue sarcoma
  - Germ cell tumors

Pulmonary Parenchymal Masses

- Bronchogenic cyst
- Bronchial atresia
- Inflammatory pseudotumor
- Pneumonia, abscess
- Pulmonary sequestration
- Cystic adenomatoid malformation

Congenital lobar emphysema (fluid-filled)
Pulmonary neoplasm
Pulmonary blastoma
Sarcoma, Rhabdomyosarcoma
Metastatic neoplasm
Osteogenic sarcoma
Wilms' tumor
Hepatoblastoma
Soft tissue sarcoma

NEUROBLASTOMA

Most common pediatric abdominal solid tumor

Pathology:

Arises from neural crest cells (precursors to sympathetic nervous system); ranges from benign ganglioneuroma to undifferentiated neuroblastoma.
Important factors in prognosis: age, stage, location, differentiation

age:	25% < 1 year
	50% < 2 years
	75% < 5 years
location:	abdomen/retroperitoneal 75% (adrenal 55%; nonadrenal 20%)
	pelvis 5%
	chest 15%
	neck 5%

Symptoms:

- Abdominal mass, anemia, fever, malaise
- Limp, paralysis (dumbbell lesion in spinal cord)
- Skin nodules
- Hypertension
- Opsoclonus-myoclonus (dancing eyes/feet)
- Horner's Syndrome

Radiology:

1) CXR/ AXR - speckled calcifications
2) CT scan abd /thorax
3) MRI – usually if concern for intraspinal extension

Work-up:

1) CBC/plts/diff
2) lytes/bun/creat/LFT's/LDH
3) Spot urine/24 hour urine for catecholamines/VMA/HVA
4) Serum ferritin/neuron specific enolase
5) Bone marrow aspirate/biopsy

Stage:

many systems in use; in US, Evans has been most common but may be replaced
Evans Staging System

- Stage I: Tumor confined to the organ or structure of origin.

- Stage II: Tumor extending in continuity beyond the organ or structure of origin but not crossing the midline. Regional nodes may be involved.
- Stage III: Tumor extends across the midline. Regional nodes bilaterally may be involved.
- Stage IV: Metastatic disease in bone cortex, distant lymph nodes, soft tissues, bone marrow.
- Stage IVS: Patients who would be stage I or II, but who have metastatic disease confined to liver, skin, or bone marrow usually in infants < 1 year.

Shimada Classification: prognosis (good or poor) based on histology of tumor and age of patient

Management:

Surgical resection if possible without sacrifice of important structures
Biopsy and neoadjuvant therapy (radiation, chemotherapy) if unresectable, followed by delayed resection after therapy

Prognosis: depends on stage, location of tumor and markers

Overall cure rate - 30%
- 90% if < 1 year of age and stage I, or IVS
- 75-80% Stage II
- 30-50% Stage III
- 15-20% Stage IV

Prognostic factors:

N-myc oncogene amplification
- (poor if > 10 copies)

DNA ploidy
Shimada classification
Age (<1yo better)

WILMS' TUMOR (Nephroblastoma)

Pathology:

Arises in kidney from metanephric blastema elements. The classic pattern is triphasic with blastemal, epithelial and stromal elements

Divides into two prognostic groups:
- Favorable Histology
- Unfavorable Histology
  - anaplastic

Renal tumors previously classified as Wilms, but now distinguished separately have worse prognosis than Wilms:
- clear cell
- sarcomatous
- rhabdoid

age: 50% < 3 years

85% < 5 years
(peak 2-4 years)

Symptoms:

Painless abdominal mass - 75-80%
Sudden enlargement suggests hemorrhage into tumor
Hematuria, hypertension

Associated anomalies:

Aniridia, hemihypertrophy, horseshoe kidneys

Radiology:

1) CXR
2) AXR
3) Abd US (IVC involvement)
4) Abdominal and Chest CT scan (lung mets still staged by CXR, not CT, unless biopsy-proven met)

Stage:

I	Limited to kidney and completely resected
II	Tumor extends beyond the capsule but is completely resected. Tumor thrombus in renal vessel.
III	Residual tumor after surgery confined to abdomen
IV	Hematogenous metastases (lung, liver, bone, brain)
V	Bilateral renal involvement

Management:

Surgery with chemotherapy/radiotherapy depending on stage.

Survival:

Stage I	> 95%
Stage II	> 90%
Stage III	> 80%
Stage IV	> 75%

EXTRACORPOREAL MEMBRANE OXYGENATION (ECMO)

ECMO provides external cardiac and pulmonary support through the use of long-term cardiopulmonary bypass utilizing a membrane lung. It has been proven to be quite effective in the treatment of several types of neonatal respiratory failure, and is moderately successful when used for post-operative cardiac support and pediatric respiratory failure. The ECMO manual outlines this process in detail and should be consulted for specific information.

ECMO is performed in the NICU/PICU with appropriate surgical equipment brought to these units from the OR. When an infant or child is being considered for ECMO, a protocol has been established so that all proper teams are notified. This protocol is outlined as a series of checklists incorporated into ECMO packets kept in the NICU or the Pediatric Surgery office. A packet should be opened for each infant and the appropriate steps followed. In particular, the Perfusion Team and the Blood Bank should be kept aware of any potential infant, as well as any changes in an infant's condition.

