

PEDIATRIC SURGERY RESIDENT MANUAL

Oregon Health & Science University
Doernbecher Children's Hospital



2009 Edition

Oregon Health & Science University/Doernbecher Children's Hospital
Pediatric Surgery Resident Manual

ACKNOWLEDGMENT

Like the care of pediatric surgical patients, which is a craft that has been generated from the combined wisdom of many people, this handbook is a combination of information from many places. Information from this book was taken from similar handbooks from Shreveport, Boston, San Francisco, Miami and Buffalo. I hope that the information contained in these pages will serve as a reference to help you better navigate your way through Children's Hospital and understand your patients. I think that taking a sick child and giving them a chance to live a normal active life is one of the most rewarding areas in surgery and I hope by the time you leave here you will agree.

Garret Zallen, MD

*One hundred years from now,
it will not matter what kind of car I drove,
what kind of house I lived in,
how much was in my bank account,
nor what my clothes looked like.
But the world may
be a little better because
I was important in the life
of a child.*

Editors:

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PEDIATRIC SURGERY MANUAL 2009

CHAPTER I: Introduction to Doernbecher

INTRODUCTION

This handbook was designed to answer many of the questions that will arise during your time at Doernbecher Children's Hospital and with the Division of Pediatric Surgery. Children's hospitals can be very intimidating places to rotate through, but with a little background and practical information many of the common pitfalls can be avoided. However, this handbook is not all encompassing and referral to the standard textbooks and literature is anticipated. We welcome any suggestions for improvements.

Education is one of the most important purposes of your time here. You will learn through a combination of teaching done on rounds, during conferences, in the operating room and in clinics. In addition to this handbook, the following references are excellent resources for additional reading in pediatric surgery. Many of these books can be found in the John Campbell Library (CDRC-West, 2nd floor), or the BICC.

Pediatric Surgery, 4th ed., Ashcraft, et al. Sanders, 2005 (note: 3rd Ed. is available in the 9 south workroom, and in surgery clinic area)

Pediatric Surgery, 5th ed., O'Neill, et al. Mosby-Year Book, Inc., 1998.

Operative Pediatric Surgery, Ziegler, Azizkhan, and Weber McGraw Hill, 2002

Pediatric Surgery Secrets, Glick, PL, Pearl, RH, Irish, MS, Caty, MG, Hanley & Belfus, 2001

DOERNBECHER AT A GLANCE:

- Lobby:** Admissions
Starbucks
- Seventh Floor:** Specialty/Surgery Clinic (Salmon)
Pediatric Radiology (Beaver)
Outpatient Pharmacy (Frog)
Pediatric Phlebotomy
General Pediatrics Clinic (Deer)
CDRC (Child Development & Rehab) Clinic (Chipmunk)
- Eighth Floor:** Short Stay Unit/Prep Clinic (Green Maple Leaf)
OR
PICU
Hallway to ED
- Ninth Floor:** Peds Acute Care Center (PACC) –9 N/S (Sun & Moon)
Call Room
- Tenth Floor:** Pediatric Hem-Onc (PHO) Clinic (Sand Dollar)
Inpatient Hem-Onc – 10 North (Starfish)
Outside doors to CDRC-West (Surgery offices)
- OHSU 12th:** DNCC (NICU)
- CDRC West:** Pediatric Surgery Offices: 2nd floor
Ann Thompson- Administrative Coordinator (Rm 231)
Nancy Jacobs, PCNS
Kelly Kiraly, Division Manager
Marvin Harrison, MD
Mark Silen, MD
Garret Zallen, MD, (Resident Conference Thurs. a.m.)
Brenda Collins, Surgery Scheduler
Adam Manwaring, Managed Care Coordinator
Penny O'Dell, Appointment Scheduler
John R. Campbell Library (Rm 223)

WEEKLY SCHEDULE:

Monday:

0700 – 0800 Grand Rounds (Old Library Audit)
0830 – 0945 Resident Conference
0830 – 1730 O.R. block time (Zallen)
1700 – 1800 Department M&M (UHS 8B-60)

Tuesday:

0800 – 1730 O.R. block time (Silen)
(O.R. Dr. Zigman – check OR schedule)

Wednesday:

0700 – 0800 Trauma Conference (DCH 11th Floor)
0800 – 0900 Tumor Board (DCH 10327)
0910 - 1210 Educational Conferences (alternate OHSU and Emanuel Hospital. Division M&M 1st and 3rd weeks)
1300 - 1600 Shriner's Pectus Clinic – every 2nd Thursday
(one resident to attend)
1300 - 1700 CLINIC: Dr. Harrison: 7th fl/Specialties

Thursday:

0700 – 0800 Peds. Resident Conference (alternates between OSHU and Emanuel)
0730–1500 O.R. block time (Harrison)
0900 - 1200 CLINIC: Dr. Zallen: 7th fl/Specialties – *every 1st, 3rd, and last Thursday.*
1300 – 1500 SHRINERS SURGERY OR CLINIC: *Surgeries on 2nd Thursday and Clinic on 4th Thursday of every month.*
Check with Ann Thompson or Dr. Zallen. These are not on the Outlook Surgery Schedule.

Friday:

0900 - 1200 CLINIC: Dr. Silen; 7th floor
0900 - 1200 CLINIC: Dr. Zallen; 7th floor – *every 2nd and 4th Friday.*

DAILY ROUTINES:

Morning rounding routines are determined by the Fellow/Chief. Afternoon rounds do not occur at a scheduled time. Ward patients are on 9 N/S (the PACC), and 10N (Hem-One);. The Chief or Fellow should routinely see PICU and NICU (DNCC) patients. **It is expected that you communicate with the appropriate attending each morning after rounds are completed.**

The OR starts at 8:30 am on Mondays and 7:30 am all other days. Block time is on Mondays, Tuesdays, and Thursdays. If operating on Monday, you are expected to report promptly to the O.R. after morning conferences.

Clinic experience is considering an important (and required) element of your training. Clinic starts at 1:00 p.m. on Wednesdays and 9:00 a.m. on Fridays. **It is expected that you are present at the beginning of clinic unless in the O.R.** It is appropriate to leave clinic for consults but have the courtesy to let the Attending know where you are going.

HELPFUL HINTS (a.k.a. Surviving and Thriving in Peds Surgery):

- **X drive, Outlook Surgery Scheduling Calendar:** If you experience difficulty accessing our specific folders when coming on service, promptly contact Kelly Kiraly (4-7115) or Ann Thompson (4-8871). The daily inpatient census needs to be updated and posted to this secure drive daily, as it is utilized and viewed by clinicians as well as office staff. Please send the updated census out to the group every day prior to multi disciplinary ward rounds at 0830, which Jenny participates in. The intern is encouraged to attend when possible.
- **Required time away:** If you must be absent for a meeting or appointment, let the fellow/chief resident know ASAP so plans for coverage can be made. The fellow & chief are responsible for the call schedules.
- **Clinic Attendance:** is an expectation. If you cannot come to clinic on time, or have to leave during clinic, have the courtesy of informing the attending surgeon of your need to be late or to leave.
- **Viewing Upcoming Surgeries:** You may view (by proxy) upcoming planned surgeries on your Groupwise calendar: Open your calendar. Click on the picture of the head on the left side. Select “proxy”; then type in “Pediatric Surgery.” Clicking on any individual case will reveal additional details and plans concerning the patient that are being coordinated by the office nurse/staff. Note that actual case order is generally not determined until the day prior to surgery. Dr. Zigman’s cases and Shriner’s cases are not listed.

- **OR:** Residents must be in the room prior to induction if they expect to perform surgery, & preferably meet the patient in pre-op. All exceptions need to be cleared with the attending prior to the case.
- **Mg/Kg Dosing:** All pediatric medication orders must include: dose/kg, dose, route, frequency and indication otherwise the order will not be filled- if the child is at the adult dose write “adult” in the dose/kg slot. This is a National Safety Standard. Non-compliance will generate pages to you, and delays in treatment for your patients.
- **TPN:** TPN is usually started at D10, 1.5g amino acid/kg and 1.0g lipid/kg- Do not go over D12.5 for a peripheral IV.
- **Epidurals are managed by the Pain Service.** Do not write for additional pain meds before consulting with them. Do not discontinue foley catheters until Epidurals are discontinued.
- **Pain in Children:** It is real, and requires appropriate treatment. Refer to the pain med guidelines in Appendix F of this manual. Tylenol is often sufficient for simple outpatient procedures in patients under 2 yrs of age. Instruct caregiver to administer around the clock for 24-48 hrs.
- **Intussusception:** All children getting an air contrast enema for intussusception must have a functioning IV and be accompanied by someone from the surgical service.
- **Routine trauma:** These patients do not need more than a CBC and a U/A. Trauma CT scans are done with IV contrast, but without oral contrast and patients do not need routine foley catheters or NG tubes.
- **Lymph Node Biopsies:** If a patient is going to the OR for a lymph node biopsy a preop chest x-ray must be obtained to rule out potentially life threatening mediastinal disease.
- **Consents:** All inpatients need to have their operative consent completed the day before. If there is a problem obtaining consent, the attending should be notified immediately.
- **Pediatric Pre-Op NPO Instructions:**

<u>Age Group</u>	<u>Solids/Milk</u>	<u>Clear Liquids</u>
0-6 months	up to 4 hrs before	up to 2 hrs before
6mo-18 yrs	up to 6 hrs before	up to 2 hrs before
- **Pre-Ops:** Complete the following on all clinic (or returning) patients:
 - H&P (EPIC)
 - Pre-Op Admitting Orders (for outpatients and a.m. admits)
 - Consent

- Medication Reconciliation Orders & Allergy Documentation (a.m. admits, also all inpatient admissions)
- Anesthesia Questionnaire (must be completed by parents PRIOR to leaving clinic)
- **Red Packets:** For convenience, the above listed paperwork is currently contained in red packets, located in the Pediatric Surgery filing cabinet. The attending surgeon must sign the H&P. Let the clinic RN/CNS know as soon as you are done pre-op'ing a patient, as she will provide specific instructions and direct them to the surgery area for a Prep Clinic visit. **Red packets are available in clinic, in Peds Hem-Onc Clinic, and in Short Stay.** For pre-oping patients that will be discharged and coming back for surgery, complete a "Red Packet", then drop off originals at the surgery front desk. Jenny, our PNP, is often available for pre-oping cases, so use your resources!
- **Wards of the State:** Children who are in foster care require DHS consent. This can be obtained by phone, if urgent. For outpatients, the RN/CNS will assist in obtaining consent in time for elective surgeries.
- **Add-Ons:** A call must be made to the Anesthesiologist On-Call (pager # 10838) or the OR Charge RN (#11947) to add-on cases. Additionally, bed control must be called if the patient is not already an inpatient, to reserve them a bed either on the ward or in the Short Stay Unit post-operatively (call 4-7000). Please provide NPO instructions and write orders accordingly, keeping in mind that cancellations may provide an opportunity for an earlier operation.
- **Patients:** Round and write notes on every patient, every day. Personally review all x-rays, laboratory values, and other important data.
- **Communications:** Communicate directly and in a timely fashion with the responsible attending about any significant changes in patient status. This shall NOT be done via email, phone messages, or any other indirect method.
- **Dictations:** It is our expectation that residents dictate all operative notes, discharge summaries, and other such documents before you leave the OR or within 24 hours of the case or discharge. If an operative report is discovered to be missing 72 hours or greater after the procedure time, OR privileges will be suspended for that resident until the documentation is completed.

- **Discharging patients:** When discharging patients home, please communicate with Nancy or office staff if they will need any tests/procedures for the follow-up appointment (this does not apply to Dr. Zigman's patients). Otherwise, instruct families to contact the office promptly after discharge so that they can get a timely follow-up appt. (503-494-7764). Most patients are seen back in 2-3 weeks.
- **Clinic Appointments:** Please, do not tell patients to come to clinic unless you notify the office--we hold clinic on the 7th floor with 13 other specialties on various days. The front desk will turn them away if there is not an appointment in the computer! (email Deanne Naylor and Brenda Collins or call 4-7764 with pt. name, MRN, and reason for visit).
- **OR Scheduling:** Scheduling of cases with the office is best done by the attending surgeon, but the Chief can email us, if necessary. Include: PT. NAME, MRN, PROCEDURE, DIAGNOSIS, LENGTH OF PROCEDURE, and whether pt. will be IP, OP, OR AM ADMIT (please cc: to the attendings as well).
- **Scheduling Tasks:** The Pediatric Surgery Office is happy to help with scheduling etc. Call or page Nancy (4-0229 or #14511) with specific needs. When Nancy is unavailable, please email Brenda Collins and Nancy Jacobs. This ensures that things are done as intended.
- **Resident Work Hours:** Observe and carry out all regulations regarding work hours and inform the responsible faculty member of any potential risk of violation of these regulations.
- **E-mail Etiquette:** Do NOT communicate medically or legally sensitive information in written form such as email, except as is required for appropriate patient charting.
- **KAISER Patients:** Be aware if a new consult is a Kaiser patient; page Dr. Zigman first when he is available (7a-5p m-Th, 7a-12p Friday) + his on call night times. He generally has cases scheduled on Tuesdays--our groupwise calendar DOES NOT reflect his cases! You must look at the daily OR schedule.
- **Call Room:** The Call Room is located on 9 South (near big white staircase). The number is 12468.
- **Pediatric Surgery Advanced Practice Nurse:**
 - Nancy Jacobs is a Clinical Nurse Specialist, primarily involved in the coordination of clinics and OR scheduling, and the phone triage world of Peds Surgery. Nancy oversees the Bowel Management Program and is also a wound and ostomy specialist, so call her if there are issues on the ward she can help with.

<u>OHSU Pediatric Surgery Office:</u>	<u>Pager</u>	<u>Phone</u>	<u>Cell</u>
CDRC West, 2 nd Fl. Rm 229	-	4-7764	
Fax	-	4-6467	
Marvin W. Harrison, M.D	11057	4-4079	
Mark L. Silen, M.D., M.B.A.	12581	4-4078	503-310-9611
Garret S. Zallen, M.D.	14856	4-0579	503-459-3375
Audrey Durrant, MD (07/09 - 06/11)	10611		
Michael Tirabassi, MD (7/08 - 6/10)	13183	971-678-7885	
Nancy Jacobs, CNS	14511	4-0229	
Peds Surgery on-call pager	17062		

Ann Thompson, Administrative Coordinator	4-8871
Brenda Collins, Surgery Scheduler	4-7434
Adam Manwaring, Managed Care Coordinator	4-1315
Penny O'Dell, Clinic Scheduler	4-4077
Kelly Kiraly, Division Manager	4-7115

Emanuel Surgery Office:

501 N. Graham, Suite 300		503-460-0065
David Bliss, M.D.	503-441-1385	
Sanjay Krishnaswami, M.D.	503-441-1079	503-367-8053
Answering Service		503-238-8400

Kaiser Pediatric Surgery:

Andrew Zigman, M.D.	503-940-0358	appts: 503-203-2176
	OHSU 10693	

DCH Peri-operative Services:

	<u>Pager</u>	<u>Phone</u>
DCH O.R. (Charge RN)	11947	8-5600
PACU		8-5650
Front Desk/Reception		8-5300
Prep Clinic (for pre-op evals)	12589	8-5303
OR Fax		8-5304
OR Rm #1		8-5608
OR Rm #2		8-5610
OR Rm #3		8-5612
OR Rm #4		8-5614
OR Rm #5		8-5616

Miscellaneous:

	<u>Pager</u>	<u>Phone</u>
Admissions	-	4-7000
Anesthesiologist on-call	10838	-
Code 99	-	4-8222
ECMO Perfusionist on-call	17234	-

Interpreter Services – Spanish	-	4-8900
Interpreter Services – All	-	4-2800
Lab	-	4-7383
Pathology	-	4-6775
Pediatric Pain Service on-call	12987	-
Peds Sedation Service Charge RN	11566	4-3136
Pediatric Surgery Resident “pager”	17062	-
Paging Operator		4-8211
Pharmacy		4-1139
Priority Paging Operator		4-9000
PICU		8-5800
9 North		8-5950
9 South		8-5900
PACC (9 N/S) Charge Nurse	10313	8-5955
Diane Lampa (PACC manager)	10553	
10 North (Hem-Onc Inpatient)		8-5100
Hem/Onc Clinic		8-5150
		8-5163
		8-5164

Other Frequently Used Numbers:

DNCC		4-8122
Lab		4-7383
Transfusion Services		4-8537
DCH Radiology (scheduling)		8-5252
CT – inpatient		4-2915
CT - outpatient		4-5792
Interventional Radiology		4-7660
MRI		4-5390
Security (emergency)		4-4444
Security (non-emergency, dispatch)		4-7744

DOCUMENTATION:

Documentation is important and expected to be done in compliance with both division and hospital requirements. The Department expects uniformity in chart notation. Following are the outlines of the forms of notation acceptable to the Department. Documentation should always be done as soon as possible. Discharge summaries and operative reports should be completed within 24 hours. Residents on our rotation who fail to complete a operative report within 72 hours will be suspended from OR privileges until documentation is completed.

Operative Note

- Date of operation
- Preoperative diagnosis
- Procedure
- Postoperative diagnosis
- Specimens –note size of resections and excisions
- Estimated blood loss
- Blood and fluids administered
- Drains used
- Anesthesia technique
- Complications
- Pre-op medications-narcotics, antibiotics, etc
- Surgeons, assistants
- Teaching physician attestation (as applicable)

Discharge Summary

- Date of admission and discharge
- Attending surgeon and Specialty
- History of present illness
- Admitting physical examination
- Admitting diagnosis
- Admitting laboratory values
- Hospital course—to include all subsequent test
- Operations, complication, etc
- Discharge medications
- Plan for surgical follow-up
- Copies to PCP/Referring MD

Documentation Tips

1. Pre-birth consults often lack an exam and other required components to select a level of service. We can default to using the length of time of the encounter. In order to do this, you must document a chief complaint and the approximate time the attending spent answering questions and counseling the patient.

2. A complete Review of Systems is defined in the 1995 CMS guidelines as follows: *“At least ten organ systems must be reviewed. Those systems with positive or pertinent negative responses must be individually documented. For the remaining systems, a notation indicating all other systems are negative is permissible. In the absence of such a notation, at least ten systems must be individually documented.”*
3. When seeing a patient for follow up, please remember to include a sign, symptom or diagnosis. If there is nothing new the patient is presenting with, then the original reason we saw the patient is acceptable.

The nature of our surgical patient population should in theory be **comprehensive** encounters that would be a higher level of service. In most cases, if we are unable to bill a high level of service, it is because a lack of proper documentation. Frequently, limited history documentation, specifically review of systems will down code an encounter. Please keep in mind when documenting outpatient clinic encounters, a comprehensive history is required to achieve a high level of service. In order to obtain a comprehensive history the following needs to be documented: 4 or more history of present illness, 3 or more past/family/social history and 10 or more review of systems.

Levels of Service

New Patient Outpatient Encounter (3 of 3 elements required)

- Level 1 problem focused history; A problem focused examination; Straightforward medical decision making
- Level 2 expanded problem focused history; expanded problem focused examination; Straightforward medical decision making.
- Level 3 detailed history; detailed examination; Medical decision making of low complexity
- Level 4 comprehensive history; comprehensive examination; Medical decision making of moderate complexity
- Level 5 comprehensive history; comprehensive examination; Medical decision making of high complexity

New Patient Outpatient Consult (3 of 3 elements required. Referring provider must be documented.)

- Level 1 problem focused history; problem focused examination; straightforward medical decision making
- Level 2 expanded problem focused history; expanded problem focused examination; straightforward medical decision making.
- Level 3 detailed history; detailed examination; Medical decision making of low complexity

- Level 4 comprehensive history; comprehensive examination; Medical decision making of moderate complexity
- Level 5 comprehensive history; comprehensive examination; Medical decision making of high complexity

Established Outpatient follow up (2 of 3 elements required)

- Level 2 problem focused history; problem focused examination; straightforward medical decision making
- Level 3 expanded problem focused history; expanded problem focused examination; Medical decision making of low complexity
- Level 4 detailed history; detailed examination; Medical decision making of moderate complexity
- Level 5 comprehensive history; comprehensive examination; Medical decision making of high complexity

Inpatient H&P (3 of 3 elements required)

- Level 1 detailed or comprehensive history; detailed or comprehensive examination; Medical decision making that is straightforward or of low complexity
- Level 2 comprehensive history; comprehensive examination; Medical decision making of moderate complexity.
- Level 3 comprehensive history; comprehensive examination; Medical decision making of high complexity

Inpatient follow up (2 of 3 elements required)

- Level 1 problem focused interval history; problem focused examination; Medical decision making that is straightforward or of low complexity
- Level 2 expanded problem focused interval history; expanded problem focused examination; Medical decision making of moderate complexity
- Level 3 detailed interval history; detailed examination; Medical decision making of high complexity

Inpatient Consult (3 of 3 elements required. Referring provider must be documented.)

- Level 1 problem focused history; problem focused examination; Straightforward medical decision making
- Level 2 expanded problem focused history; expanded problem focused examination; straightforward medical decision making
- Level 3 detailed history; detailed examination; Medical decision making of low complexity
- Level 4 comprehensive history; comprehensive examination; Medical decision making of moderate complexity
- Level 5 comprehensive history; comprehensive examination; Medical decision making of high complexity

Medical Students

1. Are expected to attend all rounds, operations, and clinics.
2. May document Chief complaint, history and review of systems.
3. May see consults and present them to the resident/ chief resident/fellow/staff.
4. Should follow 2-3 ward in-patients closely and write progress notes.
5. Should be supervised by a physician for any procedures

PATIENT ADMISSIONS:

1. The resident will consult with the chief resident/fellow/attending regarding any patient that may need admission prior to agreeing to admit the patient.
2. **Call 4-7000 to request a bed, as appropriate.**

CONSULTS:

1. All consults should be seen in an expeditious manner. When possible they should be seen with the chief resident/fellow before calling staff.
Note: Dr Zigman is to be called for Kaiser patients Monday through Friday days (Fridays until noon) , and nights when he is on call (refer to Peds. Surgery Call Schedule)
2. Non-urgent consults (i.e. gastrostomy placement) received in the evenings/nights may be seen the following morning.
3. No orders should be written as the consultant without first discussing plans with the requesting service.
4. **When writing up a consult- please put in the first sentence “Asked to evaluate patient by Dr. _____.” This is important for future communications, as well as billing.**

PROCEDURES:

1. Procedures should never be performed in a child’s room. The child’s room is considered his/her safe place. Please take them to the treatment room unless there are extenuating circumstances. Make sure an attending or chief resident has been notified that you are about to perform a procedure.
2. It goes without saying that we need consent for all procedures and many parents will want to be present. Obtain consent when you are discussing the procedure with the parent, do not plan to come back and get consent later as the parent may be gone and this may delay the operation or procedure.

3. Make sure you are getting consent from the person who has legal custody- some parents do not have custody of their children and can not sign consent forms. Our advanced practice nurses can assist you.

CLINICS:

1. Residents should wear appropriate attire to clinic. This would be shirt and tie for males and business attire for females.
2. Long-term/complex patients should be seen by the fellow or chief resident when possible. Other patients should be seen by the residents or medical students. For legal and billing reasons, medical students cannot complete New Patient H&Ps or pre-op H&Ps. All patients need to be seen by the attending before they leave clinic.
3. Residents are to document a note, which the attending will co-sign in EPIC. The attending will write a letter to the PCP. Procedure notes are generally completed by the resident.
4. Orders are selected in EPIC and must be matched with an appropriate diagnosis code. Orders that require prompt processing or scheduling should be brought to the attention of the nurse. Make sure you follow up on studies ordered on all clinic patients.
5. Pre-op patients are to be sent to Prep Clinic with their completed paperwork after you have completed your H&P, orders, and consent. Parents are to fill out the Pre-Anesthesia Questionnaire contained in the packets prior to leaving clinic. Notify the clinic nurse when you have completed the requisite paperwork so that she may provide additional pre-operative teaching and send them to pre-op clinic.
6. Pediatric Hem-Onc Clinic (PHO): You may be asked to complete a pre-op for a line placement/removal in the PHO clinic. They are located on the 10th floor. The charge nurse has Pre-op packets. Patients should be directed to the Prep Clinic (8th floor surgery area) with their packets (consent and pre-anesthesia questionnaire) if possible. If surgery is scheduled for the following day, provide with appropriate NPO instructions (see pg 20).
7. Patient Instructions for Surgery: the office staff calls all outpatients the day before surgery to provide NPO instructions and arrival times. Generally, outpatients go first, and younger children go before older ones. Add-ons for the following day are generally done at the end of the day.

KAISER PATIENTS:

Dr. Andrew Zigman is on duty Monday – Thursday from 0700-1700: Friday 0700 – 1200. Check call schedule for evenings and weekends. Contact Dr. Zigman with a major status change, even if not on duty. Pager number is 503-940-0358.

Families are to call Dr. Zigman's office at 503-203-2176 to arrange follow-up appointments. His office is located at the Mother Joseph Plaza office building, on the Providence St. Vincent Campus, across from the emergency department. He has block time in the OR on Tuesdays, and attends Wednesday morning teaching rounds with the team.

SHRINER'S PATIENTS:

1. Shriner's Pectus Clinic is held on the **4th Thursday of the month from 1:00 to 4:00 PM**. Drs. Zallen staffs this clinic. One resident is expected to participate.
2. Surgeries are scheduled on the 2nd Thursday (at Shriner's). Patients are then transferred to DCH PICU (usually one day), then must be transferred back to Shriner's.
 - Dictations: You are responsible for dictation the Operative Report and Discharge Summary.
 - **NOTE: See APPENDIX E- page 184 for specific dictation procedures at Shriners.**

OPERATING ROOM:

1. CASES:

- a. Residents and medical students are expected to be in the OR when the patient goes in the room. A patient should not go to sleep without a physician member of our team there. Emergency or critically ill or ICU patients should be escorted to the OR by the resident. Likewise, the resident should be present for extubation and return with the patient. The chief resident or attending (if no chief resident will be present for the case) needs to be notified prior to the patient being brought back to the OR.
- b. The patient's surgical site needs to be appropriately marked and the consent form needs to be signed prior to the patient going to the OR.
- c. Radiological studies should be obtained and brought to the OR if not available on PACS.

2. ADD-ON CASES:

Residents may be instructed to post a case with the OR if it is a same day or next day add-on. Call the anesthesiologist on-call (pager 10838) and Bed Control 4-7000 if the patient is not already inpatient. All cases added after noon for the next day will be posted as add-ons without a specific start time. **Otherwise, cases are to be scheduled through Nancy Jacobs (4-0229, #14511) or the Surgery Office schedulers (4-7764).** Be prepared to specify diagnosis, procedure, length of procedure, admission type (i.e. IP, OP, a.m. admit), and attending surgeon when contacting the office.

3. NPO GUIDELINES (for surgery and sedated procedures):

<u>Age Group</u>	<u>Solids/Milk/Breast</u>	<u>Clear Liquids</u>
0-6 months	up to 4 hrs before	up to 2 hrs before
6mo-18 yrs	up to 6 hrs before	up to 2 hrs before

Good rule of thumb for INPATIENTS (except for the very small infants): make NPO after midnight in case there are cancellations for the OR and their case may be moved earlier in the day.

4. INDICATIONS FOR PRE-OPERATIVE FLUIDS:

Patient with fevers, vomiting or diarrhea, or bowel preparation should have IV infusions the night prior to surgery. If surgery has been or is expected to be delayed more than 6 hours in infants (0-1 years of age) and more than 8 hours in children, an IV should be started to give replacement fluids.

ORDERING TESTS/LABS/CONSULTS:

1. Inpatient Labs: In general, our floor patients do not need daily labs. This is not only unnecessary and costly, but it is painful and traumatic to the children. TPN patients should be monitored (CMP 3x/week while advancing, weekly if on maintenance)
2. Consults: Before ordering any consults please check with the attending. Out of respect, call consultants directly with your request.
3. Pre-Op Labs: Patients do not need routine labs for most outpatient surgeries. If you have any doubt, ask attendings. Review all inpatients with the chief residents. Coags and T&C are not routinely obtained unless there is a documented need. Pregnancy tests should be considered in females who have reached menarche.
4. Blood Products: Blood products can be extended from a 3 day to a 21 day expiration if patient has not been pregnant and has not been transfused in past 90 days. Alert office RN/CNS to arrange this in advance, as a specific process must be followed to accomplish the extended outdate.

A SURGEON'S NOTEBOOK:

Surgical training is largely a self-educating process. In learning and continually refining your craft, an enormous amount of information must be assimilated. To be useful, the information must be organized and categorized. Information derived from reading journals and texts, and from attending lectures, courses, and conferences is best collated in a well thought-out file system. However the multitude of fine technical points involved in the actual conduct of an operation are more difficult to organize.

Learning to perform operations well is difficult for a variety of reasons. First, there are many fine points involved in each step of a multitude of different operations. Secondly, the multiple tricks in the armamentarium of the master surgeon are usually imparted only across the operating room table, where notes and sometimes even words are extraneous, so many of the subtle points are recognized only by the perceptive and attentive observer. Thirdly, surgical technique is often considered a simple manual skill unsuitable for intellectual analysis and refinement. It is not obvious to the novice that in the simplest step, there are a number of well-planned movements. Finally, surgical training is so long and varied that the variety of techniques used for common bile duct exploration which seemed so obvious when participating in three cases per week on a busy general surgery service may be extremely difficult to remember after several months on cardiac surgery, gynecology, and anesthesia rotations. The fine technical points involved in the less frequently performed operations are usually completely lost.

The question is how to organize and categorize technical skills. Dictated operative notes are not satisfactory because they are too wordy and cumbersome to be used as a reference and are seldom filed by category of operation. A surgical atlas is an excellent technical guide, but is not useful for recording the many technical tricks learned from different surgeons throughout surgical training. This difficulty might be overcome by a sparsely written atlas with simple line drawings to which notes and drawings can be added.

A simple but seldom used solution is to write technical notes to yourself. This should not be a big deal – a simple loose leaf notebook kept in your surgical locker will serve the purpose. One page is devoted to each operation or topic (e.g. fundoplication, bowel anastomosis). A brief outline for each operation would include: 1) Preop: Notes on surgical indications, special preparations, bowel preparation, drugs, tubes, etc.; 2) Exposure: Notes on position of patient, prepping and draping, position of the surgeon, assistants, and scrub nurse, the incision, types and position of retractors for exposure, etc. This overview is perhaps the most useful means for dissecting and analyzing an operation; 3). The operation in sequential steps: Notes on the technical maneuvers written in schematic outline – it is helpful

to divide complex operations into clearly defined steps with alternative approaches for each. Drawings are helpful. 4) Closure and postoperative care: Notes on tube and drain placement and special postoperative considerations such as when to discontinue antibiotics, remove drains, etc.

These notes should be cryptic, jotted down in telegraphic style, and embellished with drawings and doodling. Complete sentences are disallowed, and the notes need not be intelligible to any other person. But the key to the surgeon's notebook as a learning device is discipline. Notes are made immediately after an operation, as automatically as dictating an operative note or changing out of your scrub suit. When an operation is first recorded, it may take 10 to 20 minutes to rethink the various steps and details and organize them in your notes. This is an extremely useful learning device in itself, since this will fix the operative steps in your mind and you will learn as much from one operation as ten otherwise.

The major advantage of this system, however, is the ability to accumulate multiple details, and multiple alternatives for each step of each operation, gleaned from years of training at different levels with different attending surgeons in different hospitals. It now becomes easy to note on the same page variations in technique, the advantages and disadvantages of alternative maneuvers, and finally with experience, your own preference. The surgeon whose armamentarium includes a variety of techniques will have patients with fewer complications than the surgeon whose repertoire is limited to one way of dealing with a problem.