Major disease processes treated with ECMO:

Neonatal:

CDH, Meconium aspiration, persistent fetal circulation, neonatal pneumonias, pre- and post-cardiac surgery.

Pediatric:

Pneumonia with progressive respiratory failure, drowning, hydrocarbon ingestion, cardiac surgery.

ECMO CHECKLIST

"Possible ECMO" Status

1) Notify Pediatric Surgery Office (x8342),
   or Page Operator of patient's admission or transfer

2) Notify Perfusion office (x8551)
   (or ECMO call)

3) Notify Radiology (x8365 7:30-4:00 M-F)
   other times, beeper 1260
   - p CXR
   - Cranial ultrasound

4) Notify Cardiology
   - Cardiac ECHO

5) Initial LAB Tests
   - CBC + Platelets
   - Glucose
   - Lytes/Bun/Creatinine
   - Ca^{++}

- LFT's/Bilirubin
- PKU/MET Screen
- Type and Screen

Definite "ECMO" status

1) Move to ECMO Room
2) Reposition Infant (Head Reversed)
3) Notify Blood Bank (x8232)
   (See Blood Bank protocol sheet)
4) Confirm status with OR desk (x8240)
5) Confirm status with Perfusion Service (x8551) or
   Page Operator (see Perfusion schedule)
6) Pre-Bypass Labs
   - ABG (uac) (RA)
   - CBC + Platelets
   - Lytes/Bun/Creat
7) Prepare Heparin Infusion
8) Check consent form
9) Alert hospital service aide for 7th floor

BLOOD BANK GUIDELINES

1) ECMO protocols are established regarding blood product availability

TRANSFUSION PROTOCOL

ROUTINE TRANSFUSION PROTOCOL

1) Determine amount to be transfused.
2) Request amount in SYRINGE from Blood Bank - SUPER packed cells with Hct 85-90% (Leuko-Poor).
3) Transfuse packed cells via syringe pump over 1/2-1 hour.
4) Platelets transfusions are 1 UNIT of adult platelets in concentrated volume (15-20 ml's)
   - Blood Bank should deliver in syringe
   - Infuse Post - oxygenator with syringe pump over 1 hour.

EMERGENCY TRANSFUSION PROTOCOL

If patient needs to "CRASH ON" ECMO immediately upon admission, notify Blood Bank of:

Emergency Transfusion Protocol:

- Blood Bank should release 3 units of O+ packed cells
- (M.D. must sign)
- Send type and cross match

CANNULATION PROCEDURE

1) Position and secure infant
   - Shoulder roll
   - Head to left
2) Bovie pad in place

3) Personnel available - Neonatology
   - Pediatric Surgery
   - OR Team
   - NICU/PICU ECMO Nurse
   - Respiratory Therapy
4) Insure access to IV and endotracheal tube
5) Medications: Fentanyl: 1 mcg/kg
   Xylocaine: 0.5 %
6) Administer Heparin Bolus once vessels controlled; five minutes required after heparin before ligation of vessels
   100 units/kg X___kg = __Units
7) Cannulate Artery
8) Administer Succinylcholine (If not Paralyzed)
   (1 mg/kg)
   1 mg/kg X kg = _____mg
9) Increase ventilation if necessary
10) Cannulate vein
11) Hook-up and secure bypass lines
12) Stat p CXR (Call when starting cannulation)
13) Send pump bloods:
   - CBC and platelets
   - Lytes/CA^{++}
   - ACT
   - ABG

BYPASS START:

1) CHECK ACT q 15 until stable
   (Pre-oxygenator side)
2) START Heparin drip when ACT < 400
3) CHECK patient ABG q 15 until stable
4) SWITCH all IV lines to ports on pump circuit
5) REMOVE PIV'S when stable

ECMO STANDING ORDERS

GENERAL

1) DC all previous orders, except antibiotics
2) Condition Critical
3) PRE - ECMO LABS: Lytes, Ca^{++}, BUN, Cr, Alb, Mg, PO_4, Bili, CBC, plts, PT, PTT, Fibrinogen, FSP, PKU/metabolic screen