This little notebook now becomes an extremely efficient way to accumulate a mass of small pieces of information and bring them all to bear on one particular step of one particular operation. As you progress during residency and begin to perform operations with less supervision, you should frequently review and embellish your notes. It takes only a few minutes while changing into scrub clothes to brush up on the details of the different steps of a complex operation which is infrequently performed. This permits mental rehearsal before the operation which will be the actual demonstration. You will soon be improving and synthesizing the various maneuvers you have learned. Your notebook will prove useful even in this period of consolidation.

This learning system should not become a burden. It requires some discipline and only a few minutes of time to record the small technical details learned in the operating room. When nothing new is learned, nothing need be recorded. But, when small tricks are recalled and recorded, your appreciation of the surgical craft is enhanced. Operations are no longer a blur of retractors, tissues, and sutures, but a symphony of small precise steps which can be delineated, recorded, and assimilated into a masterful operation that is a technical joy. Some gifted residents may not need such a mechanism to help them remember and master the many technical nuances they are exposed to during the hundreds of operations

performed during 5 to 10 years of residency. But most will find it useful. They will follow the example of such technical masters as Leonardo da Vinci and Harvey Cushing and keep a little notebook for themselves.

RESIDENT RESPONSIBILITIES:

1. Processing: The proper care of sick infants and children depends on a logical empathetic approach to their problems using factual information to formulate treatment. This logical approach consists of: analysis, decision-making regarding management, and selection and implementation of treatment. The surgical residents play an integral role in analysis and management in all facets of care. They assemble the data, which establishes the probability that a given condition exists. Decisions for management and selection of treatment are discussed with the fellow/chief resident prior to calling staff. The fellow/chief resident should discuss the plan with staff and is responsible for contacting each attending regarding their patients.
2. Preparation: When rotating on to the service, it is essential that each child's history is reviewed, and that you have a clear understanding of the patient's issues. The surgical resident must prepare for rounds. This includes examining the patient, interviewing the night nurses as to any significant events, reviewing the previous 24 hour intake and output, labs, medications, and progress notes.
3. Re-Assessment: See patients frequently throughout the day. (Always introduce yourself and your title when entering the room.) The resident is expected to review the patient's course during the day, anticipate problems, and following discussion with staff, implement necessary treatment during the regular work day. It is inefficient to delay decision-making and treatment until late afternoon.
4. Communication: The patient's attending surgeon is to be made aware of all clinical issues and progress on a daily basis, preferably during morning hours. Ensure appropriate daily progress note is present on all patients. Communication also extends to the caregivers and nursing staff. Any patient room on 9N/S can easily be called by dialing 8-59** (** being the patient's room number).
5. Collaboration: Formulate treatment plan with staff. Delegate to members of the surgical team as appropriate.
6. Supervision: Medical students are encouraged to more closely follow 2-3 ward patients and write notes. However, for practice billing purposes, the resident must write their own H&P, consults, and follow-up notes. A co-signature of the medical student note is not acceptable.
7. Consultation: See consults in a timely fashion and discuss with staff.

8. Follow-up: Review **ALL** radiology studies with radiologist. Review all lab results that you have ordered in a timely fashion.
9. Discharge Planning: Clear discharge plans with staff prior to discharge. Involve Case Manager as early as possible if home supplies/special needs are anticipated.
10. The Inpatient Census List (clearly designate *consults*): This is an integral communication tool for the surgical team. It is to be maintained and updated daily by the intern or junior. You will be given access to the I: drive Pediatric Surgery folder when you come on service. E-mail current list the first thing each morning to all residents on the service, all attendings (Marvin Harrison, Mark Silen, Garret Zallen), advanced practice nurses (Jennifer Bevacqua, Nancy Jacobs), office support staff (Brenda Collins, Kristi Hamer, Kristal Roberts), and case manager (Debbie DeLorenzo). **Note:** To get to the census folder, go to I: drive, OHSU SOM, SURG, PDS, Census. **It is a HIPPA violation to store the census in the Transfer file or any other drive, as they are not secure.** If you have any problems with access problems, contact Kelly Kiraly (4-7115) or Ann Thompson (4-8871) in the Ped Surgery Office promptly.
11. Morbidity and Mortality Reports: A copy of completed M&M forms are to be sent to Ann Thompson, Pediatric Surgery Administrative Coordinator (fax # 4-6467) for division M&M reports.
12. Trauma Team: The pediatric trauma pager must be carried by the entire team at all times. The Junior resident on call should attend all trauma situations in the ED (all trauma). The Fellow/Chief resident should attend all Level I traumas. The Trauma Service handles all patients age 15 and above. Pediatric Surgery service admits all patients 14 years and below. On weekday evenings, the Trauma Service will initially evaluate all pediatric traumas, regardless of age. The Pediatric Surgery resident will then be called to assist only if necessary. The Trauma Team/Chief Resident will be responsible for communication with the Pediatric Surgery attending as needed. The next morning, the Trauma Service will sign-out these patients to the Pediatric Surgery service. The purpose of these changes is to take some of the load off the Pediatric Surgery residents so that your time on the service is more educational. At all times, flexibility, cooperation and good communication will be expected.
13. **The Chief Resident/Fellow is responsible for seeing all patients on rounds each day.** When possible, consults should be discussed with the Chief (or Fellow) prior to presenting to the on-call attending surgeon. Articles to the junior residents regarding patients and other topics of pediatric surgery are to be provided by the chief. Wednesday conferences are the responsibility of the fellow. The chief and/or fellow presents M&M the last Wednesday of his/her rotation..

14. **Resident Work Hours:** Each resident is responsible for monitoring his/her own hours and insuring that they do not violate them.

RN-MD COMMUNICATIONS:

Communications between disciplines can sometimes be a challenge. Our attendings, fellows, and advanced practices nurses are committed to finding ways to improve communications between the PACC RNs (Pediatric Acute Care Center, 9 North/South) and the surgery team.

Currently, the following practices have been agreed upon to improve the efficiency and predictability of communications. If you have additional suggestions for improvement, please let us know.

1. Residents will round in the a.m. **0600-0800 weekdays; 0700-0900 weekends;** and **afternoon rounds 1600-1800 weekdays.** This can be the whole team or a single designee.
2. When the Resident(s) walk onto each unit to begin rounds, they need to tell the secretary to text page the RN's that "ped surg is here". This gives the RN's opportunity to ask you questions at that time instead of paging you later. Note that 9N and 9S are two different units and you must tell the secretary of each when you arrive on the unit.
3. RN's will cluster nonurgent night calls at 0100.
4. RN's will wait near the phone for 5 minutes after paging someone.
5. Residents must change vital sign parameters if they don't want repeated calls on vitals outside written limits.
6. A Ped Surg weekly schedule will be posted on each unit, so the RN's are aware when the team is occupied elsewhere (1st call should be to PNP)
7. A preprinted order sheet of the top 10 medications prescribed is available for your use. This may help decrease nursing calls for inappropriately written orders.
8. An "Alert" sheet is located in the bedside chart right in front of the vital sign page. This is for the RN's to write down their, or the parents, nonurgent concerns. This sheet should be checked at rounds and questions/concerns verbally answered.

CHAPTER II: CARE OF THE NEONATE

CLASSIFICATION OF NEWBORNS:

Newborn infants can be classified into four groups based on their level of maturation and physical development:

- Term infant: gestational age greater than 36 weeks with a body weight greater than 2.5 kg.
- Preterm infant: gestational age less than 36 weeks with a birth weight appropriate for that age.
- Small for gestational age (SGA): gestational age greater than 38 weeks with a body weight less than 2.5 kg.
- Large for gestational age infant (LGA): weight greater than 90 percentile for gestational age or greater than 4.0 kg if term.

SGA and premature infants may weigh the same but can be distinguished by their maturity levels. (See Newborn Maturity Rating and Classification).

The special problems related to each also differ:

SGA:

- Decreased body fat leads to lack of insulation and increased risk of cold stress
- Hypoglycemia may result from decreased glycogen stores and increased metabolic activity
- Higher red blood cell and total blood volumes may lead to polycythemia and hyperviscosity

Preterm:

- Weak suck reflex
- Inadequate GI absorption
- Hyaline membrane disease
- Intravascular hemorrhage
- Hypothermia
- PDA (patent ductus arteriosus)
- NEC (Necrotizing enterocolitis)

ADMISSION AND EVALUATION OF THE NEWBORN:

1. Outside Transportation of the Surgical Neonate:
All of the transports are done by the NICU's transport team. Most if not all of the details of transfer are arranged by the NICU. If you are told of an impending transfer, please let the fellow or attending know so that appropriate plans can be made.
2. Work-up for Neonates/Pre-operative Preparation:

- a. Insert a nasogastric tube (if indicated), empty stomach and evaluate contents.
- b. Do a general but rapid examination.
- c. Examine and work on baby under radiant heat warmer.
- d. Order blood gases if cyanotic or tachypneic.
- e. Order blood as indicated.
 1. CBC
 2. Electrolytes, BUN, Ca, bilirubin, glucose
 3. Blood cultures (1 cc in each yellow top tube)
 4. Type and cross if going for surgery (20 cc/kg PRBCs)
 5. Coagulation studies
- f. Vitamin K 1 mg. (all neonates)
- g. Start IV – preferably percutaneous.
- h. Initiate x-rays and other studies and prepare for operating room, if indicated.
- i. Babies with possible cardiac anomalies need a pre-op ECHO and cardiology evaluation if indicated
- j. Interview parents and obtain history and physical, including family history.
- k. Monitor the patient continuously.
- l. If chromosomal abnormalities are suspected, blood must be drawn before any transfusions are given.
- m. **Parental consent for surgery. This is a very important facet of the pre-op preparation as not all parents are present the day of surgery. Cases can be cancelled if the consent is not done or is done improperly. If you have any questions as to what the consent should read, please ask the chief resident or attending.**
- n. Pre-operative antibiotics: Ampicillin and gentamicin or cefoxitin – usually are given in the OR unless the patient has been on antibiotics previously

3. Thermal Instability:

Heat loss can be very rapid and fatal, necessitating a number of precautions. An isolette, porta-warm mattress, or radiant-heated table is used for patient care. Heat lamps are used during procedures outside the isolette and during prepping and draping in the OR. Warmed prepping solutions are used. A temperature monitor and thermostatic warming blanket are used to evaluate thermal stability throughout. Additionally, warmed I.V. solutions are warranted for massive infusions.

4. Hypoglycemia:

Hypoglycemia is a particular risk in the infant of a diabetic mother or a small-for-gestational age baby. Symptoms can include jitteriness, seizures, apathy, hypotonia, apnea, or hypothermia; but it can be

asymptomatic. The goal is to keep glucose >40mg/100 ml. Prophylactically, one should give 4-8 mg glucose/kg/min. (e.g. 100 ml/kg/24 hours of D₁₀W). For acute hypoglycemia, administer a STAT push of D₂₅W, 1-2 ml/kg.

5. Hypocalcemia:

This is likely in low birth weight or stressed infants. Symptoms can include jitteriness, convulsions, and other nonspecific symptoms. The critical level is that of ionized calcium, which depends on serum total protein. For acute symptomatic hypocalcemia start 10% Ca gluconate at 1 ml/min to maximum dose of 3 ml/kg. Stop when clinical response is obtained. Monitor ECG continuously, in ICU or on portable cardiorespiratory monitor. Follow with Ca infusion up to 50-60 mg Ca/kg/24 hours.

NECROTIZING ENTEROCOLITIS:

1. Highly lethal disease primarily seen in low birth weight newborn infants. Characterized by ischemic necrosis of the gastrointestinal tract.
2. Clinical: Mostly seen in premature and/or low birth weight infants. The incidence of perinatal complications is high: RDS, apneic spells, low Apgar, premature rupture of the membranes, breech delivery, exchange transfusions, Cesarean section, umbilical artery catheter.
3. Onset: The time of onset is usually between the second and fifth day of life. The great majority of the infants will have been fed prior to onset of the disease. The most outstanding clinical feature is bloody diarrhea. Poor feeding, apneic spells, lethargy, abdominal distention, prolonged gastric emptying, and bile-stained emesis characterizes the disease as well as x-ray findings of pneumatosis.
4. Etiology: The etiology is unknown. Consistent contributors to the pathophysiology of NEC include: prematurity, feeding and the presence of bacteria in the GI tract.
5. Pathology: The ileum, cecum, and right colon are the most common sites of involvement. The bowel becomes dilated hemorrhagic and necrotic. Microscopically, the earliest finding is coagulation necrosis. With increasing severity there is mucosal ulceration, submucosal hemorrhage, and eventual necrosis of the entire bowel wall. A mononuclear infiltrate is present. The gas is found in the submucosa and subserosa. Thrombosis of major mesenteric arteries and veins is not present. Small blood vessels may be thrombosed, compatible with the intravascular coagulation and hemorrhagic state (frequently seen terminally).

6. Radiology: Since the clinical presentation is often nonspecific, radiology is important in early diagnosis, in addition to evaluation of progress and detection of early and late complications. The main findings are dilated bowel, intramural gas (pneumatosis), and portal venous gas.
- Dilatation: This is the earliest and most common sign. The amount of dilatation is usually related to the severity of the disease.
 - Intramural gas (Pneumatosis): In the clinical setting, this finding confirms the diagnosis. However, the amount of gas is not related to the severity of the disease, and it may disappear within 12 hours. Disappearance is not necessarily related to improvement.
 - Portal Venous Gas (PVG): Can precede or co-exist with pneumatosis. As with pneumatosis, it may appear and disappear rapidly. Those infants with PVG are usually, but not always, more severely affected. As with pneumatosis, its disappearance is not always associated with clinical improvement. Treatment: Because there is evidence NEC is infectious in nature, both prevention and therapeutic regimens are directed toward the control of microbiologic agents.
7. Management:
- NPO, NG suction,
 - Triple antibiotics (ampicillin, gentamicin, clindamycin or flagyl) for 10 days empirically.
 - KUB and left lateral decubitus films q6-8h.
 - Serial CBC, platelet count, pH.
 - Routine ID control measures, and good hand washing.
8. Surgical Indications:
- Pneumoperitoneum
 - Relative surgical indications: Abdominal wall cellulitis, RLQ mass, persisting isolated dilated loop of bowel – (“fixed loop” on AXR), failure to respond to medical therapy: Thrombocytopenia, acidosis, severe hemodynamic instability.

NEONATAL SEPSIS: RECOGNITION AND TREATMENT:

Neonatal sepsis is a major cause of death in infants with anomalies requiring surgery. The diagnosis is difficult to make because signs and symptoms are often minimal. Septic infants usually show hypothermia or temperature instability, rather than fever. Their color and general appearance are poor, representing diminished tissue perfusion. The infant is less active and may vomit as ileus develops. Urine output usually remains adequate until later stages.

The usual organisms are gram-negative bacteria and can cause disseminated intravascular coagulopathy (DIC) shortly after onset of sepsis. Therefore, serial platelet counts are a good way of detecting sepsis, whereas WBC is not. With an increasing bandemia or a platelet drop, rapid evaluation, blood cultures and appropriate therapy should be instituted. Do not delay therapy since rapid deterioration may occur - start antibiotics and wait for culture confirmation. In neonates, a lumbar puncture should be part of the sepsis work-up as these patients often have associated meningitis. With severe generalized sepsis, fluid requirements are increased.

There is often pulmonary and cardiac decompensation due to the severe toxic state with cellular injury. Ventilatory assistance may be required. Antibiotics should be chosen to provide good coverage for gram negative organisms such as Klebsiella and Pseudomonas. For prophylactic coverage in infants requiring surgery, give Ampicillin and Gentamicin.

1. Signs of sepsis:

- Hypothermia or temperature instability
- Thrombocytopenia
- Leukopenia
- Mottling
- Lethargy
- Apnea
- Poor feeding
- Left shift

2. Workup:

- Blood, urine and sputum cultures
- LP
- CXR
- INR, PTT Platelets, CBC and differential (left shift)

3. Treatment:

- Empiric antibiotics after cultures are sent.
- Support of circulation with colloid and/or pressors, if necessary.
- Administration of FFP if DIC is present.
- Respiratory support, if necessary.

Chapter III: Congenital Anomalies

ESOPHAGEAL ATRESIA

1. Embryology:

At 3 wks gestation, a respiratory diverticulum forms at the ventral aspect of the pharyngeal foregut. The diverticulum elongates and gives rise to the trachea and lung buds. The ventral trachea and dorsal esophagus are separated by a septum which forms in a cephalad direction starting at the carina. Failure of complete separation results in a tracheo-esophageal fistula (TEF). Etiology of Esophageal Atresia (EA) is unclear. Many theories exist.
2. Gross Classification:

		Frequency
Type C	EA, distal TEF	86%
Type A	EA, without TEF	8%
Type E	TEF without EA (H or N type)	4%
Type D	EA, proximal & distal TEF	1%
Type B	EA, proximal TEF	1%
3. Pathology:

EA prevents passage of saliva and feeds and results in aspiration. TEF allows for passage of liquid to the trachea via the fistula either from oral intake/saliva or reflux of gastric contents. In either case, respiratory distress, atelectasis and pneumonia result.

 - a. The esophagus of affected infants has less neural tissue in Auerbach's plexus which results in altered esophageal motility (even after repair).
 - b. The trachea is compressed by a thickened esophageal upper pouch during development and leads to tracheomalacia.
4. Symptoms and Signs:
 - a. Polyhydramnios
 - b. Prematurity in 34%
 - c. Excessive drooling
 - d. Choking, coughing, regurgitation, cyanosis with feeds
 - e. Progressive respiratory distress
5. Diagnosis:
 - a. 5 or 8 Fr relopogle sump tube is placed through the nose into the proximal esophageal pouch. If one has already been placed, remove it and attempt placement yourself. Obstruction is encountered usually at 9-13 cm. Leave this relopogle in and to suction to prevent aspiration of pharyngeal secretions. An air "pouchogram" is the first option and can be obtained by injecting around 5 cc of air into the proximal pouch and taking an x-ray immediately following. Barium is avoided due to risk of

aspiration, but on some occasions a pouchogram is obtained and barium is safer to aspirate than gastrografin, hypaque, or Omnipaque.

- b. CXR reveals:
 - i. Gas in the stomach indicates a distal TEF
 - ii. Gas in the stomach but not in the small bowel indicates an associated duodenal atresia (double bubble sign)
 - iii. Gasless abdomen indicates EA without TEF
 - iv. Lungs may be normal or show evidence of pneumonia, atelectasis, RDS; heart may be enlarged or abnormal in shape.
 - c. Bronchoscopy and esophagoscopy may be performed to clarify anatomy (done at time of operative repair at the surgeon's discretion).
6. Associated anomalies:
- a. Occur in 50%
 - b. Majority = cardiac anomalies
 - c. VACTERL (vertebral, anal, cardiac, TE fistula w/EA, renal, limb) anomalies occur as a nonrandom association. Also referred to as VATER syndrome. These infants have a higher mortality rate.
 - d. Intestinal atresias
7. Clinical approach:
- a. Healthy babies (with Type C):-> primary repair and closure of TEF
 - b. Complicated baby
 - i. Proximal pouch decompression
 - ii. Antibiotics
 - iii. Gastrostomy
 - iv. Primary Repair when lungs improve or other life threatening problems are resolved.
 - c. Ill babies: (major associated life threatening anomalies, persistent pneumonia, sepsis, RDS, small preemies).Either proceed as in 7b above or:
 - i. Division & suture of fistula may be required in patients who cannot be adequately ventilated secondary to decreased airway pressures.
8. Surgery:
- a. Division and approximation of esophageal ends. If long gap exists- defined as greater than 2 vertebral body distance between the two ends of the esophagus:

- i. Proximal pouch may be stretched with a dilator (so may distal pouch via gastrostomy) for 6 weeks or longer prior to surgery.
- ii. Baby may be allowed to grow (with proximal pouch decompression) prior to surgery.
- iii. Esophagostomy and gastrostomy may be performed to allow growth time.
- iv. At surgery, circumferential esophagomyotomy of the proximal pouch may produce enough length for anastomosis.
- v. Several creative anastomoses have been described to give added length.
- vi. Controlled suture fistula technique.
- vii. Some children may be candidates to go home with nursing care to await pouch growth to permit repair. If there are no major associated anomalies or medical problems and they have a good family with insurance/medicaid willing to cover home nursing care, they may go home at 2 weeks with gastrostomy feeds, apnea monitor, continuous suction via Replogle tube in the upper pouch, daily upper pouch bougienage, 8 hours of nursing care daily, and planned rotation of the Replogle tube between nares every 3 days with irrigation and/or replacement as needed for occlusion.

9. Complications:

- a. Anastomotic leak
- b. Recurrent TEF
- c. GERD
- d. Tracheal compression (by aorta or innominate artery)
- e. Tracheomalacia- barking cough, makes child more likely to have complications with simple viral infections

INTESTINAL OBSTRUCTION:

1. Etiology: Causes for intestinal obstruction in infants differ from those in older children. The common causes of obstruction are intestinal atresias, Hirschsprung's disease, meconium ileus or plug, and malrotation. There are several points to emphasize which are common to many infants with intestinal obstruction.
 - a. **BILIOUS EMESIS IN AN INFANT DENOTES MALROTATION- A SURGICAL EMERGENCY UNTIL PROVEN OTHERWISE.** A **rapid** evaluation with an upper-GI series is mandatory.
 - b. All infants require an adequate IV and NG tube when intestinal obstruction is suspected.
 - c. On plain films, the newborn colon cannot be distinguished from small bowel because haustral markings are not yet detectable. Only by filling the colon with contrast agent can the dilated loops be accurately identified as colon or small bowel.
 - d. Water-soluble contrast is used. Barium is contraindicated.

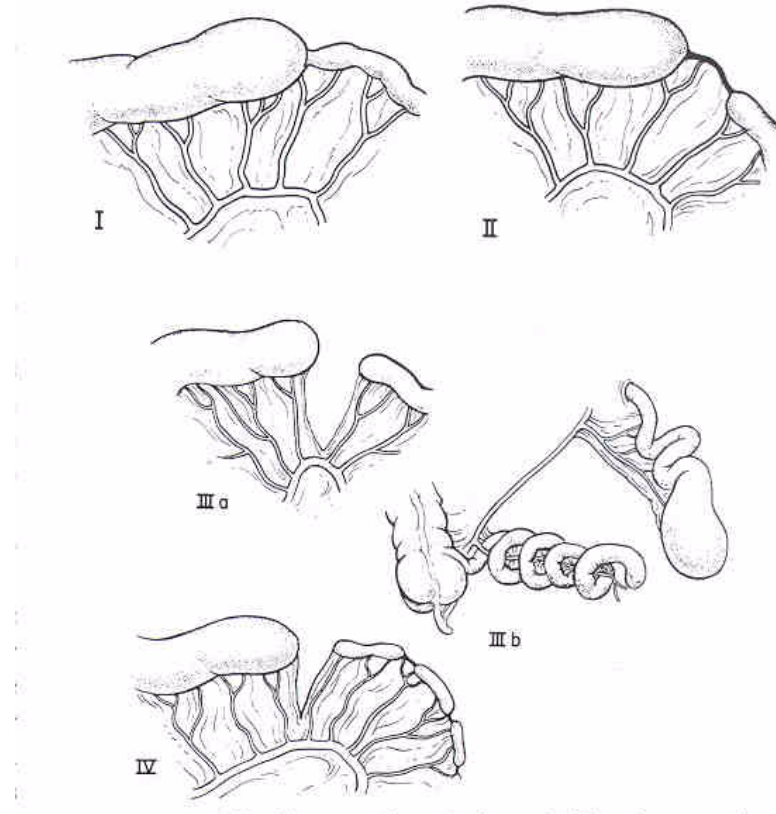
DUODENAL, JEJUNAL, ILEAL AND COLONIC ATRESIA:

1. Epidemiology:

Atresia is the most common cause of congenital intestinal obstruction in the newborn. The incidence is 1 in 2710 live births male:female ratio: 1:1
2. Classification:
 - a. **Duodenal atresia** and stenosis are described as either proximal or distal to the ampulla of Vater and are distinguished by examining the gastric contents for the presence of bile. Types include: complete, diaphragm-type, windsock web, cord-type, and absence of a duodenal segment.
 - b. **Jejunal and ileal atresia** sites are equally distributed from the Ligament of Treitz to the ileocecal junction.
 - **Type 1**: 20% of cases. An intraluminal diaphragm with continuity of the muscular layers of the bowel wall.
 - **Type 2**: 35% of cases. Atresia with a cord-like segment between the blind ends of the bowel.
 - **Type 3a**: 35% of cases. Atresia with complete separation of the blind ends and a V-shaped mesenteric defect.
 - **Type 3b**: 4 % of cases. Atresia with an extensive mesenteric defect and a distal ileum with a blood supply from a single ileocolic artery. The distal intestine coils around the vessel giving the appearance of an apple peel

deformity. Associated with unusually small distal bowel, significant shortening of overall bowel length, malrotation, and prematurity.

- **Type 4:** 6% of cases. Multiple small bowel atresias.



- c. **Colon atresia** accounts for less than 5% of all cases of intestinal atresia. The various types described for the jejunum and ileum also occur in the colon.

3. Embryology/Pathology:

At the third week of gestation when the biliary and pancreatic buds are forming, the duodenum is a solid core of epithelium. Over the next 3 to 4 weeks, the duodenum undergoes vacuolization and recanalization with reconstitution of the intestinal lumen. Failure of recanalization, results in obstruction of the lumen, often in conjunction with the developmental malformation of the pancreatic anlage and the terminal

part of the biliary tree. Jejunoileal and colonic atresias are thought to result from an ischemic injury to the bowel after the midgut has returned to the coelomic cavity.

4. Associated Anomalies:

30% of babies with duodenal atresia and 10% of those with jejunoileal atresia, have major cardiac, renal, musculoskeletal, or CNS anomalies. Trisomy 21 has been detected in 30% of infants with duodenal atresia. The VACTERL syndrome, meconium ileus, omphalocele and gastroschisis are other anomalies known to be associated with intestinal atresias.

5. Diagnosis:

a. **Prenatal:** Polyhydramnios occurs in half of newborns with duodenal and proximal jejunal atresia, and in fewer newborns with ileal or colonic obstruction.

b. **Postnatal:** NG aspirate which reveals bile-colored gastric contents or volume >25cc is highly suggestive of intestinal obstruction. Vomiting shortly after birth (bile if the obstruction is proximal), associated with abdominal distention (80% of those with obstruction distal to the jejunum), failure to pass meconium, and jaundice are all signs of intestinal obstruction. Abdominal films will help to determine the site of obstruction. The “double-bubble” sign without distal air is diagnostic of duodenal obstruction. The abdominal films of babies with proximal jejunal atresia show only a few air-fluid levels and no gas in the lower part of the abdomen. The number of air filled loops increases with the more distal the obstruction. A water-soluble contrast enema may be useful to differentiate ileal or colonic obstruction from Hirschsprung’s disease, meconium plug syndrome, or meconium ileus.

c.

6. Surgical management:

Duodenoduodenostomy is performed for duodenal atresia unless a wide gap exists between the two ends of the duodenum, in which case a duodenojejunosomy is performed. For jejunoileal atresias, the dilated bulbous tip of the proximal bowel is resected or tapered, as is the tip of the distal segment, and an end-oblique anastomosis is performed. Patients with colon atresia often have a temporary end colostomy with resection of the dilated proximal segment.

HIRSCHSPRUNG'S DISEASE:

a. Etiology:

Theories include: Failure of migration of enteric neuroblasts, failure of differentiation of precursor cells to neuroblasts/ganglion cells and, destruction of neuroblasts/ganglion cells after migration.

b. Pathophysiology:

Ganglion cells are not present to integrate the intramural cholinergic and adrenergic plexuses. As a result, the peristaltic wave of normal ganglionic bowel does not progress to the aganglionic segment, the internal rectal sphincter paradoxically contracts in response to rectal distention, and the inability to have normal bowel movements results. The involved segment is most commonly the rectosigmoid but the entire colon with or without small bowel may be involved.

c. Clinical Presentation:

- No defecation for the first twenty-four hours of life in a full term infant, abdominal distention and vomiting.
- Constipation with or without diarrhea or incontinence, in older children.
- Enterocolitis

d. Diagnosis:

- Perianal inspection: to rule out anomalies.
- Abdominal x-ray: to rule out other etiologies of bowel obstruction and contraindications to barium enema.
- Contrast enema: to identify a cone-shaped transition zone between proximal dilated and distal nondilated bowel, and to rule out other etiologies of bowel obstruction. Transition zone usually not yet developed in newborns.
- Rectal biopsy: to demonstrate neural hypertrophy and absent ganglion cells.

e. Differential Diagnosis:

- NEC
- Meconium plug syndrome
- Meconium ileus
- Intestinal atresia
- Intestinal dysmotility syndromes
- Functional constipation

f. Treatment:

- **Colonic irrigations:** These are usually sufficient to decompress the colon and allow the child to feed and grow so that they can be a candidate for a one-stage laparoscopic pull-through. Flagyl irrigations may be initiated when the child has a bout of enterocolitis. **The dose of flagyl is 50mg/kg per day dissolved into normal saline – the volume of which depends on the size of the child.** For example, most children will need around 60-100ml/irrigation and they are usually given 3 times a day. The technique is as follows- a large red rubber catheter is inserted into

the rectum and is advanced as far as possible into the colon. The catheter is then injected with 10 ml of flagyl irrigant (if the child does not have enterocolitis then saline can be used) and the syringe is detached and the effluent is allowed to run out. The catheter is then pulled out a few centimeters and the process is repeated until the catheter is all the way out of the rectum. It is important to let the catheter drain between injections.

-Ostomy: Decompression, decrease caliber of dilated bowel, improve nutrition, decrease risk of NEC. Done only in extreme cases when infant is very ill or has total colonic Hirschsprung's.

-Definitive procedure: Patient can be allowed to grow and have definitive pull-through done at several months of age. Some surgeons prefer to do these operations in the newborn period. Usually a one stage definitive procedure can be performed. Most babies are done laparoscopically.

- Swenson-distal aganglionic segment is resected, native rectum is everted, and an oblique two layer anastomosis is performed to proximal ganglionated colon.
- Duhamel/Martin – designed to avoid anterior rectal dissection. The anterior wall of the ganglionated colon is anastomosed to the native rectum to create a common wall and hence a larger rectum with ganglion cells posteriorly.
- Soave/Boley – mucosectomy of the native rectum is performed. The ganglionated colon is pulled through the rectal sleeve and anastomosed one centimeter above the anal verge. This is the technique most commonly used laparoscopically.

g. Complications:

- Anastomotic leak: 6%
- Stenosis: 9%
- Enterocolitis

MECONIUM ILEUS:

1. **Incidence:** Occurs in about 15% of infants with cystic fibrosis. The incidence of CF in Caucasians is about 1 in 2500 live births. CF is very rare in non-Caucasians. Males and females are equally affected.
2. **Diagnosis:** Plain film of the abdomen can show bowel loops of variable size with a soap bubble appearance of the bowel contents. Calcification on the abdominal films indicates meconium peritonitis resulting from an intrauterine intestinal perforation proximally. A contrast enema is contraindicated if the plain film shows calcifications.
3. **Treatment:** The initial treatment is nonoperative with enemas. Hypaque, water soluble contrast, is used. (Gastrografin, is no longer used in most institutions, as it is very hyperosmolar and can cause rapid loss of fluid into the gastrointestinal tract, leading to dehydration and shock
 - a. Under fluoroscopic control, a contrast enema is administered. This usually results in a rapid passage of semiliquid meconium which continues during the next 24 to 48 hours.
 - b. Follow-up KUB films are taken at 12 and 24 hours to evaluate progress. Multiple enemas are frequently required. Mucomyst 5% may be added to help liquefy the meconium.
 - c. Operation is indicated if the enemas fail to relieve the obstruction, if there are calcifications in the abdominal cavity, if the infant appears too ill to delay operation, or if the diagnosis of meconium ileus is in doubt.

MALROTATION

1. **Significance:** **Midgut volvulus is one of the most serious emergencies seen in the neonate or infant, and delay in diagnosis can result in loss of the entire midgut, which is uniformly fatal.** It is a common cause of intestinal obstruction in infants, which must be considered in every infant with bilious vomiting.
2. **Etiology:** Normally the mesentery attaches to the retroperitoneum from the ligament of Treitz to the cecum. However, in malrotation this fixation is shortened and can lead to a volvulus around the SMA and SMV. Sudden onset of bilious emesis is the primary presenting sign. Abdominal distention is common, but frequently is absent. Abdominal tenderness varies. On rectal exam, stool if present, may be guaiac positive.

3. Diagnosis: Plain films of the abdomen show variable findings. Definitive diagnosis requires a contrast study. An upper GI is the preferred study, but a barium enema can also be useful. The classic findings on UGI are an absence of the ligament of Treitz with the duodenum sweeping down the right side of the abdomen, often in a cork screw pattern. A cut off in the duodenum indicates obstruction due to volvulus. With an acute abdomen, emergency laparotomy is indicated.
4. Treatment: A Ladd's procedure is performed to help correct the malrotation. The steps of a Ladd's procedure are unrotation of the volvulus—usually counter clockwise, division of Ladd's bands crossing over the duodenum, separating the leaves of the mesentery, appendectomy and placing the small bowel on the right of the abdomen and the colon on the left. This operation does not correct the malrotation as it is impossible to place the intestine behind the SMA and SMV, but by broadening the mesentery and placing the colon on the left and the small bowel on the right it hopefully causes the intestine to scar into the abdomen and not volvulize.

OMPHALOCELE AND GASTROSCHISIS

1. Distinguishing Features:

<u>Feature</u>	<u>Omphalocele</u>	<u>Gastroschisis</u>
Sac:	present	absent
Umb cord:	arising from sac	lateral to defect
Size of defect:	wide range	usually < 4 cm
Malrotation:	frequent in mod to large size defect	invariably present
Anomalies:	trisomy 13, 18, 21* Prune belly Beckwith-Wiedemann	intestinal atresias

*Down's-omphaloceles often have GI, CV, CNS defects

2. Pre-natal Diagnosis: Elevated AFP, Ultrasound
3. Initial Treatment: Vaginal delivery is well tolerated at full term. Upon delivery, external abdominal organs are protected with warm saline gauze and either plastic wrap or bowel bag.

GASTROSCHISIS:

1. Management:
 - a. Bowel decompression with NGT
 - b. Baseline Hct

- c. OR for primary closure (small defect) or silo placement (large defect) for gradual reduction of abdominal contents. A Bentec silo can be placed at the bedside. This is a spring loaded silo that can be placed under the fascia
- d. Hydration: These babies have a very high fluid loss and often require fluid boluses. Intravenous hydration with balanced salt solution and colloid is essential. Infants should be administered at least 1.5-2.0x's maintenance. Urine output is not a reliable sign to follow as infants may not normally void for the first 12 to 18 hours.
- e. If the fascial opening in the gastroschisis baby is too tight it may cause infarction of the bowel as it distends with air- this may need to be emergently opened.

OMPHALOCELE

- 1. Management:
 - a. Rule out associated anomalies - OR when appropriate for closure (small defect) or delayed closure or escharification
 - b. Hypothermia is usually the immediate life-threatening problem. The baby should be placed in a bowel bag up to the axillae to minimize heat and evaporative losses. The bowel in gastroschisis can become gangrenous with compression or stretch on the blood supply. Every effort must be made to take tension off the mesentery.
 - c. Gastrointestinal decompression by NG tube is imperative to minimize further gastrointestinal distention and prevent aspiration of gastric contents.
 - d. Systemic intravenous antibiotics (ampicillin/gentamicin) are given to protect contaminated amnion and/or viscera. Infection can be a devastating problem if a prosthetic closure is necessary.
 - e. Intravenous hydration with balanced salt solution and colloid is essential. Infants should be administered at least 1.5-2.0x's maintenance. Urine output is not a reliable sign to follow as infants may not normally void for the first 12 to 18 hours.

IMPERFORATE ANUS

- 1. Epidemiology:

Incidence: 1/4000-5000 newborns.
M>F (slightly)
VACTERL Association

- 2. Classification (Peña):

Male Defects

- Cutaneous Fistula

The rectum is located within the sphincter mechanism with its lower part anteriorly mislocated. The fistula may open into the perineum or may continue at a subepithelial level and open along the midline perineal raphe, scrotum, or base of the penis. Diagnosis is made by perineal inspection.

Anal Stenosis

A congenital narrowing of the anal opening which may be associated with a mild anterior mislocation of the anal opening. Ribbon-like meconium is expelled.

- Anal Membrane

A thin membrane is present at the anal site through which meconium can be seen.

- Rectourethral fistula

The most frequent defect in males. The fistula may open into the bulbar (lower) or prostatic (higher) part of the urethra. Just above the fistula, rectum and urethra share a common wall. Lower urethral fistulas are often associated with good quality muscles, well-developed sacrum, and prominent anal dimple. Higher urethral fistulas are often associated with poor quality muscles, abnormally developed sacrum, flat perineum and no anal dimple. Frequently, patients pass meconium through the urethra.

- Rectobladder neck fistula

The rectum opens into the neck of the bladder. Prognosis is poor; the levator, muscle complex, and external sphincter are frequently poorly developed. The sacrum is frequently deformed. This defect accounts for 10% of imperforate anus cases.

- Anorectal Agenesis Without Fistula (Rare)

The rectum ends approximately 2 cm from the perineal skin. Prognosis is good. Muscle and sacral development appear normal. Rectum and urethra are separated by a thin common wall. This anomaly is associated with Down's syndrome.

- Rectal Atresia (Rare)

The rectum ends blindly as does the anal canal. The two are separated by a thin membrane or by dense fibrous tissue. Prognosis is good. Other structures are normal. This defect accounts for 1% of this group of malformations.

Female Defects

- Perineal Fistula (Cutaneous)

See cutaneous fistula described in the male.

- Vestibular Fistula
The most common defect in the female imperforate. The bowel opens to the forchette or the opening surrounding the vagina, external to the hymen. Immediately above the fistula, the rectum and vagina are separated by a thin common wall. The muscles, nerves and sacrum are usually normal and the prognosis is therefore good.
- Anorectal Agenesis Without Fistula
Same as for males. More common in females.
- Persistent Cloaca
A defect in which rectum, vagina, and urethra fuse into a single common channel with a single perineal orifice. The longer the common channel, the more complex the defect, the more involved the repair, and the worse the prognosis.

3. Surgical techniques:

Low Malformations (perineal fistula or vestibular fistula)

Simple anal dilatations may be sufficient. Minimal posterior mobilization (minimal PSARP) to place the fistula within the limits of the external sphincter may be necessary. No colostomy is required.

High Malformations

All anorectal malformations can be corrected by some form of posterior sagittal approach. 10% of male defects require the abdomen to be opened. Colostomy is required—double barrel descending colostomy is preferred, leaving enough length on mucous fistula to pull colon down later during posterior sagittal anorectoplasty. The colostomy is performed after observation for 24 hours to see if a fistula is identified on the perineum or if meconium appears in the urine. Posterior sagittal anorectoplasty (PSARP) is performed at 2 months of age after a distal colostogram is performed with water soluble contrast to identify the level of the fistula.

4. Pre-Operative Evaluation: Essential, as imperforate anus is associated with a number of other congenital malformations. Cardiac malformations are the most potentially life threatening and need to be identified.

- Echocardiogram
- Renal ultrasound
- Sacral spine xrays (usually visible on babygram)
- Physical exam
- Spinal cord ultrasound (up to 6 months) or MRI to r/o tethered cord

5. Postoperative Management:
Anal dilatations start 2 weeks postop and continue with the help of the parents, at home, until the rectum reaches the desired size for age. The colostomy may then be closed (usually about 1 month after the PSARP).
6. Complications:
Functional (diarrhea, constipation, soiling, urinary incontinence)
Anorectal strictures
Recurrent fistula
Anastomotic dehiscence

BILIARY ATRESIA:

1. Description:
Biliary atresia is an obstructive disorder of the liver and bile ducts presenting in early infancy. While “atresia” implies congenital absence of the biliary tree, it is actually a dynamic, progressive condition characterized by inflammation of the bile duct epithelium. This results in obliteration of the normal ductal system, with resulting profound cholestasis, jaundice, progressive cirrhosis and liver failure. Prompt diagnosis and surgical intervention (Kasai hepatoportoenterostomy procedure) is imperative. Successful procedure is markedly reduced in infants older than 10 weeks.
2. Epidemiology:
 - a. Incidence: 1 in 15,000 live births.
 - b. No relationship to physiologic jaundice of the newborn.
 - c. Polysplenia and intestinal malrotation in 15-20% of patients
 - d. Untreated, life expectancy less than 2 years. Most common cause of death from liver disease in children.
 - e. Female > male
3. Clinical Presentation:
 - a. Jaundice presenting slowly at 2-3 weeks of age, often not presenting to PCP until 4 to 8 weeks of life.
 - b. Normal stools progress to acholic (pale or clay colored) c. Progressive hepatosplenomegaly
4. Initial Evaluation:
 - a. Serum Bilirubin: Direct (conjugated) hyperbilirubinemia (TBili = 4 - 8 mg/dL, direct = 150-20% of total)
 - b. Liver Function (AST, ALT, GGT, Alk Phos, Albumin, PT, PTT):
 - c. Other: Hepatitis serology, TORCH titers, alpha 1-antitrypsin studies.
 - d. Abdominal Ultrasound
 - e. Hepato-Iminodiacetic Acid (HIDA) Scan: detects bile flow from liver through biliary tree and into GI tract.

- f. Liver biopsy/ Intraoperative Cholangiogram (definitive)
 - 25% with patent distal common duct to duodenum
 - 75% with entire extrahepatic biliary tree atresia

5. Treatment:

- a. Kasai procedure (hepatopertoenterostomy)
- b. Post-operative medication protocol:
 - Ursodeoxycholic acid, 20 mg/kg/day, divided BID
 - Ranitidine 4 mg/kg/day, divided BID
 - Vitamin E-TPGS, 2.5 IU/kg/day, divided BID
 - Polyvisol, 1 ml/day
 - ADEK 1 ml/day
 - TMP/SMX (greater than 2 months of age) 8 mg/kg/day of TMP and 40 mg/kg/day of SMX, divided BID
 - Nystatin oral suspension, 100,000 units/ml, 1 ml to cheek and tongue QID
 - Prednisone (total duration = 27 weeks = 6 months)
 - 4 mg/kg/day x 2 weeks
 - 3 mg/kg/day x 1 week
 - 2 mg/kg/day x 1 week
 - 1.5 mg/kg/day x 2 weeks
 - 1 mg/kg/day x 3 weeks
 - 0.75 mg/kg/day x 3 weeks
 - 0.50 mg/kg/day x 3 weeks
 - 0.25 mg/kg/day x 3 weeks
 - 0.25/0.125 mg/kg/day (alternate day) x 3 wks
 - 0.25 mg/kg/day every other day x 3 weeks
 - 0.125 mg/kg/day every other day x 3 weeks
 - D/C Prednisone

6. Outcome:

- a. Bile drainage achieved in about 90% of patients who undergo a Kasai procedure at less than 10 weeks of age.
- b. Median survival is 15 years after Kasai (bridge to transplant)
- c. 30-40% become jaundice-free, with optimistic long-term outcome
- d. 30-40% with evidence of hepatic jaundice, may eventually decompensate.
- e. 30% cirrhotic process continues uninterrupted, leading to liver failure.
- f. Bacterial cholangitis is most common complication

CONGENITAL DIAPHRAGMATIC HERNIA:

1. Embryology/pathophysiology:

The diaphragm is derived from four embryologic precursors:

- the septum transversum
- the dorsal mesentery of the esophagus
- the paired pleuroperitoneal membranes
- muscle of the lateral & dorsal body walls

The diaphragm begins to form at 3 weeks gestation. The final step in its formation is the closure of the pleuroperitoneal canals. Failure of the closure of these canals at 8 weeks gestation results in a posterolateral defect in the diaphragm (90% left sided) and a congenital diaphragmatic hernia. At 10-12 weeks gestation, the rotating midgut returns to the abdomen where it can herniate into the chest if a defect in the diaphragm persists. The mass effect of the bowel in the chest results in compression of the developing lung with resultant hypoplasia and pulmonary hypertension. The extent of lung development, degree of pulmonary hypertension and presence of other associated anomalies (most notably cardiac) determines survival.

2. Epidemiology:

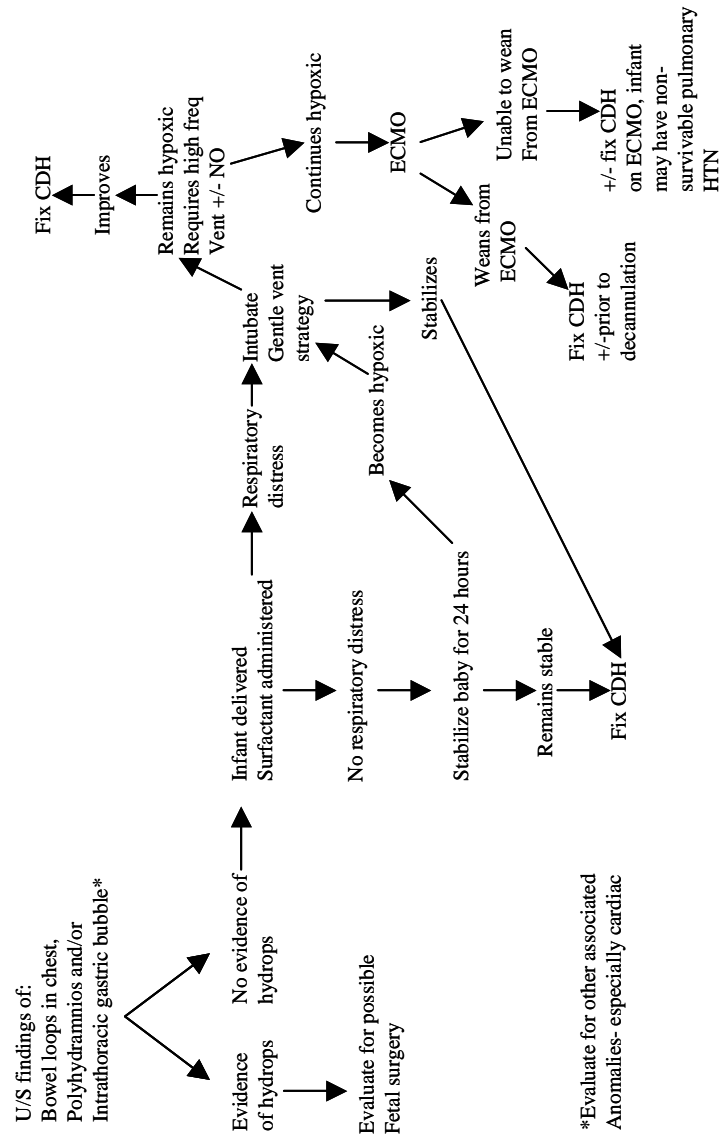
- a. Incidence: 1 in 2500 births
- b. 35-50% stillborn
- c. Mortality >50%
- d. Associated anomalies are common and include major CNS, cardiac, chromosomal abnormalities
- e. Female: male ratio 2:1

3. Diagnosis:

- a. Prenatal ultrasound
- b. After birth: scaphoid abdomen, respiratory insufficiency/cyanosis, decreased breath sounds, decreased bowel sounds, x-rays reveal bowel in the chest with mediastinum shifted and lungs compressed

4. Treatment:

- a. Insert a nasogastric tube.
- b. Do not give positive pressure mask ventilation.
- c. Do not intubate infant's respiratory status is deteriorating.
- d. Use a gentle ventilation strategy that avoid high airway pressures Permissive hypercapnea is acceptable.
- e. Start IV; insert umbilical artery line if possible
- f. If deteriorating respiratory status, intubate and use a gentle ventilation strategy
- g. Maintain systemic pressure greater than pulmonary artery pressure (Dopamine may be indicated).
- h. Watch for pneumothorax on affected as well as contralateral side
- i. Pre-op antibiotics, Ampicillin and Gentamicin



CHAPTER IV: PEDIATRIC FLUID, ELECTROLYTE & NUTRITIONAL MANAGEMENT

The most important concept to understand is why the management of a sick child is different from an adult. The child's body compartment proportions are different and compensatory responses to physiologic derangement are different. For example, a one kilogram premature infant has a blood volume of about 100 cc, so 10 cc drawn for blood work will cause a 10% depletion in blood volume. Careful attention to detail is extremely important and often determines whether the child survives.

The nutrition support team is a consultation service that can be reached to help you in providing the appropriate nutritional care.

On a service covered by many residents, in a hospital where personnel frequently rotate, it is important to have a standardized approach to parenteral fluid therapy so all members of the team can step in easily to make adjustments if necessary. Therefore, the following guidelines are presented.

The first portion of this section details pediatric fluid management and the second part lists general guidelines.

PEDIATRIC F, E&N BASICS:

1. Body Compartments: Total body water, extracellular fluid and total blood volume are all much greater in the small child than in the older child. As a percent of body weight, these values are:

<i>Age</i>	<i>Total body water</i>	<i>Extracellular fluid</i>	<i>Total blood volume</i>
Premature	80%	40%	10%
1 – 10 days	75%	40%	9–10%
1 – 6 months	70%	30%	8%
6 mo – 3 yrs	60%	25%	7%
Adult	60%	20%	5-6%

2. Caloric Requirements: Basal caloric requirements for positive nitrogen balance and growth:

<i>Patient's weight/age</i>	<i>Calorie requirements</i>
Premie	120 kcal/kg
1-10 kg	100 kcal/kg
11-20 kg	1000 cal + 50 kcal/kg over 10 kg
> 20 kg	1500 cal + 20 kcal/kg over 20 kg

These caloric requirements can be further divided as follows:

Requirements	For full term infant	For preemie
Basal	50 kcal/kg	60 kcal/kg
Activity	20 kcal/kg	10 kcal/kg
Excretion	10 kcal/kg	20 kcal/kg
For weight gain	20 kcal/kg	30 kcal/kg
	100 kcal/kg/day	120 kcal/kg/day

An infant recovering from surgery will require about 125 cal/kg/day for good wound healing and positive calorie balance. For the utilization of each 100 calories, 100 cc of water is required.

3. Glucose Metabolism: The liver in the newborn, and especially the premature infant, is relatively immature. The neonate's liver enzymes are often inadequate to utilize fat or protein for gluconeogenesis. Glycogen stores in neonates are only 1/10 of that of an older child per gram of liver tissue. Thus, one may see blood sugars of 40-50 in a stressed neonate. For blood sugar less than 40 and/or symptoms of irritability, hypotension or seizures, treat with 10% glucose. Prolonged hypoglycemia can lead to permanent brain damage. Since the premature infant does not have much fat, there are scanty energy reserves; this must be considered in the post-op period.
4. Protein Metabolism: In an adult, following surgical trauma, there is prompt protein catabolism and loss of nitrogen in the urine. In a post-op child under two years of age, this increased nitrogen excretion does not occur. The protein anabolism needs of the child are so great that even with stress and inadequate nutrition, little nitrogen is excreted. However, without proper nutrition, the child will lose weight and show a steady drop in plasma proteins.
5. Body Temperature: The smaller the neonate, the greater the relative surface area and the less the subcutaneous fat, so heat loss can occur rapidly, especially if the bowel is exposed as in gastroschisis. Severe hypothermia causes a three to fourfold increase in the minute volume of oxygen required with wasting of vital calorie reserves. There is also progressive vasoconstriction with impaired tissue perfusion, anabolic metabolism and systemic acidosis. The infant cannot shiver. Initially, the infant will be restless and then lethargic, with depressed sensorium, respiratory insufficiency, possible aspiration of gastric contents from depressed cough and gag reflex and finally death. Profound hypothermia in the premature infant is also associated with intraventricular hemorrhage.
6. Renal Function: The premature infant has significantly fewer glomeruli at birth and while the full term infant has the full adult

complement, they are less “mature” and the loops of Henle are short. Thus, the glomerular filtration rate may be only one-half that of an adult and in the first 24 hours of life, the baby may only void 30 cc with a maximum urine osmolarity of 600 (vs. maximum of 1200-1400 mosm/L in adult). During the first two to three weeks of life, urine osmolarity roughly matches serum osmolarity. Because of this poor concentrating ability, the neonate must be given adequate water to allow excretion of metabolic wastes. A BUN greater than 15 in a neonate is approaching the dry side and a BUN above 20 should prompt an evaluation for frank dehydration, drug toxicity or primary renal impairment. Renal function is related more to the baby’s postnatal age than gestational age and renal maturation involves mainly a process of tubule elongation with consequent increase in concentration ability. By 14 days, the concentrating ability is near that of an adult and at one month, the GFR is essentially that of an adult. In contrast, ADH and aldosterone both appear to be quite active from birth, so neonates undergoing surgical stress with dehydration will have a drop in urine sodium from aldosterone. Renal considerations influence decisions regarding fluid administration as follows:

- a. The newborn needs adequate hydration to excrete metabolic wastes.
 - b. The newborn has excess extracellular fluid in the first week of life, so maintenance requirements are less.
 - c. The vascular volume is small, so large fluid volumes should be given judiciously.
 - d. Because of limited ability to excrete sodium in the newborn period, sodium loads should be limited.
 - e. A rough guide of adequate urine output is 1 cc/kg/hr.
7. Potassium Therapy: In the normal newborn, very little potassium is excreted in the first few days of life and the potassium remains stable. With surgery, there is increased aldosterone release due to stress with increased potassium excretion by the kidneys. Postoperatively, there may be other sources of potassium loss such as diarrhea, high fecal loss with a colostomy, excess gastric loss with secondary alkalosis or increased excretion due to Lasix. Therefore, as long as renal function is adequate, potassium should be included in fluids in neonates (1 mEq/kg/hr is a safe dose in an emergency).
8. Calcium and Magnesium Therapy: The neonate is extremely susceptible to hypocalcemia because of immaturity of renal and parathyroid regulation and a small bone pool of available calcium. The normal newborn should have a serum calcium of about 10 mg% (3.4 mg% protein bound and 6.6 mg% diffusible of which 5.2 mg% is ionized). A stressed neonate can drop the calcium level to as low as 5 mg% with symptoms of jitteriness, irritability, frank seizures

and impaired cardiac output. Therefore, all sick neonates should receive routine calcium. There is poor correlation between serum levels and clinical manifestations (neuromuscular irritability – jitteriness to seizures).

Acute symptoms: Calcium gluconate 10% (100 mg/cc)
dose 30 mg/kg; give 0.3 cc/kg slowly, with EKG monitoring

Magnesium deficits are associated with debilitating disease, large ileostomy or fecal fistula losses, proximal jejunal resection, or with prolonged diarrhea or biliary losses. Maintenance support can be provided with 2 mEq MgSO₄ per 500 cc IV fluids.

9. Dynamics of Fluid Management: An important goal in optimal fluid management is to maintain an adequate intravascular volume in spite of fluid shifts to the extracellular fluid space, “third space” sequestration and external losses. The end result is optimal tissue perfusion, indicated by adequate urine output. Diminished tissue perfusion may result from hypothermia, dehydration, hypovolemia, or hemoconcentration. It leads to anaerobic metabolism and lactic acidosis. A loss of vascular volume initiates compensatory mechanisms, among which are stimulation of volume receptors that cause secretion of ADH in response to volume depletion. If volume is restored with a hypotonic solution, there is dilution of serum electrolytes and osmolality. Ordinarily such a dilution would activate osmoreceptors which would shut off ADH. In the presence of hypovolemia, however, ADH continues to be secreted and dilute urine cannot be excreted, so an osmolar gradient remains between the vascular space and ECF.

For example, an infant reaching the recovery room reasonably well hydrated after a two hour operation for intestinal obstruction may have a calculated third space loss of 50% of maintenance. If the fluid replacement for this is too dilute, osmoreceptors may perceive a hypotonic state and ADH, a valuable means of protecting the vascular volume, will be diminished. Furthermore, an osmotic gradient will quickly pull the hypotonic fluid into the ECF. A few hours later, hypovolemia may develop and ADH may increase in response, resulting in free water conservation and even greater hypotonicity. Therefore, for infants, the third space fluid needs should be replaced with D₅1/2 NS or NS on top of D₅1/2 NS maintenance. Careful attention to strict I/O daily weights, urine specific gravity, skin turgor, fontanelle fullness and mucous membrane moisture will help detect fluid deficiency.

10. Special Consideration for Cardiac, Hepatic and Renal Disease:

For patients with severe cardiac disease, iatrogenic cardiac failure, or severe septic stress, one should consider digitalization and diuresis only if directed by cardiology. Renal and hepatic failures require

special caution. With renal shutdown, maintenance fluid volume should be reduced. With hepatic failure, there are inadequate plasma proteins and poor aldosterone metabolism, so sodium should be restricted and albumin used in IV fluids.

11. Acidosis: In the neonate, pulmonary and renal mechanisms for correcting acidosis are much less efficient than in the adult. The pO_2 is normally low and may drop with stress resulting in the generation of large quantities of lactic and pyruvic acids through anaerobic metabolism (metabolic acidosis). If there are pulmonary difficulties, CO_2 may be retained with additional respiratory acidosis. The diameter of infant bronchi is obviously considerably smaller than those of adults. They are particularly susceptible to airway obstruction from relatively trivial amounts of edema or retained secretions.

PEDIATRIC FE&N GUIDELINES/CALCULATIONS:

1. Maintenance Fluid: **4-2-1 rule**; Intravenous – based on weight

<u>Weight (kg)</u>	<u>ml/kg/hr</u>
0 – 10	4 ml
11-20	40 ml + 2 ml/kg over 10 kg
> 20	60 ml + 1 ml/kg over 20 kg

(For example: 24kg patient : $(10 \times 4) + (10 \times 2) + (4 \times 1) = 64$ ml /hr)

***Note:** Neonates in first week of life require less fluid

<u>Day of life</u>	<u>ml/kg/24 hr</u>
1 and 2	65
3 – 7	85
8 and over	100

Electrolytes

Na + Cl: 3 mEq/kg/day
 K: 2 mEq/kg/day

Dextrose: 5% dextrose solution at maintenance rate is usually sufficient to prevent ketosis. 10% dextrose may be necessary to prevent hypoglycemia in neonates, particularly SGA babies, and infants of diabetic or toxemic mothers. Dextrose containing fluid should be continued during infusion of blood products, at reduced rates, to avoid volume overload (4-6 ml/hr).

Maintenance: D₅1/2 NS with 20 mEq KCL/liter. Add 1 unit of heparin per cc for neonates with central lines. Another option is to

write for D10 #48- this is an electrolyte solution that is specifically made for newborns.

2. Post-op Fluid:

D₅ ½NS with 20 mEq KCl/liter is given at 1x maintenance on the operative day and POD 1 if additional fluid is necessary to cover third space losses after abdominal or chest cases then ½ times maintenance of NS can be added on top of this.

3. Dehydration:

- Severity:

<u>feature</u>	<u>mild</u>	<u>moderate</u>	<u>severe</u>
% weight loss	5	10	15
Behavior	irritable	extremely irritable	lethargic
Skin turgor	↓	↓↓	↓↓↓
Skin color	pale	pale to grey	grey to mottled
Pulse	normal to ↑	↑↑	↑↑↑
Blood pressure	normal	normal	↓
Urine output	↓	↓↓	oliguria
Urine s.g.	>1.010	>1.020	>1.030
BUN	↑	↑↑	↑↑
Hematocrit	no change	↑↑	↑↑

- Type of dehydration – determined by serum Na:

- | <u>type</u> | <u>repletion</u> |
|-----------------------------|---|
| a. isotonic | balanced salt solution – Ringer’s Lactate |
| b. hypotonic
(Na <125) | <ol style="list-style-type: none"> 1. Rule out dilutional hyponatremia. 2. Replete ECF deficit with balanced salt solution. 3. Correct Na deficit after notifying Chief Resident. <p><u>Symptomatic hyponatremia:</u> (less than 120)
Give 3% NaCl – 0.5 mEq/cc, 12 cc per kg over 1-4 hrs as single dose (will raise Na by 10 mEq/cc). If seizing, give 1 cc/min until seizures stop. Treat in ICU.</p> <p><u>Severe hyponatremia:</u> Calculate Na deficit: (130 – Na) x 0.6 weight in kg. Add this amount of Na as 3% NaCl to 8 hours of regular maintenance fluid. Treat in ICU.</p> |
| c. hypertonic
(Na > 150) | <ol style="list-style-type: none"> 1. Slow administration of hypotonic saline, at 0.5 to 0.75 maintenance, correction over 48 h and maintenance over 48 h to |

avoid rapid fluid shifts and CNS effects.

2. Monitor BP – if hypertensive, decrease rate

3. Correction of free water deficit:

Volume free H₂O=

$(\text{Serum Na} - 140) \times 0.6 \text{ wt in kg} / 140$

Na < 160, replace over 24 hrs

Na = 160 – 180, 2 – 3 days

Na > 180, 3 – 4 days

4. Resuscitation in severe dehydration: – (treat in ICU)

1. Use balanced salt solution

2. Start with ¼ of daily maintenance volume over 30 minutes or 20 cc/kg bolus of NS or Ringer's Lactate

3. Repeat if response is absent or poor

4. Follow with ½ NS or Ringer's Lactate at 2 x maintenance

5. Glucose and lytes q2-4 hours

6. Dextrose-containing fluid as background maintenance – do not use dextrose in resuscitation boluses

7. Do not add KCl until urine output is established > 1 ml/kg/hr.

5. Acid/Base Disturbances:

Acidosis: Na bicarb – full strength = 1 mEq/ml;

Use ½ strength in neonates to avoid hyperosmolarity.

- Emergency 2-4 mEq/kg
- mEq bicarb = base deficit x wt in kg x

0.5 for newborns
0.4 for infants
0.3 for children

a. **Initial dose always limited** – 50% of calculated dose over 3-4 h, then reevaluate.

b. Do not treat pH > 7.35 or base deficit < -5 and remember to resuscitate the patient with fluids, as this will help to reverse the acidosis more physiologically than covering up hypovolemia with bicarb.

c. **USE ½ STRENGTH BICARB IN NEONATES** (2-4 cc/kg)

6. Replacement of Losses– Fluids and Electrolytes

Gastric: D₅ ½ NS with 20 mEq KCl/liter, ½ ml for 1 ml gastric output replaced q4-8h

Third space: Increase maintenance rate to 1.5 to 2.0 x maintenance, NS postop and POD 1. Decrease rate to maintenance and tonicity to D₅ 1/2 NS when postop diuresis begins.

Ileostomy: Early postoperative period: ½ NS with 20 mEq KCl/liter, replaced ml/ml until IV's are stopped. Oral NaCl supplementation may be required if

hyponatremia occurs. Spot check urine sodium whenever output exceeds ability to replace losses by PO intake. A low urine sodium even in the face of a normal serum sodium will inhibit normal growth of the infant. Oral Na bicarb may be necessary if hyperchloremic acidosis occurs.

Diarrhea: (For example, short gut patients): estimate daily stool loss and replace with LR with 20 mEq KCl/liter.

Pyloric Stenosis: Resuscitate with NS or LR (20 ml/kg bolus over 1 hour); repeat until urine output is established if severe dehydration is present.. Otherwise start D₅ ½ NS with 20 mEq KCl per liter at 1x maintenance and NS at ½x maintenance in addition. A 20 ml/kg bolus may be required. Goals are: normal Na; K greater than 3.5; Cl greater than 90; bicarb less than 30; and a normal or high urine pH. Paradoxical aciduria in the face of a metabolic alkalosis is an indication of continued dehydration and hypokalemia.