STAFF NURSING

1) Vital signs and blood pressure Q 1h and PRN
2) Temperature (axillary) Q 2^o, and PRN
3) HEPARINIZED PATIENT:
   NO IM MEDICATIONS
   NO VENIPUNCTURES
   NO HEELSTICKS
   NO NASAL SUCTIONING
   NO NG TUBES
4) NPO - OG (ONLY) to straight drainage
5) Blood glucose via pump line, Q 4^o and PRN
6) ROM exercises daily
7) Strict I & O q 1h
   (urine specific gravity and dipstick q shift
8) Hematest each stool
9) Suction ETT Q 2-4^o and PRN
10) SIGH BAG X 5 MIN. AFTER EACH SUCTIONING
11) Draw ABG's from the Arterial line Q 1^o and PRN
12) Arterial line: (UAC) 250 ml NS with/unit/ml Heparin at 2 ml/hr
13) IV solutions and medications are to administered via the VA ECMO circuit, unless otherwise ordered by the ECMO physician
14) D/C PIV's
15) LAB WORK
    - q Shift - CBC + Platelets
    - Electrolytes/BUN/Creatinine/Glucose/Ca^{++}, Dextrostix, pre- and post-pump ABG's
    - q AM - Total Protein, Albumin, Phosphorous, SGOT/SGPT, Total bilirubin, Mg^{++}, blood culture, PT/APTT, FSP
    - q hr - ABG's (UAC)
    - ACT's
16) Radiology (Daily)
    - p CXR
17) IVF
    - Total IVF Rate:
    - TPN
    - Intralipid
18) ECMO Perfusionist to administer PRBC's as needed to maintain Hct 38-45. ECMO technician to administer spun platelets as needed to maintain platelet count 40,000-60,000
19) Make sure there is one unit of packed cells available in the Blood Bank cooler at the beginning of each shift
20) Record I & O: Include all blood samples and infusions and blood products administered

21) Adjust the ECMO circuit to maintain appropriate parameters as per daily parameter sheet
22) Review emergency ventilator settings on the parameter sheet
23) Administer all medications and blood products into the ECMO circuit - NO IM or Subq injections
24) Under **EMERGENCY** conditions, ECMO Team may change the ventilator settings to the emergency vent settings, listed on the parameter sheet, until the respiratory therapist is available
25) Daily Weights
26) Change neck dressing - p.r.n. (weigh and record dressings)

ECMO FOLLOW-UP

ECMO follow-up should be scheduled jointly with Neonatology and Pediatric Surgery. Pediatric Neurology will be involved at the discretion of Neonatology. In the older pediatric patients, all children should have a Neurology consult while on ECMO and at the time of discharge, and should be followed jointly by Ped Surg/Pediatrics/Neurology.

Prior to discharge, baseline studies consisting of Neuro evaluation CT scan or MRI should be obtained. (BAER's should be ordered in the neonates).

Consideration should be given for a Developmental Pediatrics (Rehab) consult for the pediatric patients.

In general, follow-up should be at 2 weeks, 4 weeks, 2 months, 4 months, 6 months, and 1 year following discharge, unless specific problems warrant a different schedule.

Fibrin glue may be used for ECMO cannular site bleeding.

Fibrin Glue recipe:
10 ml cryo for fibrin glue
add 1 ml calcium chloride to
10 ml thrombin mixture
get the dual syringe from the Or

CENTRAL VENOUS ACCESS DEVICES

Central venous access devices have been used in children since the early 1970's. Their use has made a tremendous impact on the care of children requiring complex medical therapy. By minimizing the trauma associated with multiple needle sticks for blood drawing and securing peripheral venous access, central lines have significantly altered the psychological impact of hospitalization for children. The use of these devices assures a relatively safe delivery of medications, blood products and intravenous nutrition therapy. However, these devices come with their own set of associated concerns. Therefore, it is essential that everyone working with these devices have a thorough understanding of their uses, necessary care, and potential problems. The following provides information to assist in the assessment, diagnosis, and management of these concerns.

TYPES OF CENTRAL VENOUS ACCESS DEVICES

There are 4 categories of catheters used at the Hershey Medical Center. They are:

1) percutaneous (nontunneled) central venous catheters
2) silastic, tunneled catheters
3) implanted ports
4) peripherally inserted central catheters (PICC)

These devices are made of a radiopaque material which allows for easy determination of line placement. The lines are usually placed in the major veins of the neck, chest, or arm, with the distal tip in the superior vena cava or right atrium of the heart. Central lines can also be placed in veins in the lower extremities, and advanced to the inferior vena cava.

PERCUTANEOUS (nontunneled) Central Venous Catheters

Percutaneous catheters are placed into the central circulation by means of a direct puncture into a large vein (subclavian, jugular, femoral). The catheter is sutured to the skin, and the insertion site is covered with an occlusive dressing. These lines are usually indicated for relatively short-term therapy i.e. 2-3 weeks. Catheters with multiple lumens are available and are useful when the child will require simultaneous infusion of fluids and medications.

These lines are removed without the need for general anesthesia, though in children placement requires either conscious sedation or general anesthesia. Confirmation of placement is always confirmed by x-ray. Complications of placement include pneumothorax, arterial puncture, hemothorax, thrombosis of vessel and infection. When there is concern of infection or there is a need for access for a longer period of time, these catheters can be changed over a wire. Dislodgement is a real concern, as these catheters do not utilize a tunnel and antibacterial cuff system. This also means that there is a substantial risk of infection.

SILASTIC Tunneled Central Venous Catheters

Tunneled central venous catheters, most commonly the Broviac or Hickman catheter, are silastic, radiopaque catheters which are placed in a large blood vessel such as the subclavian, cephalic, internal or external jugular, or saphenous or femoral vein; they are placed either percutaneously or by cut-down. These lines are tunneled subcutaneously for several inches from the insertion site in the blood vessel to the desired exit site on the skin. In addition, one or two Dacron cuffs on the catheter are placed in the subcutaneous tunnel. These cuffs enhance tissue ingrowth and provide a protective barrier against infection by preventing tracking from the exit site into the circulation. The lines are sutured in place in the operating room during insertion, as the tissue ingrowth into the cuffs requires 2 to 4 weeks for completion. The sutures are then removed in the outpatient office in approximately 14 days. These catheters are available with either single or double lumens. If the line is placed for chemotherapy in a child with cancer, it is best to ascertain how many lumens will be necessary from the Oncologist. Most children who will go on to bone marrow transplantation will require a double lumen catheter.