Bile, pancreatic, or enteric fistula: Accurate volume measurement and electrolyte replacement required

Body Fluid Compositions - If in doubt, send fluid for electrolytes

<u>Source</u>	<u>Na</u>	<u>K</u>	<u>Cl</u>	<u>HCO₃</u>	<u>pH</u>	<u>Osm</u>
Gastric	70	5-15	120	0	1	300
Pancreas	140	5	50-100	100	9	300
Bile	130	5	100	40	8	300
Ileostomy	130	15-20	120	25-30	8	300
Diarrhea	50	35	40	50	Alk	Varies

7. Potassium Imbalance:

Hypokalemia: in maintenance fluids, 1 mEq per 50 cc (20 mEq per liter). Or can give 0.5mEq/kg IV over 1-2 hours- max 1mEq/kg/hr with EKG monitoring.

Hyperkalemia: Obtain EKG – check T waves, QRS interval. If changes are present, monitored bed required.

1. Calcium gluconate 30 mg/kg
2. Na bicarbonate
3. Glucose-insulin infusion: glucose 1.0 gm per kg as D25; regular insulin, 0.3 units per kg, IV
4. Kayexalate: 0.5 to 1.0 gm per kg in 10 percent sorbitol po or pr
5. Dialysis

Useful Numbers

Insensible Losses

0-3 years	1 ml/kg/hr
4-9 years	0.8 ml/kg/hr
> 10 years	0.5 ml/kg/hr
Blood volume	80 cc/kg

BLOOD TRANSFUSIONS:

- Use 10 cc/kg of packed red blood cells over 3 hours as a standard 1 unit transfusion to correct anemia, pre and post-op.
- Platelet transfusions – 5 cc/kg over 3 hours, usually for platelet count < 50,000.
- Estimated Blood Volume
 - a. Newborn: 90 cc/kg
 - b. Child: 80 cc/kg
 - c. Adult 70 cc/kg
 - d. To estimate the volume of packed RBC's to raise HCT from A to B:
$$\text{Volume} = \text{EBV} \times \frac{(\text{HctB} - \text{HctA})}{\text{HctPRBC}} = 80 \text{cc/kg} \times \text{wt (kg)} \times \frac{\text{HctB} - \text{HctA}}{70}$$
$$= 1.1 \times \text{wt} \times (\text{HctB} - \text{HctA})$$
 - e. Rough rules of thumb for blood replacement:
 1. 10 cc/kg of packed RBC's – Raise Hct 3-4%
 2. 0.1 unit/kg of platelets – Raise platelet count by 25,000
 - f. Safe volumes to push empirically:
 1. Packed RBC's 10 cc/kg
 2. 5% albumin 20 cc/kg
 3. Salt poor albumin (25%) 4 cc/kg

URINE OUTPUT:

- a. No ideal urine output (e.g. 1 ml/kg/hr in post surgical neonate) since osmolar load varies widely. However, most children should make 1cc/kg/hr
- b. Accurate assessment requires analysis of both urine concentration, and volume.

NUTRITION – PARENTERAL

Please refer to pediatric TPN form and this manual for ordering TPN. This is not easy, so be patient and work with nutritionist and TPN pharmacist.

A. Dextrose

Concentration: 25% maximum if catheter tip is in distal superior vena cava or right atrium

15% maximum if catheter tip is in superior vena cava, distal internal jugular or subclavian vein
 12.5% peripherally

Advancement: **gradual advancement of concentration to avoid hyperglycemia**

<u>Day</u>	<u>Premature/Newborns</u>	<u>Infant/Children</u>	<u>Older</u>
1	D 10	D 10	D 10
2	D 10	D 12.5	D 15
3	D 12.5	D 15	D 20
4	D 14	D 17.5 (if necessary)	
5-6	D 15.5	D 20	
7-8	D 17.5 (if necessary)		
9-10	D20		

Do not advance if patient has glycosuria or is hyperglycemic.
 Do not add insulin to the HAL. If the patient is hyperglycemic, a sepsis work-up should be considered.

B. Protein

<u>Requirements:</u>	<u>gm/kg/d</u>
Newborns	2.0 - 3.5
Infants	2.0 - 3.0
Older Children	1.5 - 2.0

150-200 nonprotein calories required per gm Nitrogen.
 To supply 2 g/kg/d protein, 48-64 cal/kg/d nonN cal given.
 To supply 2.5 g/kg/d protein, 60-70 cal/kg/d nonN cal given.

Advancement:

Newborns: start with 1.5% amino acids; advance by .5% QD until reach goal.

Older infants & children: start with 1.5% amino acids, advance by 1.0% QD to goal.

C. Lipid

Requirements

20% Intralipid: start with 1 gm/kg/d (minimum)

<u>maximum</u>	<u>gm/kg/d</u>
Premature/Newborns	3.5
Infants	3
Children	3

Should never account for more than 60 percent of total calories.

Requirement to prevent essential fatty acid deficiency: 0.5 – 1.0 gm/kg/d

Advancement:

Start with 1.0g/kg/day and advance 1g/kg/day to goal

Contraindications:

1. Hyperlipidemia
2. Total bilirubin greater than half of exchange level prevents use of Intralipid in 1st week in most premature infants.
3. Not required if baby is taking formula
4. Theoretic:
 - a. Severe acute respiratory failure, FiO₂ = 100%
 - b. Severe uncontrolled sepsis
 - c. Severe pancreatitis

Postoperative routine for patients previously on HAL:

1. HAL decreased in rate to 0.5 maintenance
2. Postoperative stock to supply remainder of maintenance fluid

Monitoring:

1. Daily weights
2. Daily totals of fluid and calories
3. Specific gravity and urine for sugar and acetone until concentration goal is reached, then spot check daily
4. Dextrostix daily or if glycosuria occurs
5. Lytes, BUN, glucose, CBC daily until concentration and maintenance goals are reached, then weekly
6. Bilirubin, hepatic enzymes, albumin and transferrin or prealbumin weekly when stable regimen reached
7. Serum check for lipemia in babies receiving Intralipid; check serum triglyceride and cholesterol levels initially and after any dosage alteration.
8. Septic work-up if glycosuria occurs on a previously stable regimen
9. CXR every 14 days to check line placement

NUTRITION-ENTERAL:

1. Infant Diets:

Breast milk is preferred for babies 0-6 months. Formulas simulating breast milk (lactose and milk protein) are used if infant is not breast fed, e.g. Enfamil or Similac. If cow's milk or lactose is a problem, soy protein and corn syrup solids are substituted (Isomil or Prosobee). Formulas using partially demineralized whey (SMS, PM 60/40) have lower mineral (Na, K, PO₄) levels which may benefit prematures and infants with renal or cardiovascular conditions. Infants with malabsorption or short gut require predigested formulas; predigested formulas with high osmolarity such as Pregestimil require close monitoring.

2. Caloric Density:

Infant formulas usually contain 20 kcal/30cc (0.67 kcal/cc). Most pediatric formulas i.e. Pediasure are 1kcal/cc. Caloric maintenance roughly parallels fluid maintenance, that is, 100 cal per kg per day for the 1st 10 kg. To give caloric maintenance requires 150cc of full strength formula, so enteral fluid maintenance is expressed as 150 cc/kg/day for the 1st 10 kg (6cc/kg/hr). Low birth weight and sick infants may tolerate only dilute formula and may have a fluid limitation. The total calories they receive may fall short of requirements and must be calculated periodically. Either peripheral or central parenteral nutrition may be indicated. Formulas with higher caloric density are available (24, 27 and 30 kcal per 30 cc) but have correspondingly higher osmolar loads. Changeover to these formulas should be gradual (e.g. by 2 Kcal increments). Microlipids (4.5 cal/cc) may be added, 1 cc/ounce, for extra calories.

Caloric Density of Commonly Used Nutrients

<u>Enteral</u>	<u>kcal/ml</u>
20 cal/30ml formula	0.67
24 cal	0.80
27 cal	0.90
30cal	1.00

3. Daily Requirements:

<u>Age</u>	<u>Calories</u>	<u>Protein</u>
LBW	kg x 110-150	
0-6 mo	kg x 120	kg x 2.2
6 mo-1 yr	kg x 110	kg x 2.0
1-3 yr	1300	23 grams
4-6 yr	1800	30 grams
7-10 yr	2400	36 grams

4. Illness Requirements:

Increased requirements of caloric replacement during illness:

<u>Condition</u>	<u>Percent increase</u>
Severe injury or illness	25 - 100%
Previous injury	50 - 75%
Sepsis or burn	100 -150%
Fever per degree C	12%
Cardiac failure	5 - 25%
Surgical procedure	10 - 25%
Major Surgery	20 - 30%
Long term growth failure	50 - 100%
Protein calorie malnutrition	50 - 100%

5. Assessment and Advancing Feedings:

- Weight gain goal: 10-15 mg/kg/day (~10% body weight)
- Do not change formula strength and volume at the same time. This is particularly important in malabsorption and adaptation to short gut. Patients with feeding or absorption difficulties require parenteral nutrition at maintenance volumes, in addition to enteral feeds. Start feeding with 4 cc/kg sterile water or Pedialyte. The concentration of formula is gradually advanced first, starting at 1/4 strength and increasing by 1/8 or 1/4 strength steps every 1 to 3 days or more, guided by the infant's response (water loss stool, stool volume, presence of reducing substances, weight gain). Volume is advanced once concentration approaches full strength (3/4 to full strength). Five to ten cc are added per day every 1 or more days, guided by the infant's response. When enteral feedings comprise half of maintenance requirements, (75 ml/kg/day for infants), parenteral fluid volume is decreased to avoid fluid overload. Infants on enteral feedings do not require parenteral Intralipid.
- For infant gavage feeding, a 5 or 8 French nasogastric tube is usually used. Larger infants and toddlers can use slightly larger tubes; Polyurethane tubes can be used in older children.

CHAPTER V: CLINICAL PROBLEMS

RESPIRATORY DISTRESS:

1. Initial management of respiratory distress or arrest is to ventilate the child using a mask, AMBU bag, and 100% oxygen. Virtually any degree of airway obstruction can be overcome with positive pressure ventilation. If airway obstruction is not the problem, then be careful when ventilating a baby to avoid too much pressure. Pneumothorax is easy to produce as pressure can exceed the presumably safe pressure of the “pop-off” valve. If you need assistance STAT PAGE Anesthesia and Respiratory Therapy.
 - a. Remove any secretions or other material from the patient’s mouth, nose and pharynx.
 - b. Have a proper mask fit and a hard surface behind the patient’s head.
 - c. Make certain that the tongue is not occluding the airway, either by placing an appropriate sized airway and/or by holding the mandible forward.
 - d. Observe and listen to the chest to confirm adequate ventilation.

2. Guidelines for Endotracheal Intubation
Use a face mask, airway, and AMBU bag until the resuscitation is well under control. Be ready before you attempt intubation there is usually no rush; ventilation and oxygenation come first. Ideally, you should have available: suction; suction catheters (12 or 14 gauge); proper type of bag/valve; selection of tubes; stylet if necessary; McGill forceps, if nasal tube is planned; airway manometer; benzoin; tape; and drugs.

Correct Head Position - “Sniffing Position” is best for both ventilation with bag-and-mask and for intubation. This is accomplished by FLEXION OF THE NECK and EXTENSION OF THE HEAD. Care should be taken to avoid c-spine injuries and in-line stabilization should be performed on all trauma patients until their c-spines have been cleared.

Drugs - Atropine 0.02 mg/kg, succinylcholine 1-2 mg/kg, thiopental 4 mg/kg

Insert the laryngoscope slightly to the right and use it to displace the tongue to the left. Lift up the epiglottis with the tip of the laryngoscope blade so that you can see the entire laryngeal vault. Do not traumatize the epiglottis. Introduce the endotracheal tube via the right corner of the mouth along the blade (but not through the groove of the blade) so that you have an unobscured view.

SEE THE CORDS - Don't pass the endotracheal tube blindly since this may result in esophageal perforation or perforation of the pyriform sinus.

DO NOT FORCE THE TUBE - Use a smaller one if necessary. Do not push the tube too far to avoid right mainstem bronchus intubation.

Examine the chest - look and listen as you ventilate.

AFTER INTUBATION - Ventilate with oxygen and suction via the tube. Apply benzoin to the face and tape the tube securely. Obtain CXR to confirm appropriate position of the tube.

The best way to learn intubation is in the operating room under the eye of a skilled anesthesiologist, prior to an elective operation.

3. Laryngoscope

If patient is less than 2 years of age, use a straight blade (Miller) due to anterior location of larynx and floppy epiglottis: Size (0 premature); 1 (infant); 2 (child)

NOTE THAT CURVED BLADES DO NOT FLATTEN OUT THE CURVATURE OF THE EPIGLOTTIS WELL IN INFANTS, SO USE A STRAIGHT BLADE IN THE INFANT

Age greater than 5 years, can use curved MacIntosh blade.

Ages between 2 and 5 years, either a Miller or a MacIntosh blade.

4. Endotracheal Tube

Cuffed tubes are not used in children, nor are tight fits desired (the correct fit should allow a leak at a 20mmHg inspiratory pressure). The narrowest part of the airway in a child is the subglottic area.

Note that the distance from the glottis to the carina in infants is approximately 4 cm.

“Blind” nasotracheal intubations are usually not done in older children due to the risk of bleeding from the large adenoids.

Sizes: “Rule of Thumb” is ETT size = $\text{age}/4 + 4$. Note that often one must use tubes $\frac{1}{2}$ mm smaller in ID than that calculated by this rule. Another rapid rule is that the size of the patient's little finger is equal to the size of the tracheal lumen.

A more precise guide is:

<u>Age</u>	<u>Size</u>
Preemie	2.5 - 3.0 mm ID
Newborn	3.5 mm
3-12 months	4.0 mm
1 - 2 years	4.5 mm
> 2 years	4.5 plus age (yrs)/4

Tube length (cm- tip to lip) Age (yrs) / 2 **plus** 12

5. Typical Ventilator Settings
 - a. Tidal volume 7-10 cc/kg
 - b. Pressure 25/5 cm H₂O
 - c. Rate=30 (titrate)
 - d. FiO₂ = 100% (titrate). Adjust to keep pO₂ 60-80 mm
 - e. PEEP at least 2 cm H₂O (“physiologic PEEP”)
 - f. Always humidify gases
6. Extubation in ICU
 - a. Suction mouth, nose, stomach with tube
 - b. Ventilate 2-5 minutes with high FiO₂
 - c. Extubate at maximum inspiration
 - d. High oxygen face mask
 - e. NPO x 4 hours
 - f. Racemic epinephrine for stridor or elevated pCO₂ if secondary to laryngeal edema
7. Apnea in preemies, neonates, and infants less than 1 year
 - Rule out: Hypoglycemia
Dehydration
Hypocalcemia
Temperature fluctuations
Sepsis
Brain lesions
 - Rx: Treat specific causes
Increased stimulation
Theophylline (po or IV) loading dose 5 mg/kg followed by maintenance 1 mg/kg/dose (q4-8h)
Mechanical ventilation if necessary

VASCULAR ACCESS:

1. Indications for Central Venous Catheters
 - a. Patients with chronic disease requiring frequent antibiotics, TPN, chemotherapy, or transfusions.
 - b. Patients who need IV access for hydration, antibiotics, TPN who do not have an alternate source of access.
2. Which type of line to use:
 - a. A double lumen Broviac/Hickman is needed for some oncology patients. Please ask if they are on a protocol that requires a double lumen catheter. Otherwise, a single lumen catheter is used.
 - b. A port should be used when the chemotherapy is intermittent, the family desires that the child be able to swim or when the ability of the caregivers to maintain a Broviac is questionable and the patient has suitable body habitus.
3. Options for Pediatric Access:

Peripheral veins:	antecubital Greater saphenous at ankle Umbilical vein in newborn
Antecubital PICC	
Tunneled catheter (Broviac or Portacath)	External jugular vein Internal jugular Subclavian Cephalic vein Facial vein Internal jugular vein (preemie) Saphenous vein (groin)
4. Catheter Care:
 - a. Place a Tegaderm dressing over the exit site of all central access devices. This should be changed according to the protocol of the institution. An order must be written to change dressing according to protocol.
 - b. Tape a loop of catheter onto the chest to help prevent inadvertent removal of the catheter.
 - c. **Ask the oncology service if they would like to have the port accessed after it is placed in the OR.** If it is going to be a while before it is used it should be flushed with heparin in the OR and the huber needle is removed
 - d. Prior to leaving the OR, the Broviac/Hickman or port should be flushed with approximately 1cc of 100 units/cc of heparin- less volume if it is a smaller catheter and more if it is a dialysis catheter or leave fluids infusing into the Broviac catheter when the patient leaves the OR. The

catheter will not need to be flushed with heparin as long as a continuous infusion is running. If the infusion is stopped, the catheter must be flushed with heparin flush (100 units/cc) 3 cc bid.

- e. **Always use a Huber needle when accessing a port- a standard needle will cut the silicone membrane and make the port leak.**

5. Nonfunctioning catheter:

(Note: Doernbecher IV team- pager # 17213- available to treat occlusions, repair lines and place/remove PICCS))

- a. Broviac/Hickman -

Can you obtain a blood return?

Does it flush easily? **Do not flush a catheter with any syringe that is smaller than 10cc's as the smaller syringes generate too high of a pressure and will cause the line to burst**

Is there extravasation, pain, or swelling with infusion/injection? Start with an x-ray to make sure the position of the catheter has not changed.

If there is evidence of occlusion, try TPA - see protocol below. If there is pain, swelling, or extravasation obtain a contrast study through the catheter. If the catheter is obviously cracked external to the skin, repair kits are available. Catheters should be repaired under sterile conditions following the instructions in the repair kit. You will need to know the size and manufacturer of the catheter to get the appropriate repair kit. The catheter will not be able to be used for 8-12 hours after repairing it so alternate IV access may be needed.

- b. Port

Is there pain or swelling with injection/infusion?

Are you able to withdraw blood?

Can you flush the port easily?

Most problems with ports are due to dislodgement of the Huber needle. A CXR should be obtained to make sure the tubing has not become dislodged and then the port should be re-accessed under sterile conditions using a new Huber needle. EMLA cream will make access less painful. It takes about 20 minutes for this cream to work. If the port still has no blood return and will not flush and you are **SURE** you are in the port properly, TPA may be used per protocol for Broviac catheters (this should only be done after a senior person has checked the needle placement).

6. Line Thrombosis:

- a. TPA will come from the pharmacy as 2mg of powder that

needs to be reconstituted in NS, or it will already be reconstituted. Inject using a 10cc syringe- smaller syringes will create too much pressure and can easily rupture the line. The TPA should be infused into the catheter as best as possible and left for 2 hours. After 2 hours it should be withdrawn from the catheter and the catheter flushed with heparin. **See attached protocol for 3-way stop cock technique for de-clotting lines, found in the appendix at the end of this handbook.**

- b. If aspiration of clot is not possible in one hour, repeat injection and attempt aspiration 8-12 hours later.
- c. If the catheter fails to clear after two bolus injections of TPA the line may be permanently clotted and may need to be replaced.
- d. Can add 1 unit heparin/cc TPN or IV solution to aid in preventing recurrent catheter thrombosis.

7. Catheter insertion site infection:

- a. Local infection of a CVL is defined as purulent drainage from the exit site. Tenderness or erythema along the catheter tunnel also suggests an infection.
- b. Clinical examination usually establishes the diagnosis. Cultures should be obtained from the catheter site along with peripheral and central blood cultures via the CVL. Drainage should always be gram stained.
- c. Treat with local wound care: warm soaks TID and proper dressing changes. Systemic care: empiric antibiotic coverage before cultures are back. Specific antibiotic coverage with culture results.
- d. Indications for catheter removal: continued local infection after 48 to 72 hours of specific antibiotic coverage. Development of systemic infection that will not clear per systemic infection protocol below.

8. Systemic catheter infections:

- a. Systemic catheter infection is defined as septicemia resulting from infection of a CVL. While the definition is straightforward, the diagnosis is often difficult to make. Frequently, children with central venous lines are neutropenic or have other maladies which put them at high risk for other infections. Distinguishing catheter-related sepsis from infection elsewhere can be somewhat difficult.
- b. Catheter-related sepsis is a diagnosis of exclusion. When a patient with a CVL develops a fever, all other possible causes of sepsis should be ruled out before the diagnosis of catheter sepsis is made. Candida antigen may be

helpful in cases of persistent fever without an obvious source.

- c. When catheter sepsis is suspected in a patient, at least one, and preferably two blood cultures should be obtained via venipuncture. One or two blood cultures via the line should also be obtained. The diagnosis of catheter sepsis is made if the peripheral cultures are negative and the line culture is positive.
- d. Don't forget to take down any TPN that is infusing and culture it as well.
- e. Broad-spectrum antibiotics are begun after cultures are taken. Narrow antibiotics as dictated by cultures.
- f. Indications for catheter removal: Ongoing sepsis despite appropriate coverage for 2-4 days. Continuously positive blood cultures after 2-4 days of antibiotic coverage. Positive culture for Candida.
- g. Catheter removal: Most of our catheters are removed in the OR under anesthesia. Certain attendings will remove the catheters at the bed side. Check with the chief resident or attending so that a plan for removal can be formulated and communicated to the family. If the catheter is to be removed at the bedside (treatment room) apply Emla cream for at least 20 minutes prior to removal. Remove the Emla cream and prep with Betadine and towel out area. Use lidocaine local over the cuff site. You may have to cut down over the cuff if it is far from the skin exit site. Send tip for culture.
- h. Port infections: Never I&D these. You may aspirate a pocket for culture. These do need to be removed in the OR and the whole device sent for culture.

Broviac/Hickman Catheter Sizes

Catheter Size	Age	Internal Diameter	Catheter Number	Repair Kit Number	Priming Volume
2.7	Preemie	0.5	60004	60160	.15ml
4.2	Infant	0.7	60006	60161	.3ml
6.6	Child	1.0	60012	60162	.7ml
9.6	Adult	1.6	60016	60163	1.8ml
7.0	Double Lumen		60031	60176*	
	White leg	0.8		60168	.6ml

Catheter Size	Age	Internal Diameter	Catheter Number	Repair Kit Number	Priming Volume
	Red leg	1.0		60169	.8ml
9.0	Double Lumen		60032	60170*	
	White leg	0.7		60168	.6ml
	Red leg	1.3		60169	.8ml

CHEST TUBE PLACEMENT:

1. Site: For pneumothorax, a chest tube may be placed in the anterior or anterolateral aspect of the chest wall. For pleural effusion or hemothorax, the tube should be placed posterolaterally, in the dependent position for drainage.
2. Tubes: For simple pneumothorax a Fuhrman catheter (percutaneous pig-tail) can be used. For effusions and hemothorax a straight, plastic intercostal catheters (e.g. Argyle chest tubes) are the best type to place. For an infant, a size 10-12 French is usually used. Larger tubes come in increments of 4 French (e.g. 16,20,24, 28 Fr etc).
3. Technique:
For Fuhrman catheter placement, the patient should be prepped and draped in the normal sterile fashion. Conscious sedation may be used and local anesthetic should be infused. A needle is placed in the mid-clavicular line- 2-3rd rib space or in the mid-axillary line 6-7th rib space. The needle is advanced until there is a return or air. The wire is then fed into the chest and the needle is removed. The dilator is placed over the wire to dilate the tract and then the pig-tail is inserted to the hub. The catheter is then connected to a Pleura-vac.

For an Argyle-chest tube, the site is prepped, draped and locally anesthetized with 1% Xylocaine. Estimate the proper intrathoracic length of the tube and mark the tube at that point. A small skin incision is made at a selected site over the ribs. A curved hemostat is used to tunnel superiorly and subcutaneously over the next higher rib and into the pleural cavity, the hemostat is then spread to enlarge the opening. **TROCARS ARE TO BE AVOIDED.** It is important to displace the site of entry from the skin margin by tunneling so that pneumothorax will not recur when the chest tube is removed, especially in babies with small amounts of subcutaneous tissue. In infants, care must be taken to avoid the nipple area as it can easily

be damaged by a chest tube placed to close to the nipple. Introduce the tube into the chest cavity and make certain that all sideholes are intrathoracic. Then, connect the tube to Pleura-vac suction; the amount to be determined by the indication for the tube and the size of the child.

4. Removal:

If there is no air leak, the tube is usually placed to waterseal and a chest x-ray is subsequently checked. Do not clamp chest tubes. The tube should be removed quickly after appropriate pain medication has been given. The site should be dressed with a small piece of xeroform gauze covered by a 2X2 covered with an appropriately sized Tegaderm. A large compression dressing with Elastoplast should not be used as removal is very traumatic to the skin in little children. A repeat chest x-ray is obtained. If the child is cooperative, the tube can be removed either at end inspiration or while the child performs a Valsalva maneuver. For younger children, pull the tube while the child cries.

GASTROSTOMY TUBES:

1. Balloon catheters and Malecot catheters are both used. Generally balloon catheters are used and require 3-5ml water in the balloon. In general, fill to 5 ml, except for neonates.
2. Post-operatively, a Malecot should be well taped to abdomen with retention stitch at skin level. A Hollister Drain Tube Attachment device is recommended prior to discharge, with parent teaching on how to apply. This will usually be changed to a MICKEY low profile tube after 4 weeks.
3. Laparoscopically placed MIC-KEY tubes are secured with 2 through and through sutures until day of discharge (POD #2-3). These are generally up-sized to the next longer size at 6-8 weeks post-operatively, with parent teaching to enable independent care at home.
4. Hard button low-profile tubes are placed via open technique. Generally changed in 9-12 months PRN if/when inner flapper-valve fails or cap breaks. This is done in the clinic setting, often with anxiolytic oral midazolam 0.5 mg/kg given 30 minutes beforehand.
5. MIC long tubes have a water balloon for inner retention, and an adjustable disc externally. It is important that the disc is secured down on the skin, so that tube movement occurs on top of the disc. These are generally changed out after 4-6 weeks to a low-profile MIC-KEY tube.
6. A gastrostomy tube that comes out before 10-14 days may not be able to be reinserted. If a GT comes out before 6 weeks, notify the fellow/chief resident on the service to come help you. If the tube

has been in place for over 6 weeks, then try to replace the MIC-KEY® with a new tube of the same size. Before going to replace a tube, find out what size the previous tube was- the first number is the diameter of the tube in French and the second number is the length in centimeters. Make sure they have the right size to replace the tube with. If the right size cannot be located in a relatively short amount of time, place a foley catheter of the same French size into the tract until the right sized MIC-KEY can be found. (Be sure to tape securely, so tube does not migrate and obstruct the pylorus). If the tube is unable to be passed, the tract may need to be dilated. If this is the case the resident should call a chief resident or attending for assistance. In the mean time a foley catheter that is smaller – i.e. 8-10fr may be placed into the tract to hold the tract open. These tracts can close very quickly and should not be left for any length of time without some sort of tube holding them open. If the tract requires dilation or if the g-tube has been present less than 6 weeks a contrast study with water soluble contrast should be obtained emergently. Remember, these sites will close completely in as little as 6 hours. You should take the child to radiology yourself for the study and not order it and wait for it to be done.

7. Gastrostomy Tube Care: A simple gauze dressing is cut and placed between the tube and the skin. This is changed daily and site cleaned with soap and water or ½ strength hydrogen peroxide. The balloon volume should be checked monthly, or sooner if leakage of liquid is noted around the tube. The tube should be stabilized by using a net dressing around the abdomen or by changing to a skin level tube if the timing is appropriate. Excess granulation tissue may be cauterized with silver nitrate sticks. Triamcinolone cream 0.5% TID for a short interval may also be used. Skin irritation around the GT site may be treated with nystatin, Maalox or DuoDerm. Zinc oxide-based preparations are often effective for skin protection related to gastric leakage. Contact the Enterostomal Therapists for additional recommendations, as needed for problematic sites.
8. If a g-tube is leaking do not place a tube that is larger in diameter as this will only make the g-tube tract bigger and will not solve the problem. Instead check the balloon and see if it is fully inflated. If the tube is too long, a shorter tube may be necessary or the tube may need to be dressed with more split gauze under the tube.
9. If the child has a malecot tube it should be dressed such that the tube comes straight up out of the abdominal wall. This can be accomplished using a Hollister clamp (available on most wards), by taping the tube straight up using a goal-post configuration or by placing a roll of gauze under one side and wrapping the tube over the roll of gauze. This tube can be changed for a MIC-KEY g-tube

at 6 weeks post-operatively.

10. If a new g-tube has been placed, postoperatively, the patient's gastrostomy tube is connected to a foley bag for gravity drainage overnight. If an NG is left in place, the NG tube is left to suction for the first post-operative night. It is removed on POD number one. The patient is begun on pedialyte on post-operative day number one and advance to full feeds usually by day 2 or 3. The patient can then be switched to bolus feeds. The manner in which these children are advanced is dependent on which attending placed the tube and it is important to discuss the plan with each attending. .

NG TUBES/FEEDS:

1. Nasogastric tubes are used for gastrointestinal decompression and also for gavage feedings, particularly in neonates.
2. Decompression: Use one of the small pediatric sump tubes (Salem or Replogle). The Replogle tube (8 or 10 Fr) is preferred because the two holes are at the end of the tube. The air vent on the sump tube will reduce the likelihood of occlusion of the tube - a common complication in small suction tubes. Check position daily, as these are easily carried into the duodenum. Change the tube if it is not functioning properly. Never hook the blue port to suction. This port should function as an air sump. Timing of the removal of an NG tube is not an exact science. Generally when the effluent is no longer green and the child is passing flatus, the tube can be safely removed. Placing the tube to gravity for 6-8 hours can act as a test to see if the patient is ready to have the tube removed. It is often better to wait a day longer with a tube that to have to re-place one as this can be traumatic to the child as well as the doctor. With an infant in urgent need of gastrointestinal decompression, a large red rubber catheter can be passed through the mouth as an effective means of aspirating gastric contents.
3. Gavage feedings in premature and newborn infants are indicated if the baby lacks a gag, is weak from immaturity, or has a rapid respiratory rate. A #8 French gavage tube is usually passed orally to prevent compromising the airway, since infants are nose breathers.
Tube placement is checked by auscultation of the stomach while injecting a small amount of air and then aspirating back into the syringe attached to the tube. A measured amount of fluid is poured into the syringe and passed by gravity.

4. Tube Feedings in Older Infants
When a feeding tube is to be used in an infant for continuous slow delivery of formula, one can use 6 or 8 Fr feeding tube, the 8 has a weighted tip.
When gastroesophageal reflux is a problem, the tube should be advanced into the third portion of the duodenum or upper jejunum, and the patient should be fed in the upright position.
5. Gastrostomy tube feeding regimen:
The first feeding should be small volume continuous Pedialyte in a new gastrostomy tube for the first 8-12 hours, then formula may be started. See the section on G-tube for more information.

FOREIGN BODIES:

Patients with esophageal and tracheobronchial foreign bodies are managed by both the pediatric surgery service and the ENT service. There is a call schedule in the ER for foreign bodies. The clinician must have a high index of suspicion and a very low threshold to recommend endoscopic examination if there is any question of aspiration of a foreign body, otherwise, excessive morbidity and mortality results. Foreign body problems occur most commonly in the toddler age group, but may be seen in older children (or infants) as well.

Rigid endoscopy:

In many instances foreign bodies will need to be removed with rigid endoscopes. This equipment can be difficult to use. You should make yourself familiar with the equipment prior to the child coming to the OR. There is a rigid endoscopy cart that contains all of the pieces necessary to set up the scopes. If the object is an unusual size or shape- you should have the parent bring in an identical object (if they have more than one at home) and practice which set of grabbers will work best. Sometimes a Fogarty catheter passed beyond the object and then inflated and pulled back will bring an object into the lumen so it can be grasped. Fluoroscopy can be helpful if the object is radio-opaque.