Limitations of this type of central venous access device include:

1) external positioning of the catheter, so some activity limitations occur (no showering or swimming)
2) daily care by a competent and knowledgeable caretaker is essential (dressing changes and heparin flushes)
3) infection is an ever present concern, so cleanliness and aseptic technique for catheter flushing is essential
4) home care supplies are not always covered by insurance policies.

Removal of tunneled venous catheters is usually performed in the operating room. If the catheter has only been in for a short period of time (1-2 months) removal may be attempted using a local anesthetic. Conscious sedation has been used along with a local anesthetic in older children, sparing them the need for a general anesthetic.

IMPLANTED Infusion ports

Totally implanted devices that consist of a self-sealing injection port with attachable silicon catheters provide reliable, long-term central venous access. The most commonly used devices are the Mediport, the Life Port, the Vital Port, and the Port-A-Cath brands. The dome-shaped reservoir is usually constructed of a metal or plastic casing with a raised septum made of self-sealing rubber. The reservoir may be either single or double lumen. Special noncoring needles (Huber or Gripper needles) with an angled tip (Huber tip) must be used to inject into the port. This needle has a sharply beveled tip which slices the septum when inserted. When the needle is removed, the septum reseals. **Conventional needles must never be used, as they damage the septum's self-sealing capability.**

Accessing the port does require a skin puncture with the special noncoring needle. Ethyl chloride or EMLA cream may be used to numb the skin prior to accessing the device. This may be very helpful for the child who is traumatized by frequent needle sticks. If the catheter is to be used in the immediate postoperative period, it should be accessed in the operating room and the needle left in place, as the area surrounding the incision will be quite painful, making accessing the port difficult.

Once the device has been implanted, the only care required is monthly flushing with 3 ml of heparinized saline. No dressings are required unless the line is to be accessed. Once the noncoring needle has been placed and secured with an occlusive dressing, it can be managed much like an external catheter. All fluids, pharmaceuticals, and blood products can be infused through the port, and it can be capped with an injection cap and treated as a heparin-lock.

Limitations of the device include the trauma associated with the needle sticks. There is significant risk of needle dislodgement from the reservoir during continuous infusions. This is worrisome when drugs, especially vesicants or parenteral nutrition solutions, are infused through the port. Because the port is an implanted device, troubleshooting concerns may be more difficult.

Removal of this device does require general anesthesia and a surgical procedure.

PERIPHERALLY Inserted Central Catheters (PICC Lines)

PICC lines are silastic catheters of a very small caliber, which are placed into a peripheral vein either by percutaneous stick or by cutdown. The catheters are introduced through a "break-away butterfly", then are threaded over a stylet or guide wire, into the superior vena cava. The lines are most commonly introduced in the cephalic or basilic veins in the arm (usually the non-dominant arm), then threaded into the central circulation.

PICC lines are most commonly used in children who will require parenteral nutrition or antibiotic therapy for anywhere from 2 weeks to 2 months and do not wish to undergo the surgical procedure necessary for placement of the more permanent tunneled central venous access devices. They are especially well suited for children who may not be candidates for general anesthesia but require long-term venous access.

Once in place, the PICC line is secured with an occlusive dressing. The catheter may remain in place for up to 3 months. Limitations of this type of catheter include some limitation of movement and some discomfort at the insertion site. Because of the small caliber of these lines, it is not recommended that it be used for blood sampling. Never use a syringe smaller than a 5cc with a PICC line because of the risk of rupturing the line.

COMPLICATIONS WITH VENOUS ACCESS DEVICES

Infection:

Infections include catheter sepsis, exit site infection, and tunnel infection. Prevention, through meticulous, aseptic techniques with use and care, will prolong the life of the devices. Treatment is aimed at saving the device, but if the child does not improve, the device may have to be removed.

Catheter sepsis

This is defined as fever with positive blood culture.

Symptoms include:

1) Fever
2) Chills
3) Tachycardia
4) Hypotension
5) Decreased urine output
6) Redness, swelling, tenderness at catheter site

If the device has more than one lumen, blood for cultures should be drawn from all lumens. Other sources of infection should not be overlooked, so a complete septic work-up and physical examination is done.

Treatment:

1) Prompt initiation of broad spectrum antibiotics. In the immunocompromised, neutropenic child, these antibiotics should be initiated STAT. Vancomycin and Gentamicin or Mezlocillin and Gentamicin are appropriate initially, and more specific drugs can be initiated once the culture reports are available. If the device has multiple lumens, each lumen should be infused with antibiotics, alternating doses.
2) Symptomatic relief with acetaminophen or ibuprofen.
3) Fibrinolytics like TPA should be instilled then flushed through all lumens.
4) Cultures may be drawn daily until they are negative. They should also be drawn with each temperature spike (one per 24 hour period) once treatment has begun.
5) Antibiotic therapy continues for 7-14 days after negative cultures are obtained. In the neutropenic child, therapy continues until the child's absolute neutrophil count (ANC) is greater than 500 for 5 days.
6) Catheter removal is considered if cultures remain positive for 24-48 hours after initiation of appropriate antibiotics or if deterioration of the child occurs.