Laryngeal foreign bodies:

A foreign body lodged in the oropharynx or glottis may warrant immediate attention to clear the airway, using such means as the Heimlich maneuver, finger dislodgement, direct laryngoscopy, or bronchoscopy. If possible, a mask airway should be maintained and more controlled laryngoscopy performed in the operating room. If the patient is ventilating adequately when seen, no maneuvers should be performed until the patient is in the OR where conditions and equipment are ideal. A tracheostomy set-up should be open and the appropriate size tracheostomy tube open

and ready to be used before starting any endoscopic or laryngoscopic procedure.

Tracheobronchial foreign bodies:

Less than 10% of foreign bodies are located above the carina. Most slip into the bronchus with the majority located in the right mainstem bronchus. History alone may be sufficient to warrant admission and endoscopy, even in the absence of physical and x-ray findings. Plain chest x-ray will reveal the foreign body if it is radio-opaque. However, most foreign bodies such as wood, plastic objects, peanuts, carrots, celery, or aluminum “pop-tops” are not radio-opaque. Fluoroscopy or inspiration and expiration views on CXR can detect subtle mediastinal shifts during expiration and inspiration, but cannot necessarily pinpoint the side of the foreign body. A foreign body which totally obstructs the bronchus leads to slow lung collapse and slow mediastinal shift toward the side of the offending object. Partial occlusion of the lumen causes the more common ball-valve effect, with subsequent air trapping on the side of the lesion and mediastinal shift away from the side of the foreign body. The Storz bronchoscope greatly facilitates foreign body removal from the tracheobronchial tree. A complete set of foreign body instruments is available in the OR. **All tracheal foreign bodies should be removed in the OR and it is important to make sure that all of the necessary equipment is in the room and is working properly prior to bringing the child into the OR and it is possible that the object could acutely obstruct the trachea when the child becomes agitated upon induction. The attending must be present when the child is brought into the OR.** Of note, a fine Fogarty arterial embolectomy balloon passed beyond the object can aid in its removal, particularly if the object is fragile (e.g. peanuts) and will not withstand the pressure of forceps. The consequences of the neglected foreign body are quite serious and include atelectasis, recurrent pneumonia, and eventual destruction of the segment of lobe. Since there is minimal morbidity using the miniaturized fiberoptic bronchoscope, an aggressive approach is warranted.

Esophageal foreign bodies:

An esophageal foreign body can cause respiratory distress in small children. Objects tend to lodge just below the cricopharyngeus muscle, usually behind the larynx or cervical trachea, thereby impinging or obstructing the airway.

Diagnostic tests:

A CXR will locate the object if it is radio-opaque; a KUB will determine if the object has slipped through the stomach. Esophageal foreign bodies are usually removed in the OR, but can be removed under fluoroscopy while awake (**DO NOT SEDATE**).

An AP and lateral neck/chest film should be done prior to attempting removal. If there is marked edema/esophageal narrowing or if the history suggests the object has been there more than 3 days, endoscopy will likely be needed. Removal under fluoroscopy with a Foley catheter requires an awake child, restrained on a papoose board. If they have teeth, you also need a bite block. The Foley should be passed through the mouth (use a 16 or 18Fr-they are stiffer and easier to pass) and the balloon inflated with hypaque or Conray (don't use barium, dilute gastrograffin is a last resort). Once you are past the object, inflate the balloon while watching under fluoro and gently pull the object out. If it won't come easily, let some contrast out of the balloon. Once the object is in the hypopharynx, turn the patient onto their side and rapidly pull the catheter out. The coin should pop out onto the table or be present in the mouth. Not infrequently, they swallow it again and you have to do it again. If you put the catheter down their nose, be aware you will likely pull the coin up into the nasopharynx and may have to put your finger in their mouth to get the coin out. The Foley catheter should only be used for round, dull objects. A CXR is done after removal of the foreign body. If everything went smoothly, the child can be fed and observed for 1-2 hours and discharged home with follow-up on a PRN basis. If removal is difficult or there is any concern for perforation, admit, keep NPO, and obtain swallow (gastrograffin followed by thin barium).

Gastrointestinal foreign bodies:

Once in the stomach, most ingested foreign bodies will safely transverse the gastrointestinal tract, usually within 4-5 days. The problem sites are usually the pylorus, the ligament of Treitz, and the ileocecal valve. If the object is radio-opaque, it can be followed with serial x-rays. The stools should be checked for reappearance of the object. The child should be followed for abdominal pain, vomiting, or blood in the stool. If the child has not passed the object and is otherwise doing well after 4 weeks, they should return for an x-ray. If after 4-6 weeks the object is still in the stomach, it can be retrieved by gastroscopy.

CAUSTIC INGESTIONS:

1. All patients with suspected ingestion of a caustic material are admitted for esophagoscopy under general anesthesia. Although most patients with esophageal injury show burns of the oropharynx, as well, this is not a completely reliable guideline. These patients should be admitted to the Pediatric Surgical Service or PICU.

2. Upper airway injury as well as face and hands should be assessed. Pharyngeal burns may be so severe as to require tracheostomy.
3. The alkaline substances include sodium hydroxide, phosphagens, and silicates (found particularly in drain and oven cleaners, as well as glass cleaners - Windex, detergents, and dishwasher cleaners). These chemicals cause liquefaction necrosis and may involve full thickness injury.
4. Management:
DO NOT INDUCE VOMITING. The child should be kept NPO and placed on IV fluids. In the past if circumferential burn were present at esophagoscopy, then corticosteroids (e.g. prednisone, 2-3mg/kg/day) were given for two weeks. The steroids are continued for several weeks after complete healing of the burned area, as observed by repeat esophagoscopy. This approach is controversial and should be balanced against the risk of immunosuppression in the face of a real risk of infection due to esophageal perforation secondary to transmural damage. The use of steroids should be discussed with the attending. IV antibiotics are given as long as steroids are given. Barium swallow does not adequately determine if the esophagus has been injured, but should be obtained as a baseline sometime during the first 2-3 weeks after injury. Esophagoscopy is done under general anesthesia within 24 hours of admission. Esophagoscopy is done only to the point of injury and then stopped once the diagnosis is made. Repeat esophagoscopy is usually performed in 14 days, at which time dilatations and triamcinolone steroid injections may be performed if strictures are present. Another approach that has been described is to leave an indwelling silastic stent in the esophagus to prevent stricture formation. These children will then require close follow-up with serial esophagoscopy and dilatation to prevent stricture formation.

H. OTHER TUBE SIZES

Foley Catheters

Neonate	#3.5, 5, 8 Fr feeding tubes or 6 Fr balloon catheter-DO NOT test balloon before inserting (it increases the diameter of the catheter)
Child	#8 Fr Foley
Adolescent	#16 Fr Foley

Nasogastric Tubes (Salem-sump tubes)

Premature	#6 Fr (note: 3.5 Fr special order)
Newborn	#8 Fr
Infant	#10 Fr

Toddler	#12 Fr
Child	#14 Fr
Older Child-Adoles	#16 Fr
Adult	#18 Fr

MIC-KEY Gastrostomy

Bard Buttons

(must be ordered to ward)

(must be ordered to ward)

<u>Tube</u>	<u>Logistics #</u>	<u>Tube</u>	<u>Logistics #</u>
16 Fr; 0.8 cm	(148576)		
16 Fr, 1.0 cm	(148578)	18 Fr, 1.7 cm	(123838)
16 Fr, 1.2 cm	(148575)	18 Fr, 2.4 cm	(123832)
18 Fr, 1.5 cm	(124194)	18 Fr, 3.4 cm	(123833)
18 Fr, 1.7 cm	(124197)		
18 Fr, 2.0 cm	(124199)		
18 Fr, 2.3 cm	(124203)		
18 Fr, 2.5 cm	(124204)		
18 Fr, 3.5 cm			

Cecostomy Tubes

Cecostomy tubes are placed to allow antegrade enemas for patients with fecal incontinence, with the goal of keeping them socially clean for 24 hours. Though primarily for the spina bifida population, they are also used for patients with incontinence or chronic constipation from Hirschsprung’s disease, imperforate anus, and idiopathic constipation.. Tubes used include the Bard button, the MIC-KEY tube, the MIC long tube and the Chait trapdoor low-profile cecostomy.

The latter is placed laparoscopically by Dr. Zallen. After a tube check under fluoro at 10-14 days, the families then begin home irrigations, usually with a salt water solution. These tubes are pigtail catheters only available in Dr. Zallen’s OR and in DCH Radiology. Nancy Jacobs provides the long term management. If a tube becomes dislodged, place a 10Fr Foley catheter in tract. Inflate balloon with 2-3 ml water and contact Nancy (or the surgery office) to arrange for tube replacement in radiology on the next available date.

CHAPTER VI: COMMON PEDIATRIC SURGERIES

APPENDICITIS:

The most frequent surgical problem evaluated in the emergency room is “abdominal pain, rule out appendicitis”. Appendicitis is by far the most common abdominal surgical problem in childhood. If an appendectomy is appropriately performed soon after the onset of symptoms, the child is usually discharged from the hospital the next day. However, if the diagnosis and treatment are not accomplished early, an unforgettable clinical nightmare can result.

1. Guidelines:

Abdominal pain begins in the periumbilical area and usually (but not invariably) later shifts to the right lower quadrant. Remember that the peritoneal cavity is six-sided and localized pain will reflect where the appendix or its inflammatory fluid resides (e.g. retrocecal or pelvic appendix). Perforation commonly occurs at approximately 36 hours after the pain begins.

Abdominal pain usually PRECEDES vomiting.

Appendicitis is at times accompanied by anorexia, nausea, and vomiting, but these are not discriminating signs.

Consistent, localized point tenderness is the cardinal reliable sign of appendicitis, whereas other physical findings tend to be variable.

Fever and leukocytosis tend to be minimal in early appendicitis.

Rectal examination should be done in all cases of abdominal pain since this may be the only way to detect the tenderness associated with the retrocecal appendix, or to feel a pelvic mass, phlegmon, or abscess.

A calcified fecalith on KUB is strong evidence for appendicitis, but is found in only 10-15% of the cases. If the patient has RLQ pain and a fecalith, the appendix comes out. An incidental finding of a fecalith during work-up of another problem may warrant elective appendectomy. A flat and upright AXR should be obtained if it is not absolutely obvious that the patient has appendicitis.

In the child less than 2 years of age, the appendix is usually perforated by the time the child is brought to the emergency ward. Fortunately, appendicitis in this age group is infrequent (approximately 2% of all cases).

If one suspects perforated appendicitis, it should be ascertained whether it is generalized peritonitis or localized. Localized perforations with abscess and without bowel obstruction may be

drained by laparoscopy or CT guided drainage and an interval appendectomy performed 6-8 weeks later. Patients with generalized peritonitis and recent perforation are fluid resuscitated, given rectal Tylenol to get their temperature under 101, and started on broad spectrum antibiotics before going to the operating room. Perforated appendicitis in children is not an emergency. The child needs to be stabilized before proceeding to operation or can be treated non-operatively on triple antibiotics and have an interval appendectomy performed in 6 to 8 weeks.

In some children, the diagnosis will be equivocal. CT and ultrasound are good adjunctive tests and should be reserved for difficult cases or cases of suspected abscess. Rectal contrast should be used on CT scans for appendicitis. Adolescent girls should have a pelvic exam by you with cervical cultures. If they have already had a speculum exam before you see them, ask if there was pus coming from the os and you must do a bimanual exam yourself (or someone on our team-not the medical student). DO NOT fall into the trap of believing adolescent girls that tell you they haven't had sex. All girls that have gone through menarche need a pregnancy test as well. Many of these girls will need a pelvic ultrasound to rule out ovarian cysts/TOA's as a cause of their problem. Ask the ultrasonographer to look specifically for fluid in the cul-de-sac as well. Their bladder needs to be full for this test, so consider this when ordering urine tests (tell them not to void until after the ultrasound), they may need a Foley to fill their bladder or fluid bolus IV (avoid po fluids unless you are reasonably sure they are not going to need an operation in the next 8 hours).

Clinical Management Guidelines for OHSU/Doernbecher:

CT scans should be used in the small percentage of patients where there is a true diagnostic dilemma, or in those where the diagnosis is late and there may be a phlegmon or abscess. That being said, the following is the protocol currently followed for patients in whom a CT is deemed appropriate:

- 1. ED will place IV, place order, and call Radiology resident to arrange CT*
- 2. Pediatric surgeons will NOT be required to see the patient before the CT, unless desired by the ED attending*
- 3. Children with symptoms for less than 36 hours will be given PO contrast with a long (2 hour) prep and IV contrast*
- 4. Children with symptoms for more than 36 and suspicion of abscess will be given PO contrast (2 hr oral prep), rectal contrast (if tolerated), and IV contrast*

Because variations in protocol are sometimes warranted based upon patient age, condition, and body habitus, the protocol may be tailored on a case by case basis as necessary.

2. Antibiotics:

Mefoxin (cefoxitin) preop for simple appendicitis, ampicillin, gentamicin and flagyl preop for allergic patients and patients suspected of having perforated appendicitis. Patients with gangrenous or perforated appendicitis will get IV antibiotics until they are afebrile for 48 hours, eating, and WBC is <12,000. At this time they can be D/C'd home without antibiotics.

PYLORIC STENOSIS:

1. Suspect the diagnosis in any infant 2 weeks to 3 months with projectile, nonbilious vomiting. There is an increased incidence in first born males and in patients whose mom had it.
2. You should attempt to palpate an epigastric olive to confirm the diagnosis. To do this, the stomach must be empty and the baby must be quiet. Place a 8 fr. replegle tube as an NG or OG tube and suction the stomach. Leave this tube in place on low continuous wall suction if they have had a barium study before arriving here. Once you have decompressed the stomach, put some sugar on the infant's pacifier and have parent try to console the child. With the child lying on the exam table and you on the right side of the patient, place your right hand on the abdomen and your left hand should lift the legs towards the abdomen, making sure you are under the liver edge and press down to the spine in the epigastrium. Then roll your right hand toward the umbilicus and the olive should roll beneath your fingers and "pop up" as you release it. It feels hard and is literally the size of an olive. This is usually hard to feel unless the child has been sick long enough to be significantly dehydrated. Don't spend more than 10-15 minutes trying this or more than 3-4 attempts.
3. Ultrasound is the diagnostic study of choice if an olive is not palpable. Normal channel length is 1.4 cm; normal muscle thickness is less than 4mm. Perhaps more importantly than the measurements of the pylorus is the mechanics of the muscle itself. If the pylorus does not open and allow the stomach contents to empty while it is being watched on real time ultrasound (no exact time for this has been published, but a minimum of 10minutes is used at WCHOB) than one can be relatively certain of the diagnosis of pyloric stenosis.

4. Significant dehydration is a common finding. Gastric losses cause hypochloremic, hypokalemic alkalosis. Fluid boluses of NS 10-20cc/kg should be given until urine output is established. Then start an IV of D5 ½ NS with 10-20mEq KCL/liter at maintenance and follow electrolytes in 8-12 hours. Rapid rehydration and correction of electrolytes is not warranted. Pyloric stenosis is not an emergency. Operation is carried out once dehydration and electrolyte abnormalities are corrected. These should not be corrected rapidly and under most circumstances the infant does not need labs more frequently than every 8-12 hours. Some exceptions do occur and should be discussed with staff. Pre-printed admission orders are available.
5. Pyloromyotomy is carried out laparoscopically for most patients. If the patient is a very small ex-premie or if the umbilicus has recently fallen off and is still wet then they may be a candidate for an open procedure.
6. Post-op management used to be a very complicated regime of increasing the strength and then increasing the volume over a period of days. We now simplify this routine. The babies can start on regular feeds at either ad lib or one ounce depending on which attending is covering the patient. If the infant starts on one ounce, he/she can advance one ounce/feed until they are taking what they used to take at home and then they can go to ad lib feeding. If the babies vomit, which they frequently do, the feedings are continued as before. They do not need to be made NPO- keep feeding them and they will do just fine. No antibiotics are needed post-op. Marcaine is put in the wounds at operation, so only Tylenol is needed post-op and is given rectally or orally (10-15mg/kg) around the clock. Pre-printed orders are online.
7. In the event the patient has a mucosal injury repaired at operation, an NGT is left in place on low continuous wall suction for 1 to 3 days. Depending on the severity and extent of repair, a contrast study may be obtained prior to removing the NGT and starting feeds. Each case will be unique.

INTUSSUSCEPTION:

1. Intussusception occurs most frequently between the ages of 3 months and 3 years. Peak incidence is 4-10 months. If the patient is younger or older, one should be suspicious of a lead point for the intussusception such as a Meckel's diverticulum, or an intestinal polyp.
2. Suspect the diagnosis in any infant or child with a history of episodic paroxysmal abdominal pain with straining, crying,

- drawing up of knees and inconsolability.
3. Abdominal exam may be normal or a tender right-sided, “sausage-shaped” mass may be palpable. Heme positive or currant jelly stools are a late finding.
 4. Obtain a flat and upright abdominal x-ray (which may show a mass or paucity of gas in the RLQ) and start a bolus with 20cc/kg of NS if the child is dehydrated
 5. Have a low threshold for obtaining an air contrast enema for diagnosis and non-operative reduction. Evidence of peritonitis, perforation, gangrenous bowel or sepsis dictates operative exploration with rapid pre-op resuscitation and triple antibiotics (Ampicillin, Gentamicin, flagyl).
 6. A resident should accompany the patient to radiology and be present for the air contrast reduction. Pressure is not to exceed 120 mmHg. Reduction is not considered successful unless there is free reflux of air into the ileum. Repeat attempts can be made if the baby’s condition will permit.
 7. **If there is a perforation during the air contrast enema, the child may go into extremis due to the high pressure in the peritoneal cavity. If this is the case the abdomen should be rapidly decompressed with a large angiocath placed into the RLQ. This may need to be done quickly as the rapid and high pressure pneumoperitoneum can cause rapid respiratory embarrassment and abdominal compartment syndrome.**
 8. If there is a successful reduction with the air contrast enema, the patient is usually admitted for 23 hour observation after reduction. Keep NPO for the first 8 hours, then gradually advance the diet. Warn parents and staff that high temperatures can follow reduction. If symptoms recur, air contrast reduction should be performed again. Operation is mandatory if reduction cannot be accomplished. The recurrence rate after either barium enema reduction or surgical reduction is approximately 5%.
 9. Intussusception is known to occur in children after any type of operation but usually a laparotomy. For example, if a purse string turn-in of too large a segment of cecal wall is performed during appendectomy, this can serve as a lead point. A small bowel-to-small bowel intussusception (ileoileal, etc) can be particularly difficult to diagnose in the postoperative period after intra-abdominal procedures. Intussusception should be a primary consideration in a patient who has a prolonged “ileus” with bilious NG drainage. Ileoileal intussusception is not usually diagnosed or treated non-operatively (it can be seen on U/S or CT scan- but will not be detected on air contrast enema). It is usually found on exploration for small bowel obstruction in the post-operative period.

INGUINAL HERNIAS, INCARCERATED INGUINAL HERNIA:

1. Embryology and Anatomy: During the 7th and 8th month of gestation, the testes descend into the scrotum and carry a diverticulum of peritoneum with them called the processus vaginalis. The portion of the processus immediately surrounding the testis becomes the tunica vaginalis and the remainder of the processus usually obliterates. In a significant number of patients, the processus remains patent leaving a communication between the peritoneal cavity and the scrotum. A patent processus through which the bowel or other intraabdominal contents protrude is called a hernia, one which contains peritoneal fluid alone is called a hydrocele. A hydrocele that involves a small fluid collection around the testicle and does not track up to the internal rings and does not change in size may represent a non-communicating hydrocele. These hydroceles are usually present at birth. Since there may not be an opening at the internal ring, these may not develop into a hernia and are often watched for a period of time. A hydrocele that appears later in life or changes in size is a communicating hydrocele. This likely represents a patent process that opens into the peritoneal cavity, but is not quite large enough for bowel to pass through the ring.
2. Epidemiology: The incidence of indirect inguinal hernia in infants and children is approximately 1-5% with a 10:1 male to female ratio and a sixfold increase in incidence with prematurity. Direct and femoral hernias are rare. The incidence of bilateral inguinal hernias is influenced by age at initial presentation, the sex of the patient, the presence of increased intraabdominal pressure, and the presence of associated disorders.
3. Presentation: A groin bulge visible, usually intermittently, during times of increased intraabdominal pressure (i.e. crying in an infant).
4. Treatment: Inguinal hernias are repaired through a small groin incision or laparoscopically depending on the attending. Communicating hydroceles are usually repaired in a similar fashion as they usually represent a small hernia.

Noncommunicating hydroceles are usually self limiting and may be observed for a period of time to see if they will resolve spontaneously.

Incarcerated

1. This condition is age-related occurring most often in infants during the first year of life. Most, if not all, can be reduced manually, which obviates the need for emergency surgery.

2. Reduction techniques:

Occasionally, simply holding the baby in very steep Trendelenburg position (by propping up on sheets or towels) reduces the hernia, due to the pull of the mesentery.

Have an assistant hold the infant above the knees in a frog leg position to relax the abdominal wall.

Fingers of one hand should attempt to fix the hernia and align the internal and external rings while the other hand should press the incarcerated mass upward toward the canal. So it is a push up toward the umbilicus with your dominant hand while the nondominant hand stabilizes the ring alignment then push straight in with your dominant hand using continuous, steady pressure (5 minutes). You should be patient and comfortable when attempting reduction, as this may take 5 minutes of steady pressure to get it to budge and may take several tries. Try to milk the bowel contents out of the incarcerated bowel, until it “pops” back into the abdomen.

3. If successful, the patient is always admitted, and the repair is performed electively within the next 12-48 hours, once the edema has resolved. The infant should have serial examinations to rule out re-incarceration.
4. Emergency surgical intervention is required if the hernia cannot be reduced, or if there is postreduction evidence of persistent intestinal obstruction, or nonviable bowel. This is a rare, but a possible event.
5. It is imperative to differentiate an incarcerated hernia from a hydrocele of the cord. Hydrocele of the cord is often tense. One can distinguish the end of the hydrocele from the testes itself and can also appreciate the proximal end of the cord (the internal ring is flat with no hernia sac coming through it). These infants can be sent home and subsequently be evaluated in the office.
6. An unfortunate complication of an incarcerated hernia is hemorrhagic infarction of the testicle. Reduction will usually reinstitute blood flow to the testis.
7. Incarcerated inguinal hernias in girls are often sliding hernias containing ovary and tube. These need to be repaired within 24 hours of diagnosis if they cannot be reduced as some episodes of ovarian torsion and loss have been documented. If they can be reduced, they need to be repaired on a semi-elective basis.
8. Non-incarcerated hernias in premature infants:
If consulted on an inpatient preemie with multiple problems, it can be repaired just prior to discharge home.

Babies up to 60 weeks gestational age with a history of prematurity (gestational age less than 36 weeks at birth) must be admitted overnight after hernia repair for apnea monitoring.

UNDESCENDED TESTICLES (Cryptorchidism):

1. General

- Observe and follow children with undescended testes until 1 year unless symptomatic inguinal hernia is present.
- 90% have associated inguinal hernia
- Testes may be undescended, ectopic (suprapubic, perineal, femoral), retractile, or absent. Always document position of palpable testes at pubis, external ring, perineum, or internal ring, or non-palpable.
- Always mention the possibility of orchiectomy in the event the testis is atrophic.
- Always mention the possibility that the testis may be absent.
- A female presenting with bilateral groin masses should be evaluated for testicular feminization. This may include vaginotomy and an endocrine work-up.

2. Embryology

The testes form from the medial portion of the urogenital ridge and descend, with the help of hormonal influence and the gubernaculum, through the internal inguinal ring to eventually lie in the scrotum. Failure of descent is thought to be due to inadequate hormone levels or failure of the end organ to respond.

3. Incidence

The incidence of undescended testes in term infants is 0.5%, and is 5% in premature infants. 50% occur on the right, 25% on the left, and 25% bilaterally, an incidence which parallels that of inguinal hernias in children. Undescended testes are invariably associated with ipsilateral inguinal hernias.

4. Diagnosis

- The differential diagnosis of an empty hemiscrotum includes agenesis, atrophy, undescended, ectopic, and retractile testes.
- An undescended testis must be differentiated from a retractile testis, which can be milked into a scrotal position, and does not require surgery.

5. Treatment

Indications

- Decreased fertility due to increased environmental temperature.
- Increased incidence of testicular malignancy (40x)
 - Malignancy risk is unchanged with surgery. Testicle is brought down so it can be properly monitored for

development of malignancy as well as other reasons below.

- Risk of malignancy in contralateral descended testis is increased.
- Malignancy (most commonly seminoma) usually occurs after 20 years.
- Increased incidence of trauma due to abnormal position.
- Increased incidence of torsion.
- Cosmesis

6. Surgery

- Optimal timing: one year of age.
- Dartos pouch technique vs. transseptal fixation of testis.
- High retroperitoneal or intraabdominal testes may require transposition of the internal ring to obtain adequate length of the vasculature and vas deferens.
- If adequate length cannot be obtained, the testicular vessels can be divided and the collateral supply from the inferior epigastric artery is allowed to supply the testis.
- Orchiectomy may be indicated if the testis does not reach the scrotum after all attempts have failed, provided a second testis is present.
- Bilateral undescended testes must be worked up for urinary tract anomalies and intersex.

UMBILICAL HERNIA:

Many of these will close spontaneously. This is the only hernia that may spontaneously resolve. The probability that a hernia will close is directly related to its size during infancy. If the defect is large enough to only admit the tip of the pinky finger there is approximately a 90% chance it will close. If the tip of the index finger will fit in it there is around a 50% chance it will close and if the thumb will fit into the defect then there is only around a 10% chance the defect will spontaneously close (rule of thumb). For this reason, large hernias may be repaired at a younger age.

TESTICULAR TORSION:

1. This problem is usually dealt with by the urologists, but is an important pediatric surgical emergency. The peak age group for this condition is adolescent boys ages 12-18 with an annual incidence of 1 in 4000 males below the age of 25. Torsion also can occur in the infant particularly in the undescended testis.
2. Major considerations:

- a. Early operative intervention if diagnosis is suspected in order to save the testicle (6 hour golden period).
 - b. The differential diagnosis includes torsion of the appendix testis or epididymitis.
 - c. Consent for operation should include fixation of the contralateral testis.
3. Torsion of the appendix testis
- a. The symptoms are very similar to testicular torsion, but the child is often pre-teen.
 - b. Examination may reveal localization of the pain to the upper pole of the testis, and transillumination may reveal the “blue dot” sign.
 - c. Later presentation may reveal erythema, diffuse tenderness, and reactive hydrocele, making differential impossible. Testicular scan is often helpful, but may be misleading. Doppler ultrasound is usually helpful, but not 100% sensitive.
 - d. If one is certain of the diagnosis, this condition can be treated with analgesia and scrotal support. Symptoms usually subside after 5-12 days.
4. Epididymitis
- a. It is rare in children <14 years, except in association with mumps. If present in the younger child without mumps, a urinary tract evaluation with IVP and VCUG, is essential to rule out ectopic ureter to vas, incompetent ejaculatory ducts, or other urologic conditions.
 - b. Examination should include a rectal examination to rule out associated prostatitis. Urinalysis is essential because urinary tract infection is almost invariably present.
 - c. Elevation of the testis sometimes gives relief with epididymitis, but usually not with torsion.
5. A doppler ultrasound scan can usually make the diagnosis of torsion and should be the test of choice in a patient who presents with testicular pain in which you suspect a torsion.

GYNECOLOGIC CONDITIONS:

1. A young girl presenting with a history of vaginal bleeding may require exam under anesthesia to rule out foreign body, sexual abuse or urethral prolapse. Many of these children can be carefully examined awake or with appropriate sedation. One note of caution, it is usually only possible to get ONE GOOD EXAM on these children, so talk to your staff or chief resident before the first exam to be sure you know everything to consider on your exam.

2. Ovarian torsion should be considered in the differential of abdominal pain in all girls. It typically comes on suddenly and classically is associated with vomiting. The physical findings may be relatively minimal unless the ovary is already infarcted. A pelvic ultrasound should be obtained immediately - DO NOT DELAY! The same 6 hour rule applies as for all vascular ischemia. The bladder must be full so consideration must be given to placing a Foley and filling the bladder with sterile water/saline to facilitate timely performance of an ultrasound. Remember, ultrasound is not very good for this problem and an exploration may still need to be done to completely rule out ovarian torsion. However, sometimes, ultrasound may make a diagnosis that does not require surgery (hemorrhagic corpus luteum cyst) or may actually show good flow to the ovaries. If a diagnosis of hemorrhagic cyst is made, it is imperative that through Doppler signals are seen on ultrasound, otherwise, ovarian torsion cannot be ruled out. Laparoscopic or open exploration is the best way to be 100% sure.

GASTROESOPHAGEAL REFLUX DISEASE:

1. General
 - The lower esophageal sphincter (LES) is a physiologic valve comprised of three parts: the esophageal hiatus, the angle of His, and the high pressure zone of the distal esophagus.
 - The LES allows for the passage of food into the stomach and prevents its regurgitation as well as that of acid.
 - A certain amount of reflux is normal.
 - Symptomatic reflux is pathologic and can result in: respiratory, nutritional and esophageal inflammatory symptoms.
2. Complications:
 - Respiratory manifestations include: apnea, aspiration pneumonitis, coughing, choking, reactive airway disease, and bronchopulmonary dysplasia.
 - Nutritional manifestations. An incompetent LES can result in significant loss of calories and a delay in growth and development.
 - Esophageal manifestations include: esophagitis, which may be complicated by stricture, anemia, shortening of the esophagus, or Barrett's esophagus. An unusual manifestation of reflux esophagitis, Sandifer's syndrome, in which voluntary contortions

of the head, neck and trunk are performed, in order to improve distal esophageal peristalsis, is improved by fundoplication.

3. Causes of GERD

Delayed gastric emptying may be the cause of reflux in neurologically impaired patients. Reflux is seen in 10% of patients who have had pyloric stenosis, 75% of those surviving CDH, most patients who have undergone repair of EA and TEF, and those with intestinal malrotations.

4. Diagnosis

Studies are instituted only when symptomatic GER is suspected. Barium esophagram should be obtained on all patients to evaluate for anatomical defects such as malrotation. Upper GI is not a good study to evaluate for reflux as it is a very short study and it may be difficult to differentiate physiologic reflux from pathological reflux. A 24 hour pH probe is the study of choice when there is any question as to the severity of the reflux. The study should be performed 72 hours after any antireflux medications have been stopped. A 24 hour pH probe placed in the proximal and distal esophagus is used to detect reflux episodes in which the pH drops to 4 or below. The number of episodes, the number lasting more than 5 minutes, the duration of the longest episode, the mean duration of reflux, and the percentage of time that the pH is less than 4 are determined and given a score in order to quantitate the GER. Sensitivity and specificity are in the 90% to 95% range. With a strong reflux history a confirmatory study is not necessary. Esophageal manometry and scintigraphy are reputed as useful by some. Esophagoscopy and possible biopsy are used when esophagitis is suspected.

5. Therapy

Positional therapy includes: assuming a more upright position, reducing the volume of gastric content and increasing its viscosity, and altering lifestyle, including choices of food. Medical therapy includes: the administration of antacids and medications that increase esophageal peristalsis, increase the activity of the LES, and increase gastric emptying (i.e. metaclopramide). Surgical therapy consists of a Nissen fundoplication with or without gastrostomy tube placement, which establishes an intraabdominal portion of esophagus and creates an angle of His.

6. Nissen Fundoplication-Indications

Absolute indications include: patients with apnea who have required resuscitation, patients with recurrent or continuous pneumonitis, patients with esophagitis, and patients who have failed nonoperative therapy.

Relative indications include: patients with atypical asthma, croup, coughing at night, choking episodes, positive pH probe and chronic vomiting.

Pre-op work up patients need upper GI to evaluate anatomy, a pH probe unless there has been good evidence of reflux, i.e. recurrent pneumonias, dying spells, esophagitis etc.

7. Postop Course

Gastrostomy:

Postoperatively, the patient's gastrostomy tube is connected to a foley bag for gravity drainage for the first 48 hours. The NG tube is left to suction for the first post-operative night. It is removed on POD number one. On POD 2 the g-tube is elevated (left open to vent). If the patient tolerates his/her own secretions, feeds are begun on POD 3 starting with pedialyte or clears, leaving the tube open to vent (elevated) at all times. See previous section on gastrostomy tube feedings. **The exact regimen is attending dependent and should be verified with individual attending.**

No Gastrostomy:

Start clears on POD 1 and advance to regular diet as tolerated; with discharge POD 1 if tolerating regular diet.

BOWEL PREP:

- Start day prior to OR, patient should arrive early to mid-morning.
- Go to: ozone.ohsu.edu/healthsystem/HIS for a copy of: **Pediatric Surgery: Pre-surgery Bowel Preparation Orders**. These have been approved by all of our attending surgeons.
- Clear Liquid diet during bowel prep. NPO as appropriate for planned OR time. Golytely: 50 cc/kg/hr for four hours. **If this is a small baby, it is important to begin IV hydration during the bowel prep.** Most children will need an NG (small feeding tube) tube as they will not tolerate drinking the Golytely. Do not delay too long in placing the NG tube in the hopes the child will drink the Golytely.
- **Be sure infusion gets stopped in time to meet NPO guidelines.**
- Saline enemas should be given to patients with a Hartman's pouch or distal defunctional ostomy. Some attendings prefer to do in the OR. Oral antibiotics are not routinely given.
- **Patients must be clear from below. You should check the effluent from their stoma or rectum to determine the effectiveness of the bowel prep- if the effluent is not clear then a repeat dose of Golytely or enemas may be necessary.**
- **If there are problems with the bowel prep, the chief resident or attending must be notified**

CHAPTER VII. TUMORS

NEUROBLASTOMA:

General Information

This information has been taken from the NIH web site for neuroblastoma. The references have been removed to save space. If you would like to see any of the references please go to www.cancer.gov/cancerinfo/pdq/pediatric/treatment and click on neuroblastoma. These recommendations were current as of 7/2003

This cancer treatment information summary provides an overview of the prognosis, diagnosis, classification, and treatment of neuroblastoma.

The National Cancer Institute provides the PDQ pediatric cancer treatment information summaries as a public service to increase the availability of evidence-based cancer information to health professionals, patients, and the public. An Editorial Board of pediatric oncology specialists updates these summaries regularly according to the latest published research findings.

Cancer in children and adolescents is rare. Children and adolescents with cancer should be referred to medical centers that have a multidisciplinary team of cancer specialists with experience treating the cancers that occur during childhood and adolescence. This multidisciplinary team approach incorporates the skills of the primary care physician, pediatric surgical subspecialists, radiation oncologists, pediatric medical oncologists/hematologists, rehabilitation specialists, pediatric nurse specialists, social workers, and others in order to ensure that children receive treatment, supportive care, and rehabilitation that will enable them to achieve optimal survival and quality of life. Refer to the PDQ Supportive Care summaries for specific information about supportive care for children and adolescents with cancer.

Guidelines for pediatric cancer centers and their role in the treatment of pediatric patients with cancer have been outlined by the American Academy of Pediatrics. [1] At these pediatric cancer centers, clinical trials are available for most of the types of cancer that occur in children and adolescents, and the opportunity to participate in these trials is offered to most patients/families. Clinical trials for children and adolescents with cancer are generally designed to compare potentially better therapy with therapy that is currently accepted as standard. Most

of the progress made in identifying curative therapies for childhood cancers has been achieved through clinical trials. Information about ongoing clinical trials is available from the NCI Cancer.gov Web site. Neuroblastoma is predominantly a tumor of early childhood, with two thirds of the cases presenting in children younger than 5 years of age. In rare cases, neuroblastoma can be discovered prenatally by fetal ultrasonography. [2] Neuroblastoma originates in the adrenal medulla or the paraspinal sites where sympathetic nervous system tissue is present. The most common symptoms are due to a tumor mass or to bone pain from metastases. Proptosis and periorbital ecchymosis are common and arise from retrobulbar metastasis. Extensive bone marrow metastasis may result in pancytopenia. Abdominal distention with respiratory compromise due to massive liver metastases occurs in infants. Because they originate in paraspinal ganglia, neuroblastomas may invade through neural foramina and compress the spinal cord, causing paralysis. Fever, anemia, and hypertension are found occasionally. Multifocal neuroblastoma occurs rarely, usually in infants, and generally has a good prognosis.[3] Rarely, children may have severe watery diarrhea due to the secretion of vasoactive intestinal peptide by the tumor.

Children with neuroblastoma rarely present with paraneoplastic neurologic findings including cerebellar ataxia or opsoclonus/myoclonus.[4] The opsoclonus/myoclonus syndrome appears to be caused by an immunologic mechanism that is not yet fully defined.[5,6] Unlike other neuroblastomas, the primary tumor usually is diffusely infiltrated with lymphocytes.[7] Patients who present with this syndrome often have neuroblastomas with favorable biologic features and are likely to survive, although tumor-related deaths have been reported. Neurologic dysfunction is most often a presenting symptom but may arise after removal of the tumor. Opsoclonus/myoclonus is frequently associated with pervasive and permanent neurologic and cognitive deficits, including psychomotor retardation.[6,8,9] Some patients may clinically respond to removal of the neuroblastoma, but improvement may be slow and partial; symptomatic treatment is often necessary. Adrenocorticotropic hormone (ACTH) treatment is thought to be effective, but some patients do not respond to ACTH.[5,8] Various drugs, plasmapheresis, and intravenous gamma-globulin have been reported to be effective in selected cases.[8,10] It has been suggested that the long-term neurologic outcome may be superior in patients treated with chemotherapy, possibly by means of its immunosuppressive effects.[10]

The diagnosis of neuroblastoma requires the involvement of pathologists who are familiar with childhood tumors. Some neuroblastomas cannot be discriminated from other small round blue cell tumors of childhood (such as lymphomas, primitive neuroectodermal tumor, and rhabdomyosarcoma) by conventional light microscopy. Evidence for sympathetic neuronal differentiation may be demonstrated by immunohistochemistry, electron microscopy, or by finding elevated levels of serum catecholamines (dopamine, norepinephrine) or urine catecholamine metabolites: vanillylmandelic acid (VMA) or homovanillic acid (HVA). The minimum criterion for a diagnosis of neuroblastoma that has been established by international agreement is based on 1 of the following: 1) An unequivocal pathologic diagnosis made from tumor tissue by light microscopy (with or without immunohistology, electron microscopy, or increased levels of serum catecholamines or urinary catecholamine metabolites); 2) The combination of bone marrow aspirate or trephine biopsy containing unequivocal tumor cells (e.g., syncytia or immunocytologically-positive clumps of cells) and increased levels of serum catecholamines or urinary catecholamine metabolites as described above.[11]

Approximately 70% of all patients with neuroblastoma have metastatic disease at diagnosis. The prognosis for patients with neuroblastoma is related to their age at diagnosis, clinical stage of disease, and (in patients older than 1 year of age) regional lymph node involvement. Other conventional prognostic variables include the site of the primary tumor and tumor histology (see Cellular Classification section). [12-15]

Children of any age with localized neuroblastoma and infants younger than 1 year of age with advanced disease and favorable disease characteristics have a high likelihood of long-term, disease-free survival. [12,16] Older children with advanced-stage disease, however, have a significantly decreased chance for cure despite intensive therapy. As an example, aggressive multiagent chemotherapy has resulted in a 2-year survival rate of approximately 20% in older children with stage IV neuroblastoma. [17,18] Neuroblastoma in the adolescent or adult has a worse long-term prognosis regardless of stage or site and, in many cases, a more prolonged course. [19]

A number of biologic variables have been studied in children with this tumor. Of particular importance are Shimada histology, aneuploidy of tumor DNA, and amplification of the *N-myc* oncogene within tumor tissue, since treatment decisions may be based on these factors. [15,16,20-23] An open biopsy is usually needed to obtain adequate tissue for determination of these biological characteristics. Hyperdiploid tumor DNA is associated with a favorable prognosis, [24]

especially in infants with neuroblastoma, [20] while N-myc amplification is associated with a poor prognosis regardless of patient age. [16,20,25] In contrast to N-myc gene amplification, the degree of expression of the N-myc gene in the tumor does not predict prognosis. [26,27] Amplification of N-myc is associated with deletion of chromosome 1p and gain of the long arm of chromosome 17(17q), the latter of which independently predicts a poor prognosis. [28,29] A higher proportion of proliferating tumor cells may independently predict poor prognosis. [30] Expression of the gene encoding one of the high-affinity neurotrophin receptors (termed TRK-A) is associated with good prognosis tumors. [31] Increased levels of telomerase RNA, [32] elevated serum ferritin, [33] elevated serum lactate dehydrogenase, [34] and the persistence of neuroblastoma cells in bone marrow during or after chemotherapy are each associated with poor prognosis. [22,33-40] Biologic staging consisting of N-myc copy number and age is useful in defining prognosis and treatment of stage III neuroblastoma. [23] Neuroblastoma has been categorized into three biological groups. One type expresses the TRK-A neurotrophin receptor, is hyperdiploid, and tends to spontaneously regress. Another type expresses the TRK-B neurotrophin receptor, has gained an additional chromosome, 17q, has loss of heterozygosity of 14q, and is genomically unstable. In a third type, chromosome 1p is lost and the N-myc gene becomes amplified. [41,42]

Many of the improvements in survival in childhood cancer have been made using new therapies that have attempted to improve on the best available, accepted therapy. Clinical trials in pediatrics are designed to compare potentially better therapy with therapy that is currently accepted. This may be done in a randomized study of 2 treatment arms or by evaluating a single new treatment and comparing the results with those previously obtained with standard therapy.

The current data do not support neuroblastoma screening. Screening infants for neuroblastoma by assay of urinary catecholamine metabolites was initiated in Japan. [43] A large population-based North American study in which most infants in Quebec were screened at ages 3 weeks and 6 months has shown that screening detects many neuroblastomas with favorable characteristics [44,45] that would never have been detected clinically, apparently because the tumors would have spontaneously regressed. Another study of infants screened at 1 year of age shows similar results. [46] Screening at 3 weeks, 6 months, or 1 year of age caused no reduction in the incidence of advanced-stage neuroblastomas with unfavorable biological characteristics in older children nor did it reduce the number of deaths from neuroblastoma in

infants screened at any age. [45,46] Thus, there appear to be no public health benefits from screening infants for neuroblastoma at these ages.

Spontaneous regression of neuroblastoma has been well described in infants, especially in those with the 4S pattern of metastatic spread. [47,48] Regression generally occurs only in tumors with a near triploid number of chromosomes that also lack N-myc amplification and loss of chromosome 1p. Features associated with spontaneous regression include the lack of expression of telomerase, [49,50] expression of Ha-ras, [51] and expression of the neurotrophin receptor TrkA, a nerve growth factor receptor. Recent studies have suggested that selected infants who appear to have asymptomatic, small, low-stage neuroblastoma detected by screening, often have tumors that spontaneously regress and may be observed safely without surgical intervention or tissue diagnosis. [52] Currently, the Children's Oncology Group (COG) is studying whether it is safe to simply observe neonates with small adrenal masses that are presumed to be neuroblastomas (COG ANBL00P2). These masses are usually found on prenatal or incidental ultrasound.

Cellular Classification

One clinicopathologic staging system involves evaluation of tumor specimens for the amount of stromal development, the degree of neuroblastic maturation, and the mitosis-karyorrhexis index of the neuroblastic cells. [1-3] Favorable and unfavorable prognoses are defined on the bases of these histologic parameters and on patient age. The prognostic significance of this classification system, and of related systems using similar criteria, has been confirmed in several studies. [2-4] Neuroblastomas containing many differentiating cells, termed ganglioneuroblastomas, tend to have favorable biological properties and have a better prognosis. [5,6]

Stage Information

The treatment section of this document is organized to correspond to the Children's Oncology Group (COG) risk-based schema for treatment of neuroblastoma. This schema is based on 3 factors: patient age at diagnosis, certain biological characteristics of the patient's neuroblastoma tumor, and the stage of the tumor as defined by the International Neuroblastoma Staging System (INSS). The INSS has replaced the previously used Children's Cancer Group (CCG) and Pediatric Oncology Group (POG) staging systems. The INSS is described below, and the COG risk-based treatment schema is described in Table 1.

A thorough evaluation for metastatic disease should be performed prior to therapy initiation. The following investigations are recommended:

[1]

1. Bone marrow should be assessed by bilateral posterior iliac crest marrow aspirates and trephine (core) bone marrow biopsies to exclude bone marrow involvement. To be considered adequate, core biopsy specimens must contain at least 1 cm of marrow (excluding cartilage).
2. Bone should be assessed by metaiodobenzylguanidine (MIBG) scan (applicable to all sites of disease) and by technetium 99 scan if the results of the MIBG scan are negative or unavailable. Plain radiographs of positive lesions are recommended.
3. Palpable lymph nodes should be clinically examined and histologically confirmed. Nonpalpable lymph nodes should be assessed by computerized tomography (CT) scan with three-dimensional (3D) measurements.
4. The abdomen and liver should be assessed by CT scan and/or magnetic resonance imaging (MRI). Ultrasound is considered suboptimal for accurate 3D measurements.
5. The chest should be examined by anteroposterior and lateral chest radiography. CT scans and/or MRI are necessary if the results are positive or if abdominal disease extends into the chest.
6. Lumbar puncture should be avoided as CNS metastasis at diagnosis is rare and lumbar puncture may be associated with an increased incidence of subsequent development of CNS metastasis. [2]

International Neuroblastoma Staging System

The International Neuroblastoma Staging System (INSS) combines certain features of the previously used POG and CCG systems. [1,3] It has been shown to identify distinct prognostic groups. [1,3-5]

- **Stage 1:** Localized tumor with complete gross excision, with or without microscopic residual disease; representative ipsilateral lymph nodes negative for tumor microscopically (nodes attached to and removed with the primary tumor may be positive).
- **Stage 2A:** Localized tumor with incomplete gross excision; representative ipsilateral nonadherent lymph nodes negative for tumor microscopically.
- **Stage 2B:** Localized tumor with or without complete gross excision, with ipsilateral nonadherent lymph nodes positive for

tumor. Enlarged contralateral lymph nodes must be negative microscopically.

- **Stage 3:** Unresectable unilateral tumor infiltrating across the midline, with or without regional lymph node involvement; or localized unilateral tumor with contralateral regional lymph node involvement; or midline tumor with bilateral extension by infiltration (unresectable) or by lymph node involvement. The midline is defined as the vertebral column. Tumors originating on 1 side and crossing the midline must infiltrate to or beyond the opposite side of the vertebral column.
- **Stage 4:** Any primary tumor with dissemination to distant lymph nodes, bone, bone marrow, liver, skin, and/or other organs (except as defined for stage 4S).
- **Stage 4S:** Localized primary tumor (as defined for stage 1, 2A, or 2B), with dissemination limited to skin, liver, and/or bone marrow (limited to infants less than 1 year of age). Marrow involvement should be minimal (<10% of total nucleated cells identified as malignant by bone biopsy or by bone marrow aspirate). More extensive bone marrow involvement would be considered to be stage 4 disease. The results of the meta-iodobenzylguanidine (MIBG) scan (if performed) should be negative for disease in the bone marrow.

Children’s Oncology Group Neuroblastoma Risk Grouping

In North America, the COG is investigating a risk-based neuroblastoma treatment plan that assigns all patients to low-, intermediate-, and high-risk groups based on age, INSS stage and tumor biology.

The following table outlines the COG neuroblastoma risk group assignment schema. The risk group assignment determines the treatment plan for each patient. Patients assigned to the low-, intermediate-, and high-risk groups have an overall survival of greater than 90%, 70% to 90%, and greater than 30% respectively, 3 years after diagnosis.

INSS Stage	Age	MYCN Status	Shimada Histology	DNA Ploidy	Risk Group
1	0-21y	Any	Any	Any	Low
2A/2B*	<365d	Any	Any	Any	Low
	>or=365d-21y	NonAmp	Any	-	Low

INSS Stage	Age	MYCN Status	Shimada Histology	DNA Ploidy	Risk Group
	>or=365d-21y	Amp	Fav	-	Low
	>or=365d-21y	Amp	Unfav	-	High
3***	<365d	NonAmp	Any	Any	Intermediate
	<365d	Amp	Any	Any	High
	>or=365d-21y	NonAmp	Fav	-	Intermediate
	>or=365d-21y	NonAmp	Unfav	-	High
	>or=365d-21y	Amp	Any	-	High
4***	<365d	NonAmp	Any	Any	Intermediate
	<365d	Amp	Any	Any	High
	>or=365d-21y	Any	Any	-	High
4S**	<365d	NonAmp	Fav	>1	Low
	<365d	NonAmp	Any	=1	Intermediate
	<365d	NonAmp	Unfav	Any	Intermediate
	<365d	Amp	Any	Any	High
Biology Defined By:	MYCN Status: Amplified (Amp) vs. NonAmplified (NonAmp)				
	Shimada Histopathology: Favorable (Fav) vs. Unfavorable (Unfav)				

INSS Stage	Age	MYCN Status	Shimada Histology	DNA Ploidy	Risk Group
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DNA Ploidy: DNA Index (DI) ≥ 1 ; hypodiploid tumors (with DI < 1) will be treated as a tumor with a DI > 1 (DNA index < 1 (hypodiploid) to be considered favorable ploidy).

** INSS 2A/2B symptomatic patients with spinal cord compression, neurologic deficits or other symptoms will be treated on the LOW RISK NB Study with immediate chemotherapy for 4 cycles (Course 1).*

*** INSS 4S infants with favorable biology and clinical symptoms will be treated on the LOW RISK NB Study with immediate chemotherapy until asymptomatic (2-4 cycles). Clinical symptoms defined as: respiratory distress with or without hepatomegaly or cord compression and neurologic deficit or inferior vena cava (IVC) compression and renal ischemia; or genitourinary obstruction; or gastrointestinal obstruction and vomiting; or coagulopathy with significant clinical hemorrhage unresponsive to replacement therapy.*

**** INSS 3 or 4 patients with clinical symptoms as above (or in the investigator's opinion it is in the best interest of the patient) will receive immediate chemotherapy.*

Treatment Option Overview

The treatments described in this summary are based on the Children's Oncology Group (COG) Risk Stratification Schema, which is described in the [Stage Information](#) section. The risk of progression of the tumor causing morbidity and mortality is gauged based on the stage of the tumor, the age of the child at diagnosis, and tumor biology. The biological features considered are the Shimada histology, amplification of the *N-myc* gene, and the number of chromosomes in tumor cells. Treatment information is presented in this format because most children with neuroblastoma in North America are treated according to the COG schema. Accurate determination of biological characteristics, such as Shimada histology, usually requires an open biopsy. Urinary excretion of the catecholamine metabolites vanillylmandelic acid (VMA) and homovanillic acid (HVA) per mg excreted creatinine should be measured prior to therapy. If elevated, these markers can be used to determine the persistence of disease.

This risk-based neuroblastoma treatment plan assigns each patient to a low-, intermediate-, or high-risk group. (Risk groups are defined in Table 1 of the [Stage Information](#) section of this summary). In patients without metastatic disease, initial surgery is performed to establish the diagnosis, to resect as much of the primary tumor as safely possible, to accurately stage disease through sampling of regional lymph nodes that are not adherent to the tumor, and to obtain adequate tissue for biologic studies.

Low Risk

Treatment for patients categorized as low risk (Table 1) consists most commonly of surgery alone but in some cases surgery combined with 6 to 12 weeks of chemotherapy. The chemotherapy consists of carboplatin, cyclophosphamide, doxorubicin, and etoposide. The cumulative dose of each agent is kept low in order to minimize permanent injury from the chemotherapy regimen.[1]

Intermediate Risk

Patients categorized as intermediate risk (Table 1) are treated with surgery and 12 to 24 weeks of the same chemotherapy regimen described above.[2]

High Risk

In contrast, patients categorized as high risk (Table 1) are generally treated with aggressive multiagent chemotherapy consisting of very high doses of the drugs listed above but often also including ifosfamide and high-dose cisplatin. After a response to chemotherapy, resection of the primary tumor should be attempted, followed by myeloablative chemotherapy, sometimes total-body irradiation, and autologous stem cell transplantation. Irradiation of residual tumor and original sites of metastases is often performed before, during, or after myeloablative therapy. After recovery, patients are treated with oral 13-cis-retinoic acid for 6 months. Both myeloablative therapy and retinoic acid improve outcome in patients categorized as high risk.[3]

Radiation

Radiation therapy is reserved for patients with symptomatic life- or organ-threatening tumor that does not respond rapidly enough to chemotherapy, or for intermediate-risk patients whose tumor has responded incompletely to both chemotherapy and attempted resection and also has unfavorable biologic characteristics. Radiation therapy to the primary site is recommended for high-risk patients even in cases of complete resection.

Relapse Therapy for Low and Intermediate Risk Patients

As part of the COG treatment plan, specific relapse therapy is defined for low- and intermediate-risk patients determined by patient age at recurrence, stage and biology of the recurrence.

Urgent Chemotherapy

Immediate treatment should be undertaken for symptomatic spinal cord compression. Neurologic recovery is more likely the less the severity of compromise and the shorter the duration of symptoms. Neurologic outcome appears to be similar whether cord compression is treated with chemotherapy, radiation therapy, or surgical laminectomy. However, laminectomy may result in later scoliosis, and chemotherapy is often needed whether or not surgery or radiation is used.[4,5] The COG

neuroblastoma treatment plan recommends immediate chemotherapy for cord compression in patients classified as low or intermediate risk.[1,2]

Observation without Surgery of Localized, Presumed Adrenal Neuroblastoma in Infants

Studies suggest that selected presumed neuroblastomas detected in infants by screening may safely be observed without obtaining a definitive histologic diagnosis and without surgical intervention, thus avoiding potential complications of surgery in the newborn.[6,7] The experience with tumors detected by mass urinary catecholamine metabolite screening in Japan appears to be applicable to tumors detected by prenatal or perinatal ultrasound in the United States. Twenty-six infants who had presumed Evans stage I, II, or IVS by imaging, urinary VMA and HVA levels less than 50 mcg per mg creatinine, no tumor involvement of great vessels or invasion into the spinal canal, and tumor size less than 5 cm were observed frequently with imaging. Biopsy and tissue diagnosis were not obtained initially. The tumor increased in size in about one third of the infants and was resected without any apparent increase in stage. All had favorable biologic features. In two thirds, after observation for 6 to 73 months, no surgery had been performed, the VMA and HVA had normalized, and in several cases the tumors had become undetectable by imaging.[6] The COG is currently investigating systematic observation without surgery for infants with presumed small Evans stage I adrenal neuroblastoma detected by prenatal or perinatal ultrasound.

Treatment of Low Risk Neuroblastoma

In North America, the Children's Oncology Group (COG) is investigating a risk-based neuroblastoma treatment plan that assigns all patients to low-, intermediate-, and high-risk groups based on age, International Neuroblastoma Staging System (INSS) stage, and tumor biology (*N-myc* gene amplification, Shimada histology, and DNA ploidy) [1] (Risk groups are defined in [Table 1](#) of the Stage Information section of this summary).

Patients with low-risk neuroblastoma have a cure rate greater than 90%.[2-5] The following tumors are categorized as low risk (see [Table 1](#)):

1. INSS stage 1 tumors in patients of any age. Stage 1 is defined as gross complete resection.
2. INSS stage 2A and 2B tumors in infants.
3. INSS stage 2A and 2B tumors in children over 1 year of age and in whom the tumor demonstrates either favorable Shimada histopathology or nonamplification of *N-myc*.
4. INSS stage 4S tumors in infants less than 1 year of age with all favorable biological features (i.e., *N-myc* not amplified, favorable Shimada histopathology, and hyperdiploid DNA).

Low-risk neuroblastomas are generally treated with surgical resection and observation or observation alone.[1]

Stage 2 low-risk tumors are treated with chemotherapy only if less than 50% of the tumor has been resected. In the other low-risk patients, chemotherapy is recommended only for life- or organ-threatening symptoms that cannot be relieved by safe surgical resection of the mass. Such symptoms include respiratory distress, renal or bowel ischemia, spinal cord compression, gastrointestinal or genitourinary obstruction, or coagulopathy.[1] Chemotherapy is given for 6 to 24 weeks and consists of moderate doses of carboplatin, cyclophosphamide, doxorubicin, and etoposide. The cumulative dose of each agent is kept low in order to minimize permanent injury from the chemotherapy regimen.[1] Radiation therapy is reserved for patients with symptomatic life- or organ-threatening tumor that does not respond rapidly enough to chemotherapy.

Studies suggest that selected presumed neuroblastomas detected in infants by screening may be safely observed without surgical intervention.[6,7] The COG is investigating systematic observation without diagnostic surgery for selected infants with presumed INSS stage 1 adrenal neuroblastoma detected by prenatal or perinatal ultrasound (COG-ANBL00P2).

The treatment of children with low-risk stage 4S disease is dependent on clinical presentation.[8,9] Children who are clinically stable with this special pattern of neuroblastoma may not require therapy. The development of complications, such as functional compromise from massive hepatomegaly, is an indication for intervention, especially in infants younger than 2 to 3 months of age.[8,10,11] In a study of 80 infants with 4S disease, those who were asymptomatic had 100% survival with supportive care only, and patients with symptoms had an 81% survival rate when they received low-dose chemotherapy.[10] Resection of primary tumor is not associated with improved outcome.[8-10]

The COG neuroblastoma treatment plan also defines the treatment for progression or recurrence of low-risk neuroblastoma. The treatment is dependent on the characteristics of the progression or recurrence (see Recurrent Neuroblastoma section).

Treatment of Intermediate Risk Neuroblastoma

In North America, the Children's Oncology Group (COG) is investigating a risk-based neuroblastoma treatment plan that assigns all patients to low-, intermediate-, and high-risk groups based on age, International Neuroblastoma Staging System (INSS) stage, and tumor biology (*N-myc* gene amplification, Shimada histology, and DNA ploidy). (Risk groups are defined in Table 1 of the Stage Information section of this summary)

Patients with Intermediate-Risk neuroblastoma generally have a cure rate of 70% to 90%. The following patients are categorized as intermediate risk (see [Table 1](#)):

1. INSS stage 3 tumors in infants less than 1 year of age and in whom the tumor lacks N-myc gene amplification.
2. INSS stage 3 tumors in children 1 year of age or older and in whom the tumor lacks N-myc gene amplification and has favorable Shimada histopathology.
3. INSS stage 4 tumors in infants less than 1 year of age and in whom the tumor lacks N-myc gene amplification.
4. INSS stage 4S tumors in infants less than 1 year of age and in whom the tumor lacks N-myc gene amplification and has either unfavorable Shimada histopathology or is near diploid in chromosome number, or both.

There is considerable variation in outcome, and, therefore, in treatment for children with stage 3 disease (tumor involving both sides of the midline by virtue of either invasion into normal tissues or lymph node metastasis). Infants younger than 1 year of age have a greater than 80% chance of cure while older children have a cure rate of 50% to 70% with current, relatively intensive therapy.^[1-4] In one study, those with favorable compared to unfavorable biologic features (Shimada histology and *N-myc* gene amplification) had event-free survival rates of almost 100% and about 50%, respectively.^[5] In cases of abdominal neuroblastoma thought to involve the kidney, nephrectomy should not be undertaken before a trial of chemotherapy has been given.^[6]

Patients classified as intermediate risk with stage 3 tumors with favorable or unfavorable Shimada histology are treated with 12 weeks and 24 weeks of chemotherapy respectively. In patients classified as intermediate risk with favorable biology, radiation therapy is reserved for patients with symptomatic life- or organ-threatening tumor that does not respond rapidly enough to chemotherapy. In patients classified as intermediate risk with unfavorable biologic features, radiation therapy is given if residual viable tumor remains after 24 weeks of chemotherapy and second-look surgery.

Survival of patients with INSS stage 4 disease is strongly dependent on age. Children younger than 1 year of age at diagnosis have a good chance of long-term survival (5-year disease-free survival rate of 50% to 80%), ^[7,8] with outcome particularly dependent on tumor cell ploidy (hyperploidy confers a favorable prognosis while diploidy predicts early treatment failure).^[2,9]

Infants less than 1 year of age at diagnosis with INSS stage 4 neuroblastoma that does not have *N-myc* gene amplification are categorized as intermediate risk.[10] These infants are treated with 12 weeks of chemotherapy if the tumor has both favorable Shimada histology and hyperdiploidy, and if not, with 24 weeks of chemotherapy.

Infants less than 1 year of age at diagnosis with INSS stage 4S neuroblastoma without amplification of the *N-myc* gene, but with unfavorable Shimada histology, diploid DNA or both, are classified as intermediate risk. These infants are treated with 24 weeks of chemotherapy.

Chemotherapy for intermediate-risk patients consists of moderate doses of carboplatin, cyclophosphamide, doxorubicin, and etoposide given for 12 to 24 weeks. The cumulative dose of each agent is kept low in order to minimize permanent injury from the chemotherapy regimen.[11]

The COG Neuroblastoma Treatment Plan also defines the treatment for progression or recurrence of intermediate-risk neuroblastoma. This treatment depends on the characteristics of the progression or recurrence (see the [Recurrent Neuroblastoma](#) section of this summary).

Treatment of High Risk Neuroblastoma

In North America, the Children's Oncology Group (COG) is investigating a risk-based neuroblastoma treatment plan that assigns all patients to low-, intermediate-, and high-risk groups base on age, International Neuroblastoma Staging System (INSS) stage, and tumor biology (*N-myc* gene amplification, Shimada histology, and DNA ploidy).[1] (Risk groups are defined in [Table 1](#) of the Stage Information section of this summary)

The following patients are considered to have high-risk neuroblastoma (see [Table 1](#)):

1. INSS stage 2A/2B tumors in children over 1 year of age and in whom the tumor has both unfavorable Shimada histology and *N-myc* gene amplification.
2. INSS stage 3 tumors in infants less than 1 year of age and in whom the tumor demonstrates *N-myc* gene amplification.
3. INSS stage 3 tumors in children over 1 year of age and in whom the tumor demonstrates either *N-myc* gene amplification or unfavorable Shimada histology.

4. INSS stage 4 tumors in infants less than 1 year of age at diagnosis and in whom the tumor demonstrates *N-myc* gene amplification.[2]
5. INSS stage 4 tumors in children over 1 year of age.
6. INSS stage 4S tumors in infants less than 1 year of age at diagnosis and in whom the tumor demonstrates *N-myc* gene amplification.