Exit site, Resevoir site, or Tunnel Infection

This is defined as erythema, phlebitis, or drainage of the exit site, subcutaneous reservoir, or along the catheter insertion tunnel. Fever may be present.

Symptoms include:

1) Erythema at exit site, or red streak along route of catheter.
2) Tenderness at exit site
3) Swelling at exit site
4) Purulence within 2 cm of exit site
5) Warmth and swelling along the tunnel site
6) Induration along the subcutaneous tract, greater than 2 cm from the exit site.
7) Fever

Diagnosis is made from the physical exam, positive cultures, and gram stain. Blood cultures should also be drawn from all lumens of the device.

Treatment:

1) Obtain blood and exit site cultures.
2) IV antibiotics may be considered, especially in the neutropenic patient.
3) Dressing changes should be done daily instead of every other day. If drainage is excessive, dressing changes may be done as many as 4 times daily.
4) Hydrogen peroxide swabs should be added to the cleaning regimen of the dressing change if drainage is present.
5) Add an antibiotic ointment (bacitracin, Bactroban, triple antibiotic ointment, Silvadene cream) or antifungal ointment (Mycostatin, nystatin ointment, Lotrimin) to site care.
6) Cover the exit site with a gauze dressing and tape frame instead of an occlusive dressing.
7) The catheter is at increased risk of dislodgement, so extra care in securing the catheter should be undertaken.
8) If symptoms do not resolve after initiation of antibiotic therapy, the catheter should be removed.

Cutaneous Reaction

Cutaneous reactions can range from mild to severe, and may be caused by sensitivities to any of the cleaning solutions or dressing materials used in the daily care of central venous access devices. Gauze dressings with a tape frame may be better tolerated than occlusive dressings.

Cutaneous reactions to the materials used in daily care must not be dismissed by the house officer. Skin breakdown may predispose the child to local infection and discomfort.

Occlusion

Total or partial occlusion of the catheter is the second most common problem associated with central venous access devices. Occlusion is the result of intraluminal clot formation, drug precipitate or occlusion of the tip of the catheter by the fibrin sheath that develops around all

catheters within days of placement. In an implanted port, changing the needle in the port and again attempting to flush may clear the occlusion.

Intraluminal Clot Formation

This type of catheter occlusion may occur gradually or may develop suddenly.

Symptoms:

1) Inability or resistance to flushing the catheter
2) Inability to aspirate from the catheter
3) Slow or stopped gravity flow
4) Catheter leaking
5) Occlusion alarm from the infusion pump

Diagnosis:

Breakage, catheter migration, and venous thrombosis must be ruled out. Observe for edema of the extremity, neck, or upper chest, with venous distention and pain. These symptoms will occur on the same side of the body as the catheter. A radiopaque dye study may be helpful to ascertain the size of the clot and the integrity of the catheter. Ultrasound or ECHO cardiography can also be utilized to confirm thrombosis of the catheter.

Treatment:

If repositioning the patient has failed to clear the catheter, infusing 10 ml of normal saline using a push/pull motion in flushing and aspirating may work. Connecting the syringe directly to the hub of the catheter instead of using a needle through an injection cap may work better. If resistance is still met, **do not use force**. Fibrinolytic agents may be used.

TPA 2mg/2cc is used to try to clear the catheter. As much of the dose that can be instilled into the catheter should be instilled. Allow the TPA to sit in the catheter for an hour or two. It can then be flushed through with normal saline. If patency has not been restored, the TPA may be repeated once.

Precipitate Occlusion

Precipitate occlusion of a catheter or reservoir can occur in any child who receives frequent medications, hyperalimentation or intralipids. This type of occlusion is much easier to prevent than it is to treat, as it does not resolve with the use of fibrinolytic agents. Flushing the lines with 10-20 ml of normal saline in between medications or after hyperalimentation solution is stopped prior to heparin locking will adequately flush the line and should prevent precipitate occlusion. The catheter may have to be removed if the precipitate cannot be dissolved.

Treatment:

Some success has been documented in clearing drug precipitates with the infusion of fluids without any additives (D5W or NS) at a rate of 20-40 ml/hour. Should this fail, using a 0.1 N solution of hydrochloric acid may be tried with the attending physician's permission. Inject 0.2-1.0 ml into the occluded catheter using a glass syringe, allow it to dwell for 5-15 minutes, then aspirate. Repeated attempts should be made to aspirate every 5 minutes for 30 minutes. After that, as much of the hydrochloric acid as possible should be aspirated from the catheter. This process can be repeated once.

Catheter Misplacement

These problems are infrequent. They include pneumothorax, hemothorax, perforation of the vessel, and catheter dislodgement during surgery. A chest x-ray to confirm placement should be done prior to using a newly placed catheter. Patients are also monitored for chest pain, shortness of breath, cyanosis, and bleeding.