For children with high-risk neuroblastoma, long-term survival ranges from 10% to 40%. Children with aggressively treated, high-risk neuroblastoma may develop late recurrences, some more than 5 years after completion of therapy.[3,4] A randomized study was performed comparing high-dose therapy with purged autologous bone marrow transplantation versus 3 cycles of intensive consolidation chemotherapy. The 3-year event-free survival was significantly better in the autologous bone marrow transplantation arm (34%) compared to the consolidation chemotherapy arm (18%). In addition, patients on this study were subsequently randomized to stop therapy or to receive 6 months of 13-cis-retinoic acid.[5] Patients who received 13-cis-retinoic acid had significantly better 3-year event-free survival than patients receiving no maintenance therapy. This was true for all patient subgroups. Based on these results, future clinical trials will build upon autologous stem cell transplantation and cis-retinoic acid for high-risk neuroblastoma.[5]

The potential benefit of aggressive surgical approaches in high-risk patients with metastatic disease in order to achieve complete tumor resection, either at the time of diagnosis or following chemotherapy, has not been unequivocally demonstrated. Two studies reported that complete resection of the primary tumor at diagnosis improved survival; however, the outcome in these patients may be more dependent on the biology of the tumor, which itself may determine resectability, than on the extent of surgical resection.[6-9]

Standard Treatment

Patients classified as high risk receive treatment with an aggressive regimen of combination chemotherapy consisting of very high drug doses. Drugs often used include cyclophosphamide, ifosfamide, cisplatin, carboplatin, vincristine, doxorubicin, and etoposide. After a response to chemotherapy, resection of the primary tumor should be attempted, followed by myeloablative chemotherapy and stem cell rescue (bone marrow and/or peripheral blood stem cell transplantation). The use of purged stem cells is under investigation. Radiation to the primary tumor site should be undertaken whether or not a complete excision was obtained. Radiation of

sites of metastatic disease is determined on an individual case basis. After recovery, patients are treated with oral 13-cis-retinoic acid for 6 months. Both myeloablative therapy and postchemotherapy retinoic acid improve outcome in patients categorized as high risk.[5]

Recurrent Neuroblastoma

The prognosis and treatment of recurrent or progressive neuroblastoma depends on many factors including initial stage, tumor biological characteristics at recurrence, the site and extent of the recurrence or progression, and on the previous treatment as well as individual patient considerations. When recurrence is widespread, the prognosis is usually poor despite additional intensive therapy.[1,2] In selected cases, recurrence may be treated successfully with limited interventions. The combination of cyclophosphamide plus topotecan has been shown to be active in patients with recurrent or refractory disease.[3]

Unlike at initial presentation, central nervous system involvement is common. Inward compression of the brain from cranial metastases can occur, and rarely meningeal and isolated intracranial metastasis occur. Early recognition and treatment of central nervous system involvement may result in reduced neurologic impairment.[4,5]

Clinical trials are appropriate and should be considered.[6-11] Information about ongoing clinical trials is available from the [NCI Cancer.gov Web site](http://www.nci.nih.gov).

Children's Oncology Group Treatment Plan

In North America, the Children's Oncology Group is investigating a risk-based neuroblastoma plan that assigns all patients to Low-, Intermediate-, and High-Risk groups based on age, INSS stage, and tumor biology (*N-myc* gene amplification, Shimada histology, and DNA ploidy).[12] Treatment of recurrent disease is determined by the risk group at the time of diagnosis (Table 1), extent of disease at recurrence, patient age at recurrence, and the tumor biology at recurrence. If tumor is unavailable for biological studies at recurrence, the biology of the tumor at the time of diagnosis is used to help determine treatment.

Recurrent Neuroblastoma in Patients Initially Classified as Low Risk

(Risk categories are defined in [Table 1](#) in the Stage Information section of the summary).

Local/Regional Recurrence is Resected if Possible

1. Those with favorable biology are observed if resection is total or near total, while those with favorable biology and a less than near-total resection are treated with 12 weeks of chemotherapy.
2. Infants less than 1 year of age at the time of local/regional recurrence whose tumors have any unfavorable biologic properties are observed if resection is total or near total. If the resection is less than near total, these same infants are treated with 24 weeks of chemotherapy.

Chemotherapy consists of moderate doses of carboplatin, cyclophosphamide, doxorubicin, and etoposide. The cumulative dose of each agent is kept low in order to minimize permanent injury from the chemotherapy regimen.[13] Older children with local recurrence with either unfavorable Shimada histology or *N-myc* gene amplification have a poor prognosis and should be treated with an aggressive regimen of combination chemotherapy consisting of very high doses of the drugs listed above but often also including ifosfamide and high-dose cisplatin. Both myeloablative therapy and postchemotherapy retinoic acid may improve outcome of patients with a poor prognosis.[14]

Metastatic Recurrence

Metastatic recurrent or progressive neuroblastoma in an infant initially categorized as low risk (Table 1) and less than 1 year of age at recurrence, whether the patient has INSS stage 1, 2, or 4S at the time of diagnosis is treated according to tumor biology:

1. If the biology is completely favorable, metastasis is in a 4S pattern, and the recurrence or progression is within 3 months of diagnosis, the patient is observed systematically.
2. If the metastatic progression or recurrence with completely favorable biology occurs greater than 3 months after diagnosis or not in a 4S pattern, then the primary tumor is resected if possible and 12 to 24 weeks of chemotherapy is given, depending on response.
3. If the tumor in the infant with metastatic recurrence or progression has unfavorable Shimada histology and/or is diploid, the primary tumor is resected if possible and 24 weeks of chemotherapy is given.

Chemotherapy consists of moderate doses of carboplatin, cyclophosphamide, doxorubicin, and etoposide. The cumulative dose of each agent is kept low in order to minimize permanent injury from the chemotherapy regimen.[13]

Any child initially categorized as low risk who is over 1 year of age at the time of metastatic recurrent or progressive disease usually has a poor prognosis and should be treated with an aggressive regimen of combination chemotherapy consisting of very high doses of the drugs listed above but often also including ifosfamide and high-dose cisplatin. Both myeloablative therapy and post-chemotherapy retinoic acid may improve outcome of patients with a poor prognosis.[14]

Wilms' Tumor (nephroblastoma) and other renal tumors

This information has been taken from the NIH web site for Wilm's tumor. The references have been removed to save space. If you would like to see any of the references please go to www.cancer.gov/cancerinfo/pdq/pediatrictreatment and click on Wilm's Tumor. These recommendations were current as of 7/2003

General Information

This cancer treatment information summary provides an overview of the prognosis, diagnosis, classification, staging, and treatment of Wilms' tumor and other childhood kidney tumors (clear cell sarcoma of the kidney [CCSK], rhabdoid tumor of the kidney, and neuroepithelial tumor of the kidney [NETK]). (Refer to the PDQ summary on Unusual Cancers of Childhood for more information about childhood renal cell carcinoma treatment).

The National Cancer Institute provides the PDQ pediatric cancer treatment information summaries as a public service to increase the availability of evidence-based cancer information to health professionals, patients and the public. These summaries are updated regularly according to the latest published research findings by an Editorial Board of pediatric oncology specialists.

Cancer in children and adolescents is rare. Children and adolescents with cancer should be referred to medical centers that have a multidisciplinary team of cancer specialists with experience treating the cancers that occur during childhood and adolescence. This multidisciplinary team approach incorporates the skills of the primary care physician, pediatric surgical subspecialists, radiation oncologists, pediatric medical oncologists/hematologists, rehabilitation specialists, pediatric nurse specialists, social workers, and others in order to ensure that children receive treatment, supportive care, and rehabilitation that will achieve optimal survival and quality of life. Refer to the PDQ Supportive Care summaries for specific information about supportive care for children and adolescents with cancer.

Guidelines for pediatric cancer centers and their role in the treatment of pediatric patients with cancer have been outlined by the American Academy of Pediatrics.[1] At these pediatric cancer centers, clinical trials are available for most of the types of cancer that occur in children and adolescents, and the opportunity to participate in these trials is offered to most patients/families. Clinical trials for children and adolescents with

cancer are generally designed to compare potentially better therapy with therapy that is currently accepted as standard. Most of the progress made in identifying curative therapies for childhood cancers have been achieved through clinical trials. Information about ongoing clinical trials is available from the [NCI Cancer.gov Web site](http://www.nci.nih.gov).

Wilms' tumor is a curable disease in the majority of affected children. Approximately 500 cases are diagnosed in the United States annually. More than 90% of patients survive 4 years after diagnosis which is an improvement over the 80% survival observed from 1975 to 1984.[2] The prognosis is related not only to the stage of disease at diagnosis, the histopathologic features of the tumor, patient age, and tumor size but also to the team approach to each patient by the pediatric surgeon, radiation oncologist, and pediatric oncologist.[2-4] Previous clinical trials have, in part, evaluated whether reduced therapy is sufficient to control disease in patients with early-stage, favorable-histology Wilms' tumor with some success.[5,6] Ongoing clinical trials are evaluating biologic factors in the development of Wilms' tumor, clear cell sarcoma, and rhabdoid tumor of the kidney.[2]

Wilms' tumor normally develops in otherwise healthy children but just under 10% occur in individuals with recognized malformations. The phenotypes associated with Wilms' tumor can be classified as overgrowth or nonovergrowth syndromes. Overgrowth syndromes are the result of excessive prenatal and postnatal somatic growth, and result in macroglossia, nephromegaly, and hemihypertrophy. The two most common overgrowth disorders associated with Wilms' tumor are Beckwith-Wiedemann syndrome and isolated hemihypertrophy.[7-11] Others include Perlman syndrome, Sotos syndrome, and Simpson-Golabi-Behemel syndrome. Nonovergrowth disorders associated with Wilms' tumor include isolated aniridia, trisomy 18, aniridia in combination with genitourinary malformations, and mental retardation (AGR) syndrome, Bloom syndrome, and Denys-Drash syndrome.[12] Children with a predisposition to develop Wilms' tumor (e.g., Beckwith-Wiedemann, hemihypertrophy) should be screened with ultrasound every 3 months until they reach 8 years of age.[7-11]

Wilms' tumor (hereditary or sporadic) appears to result from changes in one or more of several genes. Specific germ-line mutations in one of these genes (Wilms' tumor gene-1, *WT1*) located on the short arm of chromosome 11 (band 11p13) are not only associated with Wilms' tumor but also cause a variety of genitourinary abnormalities such as cryptorchidism and hypospadias,[13] and the rare Denys-Drash syndrome. A gene that causes aniridia is located near the *WT1* gene on chromosome 11p13, and deletions encompassing the *WT1* and aniridia genes may

explain the association between aniridia and Wilms' tumor. Patients with aniridia or hemihypertrophy should be screened with ultrasound every 3 months until they are 6 years of age.[7] Children with Wilms' Aniridia-Genitourinary abnormalities-(mental) Retardation (WAGR) syndrome are at increased risk of eventually developing renal failure and should be monitored. Patients with Wilms' tumor and aniridia without genitourinary abnormalities are at lesser risk but should be monitored.[14] There appears to be a second Wilms' tumor gene at or near the Beckwith-Wiedemann gene locus on chromosome 11p15, and children with Beckwith-Wiedemann syndrome are at increased risk for developing Wilms' tumor. Approximately one fifth of patients with Beckwith-Wiedemann syndrome who develop Wilms' tumor present with bilateral disease, primarily at diagnosis, although metachronous recurrence is also observed.[7-9]

Approximately one third of Wilms' tumors have loss of genetic material in the tumor cells from the short arm of chromosome 11, encompassing 1 or both of the Wilms' tumor gene regions on this chromosome. Genes on other chromosomes may also have an etiologic role in Wilms' tumor, and loss of genetic material from chromosome 16 and/or chromosome 1p occurs in some tumors.[15,16] Many Wilms' tumors appear to arise from abnormally retained embryonic kidney precursor cells arranged in clusters termed nephrogenic rests. The different genetic lesions are associated with different subtypes of nephrogenic rests.[17] Wilms' tumors that develop from intralobar nephrogenic rests generally contain heterologous elements such as smooth muscle, cartilage, and fat cells, and are associated with loss of DNA on the short arm of chromosome 11p and occasionally with WT1 gene mutation. In contrast, Wilms' tumors that develop from perilobar nephrogenic rests, which appear to reflect a slightly later stage in renal embryonic development and are generally found in older children are associated with loss of imprinting of the IGF2 gene, which stimulates cell proliferation. Usually the maternal copy of IGF2 is imprinted, that is, not expressed in the embryo, and when it is expressed in the tumor, twice as much IGF2 RNA is made. Perilobar rests are also associated with Wilms' tumors in children with Beckwith-Wiedemann syndrome.[18]

Despite the number of genes that appear to be involved in the development of Wilms' tumor, hereditary Wilms' tumor is uncommon, with 1% to 2% of patients having a positive family history for Wilms' tumor.[19,20] The risk of Wilms' tumor among offspring of persons who have had unilateral (i.e., sporadic) tumors is quite low (< 2%).[21] Siblings of children with Wilms' tumor have a low likelihood of developing Wilms' tumor.[19] About 4% to 5% of patients have bilateral Wilms' tumors, but these are not usually hereditary.[19,20] Many bilateral tumors are present at the time Wilms' tumor is first diagnosed (i.e., synchronous), but a second Wilms' tumor may also develop later in the remaining kidney of 1% to 3% of children treated

successfully for Wilms' tumor. The incidence of such metachronous bilateral Wilms' tumors is much higher in children whose original Wilms' tumor was diagnosed at less than 12 months of age and/or whose resected kidney contains nephrogenic rests. Periodic abdominal ultrasound is recommended for early detection of metachronous bilateral Wilms' tumor as follows: children with nephrogenic rests in the resected kidney (if < 48 months of age at initial diagnosis) - every 3 months for 6 years; children with nephrogenic rests in the resected kidney (if > 48 months of age at initial diagnosis) - every 3 months for 4 years; other patients - every 6 months for 2 years, then yearly for an additional 1 to 3 years.[22,23]

Clear cell sarcoma of the kidney, rhabdoid tumor of the kidney, and neuroepithelial tumor of the kidney (see descriptions in the Cellular Classification section) are childhood renal tumors unrelated to Wilms' tumor. Because of their renal location, they have been treated on clinical trials developed by the National Wilms' Tumor Study Group. The approach to their treatment, however, is distinctive from that of Wilms' tumor, and requires timely and accurate diagnosis.

Cellular Classification

Wilms' tumor

Although most patients with a histologic diagnosis of Wilms' tumor fare well with current treatment, approximately 10% of patients have histopathologic features that are associated with a poorer prognosis, and, in some types, with a high incidence of relapse and death. Wilms' tumor can be separated into 2 prognostic groups on the basis of histopathology:

- **Favorable histology:** Histologically mimics development of a normal kidney consisting of 3 components: blastema, epithelium (tubules) and stroma. There is no anaplasia.
- **Unfavorable histology:** Characterized by anaplasia (extreme cellular pleomorphism and atypia, diffuse).[1] Focal anaplasia may not confer nearly as poor a prognosis as diffuse anaplasia. Anaplasia is associated with resistance to chemotherapy and may still be detected after preoperative chemotherapy.[2,3]

Clear Cell Sarcoma

Clear cell sarcoma (CCSK) of the kidney is not a Wilms' tumor variant, but it is an important primary renal tumor associated with a significantly higher

rate of relapse and death than favorable histology Wilms' tumor. In addition to pulmonary metastases, clear cell sarcoma also spreads to bone, brain and soft tissue. The classic pattern of CCSK is defined by nests or cords of cells separated by regularly spaced arborizing fibrovascular septa.[4]

Rhabdoid Tumor of the Kidney

Initially thought to be a rhabdomyosarcomatoid variant of Wilms' tumor, it is a distinctive and highly malignant tumor type. A distinct clinical presentation with fever, hematuria, young age (mean 11 months), and high tumor stage at presentation suggests a diagnosis of rhabdoid tumor of the kidney.[5] Rhabdoid tumor of the kidney (RTK) tends to metastasize not only to the lungs, but also to the brain. As many as 10% to 15% of patients with RTK also have central nervous system lesions, now designated atypical teratoid tumors.[6,7] The most distinctive features of rhabdoid tumors of the kidney are rather large cells with large vesicular nuclei, a prominent single nucleolus, and in some cells, the presence of globular eosinophilic cytoplasmic inclusions.

The characteristic molecular lesion found in rhabdoid tumors of the kidney is loss-of-function mutations of the hSNF5/INI1 gene, which is located at chromosome band 22q11.2.[8,9] This same molecular abnormality is found in tumors of the central nervous system termed atypical teratoid and rhabdoid tumors.[8,9] Some patients with rhabdoid tumors have constitutional mutations of the hSNF5/INI1 gene,[8,10] and these children are at increased risk for second primary brain tumors.[11]

Neuroepithelial Tumors of the Kidney

Neuroepithelial tumors of the kidney (NETK) is an extremely rare entity demonstrating a unique proclivity for young adults. It is a highly aggressive neoplasm, more often presenting with penetration of the renal capsule, extension into the renal vein, and metastases.[12,13] Primary neuroepithelial tumors of the kidney consist of primitive neuroectodermal tumors (PNET) characterized by CD99 (MIC-2) immunostaining and the EWS/FLI-1 or closely related gene fusion products and small cell carcinomas characterized by chromogranin positivity. The two subtypes may be difficult to distinguish. Within both types of NETK, focal, atypical histologic features have been seen including clear cell sarcoma, rhabdoid tumor, malignant peripheral nerve sheath tumors, and paraganglioma.[12]

Stage Information

Wilms' tumor

A clinicopathologic staging system has been devised by the National Wilms' Tumor Study Group. The clinical stage is determined by the pediatric surgeon in the operating room and is confirmed by the pathologist. Staging, which is based on the degree of gross and microscopic tumor extension, is the same for tumors with favorable or unfavorable histologic features. Thus, patients should be characterized by a statement of both criteria (for example, stage II, favorable histology or stage II, unfavorable histology).[1,2]

A description of the staging system and incidence from the Third National Wilms' Tumor Study (NWTS-3[2]) is given below.

Stage I (43% of patients)

Stage I Wilms' tumor is defined as tumor limited to the kidney and completely excised. The surface of the renal capsule is intact. The tumor is not ruptured before or during removal. The vessels of the renal sinus are not involved. There is no residual tumor apparent beyond the margins of excision.

Stage II (23% of patients)

Stage II Wilms' tumor is defined as tumor that extends beyond the kidney but is completely excised. No residual tumor is apparent at or beyond the margins of excision. Any of the following conditions may exist:

1. Regional extension of the tumor, i.e., penetration through the outer surface of the renal capsule into the perirenal soft tissue or more than 1 to 2 mm of tumor invasion into the renal sinus.
2. Vessels outside the kidney are infiltrated or contain tumor thrombus.
3. The tumor was biopsied or there was local spillage of tumor confined to the flank.
4. In the National Wilms' Tumor Study 5 (NWTS-5), tumors with evidence of invasion of the vessels in the renal sinus (without any other reason to classify as stage II) were classified as stage II versus the stage I classification they were given in NWTS-1 through 4.[3]

Stage III (23% of patients)

Stage III Wilms' is defined as residual tumor confined to the abdomen. One or more of the following conditions may exist:

1. Lymph nodes in the renal hilus, the periaortic chains, or beyond are found to contain tumor on biopsy. Lymph node involvement in the thorax or other extra-abdominal sites would be a criterion for stage IV.
2. There has been diffuse peritoneal contamination by the tumor, such as by spillage of tumor beyond the flank before or during surgery or by tumor growth that has penetrated through the peritoneal surface.
3. Implants are found on the peritoneal surfaces.
4. Tumor extends beyond the surgical margins either microscopically or grossly.
5. Tumor is not completely resectable because of local infiltration into vital structures.

Stage IV (10% of patients)

Stage IV Wilms' tumor is defined as the presence of hematogenous metastases. There are metastatic deposits beyond stage III, e.g., to the lung, liver, bone, or brain or to a combination of these sites.

Stage V (5% of patients)

Stage V Wilms' tumor is defined as bilateral renal involvement at the time of initial diagnosis.

For patients with bilateral involvement, an attempt should be made to stage each side according to the above criteria on the basis of extent of disease prior to biopsy. The 4-year survival was 94% for those patients whose most advanced lesion was stage I or stage II; 76% for those whose most advanced lesion was stage III.[4]

Stage I-IV Anaplasia (unfavorable histology)

Children with stage I anaplastic tumors have an excellent prognosis. They can be managed with the same regimen given to stage I favorable histology patients. Children with stage II through stage IV diffuse anaplasia, however, represent a higher-risk group. These tumors are more resistant to the chemotherapy traditionally used in children with Wilms' tumor (favorable histology).[4]

Treatment Option Overview

Wilms' tumor

Because of the relative rarity of this tumor, all patients with Wilms' tumor should be considered for entry into a clinical trial. Treatment planning by a multidisciplinary team of cancer specialists (pediatric surgeon or pediatric urologist, pediatric radiation oncologist, and pediatric oncologist) with experience treating Wilms' tumor is required to determine and implement optimum treatment.

The National Wilms' Tumor Study Group, which is now part of the Children's Oncology Group (COG), has established standard treatment for Wilms' tumor in North America which consists of surgery followed by chemotherapy and, in some patients, radiation therapy.[1,2] Pulmonary nodules not detected on chest radiographs but visible on computed tomography of the chest do not mandate treatment with whole-lung irradiation.[3,4] The major treatment conclusions of the National Wilms Tumor Studies (NWTS 1-4) are:

1. Routine, postoperative radiation therapy of the flank is not necessary for children with stage I tumors or stage II tumors with favorable histology (FH) when post nephrectomy combination chemotherapy consisting of vincristine and dactinomycin is administered.
2. The prognosis for patients with stage III/FH is best when treatment includes a) dactinomycin, vincristine, doxorubicin, and 1,000 cGy of radiation therapy to the flank; or b) dactinomycin, vincristine, and 2,000 cGy of radiation therapy to the flank.
3. The addition of cyclophosphamide to the combination of vincristine, dactinomycin, and doxorubicin does not improve prognosis for patients with Stage IV/FH tumors.
4. Single-dose ("pulse-intensive") treatment with dactinomycin (stages I-II/FH, stage I anaplastic), and doxorubicin (stage III/FH, stages III-IV, or stages I-IV clear cell sarcoma of the kidney) is equivalent to the divided-dose courses, and results in the same event-free survival, greater dose-intensity, and is associated with less toxicity and expense.[5]
5. 18 weeks of therapy is adequate for patients with stage I/FH whereas other patients can be treated with 6 months of therapy instead of 15 months.[1,5-8]

Operative principles have evolved from NWTS trials. The most important role for the surgeon is to ensure complete tumor removal without rupture

and perform an assessment of the extent of disease. Radical nephrectomy-lymph node sampling via a transabdominal incision is the procedure of choice. Partial nephrectomy is currently not recommended, although it may be considered for very small tumors discovered by ultrasound screening.[9] The contralateral kidney must be palpated and inspected through an opening in the fascia. Hilar, periaortic, iliac and celiac lymph node sampling is mandatory. Furthermore, any suspicious node basin should be sampled. Margins of resection, residual tumor, and any suspicious node basins should be marked with titanium clips. Liver wedge resection or partial duodenal or colonic resections are acceptable for complete en bloc excision. Wilms' tumor arising in a horseshoe kidney is rare and accurate preoperative diagnosis is important in planning the operative approach. Primary resection is possible in the majority of cases. Inoperable cases can usually be resected after chemotherapy.[10]

Patients with massive, nonresectable unilateral tumors, bilateral tumors, or venacaval tumor thrombus above the hepatic veins are candidates for preoperative chemotherapy because of the risk of initial surgical resection.[11] Preoperative chemotherapy should follow a biopsy, which may be performed percutaneously.[12-17] Preoperative chemotherapy makes the tumor removal easier and may reduce the frequency of surgical complications.[11,17,18]

Newborns and all infants less than 12 months of age require a reduction in chemotherapy doses to 50% of those given to older children.[19] This reduction diminishes toxic effects reported in children in this age group enrolled in NWTs studies while maintaining an excellent overall outcome.[20] Liver function tests in children with Wilms' tumor should be monitored closely during the early course of therapy based on hepatic toxic effects (veno-occlusive disease) reported in those patients.[21,22] Dactinomycin should not be administered during radiation therapy. Children treated for Wilms' tumor are at increased risk for developing second malignant neoplasms. This risk depends on the intensity of their therapy, including the use of radiation and doxorubicin, and on possible genetic factors.[23] Congestive heart failure has been shown to be a risk in children treated with doxorubicin with the degree of risk influenced by cumulative doxorubicin dose, irradiation to the heart, and gender (females at increased risk).[24] Efforts, therefore, have been aimed toward reducing the intensity of therapy when possible.

Clear Cell Sarcoma of the Kidney:

The approach for treating clear cell sarcoma of the kidney (CCSK) is different from Wilms' since the overall survival of children with CCSK

remains considerably lower than patients with favorable histology Wilms' tumor. In the NSTS-3 study, the addition of doxorubicin to the combination of vincristine, dactinomycin, and radiation therapy resulted in an improvement in disease-free survival for patients with clear cell sarcoma of the kidney.[25]

Under the current study, children with stage II-IV diffuse anaplasia and stage I-IV clear cell sarcoma of the kidney are currently treated with a new chemotherapeutic regimen combining vincristine, doxorubicin, cyclophosphamide, and etoposide in an attempt to further improve the survival of these high-risk groups. All these patients will receive radiation therapy to the tumor bed.

Rhabdoid tumor of the kidney

Patients with rhabdoid tumor of the kidney continue to have a poor prognosis despite aggressive therapy.

Stage I Wilms' Tumor

Regardless of histology, all stage I Wilms' tumor patients have an excellent prognosis with the same treatment.[1]

Favorable histology tumors (94.9% 2-year relapse-free survival [RFS]; 98.7% 2-year survival):[2]

- Nephrectomy with lymph node sampling and 18 weeks of chemotherapy with vincristine and pulse-intensive dactinomycin.

Focal or diffuse anaplasia (87.5% 2-year RFS, 85.5% 2-year survival):[2]

- Nephrectomy with lymph node sampling and 18 weeks of chemotherapy with vincristine and pulse-intensive dactinomycin.

It may be possible to treat a subset of Stage I Wilms' tumor patients with surgery alone without chemotherapy. The Children's Oncology Group (COG) is planning a large study to address this question. A pilot study was performed in National Wilms' Tumor Study Group-5 (NWTS-5) infants with stage I/FH (favorable histology) Wilms' tumor who were less than 24 months of age and whose nephrectomy specimen weighed less than 550 grams.[3] Even though there was 2 times the recurrence rate in this trial, it's

possible that most of these patients may be successfully salvaged with chemotherapy.

Stage II Wilms' Tumor

Favorable histology tumors (85.9% 2-year relapse-free survival [RFS]; 97% 2-year survival):[1]

- Nephrectomy with lymph node sampling and 18 weeks of chemotherapy with vincristine and pulse-intensive dactinomycin.

Focal anaplasia:

- Nephrectomy with lymph node sampling, abdominal irradiation, and 24 weeks of chemotherapy with vincristine, doxorubicin, and pulse-intensive dactinomycin.

Diffuse anaplasia (70% 4-year survival):

- Nephrectomy with lymph node sampling, abdominal irradiation, and 24 weeks of chemotherapy with vincristine, doxorubicin, etoposide, cyclophosphamide, and mesna.[2]

Stage III Wilms' Tumor

Favorable histology tumors (91.1% 2-year relapse-free survival [RFS]; 98.2% 2-year survival):[1]

- Nephrectomy with lymph node sampling, abdominal irradiation, and 24 weeks of chemotherapy with vincristine, doxorubicin, and pulse-intensive dactinomycin.

Focal anaplasia:

- Nephrectomy with lymph node sampling, abdominal irradiation, and 24 weeks of chemotherapy with vincristine, doxorubicin, and pulse-intensive dactinomycin.

Diffuse anaplasia (56% 4-year survival):

- Nephrectomy with lymph node sampling, abdominal irradiation, and 24 weeks of chemotherapy with vincristine, doxorubicin, etoposide, cyclophosphamide, and mesna.[2]

Inoperable tumors

Treatment options

Patients who have tumors with caval extension above the hepatic veins or that are so massive that their surgeons consider the risk of initial surgical removal too great should be biopsied and treated with preoperative chemotherapy.[3] If surgery is performed in a patient with caval or atrial extension, care should be taken to ensure that appropriate resources are available for pediatric cardiopulmonary bypass.[4,5] On the National Wilms' Tumor Study-5 (NWTS-5), these patients are treated after biopsy by initial chemotherapy with vincristine and dactinomycin with or without doxorubicin. If no reduction in tumor size has occurred after using 3 drugs, then radiation therapy should be used.[6] Surgery is performed as soon as sufficient tumor shrinkage has occurred, generally within 6 weeks of diagnosis. Patients are subsequently treated as for stage III tumors, which includes postoperative radiation therapy. Because of the 5% to 10% error rate in preoperative diagnosis of renal masses after radiographic assessment, confirmation of the diagnosis by biopsy (which may be performed percutaneously) should be obtained prior to chemotherapy.[3]

Stage IV Wilms' Tumor

Favorable histology tumors (80.6% 2-year relapse-free survival [RFS]; 89.5% 2-year survival):[1]

- Nephrectomy with lymph node sampling, abdominal irradiation according to local stage of renal tumor, bilateral pulmonary irradiation for patients with chest x-ray evidence of pulmonary metastases, and 24 weeks of chemotherapy with vincristine, doxorubicin, and pulse-intensive dactinomycin.

Focal anaplasia

- Nephrectomy with lymph node sampling, abdominal irradiation according to local stage of renal tumor, bilateral pulmonary irradiation for patients with chest x-ray evidence of pulmonary metastases, and 24 weeks of chemotherapy with vincristine, doxorubicin, and pulse-intensive dactinomycin.

Diffuse anaplasia (17% 4-year survival)

- Nephrectomy with lymph node sampling, abdominal irradiation, whole-lung irradiation for patients with chest x-ray evidence of pulmonary metastases, and 24 weeks of chemotherapy with vincristine, doxorubicin, etoposide, cyclophosphamide, and mesna.[2]

Stage V Wilms' Tumor

Previously, the surgical approach to bilateral Wilms' tumor was nephrectomy of the side with the larger lesion. However, initial unilateral nephrectomy may predispose patients with bilateral disease to late renal failure.[1] Furthermore, studies demonstrate no difference in survival for children who undergo initial bilateral biopsy followed by chemotherapy and then surgical resection compared with patients who have initial resection followed by chemotherapy. Surgical strategy therefore attempts to preserve renal mass to minimize the risk of late renal failure. Surgical stage should be assigned to both kidneys. Thus, bilateral biopsies and lymph node sampling should be performed. Following 6 weeks of chemotherapy the patient is reassessed. If serial imaging studies show no further reduction in tumor, a second-look surgical procedure should be performed (partial nephrectomy or wedge excision) if negative margins can be obtained, otherwise another biopsy to confirm viable tumor.[2] Chemotherapy and/or radiation therapy following the second-look operation is dependent on the response to initial therapy, with more aggressive therapy required for patients with inadequate response to initial therapy observed at the second procedure.[2-7]

Approximately 10% of patients with bilateral tumors have unfavorable (anaplastic) histology and may benefit from more aggressive chemotherapy and radiation therapy and an aggressive surgical approach at the second-look operation.[4]

Clear Cell Sarcoma of the Kidney

Clear cell sarcoma of the kidney (stages I-IV 84% 2-year relapse-free survival [RFS]; 100% 2-year survival, but later relapses have been known to occur):[1]

Treatment options under clinical evaluation

- Nephrectomy, abdominal irradiation using 1,080 cGy for all patients, whole-lung irradiation for patients with pulmonary metastasis, and 24 weeks of chemotherapy with vincristine, doxorubicin, etoposide, and cyclophosphamide.[2]

Rhabdoid Tumor of the Kidney

Rhabdoid tumor of the kidney (Stages I-IV 25% 4-year disease-free survival)

- No satisfactory treatment has been developed for these children. National Wilms' Tumor Study-5 (NWTS-5) is studying the use of cyclophosphamide, etoposide, and carboplatin in these patients following nephrectomy. Combinations of etoposide and cisplatin, and etoposide and ifosfamide, have been used.[1,2]

Neuroepithelial Tumor of the Kidney

Optimal treatment has not been established for these tumors. Treatment according to Ewing's/PNET protocols should be considered.[1]

Recurrent Wilms' Tumor and Other Childhood Kidney Tumors

The prognosis and selection of further treatment for patients with recurrent Wilms' tumor depend on many factors, including the site of recurrence, tumor histology, length of initial remission, and initial chemotherapy regimen (2 versus 3 drugs).[1,2]

Patients with anaplastic/unfavorable histology tumors, tumor recurrence in the abdomen after treatment with radiation therapy, recurrence within 6 months of nephrectomy, or recurrence after initial 3-drug therapy, have a poor prognosis.[2] The 2-year survival rate for children after local recurrence is 43%,[3] but the prognosis appears to have improved in the last several years.[2] The combination of ifosfamide, etoposide and carboplatin has demonstrated activity in this group of patients, but significant hematologic toxic effects have been observed.[4,5] While high-dose chemotherapy followed by autologous bone marrow has been utilized in the

past [6,7], a recent POG/CCG intergroup study used a salvage induction regimen of cyclophosphamide and etoposide (CE) alternating with carboplatin and etoposide (PE) followed by delayed surgery. Disease-free patients were assigned to maintenance chemotherapy with 5 cycles of alternating CE and PE, and the remainder of patients to ablative therapy and autologous marrow transplant. All patients received local radiation therapy. The 3-year survival was 52% for all eligible patients, while the 3-year survival was 64% and 42% for the chemotherapy consolidation and autologous marrow transplant subgroups, respectively.[8] Patients in whom such salvage attempts fail should be offered treatment on available phase I or phase II studies.