Catheter Perforation into a Body Cavity

This can present with life threatening symptoms of shock, requiring immediate resuscitation efforts.

Hydrothorax symptoms:

1) Decreased breath sounds
2) Respiratory distress
3) Dullness to percussion
4) Rapidly advancing symptoms of shock (decreased BP, increased HR progressing to decreased HR, increased RR)

Superior Vena Cava Syndrome

This is a rare concern in children, however, it may occur at any time following placement of a central venous access device. It is caused by thrombus formation in the vessel in which the catheter has been placed, and results in obstruction of venous return to the superior vena cava.

Symptoms:

1) Edema of the neck, chest, head, or upper extremities
2) Distention of the neck or chest veins on the same side as the catheter is placed
3) Respiratory distress

Diagnosis:

Ultrasound or ECHO cardiography may be helpful in ascertaining venous thrombosis.

Treatment:

Surgical removal of the catheter is usually necessary. Heparin or fibrinolytic therapy can be used, with appropriate monitoring in the PICU or PIMC.

Air Embolism

Air can enter the system through inadvertent opening of the catheter system without clamping, through accidental disconnection of the system, or if the catheter is accidentally severed or damaged. It can be introduced during surgical placement or at anytime that the catheter is used.

Symptoms:

1) Sudden onset of chest pain
2) Shortness of breath
3) Light-headedness
4) Tachycardia
5) Pleuritic pain
6) Anxiety

7) Course mechanical noise heard over the pericardium

These symptoms may be transient, and signs are often clinically absent.

Diagnosis:

Diagnosis is based on clinical symptoms. A lung perfusion scan can assist in determining whether symptoms are due to air or thrombus.

Treatment:

The catheter should be clamped immediately. Place the child on the left side and place the bed in Trendelenburg position. Administration of oxygen may make the child more comfortable. Supportive measures may alleviate some symptoms.

Catheter Breakage

It requires immediate attention, as other problems such as contamination of the line, air embolism, or exsanguination may result. Special care should be used in an attempt to prevent this problem. However, forceful flushing, inadvertently clamping the catheter with a hemostat, accidentally cutting or puncturing the catheter with a needle all are plausible causes. It is also possible to crack the hub of the catheter - usually when attempting to disconnect tubing using a hemostat.

External Breakage

Symptoms:

1) Fluid leaking from the catheter
2) "Bubble" or rupture of the catheter
3) Catheter is severed
4) Fluid leaking from the hub
5) Hearing a "pop" or rupture while flushing
6) Burning or pain with flushing
7) Resistance to flush

Treatment:

Clamp the catheter proximal to the break. Temporary repair of a hole may be performed using "Microfoam" tape over the hole. Do not use the catheter until a permanent repair has been made.

Obtain a repair kit from Central Stores in the appropriate size. Most catheters have the size printed on the external portion of the device. Repair is performed by the surgical resident or a nurse who has been specially educated in catheter repair. Directions for the repair are included in each kit. It should be repaired using sterile technique.

If the hub of the catheter is cracked or the catheter is accidentally severed, a temporary repair can be done using a blunt-end peripheral IV catheter (14- or 16-gauge). Using sterile technique, the severed end of the catheter is cleaned, then the blunt end of the IV catheter is inserted into the open lumen, then taped securely. This may allow the catheter to be used until a more permanent repair can be made.

Internal Breakage

Symptoms may include:

1) Dysrhythmia
2) Swelling along the tunnel
3) Pain along the tunnel
4) Numbness along the tunnel
5) Loss of blood return
6) Resistance to flush

In subcutaneous ports, the catheter can become disconnected from the reservoir internally. This may be asymptomatic or cause some of the above symptoms. Catheter embolization may be asymptomatic with the exception of the loss of blood return and slight resistance to flush.

Diagnosis:

A chest x-ray may often identify catheter disconnection and migration, but a dye study is more definitive for internal breakage. A lateral view may more clearly identify a catheter that has separated from the port reservoir.

Treatment:

Pressure should be applied to the point at which the internal breakage is suspected. The embolized portion of the catheter can then be removed in the Cath Lab, Operating Room, or Interventional Radiology.

Subcutaneous Extravasation

This concern is limited to children with implanted ports, and is usually caused by accidental dislodgement of the Huber or Gripper needle from the port, with extravasation of fluid or medications into the subcutaneous tissue surrounding the reservoir of the port.

Symptoms:

1) Pain in the chest at the port insertion site
2) Swelling at the site and surrounding tissue
3) Tenderness over the port
4) Slight resistance to flush
5) Loss of blood return

Diagnosis:

The above symptoms should alert the practitioner to a problem. Pressure may be applied over the needle and another attempt to aspirate is made. One should feel the needle hit the back of the reservoir chamber.

Treatment:

Stop the infusion. If possible, re-access the device and flush with 5 ml of heparin 1:100 ml. Warm compresses can be applied until the edema has subsided enough to allow for re-accessing the port.

Some tissue damage may result if medications or hyperalimentation solution extravasate. This reaction may be minimal or may cause a large tissue slough. These

wounds are treated like burns, and may require tissue debriding, dressings, and antibiotic therapy. The subcutaneous port may or may not need to be removed.

If a vesicant drug has extravasated, the procedure for vesicant extravasation (Nursing Practice Manual, Standard M-40) should be initiated immediately. The "Guide to Injectable Oncology Chemotherapeutic Agents" found in the 7th floor treatment room and the pharmacy also has protocols for vesicant drug extravasation.

IMMUNIZATION-RELATED ISSUES

RECOMMENDED CHILDHOOD IMMUNIZATION SCHEDULE (2003)

Hepatitis B (3 doses): birth, 1-4 mo, 6-18 mo

Diphtheria, tetanus, pertussis (DTaP) (4 doses): 2 mo, 4 mo, 6 mo, 4-6 yo

Tetanus toxoid (Td): 11-12 yo and every 10 y after that

H influenza (4 doses): 2 mo, 4 mo, 6 mo, 12-15 mo

Polio (IPV) (4 doses): 2 mo, 4 mo, 6-18 mo, 4-6 yo

Measles, mumps, rubella (MMR)(2 doses): 12-15 mo, 4-6 yo

Varicella (1 dose): 12-18 mo

Pneumococcal (PCV7) (4 doses): 2 mo, 4 mo, 6 mo, 12-15 mo

For selected, high-risk patients, additional vaccines may be indicated:

Hepatitis A series: 24 mo - 18 yo

Influenza: yearly starting at 6 mo

additional Pneumococcal (23PS): > 24 mo

RSV immunoprophylaxis (Palivizumab, a monoclonal RSV-Ig): if < 2yo and with chronic lung disease or premature infants with high exposure risk

*Please consult our pediatric or neonatal colleagues for assistance if necessary.

SPLENECTOMY IMMUNIZATION AND ANTIBIOTIC PROPHYLAXIS

Children who present for elective splenectomy should receive the following immunizations pre-operatively. If possible, immunizations should be given at least 2 weeks before splenectomy. If not possible, as in trauma patients, the immunizations can be given prior to discharge.

Pneumococcal vaccine: Pneumovax (23PS) 0.5 ml SC or IM

If possible, 1 dose should be given 2 weeks before surgery. A second dose is recommended 3-5 years after the first dose. This vaccine is less effective in patients < 2yo, and PCV7 is therefore given according to the standard schedule, followed by the first dose of 23PS at 2 years old.

Meningococcal vaccine: 0.5 ml SC or IM

Recommended for all asplenic children age 2 or older

Haemophilus influenza type b vaccine: Hib 0.5 ml SC or IM

Antibiotic Prophylaxis:

For antimicrobial prophylaxis, oral penicillin V(125 mg twice a day for children younger than 5 years of age and 250mg twice a day for children 5 years of age and older) usually is recommended. Some experts recommend amoxicillin (20mg/kg per day). Penicillin is given into the teenage years, and instructions given to report to their pediatrician for high fevers.

Whenever possible, alternatives to splenectomy should be considered. Management options include postponement of splenectomy for as long as possible in congenital hemolytic anemias, preservation of accessory spleens, performance of partial splenectomy for benign tumors of the spleen, conservative (nonoperative) management of splenic trauma, or when feasible, repair rather than removal, and if possible, avoidance of splenectomy when immunodeficiency is present (eg, Wiskott-Aldrich syndrome). Post splenectomy children should wear a medic-alert badge stating that they are asplenic.

TETANUS AND WOUNDS

WOUND CLASSIFICATION		
Clinical Features	**Tetanus-Prone Wounds**	**Nontetanus-Prone Wounds**
Age of wound	> 6 hours	≤ 6 hours
Configuration	Stellate wound, avulsion	Linear wound, abrasion
Depth	> 1 cm	≤ 1 cm
Mechanism of injury	Missile, crush, burn, frostbite	Sharp surface (for example, knife, glass)
Signs of infection	Present	Absent
Devitalized tissue	Present	Absent
Contaminants (dirt, feces, grass, saliva, and so on)	Present	Absent
Denervated and/or ischemic tissue	Present	Absent

PROTOCOL FOR TETANUS PROPHYLAXIS

Obtain a history of tetanus immunization from medical records so that appropriate tetanus prophylaxis can be accomplished. Individuals with risk factors for inadequate tetanus immunization status (immigrants, rural or urban poor, elderly without known interval booster shots) should be treated as unknown.

History of adsorbed tetanus toxoid (doses)	Tetanus-prone wounds		Nontetanus-prone wounds	
	Tt[1]	TIG	Tt[1]	TIG
Unknown or < 3	Yes	Yes	Yes	No
≥ 3[2]	No[3]	No	No[4]	No

Tt = Tetanus toxoid adsorbed (for adult use); TIG = tetanus immune globulin (human).
[1]For children less than seven years old, DPT may be considered.
[2]If only three doses of fluid toxoid have been received previously, a fourth dose, preferably an adsorbed toxoid, should be given.
[3]Yes, if more than five years since last dose.
[4]Yes, if more than 10 years since last dose (more frequent boosters are not needed and can accentuate side effects).

APPENDIX I: PROCEDURE FOR REPAIR OF DAMAGED EXTERNAL CATHETERS

For temporary repair:

1) Obtain:
   - Sterile suture removal kit (containing scissors)
   - 14- or 16-gauge peripheral IV catheter
   - Injection cap
   - Silk suture
2) Temporarily clamp the catheter proximal to the patient.
3) Leaving as much external tubing as possible, cut off the damaged portion and discard.
4) Insert a peripheral IV catheter (14- or 16-gauge) into the remaining length of the catheter.
5) Place an injection cap on the newly attached connection, and flush the catheter with appropriate solution.
6) Tie a silk suture around the new connection.
7) The catheter may now be used temporarily until a permanent repair can be done.

For permanent repair:

1) Use repair kit from appropriate manufacturer. Repair kits usually contain a sterile hub, with attached tubing, a connector, silastic glue, and instructions. You will also need a sterile scissors.
2) Clamp the catheter between the patient and the damaged portion of the catheter.
3) Cut off the damaged portion, leaving as much external tubing as possible, maintaining sterility of cut end.
4) Attach the new hub and tubing connector with glue.
5) Tape a tongue depressor to both sides of the hub joint for 24 hours to stabilize it while the glue sets.
6) Allow 1 to 2 hours for the glue to set before flushing and using the catheter.

APPENDIX II: OPERATING ROOM LIST FOR NICU PROCEDURES

CUTDOWN CENTRAL VENOUS CATHETER PLACEMENT IN THE NEONATAL ICU

1) Equipment from the Operating Room
   - 5-0 Nylon
   - Jewelers forceps (2)
   - Jakes hemostats (2)
   - 5-0 Polysorb on P3 cutting needle
   - Headlight
   - Loupes

2) Equipment from the Neonatal ICU
   - Gowns
   - Gloves
   - Extra towels
   - Cutdown tray

3) Catheters
   - 1.9 French silastic catheter kit - includes additional equipment, such as Betadine for prep and Steri-Strips
   - 28-gauge silastic catheter requires additional Betadine swabs and Steri-Strips
   - Tegaderm for final dressing
   - Introducer needle is separate but available in NICU

ECMO CANNULA REPOSITION/CANNULA REMOVAL/WOUND EXPLORATION FOR BLEEDING

1) Material from Operating Room
   - ECMO tray
   - ECMO cart
   - Vesiloop
   - Loupes
   - Electrocautery with grounding pad for neonate

2) Equipment from the Neonatal ICU
   - 1/2% Xylocaine local
   - Pavulon, Morphine, or Fentanyl
   - Gowns
   - Towels
   - Gloves
   - Betadine swabs

APPENDIX III: PROPHYLACTIC ANTIOBIOTIC REGIMENS

SURGICAL WOUND CLASSIFICATION

Class I/Clean: An uninfected operative wound in which no inflammation is encountered and the respiratory, alimentary, genital, or uninfected urinary tract is not entered. In addition, clean wounds are primarily closed and, if necessary, drained with closed drainage. Operative incisional wounds that follow nonpenetrating (blunt) trauma should be included in this category if they meet the criteria.

Class II/Clean-Contaminated: An operative wound in which the respiratory, alimentary, genital, or urinary tracts are entered under controlled conditions and without unusual contamination. Specifically, operations involving the biliary tract, appendix, vagina, and oropharynx are included in this category, provided no evidence of infection or major break in technique is encountered.

Class III/Contaminated: Open, fresh, accidental wounds. In addition, operations with major breaks in sterile technique (e.g., open cardiac massage) or gross spillage from the gastrointestinal tract, and incisions in which acute, nonpurulent inflammation is encountered are included in this category.

Class IV/Dirty-Infected: Old traumatic wounds with retained devitalized tissue and those that involve existing clinical infection or perforated viscera. This definition suggests that the organisms causing postoperative infection were present in the operative field before the operation.

ANTIBIOTIC PROPHYLAXIS FOR SURGERY

Class I: no prophylaxis.
Common procedures in this class include:
- Subcutaneous lump and bump removal
- Hernias and hydroceles
- Orchiopexy

Class I with implant: prophylaxis with cefazolin recommended
- Placement of central venous access device
- CDH repair (may require PTFE patch)
- Nuss procedure/Lorenz bar placement

Class II/III: prophylaxis based on surgical site
- Lung and upper GI tract – cefazolin
  - Common procedures:
    - Thoracic procedures, including esophagus
    - Nissen fundoplication
    - Gastrostomy tube
- Lower GI tract - cefoxitin or cefotetan

Common procedures:

Appendectomy (non-ruptured)
Colectomy

Many neonatal patients with Class II/III procedures receive Gentamicin with Ampicillin or Vancomycin.

Class IV: no prophylaxis since already infected, but do require treatment, usually with broad spectrum IV antibiotics

Patients with penicillin or cephalosporin allergy can receive alternative antibiotics, such as vancomycin, clindamycin, ciprofloxacin, aztreonam, gentamicin and metronidazole.

SBE PROPHYLAXIS

- Ampicillin 50 mg/kg + Gentamicin 2 mg/kg (IV or IM)
- One dose before surgery, one dose 8 hours postoperatively