Patients with recurrent clear cell sarcoma of the kidney are eligible for treatment on the National Wilms' Tumor Study Group protocol for relapsed Wilms' tumor (NWTS 5/R). Patients with recurrent rhabdoid tumor of the kidney and neuroepithelial tumor of the kidney should be considered for treatment on available phase I and phase II clinical trials.

CHAPTER VII. HEAD AND NECK LESIONS

CERVICAL LYMPHADENOPATHY:

An enlarged lymph node is the most common neck mass in children. Most are anterior to the sternocleidomastoid muscle. Infection is the usual cause of enlargement; viral etiology, that may persist for months. Acute suppurative submandibular adenitis occurs in early childhood (6 mo – 3 yrs), is preceded by pharyngitis or URI, the child develops erythema, swelling and cellulitis, and management is antibiotics and drainage. Chronic adenitis: persistent node (>3 wk, tonsillar), solitary, non-tender, mobile and soft. Generally no tx if < 1 cm, for nodes above 2 cm sizes with rapid growth, clustered, hard or matted do biopsy. Other causes are: (1) Mycobacterial adenitis-atypical (MAIS complex), swollen, non-tender, non-inflamed, positive skin test, excision is curative, chemo tx is of no value. (2) Cat-Scratch adenitis-caused by Bartonella henselae, transmitted by kittens, positive complement fixation test, minimally tender, fluctuant regional nodes, spontaneous resolution. (3) Hodgkin's disease mostly teenager and young adults, continuing growth, non-tender node, associated to weight loss, biopsy is diagnostic. **If a patient is going to the OR for a node biopsy a preoperative chest x-ray must be obtained to rule out potentially life threatening mediastinal disease.**

CONGENITAL TORTICOLLIS:

Congenital muscular torticollis is a disorder characterized by shortening of the cervical muscles, most commonly the sternocleidomastoid (SCM) muscle, and tilting of the head to the opposite side. This is the result of endomysial fibrosis of the SCM muscle. There is a relationship between birth position and the side affected by the contracture. Congenital torticollis is associated with: plagiocephaly (a craniofacial deformity), fascial asymmetry (hemihypoplasia), scoliosis and atrophy of the ipsilateral trapezius muscle if not corrected. Torticollis can develop at any age, although is more common during the first six months of life. The SCM muscle can be a fibrous mass, or a palpable tumor 1-3 cm in diameter within the substance of the muscle may be identified by two to three weeks of age. Management is conservative in most cases using early physiotherapy exercises a mean duration of three months to achieve full passive neck range of motion. The severity of restriction of motion is the strongest predictor of treatment duration. Those children with failed medical therapy or the development of facial hemihypoplasia should undergo surgical transection of the SCM muscle. Consider cervical subluxation or brain stem tumor in older children with the acute onset of torticollis.

THYROGLOSSAL DUCT CYSTS:

Thyroglossal duct cyst (TDC) is the most common congenital anterior midline neck mass usually (2/3 of cases) presenting before the second decade of life. Symptoms appear at an average age of four with the sudden appearance of a midline cystic mass at the level of the angle of the neck, moving with tongue protrusion and swallowing. Males are more commonly affected than females. TDC is an embryologic anomaly arising from epithelial remnant left after descent of the developing thyroid from the foramen cecum. The lining is cuboidal, columnar or pseudostratified epithelium. TDC is associated with discomfort, infection and a slight probability of malignancy. A legally protective requirement is to document that the mass is not ectopic thyroid gland by performing a thyroid scan. Diagnosis is physical. Sonograms will show a cyst between 0.4 and 4 cm in diameter, with variable sonographic appearance and no correlation with pathological findings of infection or inflammation. Once infected, surgical excision is more difficult and recurrence will increase. Management is Sistrunk's operation: Excision of cyst with resection of duct along with the central portion of hyoid bone (a minimum of 10-15 mm of hyoid bone should be removed) and some muscle surrounding the proximal ductules (the length of single duct above the hyoid bone spreads into many ductuli as it approaches the foramen cecum). Extensive dissection can cause pharyngodynia. The greatest opportunity for cure is surgery at initial non-inflamed presentation. Inadequate excision is a risk factor for further recurrence.

BRANCHIAL CLEFT SINUSES:

Branchial cleft sinuses originate from the 1st, 2nd and 3rd branchial apparatus during embryogenesis of the head and neck. Anomalies of the 2nd branchial cleft are by far the most commonly found. They can be a cyst, a sinus tract or fistula. Fistulae (or sinus tract if they end blindly) display themselves as a small cutaneous opening along the anterior lower third border of the sternocleidomastoid muscle, communicates proximally with the tonsillar fossae, and can drain saliva or a mucoid secretion. Management consists of excision since inefficient drainage may lead to infection. Dissection along the tract (up to the tonsillar fossa) can be safely and easily accomplished after probing the tract with a small guide wire or probe in place. This will prevent injury to nerves and vessels and accomplish a smaller scar. Occasionally a second stepladder incision in the neck will be required. 1st branchial cleft sinuses are uncommon, located at the angle of the mandible, and communicating with the external auditory canal. They have a close association with the facial nerve. 3rd branchial cleft sinuses are very rare, run into the piriform sinus and may be a cause of acute thyroiditis or recurrent neck infections.

LYMPHATIC MALFORMATIONS:

Lymphatic malformations is an uncommon congenital lesion of the lymphatic system appearing as a multilocular fluid filled cavity most commonly in the back/neck region, occasionally associated with extensive involvement of airway or vital structures. The etiology is intrauterine failure of lymphatics to communicate with the venous system. Prenatal diagnosis can be made during the first trimester of pregnancy as a huge neck tumor. Differential diagnosis includes teratomas, encephalocele, hemangiomas, etc. There is a strong correlation between prenatal dx and Turner's syndrome (> 50%), structural defects (Noonan's syndrome) and chromosome anomalies (13, 18, 21). Early diagnosis (< 30 wk gestation) is commonly associated with non-immune hydrops and dismal outcome (fetal death). Spontaneous regression is unlikely but can explain the webbed neck of Turner's and Noonan's children. Prenatal dx should be followed by cytogenetic analysis: chorionic villous sampling, amniocentesis, or nuchal fluid cell obtained from the CH itself to determine fetal karyotype and provide counseling of pregnancy. Late diagnosis (>30 wks) should be delivered in tertiary center prepared to deal with dystocia and postnatal dyspnea of newborn. The airway should be secured before cord clamping in huge lesions. Intracystic injection of OK432 (lyophilized product of *Streptococcus pyogenes*) caused cystic (hygromas) lymphangiomas to become inflamed and led to subsequent cure of the lesion without side effects.

CHAPTER IX: ECMO (extracorporeal membrane oxygenation)

1. General:

Any patient failing CMV (conventional mechanical ventilation). This is defined as PaCO₂ > 60, or postductal PaO₂ < 60 torr for 4 hours documented on three blood gases with FI_{O2} = 1.0 and PIP < 35 cm water (to minimize barotrauma). Any patient placed on ECMO prior to repair of CDH may be maintained on the ECMO circuit for as long as several weeks. During this time if the infant meets criteria for removal from ECMO, the patient may be taken off the circuit prior to or immediately after repair of CDH. Patients may be maintained on the ECMO circuit for additional days. Repair of CDH is not urgent but should be done once the pulmonary status is stabilized and the child is off or ready to come off ECMO.
2. Indications:
 - CDH babies as part of CDH protocol.
 - Meconium aspiration and persistent fetal circulation (PFC) unresponsive to medical management.
 - Pediatric patients with pulmonary failure.
 - Postoperative cardiac surgery patients with transient cardiopulmonary failure.
 - Myocarditis
 - Sepsis
 - Generally any condition that has a reversible cause or operative cure that requires temporary cardio-pulmonary support
3. Contraindications:
 - Pre-existing significant intracranial hemorrhage.
 - Weight < 2 kg (relative).
 - Congenital and/or neurologic abnormalities incompatible with good outcome.
4. Venoarterial ECMO

Involves cannulation of the right atrium via the right internal jugular vein and cannulation of the aortic arch via the right common carotid artery (which is usually ligated). If possible, the artery is reconstructed at the conclusion of the ECMO.
5. Venovenous ECMO:

Involves cannulation of the right internal vein with a 14 French double lumen cannula. This is the preferred method of cannulation for most infants with adequate cardiac stability. Since venovenous ECMO offers no cardiac support, post-cardiac surgery patients and those patients requiring epinephrine support will receive venoarterial ECMO.

6. ECMO Circuit: The ECMO circuit is simple and consists of the following.
 - Polyvinylchloride tubing.
 - Roller pump with a servo-regulated 10 cc venous reservoir bladder.
 - Membrane oxygenator.
 - Heat exchanger that works via countercurrent principle.
 - The priming volume of the circuit is 450 cc. The circuit is primed with a mixture of packed rbc, fresh frozen plasma, calcium, and buffer solutions.
7. Patient Management:
 - Ventilator settings depend on whether the patient is on VA or VV ECMO. With VA ECMO, lung rest is achieved by placing FIO₂ at 21% and pressures and rate low. Paralysis is reversed and vasoactive drugs are weaned if possible. On VV ECMO, additional pulmonary support is required and higher ventilator settings are used.
 - Drugs: Antibiotics. Hydralazine is given for hypertension, unresponsive to volume unloading, at 0.1-0.4 mg/kg q4h. Phenobarbital is given for perinatal hypoxia or overt seizure activity at 20 mg/kg to load and 2.5 mg/kg q12h.
 - TPN: Can be given via system beginning on day #2 at 4-5 cc/kg/hr.
 - Routine U/S to rule out head bleed.
 - Hypertension: This is the leading cause of morbidity in ECMO patients. Any sustained mean arterial pressure over 70 cannot be tolerated, and must be treated emergently with Hydralazine or Nitroprusside.
 - Coagulopathy: Coagulopathic patients receive aggressive exchange transfusions until coagulopathy is normalized. Usually these patients are suffering from sepsis.
 - AMICAR: Patients considered at high risk for bleeding (premature, anticipated surgical procedure such as CDH repair) may receive aminocaproic acid prophylactically. The dose is 100 mg/kg administered as an IV bolus and then 30 mg/kg/hr as a continuous drip thereafter.
8. ECMO/Management:
 - Blood Flow: To regulate pO₂ (60 mmHg). Usual flow is 100-120 cc/kg/min. ECMO flow is usually limited by venous return.
 - Gas Flow: To regulate pCO₂ (35-45 mg Hg). The sweep gas flow ventilating the oxygenator is usually 100% O₂ at 1-2 liters/min. May need to blend in carbogen.

- Blood Volume: To regulate perfusion. Maintain Hct > 45. Use FFP liberally. Ultrafiltration using an Amicon Minifilter is occasionally needed if the patient fails to diurese.
 - Anticoagulation: Continuous heparin I.V. to maintain ACT at 180-220 sec (normal 90-150 sec). All irrigation solutions should be changed to nonheparinized solutions. No IM meds, venipuncture, or heelsticks.
 - Platelets: Maintain platelets > 50,000, higher if bleeding complications are a problem. This usually requires 1-2 units of platelets per day.
9. Test Period off ECMO:
- FIO₂ = 1.0, pressure 24/2, rate = 30.
 - D/C gas flow into oxygenator.
 - Decrease water bath temperature.
 - Stop all infusions into ECMO circuit including heparin, and come off bypass by clamping arterial and venous catheters and unclamping A-V bridge. Monitor infant and circuit ACT and administer heparin as necessary to circuit to keep ACT 180-220.
 - Check ABG's at 5, 10, and 15 minutes.
 - Return to bypass if deterioration occurs.
 - Indications for Discontinuation of ECMO Support
 - Adequate oxygenation during trial off on reasonable FIO₂ (<50%)
 - Ventilator wean to low settings (MAP < 10).
 - Failure to improve after 10-14 day course or uncontrolled bleeding may necessitate discontinuation of ECMO on high ventilator settings.
 - Neurologic (e.g. ICH) or other oxygen system failure incompatible with meaningful life is an occasional, unfortunate indication.

CHAPTER X: TRAUMA

OHSU Trauma Team:

The pediatric trauma pager must be carried by the entire team at all times. The Junior resident on call should attend all trauma situations in the ED (all trauma). The Chief resident should attend all Level I traumas. The Trauma Service handles all patients age 15 and above. Pediatric Surgery service admits all patients 14 years and below. On weekday evenings, the Trauma Service will initially evaluate all pediatric traumas, regardless of age. The Pediatric Surgery resident will then be called to assist only if necessary. The Trauma Team/Chief Resident will be responsible for communication with the Pediatric Surgery attending as needed. The next morning, the Trauma Service will sign-out these patients to the Pediatric Surgery service. The purpose of these changes is to take some of the load off the Pediatric Surgery residents so that your time on the service is more educational. At all times, flexibility, cooperation and good communication will be expected.

Transfer of Trauma Patients from PICU to Floor:

In general, isolated injuries involving another single subspecialty should be transferred to the appropriate service upon discharge from the PICU. Multiple trauma stays on pediatric surgical service.

Currently, isolated head or spinal injury operated on by peds neurosurgery goes from the PICU/peds surgery service to peds neurosurgery floor service. All other neurotrauma patients will stay on the peds. surgery service.

Special Cases:

- 1) Severe non-operative head injury (bolt, ventriculotomy) stay in PICU until they are awake or they receive trachs/Gtubes (i.e. no longer acute). These patients would then transfer to peds surgery floor service to await rehab. Peds neurosurgery will sign off while in PICU or on the floor based on attending opinion that any acute head injury issues are over. (Peds neurosurgery is always available to re-consult as needed).
- 2) Children with neurosurgical operation plus significant other system injuries and /or operations will continue to be followed closely on the floor as consultants.

I. INTRODUCTION

A. MAGNITUDE OF THE PROBLEM

Injuries are the most common cause of death and disability in the pediatric age group. The magnitude of this problem makes its treatment and prevention exceedingly important. More than 22,000 children in the United States died of accidental injury in 1986. Over half a million children are hospitalized for injuries each year and an additional 16 million children are estimated to be seen in emergency rooms for treatment of accidental injuries. i The problem can be emphasized in two ways: by analyzing historical trends in the United States and by comparing U.S. statistics to international standards. In the past sixty years, childhood deaths in the United States as a result of infection have decreased 90%, while those due to injury have only been reduced by 40%. 1 During the period from 1968 to 1986, death rates in the U.S. from injury were reduced by 25%, while death rates as a result of other diseases fell 56%. ii Williams and Kotch compared World Health Organization data for childhood death rates in the United States to those of Canada, England, Wales, France, Norway, and the Netherlands and found the U.S. to have the highest overall injury mortality rates. Moreover, while injury mortality rates in other countries were decreasing, those in the U.S. were rising. These differences were thought to result from increasing death rates due to homicide and motor vehicle accidents. iii Pediatric injury and death has a significant economic impact on our society. This can be estimated by analyzing both future-lost productivity as well as direct and indirect costs. An estimate of the cost of lost productivity due to 22,000 childhood injury deaths in 1986 was \$ 8.3 billion dollars. 1 Approximately 600,000 children are hospitalized per year in the United States for injuries. This is the second most common reason for pediatric admission and results in the most hospital days per patient. Direct and indirect costs of injury to children 0-19 years of age have been estimated at 7.5 billion dollars per year.

B. UNIQUE ASPECTS OF CHILDHOOD INJURY

Childhood injuries differ from their adult counterparts in many respects. First, there exist within the pediatric age group heterogeneity in type of injury as function of age. This results from both susceptibility to injury on an anatomic and behavioral basis and from age-related exposure to different mechanisms of injury. Second, children are increasingly being victims of violent behavior from their caretakers and by being accidental victims of gunfire. Childhood injuries are also unique because of the extraordinary number of injuries that are potentially preventable through public education and legislation. The types of accidental injuries children sustain reflect a combination of the child's cognitive and physical capabilities, their environmental exposures and preventative measures applied by caretakers and society at large.

Anatomic and behavioral basis for childhood injury

Unlike the adult who has already had complete physical and cognitive development, a child at any age represents a dynamic process of physical, intellectual and behavioral development. Anatomic differences between adults and children result in different patterns of injury despite similar mechanisms of injury. Behavioral and intellectual development of the child and adolescent make them susceptible to many dangerous situations that adults are not. The size of the child and age related body proportions has direct impact on the types of injuries children sustain. The infant has a proportionately larger head than older children and adults. This raises their center of gravity and makes the unrestrained infant more susceptible to ejection during a motor vehicle accident. Their relatively large head and weak neck and shoulder muscles also make the infant more susceptible to cervical spine injury during rapid deceleration. At all times in childhood the center of gravity is higher than in the adult. This means that the young child being restrained by only a lap belt may "jackknife" over the seat belt during rapid deceleration. Hollow visceral injuries and thoracic and lumbar spine injuries are known to occur after this type of "seat belt injury". Children also display significant differences in muscle mass and bone density. The decreased muscle mass of children and the relatively large size of solid viscera are thought to make these organs more vulnerable to blunt trauma. Children's bones are less ossified than adults. As a result, significant trauma may not result in bony fracture.

In the child with chest trauma, the absence of injury to the chest wall may result in an underestimation of associated intrathoracic injuries. The diminutive size of some bony structures such as the pelvis results in minimal protection for intrapelvic structures such as the urinary bladder. The most obvious difference between children and adults is that they are smaller. While this is readily apparent, it does have important implications for the type of injuries children sustain and their subsequent treatment. Because children are lighter and smaller they are more susceptible to ejection from moving vehicles. An adult pedestrian struck by a car will be initially hit in the lower extremities and then thrown onto the hood or road. The young child will initially be struck in the upper torso or head, often resulting in a fatal outcome. Anatomic differences between adults and children result in important differences in treatment after injury. The pediatric airway is uniquely different and inability to access the airway in the severely injured child is often the major impediment to successful resuscitation. Size limitations often constrain the ability of health care workers to obtain vascular access and special techniques must be learned to adapt to small children.

The behavioral and cognitive development of the child results in different injury patterns in different age groups. The innate curiosity of the infant and

toddler and their relative absence of fear result in injuries such as caustic ingestions and electrical injuries. The preschool child has similar limitations on his or her ability to perceive danger. This limitation, combined with well developed motor skills, exposes them to the hazards of drowning and pedestrian accidents. The adolescent's sense of invulnerability and need for experimentation can lead to drinking and driving and other risky behavior.

Injury patterns due to mechanism of injury

A child or adolescent's physical, cognitive and emotional state interact with their environmental exposures to place them at risk for certain types of injury. This risk can be modified by existing laws and parental influence. A child's environment increases the risk of certain types of injury. The increasing use of the automobile in our society has resulted in significant increases in motor vehicle related fatalities and injuries. Children have not been spared these injuries. Pediatric injuries sustained in rural areas differ from those in urban areas. Children in coastal states such as California and Florida have higher rates of drowning and near drowning episodes.

The important role the automobile plays in our society is reflected in its role in injuries to children. Deaths due to motor vehicle accidents are the most common cause of traumatic death in children. Predictably, young men in the age group 15-19 years comprise most of these fatalities. Motor vehicle fatalities in adolescents involve alcohol in close to 50% of accidents. Deaths as a result of motor vehicle accidents in children results in the greatest number of years of life lost of any childhood or adult illness. Injuries to pedestrians are another important type of motor vehicle related accident. Most injuries and fatalities occur in children age 4 to 9 years old. It is the most common cause of injury related death in this age group. Pedestrian fatalities accounted for 1,787 deaths in 1986. Most pedestrian accidents in children occur during daylight hours and in urban areas. Non-white children have a 1.5 increased fatality rate from pedestrian accidents. The majority of pedestrian accidents result from children running into traffic and a minority from children being backed over.

Homicide is the second leading cause of accidental death to children. In 1986, 2877 pediatric deaths were attributed to homicide. Homicide occurs predominantly in two pediatric age groups. Intentional violence resulting in death in the 0 to 3 year age group is usually related to child abuse. The other peak rate occurs in the 14 and over age group. Demographic data indicates that most deaths occur in males and close to half in blacks. Suicide is the third leading cause of death among persons less than 19 years of age. In 1986, 2151 deaths were attributed to suicide. While suicide rates in adults have decreased, pediatric suicide rates have continued to increase.¹

Adolescent males are more likely to attempt and be successful at taking their own lives. This is likely due to the use of firearms by males as the method of choice of committing suicide.

No discussion of homicide and suicide in children would be complete without discussing the implications of firearms on the children of our society. Approximately 30,000 people die each year from firearm injuries in the United States. Ten to 12% of these fatalities are in children. Firearm related fatalities occur 100 to 1000 times more frequently in U.S. children than in children of other developed countries.³ In young black males, death from gunfire is the most common cause of mortality. Firearm mortality in children has risen and will continue to rise because of the absolute amount of firearms in our society and because of their availability to children. There are reported to be more than 200 million firearms currently in the United States. Approximately one in four households in the United States have a handgun. Availability of guns has had direct effects on pediatric homicide, suicide and unintentional shootings. Domestic homicide is three times as likely when there is a gun in the home. Adolescent suicide rates have doubled in the last decade. Availability of a gun in the home has been shown to increase the rate of successful suicide in the adolescent. Equally tragic is the fact that the majority of unintentional firearm fatalities in children result from access to a loaded gun in the home. Half of the fatalities occur in the child's own home. Fatalities due to firearms do not comprise the entire picture. It is estimated that for every fatality, 4 to 7 children are wounded by firearms. This results in an enormous cost to our society and an immeasurable personal cost to the individuals effected by this violence.

Child abuse

The unique aspects of child abuse warrant it's separate treatment. Approximately 2-3000 children a year are fatally injured as a result of child abuse. Modern awareness of the problem of child abuse was established in 1962 by Kempe with the publication of "The battered child syndrome".^{iv} Approximately three million reports of child abuse were filed with childhood protective agencies in the United States in 1993. One third of these reports were confirmed.^v The majority of cases of child abuse result from someone the child knows. Factors that increase the risk of abuse include substance abuse in the family, lower socioeconomic status, single parenthood, and abuse of the parent. While those living at or below the poverty level have a higher risk of abuse, child abuse is not restricted to any socioeconomic class. The most common etiology of fatal child abuse is head injury from striking or throwing a child. Child abuse may present as burns, abdominal injuries, unusual or severe bruising, long bone fractures, vaginal lacerations and anal tears. The "shaken baby" syndrome is a unique form of child abuse. This form of abuse, usually occurring in infants, results in inflicted craniocerebral trauma. Clinical findings include subdural or

subarachnoid hemorrhages and retinal hemorrhages with minimal external findings. The mechanism of the injury is thought to be the tearing of bridging veins from the cortex during vigorous shaking. There is evidence that violent contact of the skull with a hard surface during deceleration is necessary for these injuries. Affected infants present with vomiting, lethargy, somnolence or coma.

The ability to diagnose child abuse results from knowledge of the manifestations of child abuse and an understanding of parental dynamics, which put the child at risk. Children with injuries suspected to be due to child abuse should be admitted to the hospital and Childhood Protective Services should be contacted. Injuries should be carefully documented and external injuries should be photographed. Occult injuries should be sought by performance of skeletal survey radiographs and by ophthalmologic exam. The physicians's role is to document and treat injuries and not to judge the behavior of the caretakers.

Prevention

The term "accidental" injury implies a random event in which victims are unable to avoid injury as a result of chance or bad luck. Unfortunately many "accidental" injuries in children are either avoidable or can be minimized by the appropriate use of preventative measures. Optimal strategies to prevent childhood injury involve education, behavior modification and legislation. Several groups in the United States have been active in promoting injury prevention in children. Among them has been the American Academy of Pediatrics. They have been integral in the passage of legislation such as the Flammable Fabrics Act, the Refrigerator Safety Act, and the Federal Hazardous Substance Act. Preventative strategies will be discussed in relation to mechanisms of injury.

Occupant and pedestrian injuries from motor vehicles are the greatest source of pediatric morbidity and mortality. Strategies to reduce injuries include promoting uniformity and enforcement of seat belt laws, educating parents about seat belt utilization, and improved vehicle and road design. As was previously noted, close to half of all motor vehicle fatalities involving adolescents involve alcohol. Attempts to influence attitudes of adolescents toward drinking and driving must be included in any approach to reduce motor vehicle fatalities. Motor vehicle-pedestrian injuries usually occur in urban areas during daylight hours. In order to prevent injuries in heavily trafficked areas, safe playgrounds must be available to young children. In addition, children must be taught traffic safety skills early in elementary school.

The United States leads all industrialized nations in pediatric homicide and suicide rates. The reasons for the increase in interpersonal violence among children are complex. Certainly poverty, urban decay, drug abuse and gang behavior are important reasons. Accessibility to hand guns has a central role

in the escalation of personal violence. A survey of students in middle school and high school conducted by the Harvard School of Public Health revealed that 59% of the students knew where they could obtain a gun if they needed one. vi Strategies to reduce violent deaths include teaching young people conflict resolution techniques, reducing gun availability through gun restriction legislation, and educating gun owners about gun safety.

A number of effective strategies exist for the prevention of burn injury in children. Mortality can best be avoided by the appropriate installation and maintenance of smoke detectors and by fire safety education. Morbidity from scald burns can be avoided by reduction of water temperature in hot water heaters and by parental education.

Many other specific recommendations can be made to improve childhood safety. Areas that have implemented bicycle helmet laws and education have seen reductions in head injury. Urban areas that have installed window grates in high rise apartment buildings have seen mortality and morbidity from falls reduced. It is clear that the magnitude of problem of accidental injury in children is enormous. Preventative strategies to avoid injury and death have been shown to be effective when applied. The success of injury prevention is dependent on public recognition of the problem, parental education, and appropriate funding of agencies and organizations responsible for these programs.

II. ORGANIZATIONAL ASPECTS OF PEDIATRIC TRAUMA CARE

A systematic approach to the management of injuries to children reduces morbidity and mortality. This approach integrates activation of Emergency Medical Services (EMS), immediate recognition of injuries, resuscitation and transport to the appropriate trauma facility. Organized approaches to care of the injured adult and child are relatively recent events. Analysis of battlefield casualties in Korea and Vietnam demonstrated the survival advantage of immediate stabilization, rapid transport and early definitive care of the severely injured. National attention of the need for an organized approach to civilian injuries began with the publication of "Accidental Death and Disability: The Neglected Disease of Modern Society" by the National Science Foundation in 1966. vii The American College of Surgeons Committee on Trauma documented the necessary components of a trauma center in 1983 with the revised "Hospital and Pre Hospital Resources for Optimal Care of the Injured Patient." viii The specific characteristics of a pediatric trauma center are defined in an Amendment to this document.

Impressive gains have been made in the systematic care of adults. Emergency medical systems for children have not realized the same success. This is due to a number of factors, including delayed recognition of the problem of pediatric trauma, inadequate funding of trauma systems,

lack of definitive research on pediatric trauma, and regional issues surrounding the optimum location for the definitive care of children.

A. PRE-HOSPITAL CARE

The child enters the Emergency Medical Service for Children (EMS-C) soon after sustaining an injury. Rapid entry into the system is facilitated by use of the 911 calling system. The effectiveness of the 911 system is enhanced by education of primary care providers, parents, babysitters, teachers and day-care workers as to its appropriate use. The first responder to the injured child varies among communities and EMS systems. This may range from a highly trained Paramedic in urban areas to a volunteer fireman with little pediatric exposure in rural areas. It is very difficult for field personnel to establish and maintain skills for the initial response to the pediatric patient. Skill maintenance is difficult because pediatric emergency calls comprise only approximately 10 % of all calls to an EMS system.^{ix} Of these pediatric calls, most are for medical emergencies.

The particular skills most difficult for the EMS provider to maintain are assessment of cardiorespiratory stability and airway and intravenous access. A lack of consistent

exposure to children makes recognition of normal and abnormal responses to injury difficult. EMS providers should have ready access to tables that describe normal values for pediatric respiratory rates, blood pressure and heart rate. Airway access is difficult in the small child due to diminished size and alterations of glottic anatomy. These factors, combined with the adverse conditions the EMS provider must contend with, result in low success rates for pediatric intubation. Prolonged attempts at intubation should not be attempted if bag-valve-mask ventilation is successful. Intravenous access can be very difficult in the infant and child. Transportation of a child to a trauma center should not be delayed while multiple attempts at intravenous access are made. These should be done en route and by an intraosseous technique if rapid intravenous access is not established. In order to provide optimal care of the injured infant and child, equipment modifications to ambulances must be made. Supplies should include appropriate size endotracheal tubes, laryngoscopes, medication vials and ventilation masks. The ambulance should have the capacity to be rapidly warmed to compensate for the inevitable hypothermia that the child sustains at the scene of the accident.

B. PEDIATRIC TRAUMA SYSTEMS

The optimal care of injured children requires integration of the pre-hospital phase of treatment into a pediatric trauma system. Significant reduction in injury-related mortality has resulted from this approach in adults. The central component of a regional pediatric trauma system is the trauma center. The American College of Surgeons have described the necessary

components of a Level I pediatric trauma center. To summarize, it must be either a freestanding children's hospital or have a pediatric center within an general hospital. It must have 24-hour coverage by appropriate personnel including pediatric surgeons, orthopedists, emergency room physicians, neurosurgeons, anesthesiologists and radiologists. There must also be an ongoing quality improvement process and a trauma nurse coordinator must oversee the education programs of the trauma center. 8 Unlike adult hospitals, which have a three tier trauma designation, children's programs are either designated as a Level I or II.

Assessment of patient care outcomes and triage criteria are dependent upon methods of quantifying or stratifying injuries in children. The most widely applied system for triage scoring in children is the Pediatric Trauma Score (PTS). x This scoring system was developed in 1984 and incorporates those assessments which take into consideration the unique spectrum of childhood injuries. Six "areas" are graded according the size of the child, ability to maintain an airway, blood pressure, level of consciousness, presence of a cutaneous wound and presence of a skeletal wound are graded from +2 to -1. A score of 9 or greater implies close to 100% survival while a lower score is associated with a linear reduction in survival. Adult scoring systems such as the Injury Severity Score (ISS) underestimate the influence of head injuries on survival in pediatric populations. Most other systems including the Modified Injury Severity Score and Anatomic Index of Injury Severity and Trauma Score have little application to pediatric triage.

III. RESUSCITATION

Resuscitation of the injured child begins at the site of injury and occurs continuously during transportation to the hospital environment. The importance of early and effective resuscitation cannot be overestimated. The completeness of the initial assessment at the scene of the accident is dependent upon the training of the first responders, severity of the injury, and proximity to the pediatric trauma center. The focus at the scene of the accident should be to perform immediate stabilization and transport. Reasonable activities at the scene of the accident include obtaining vital signs, cervical and lumbar spine immobilization, obtaining an airway, establishing intravenous access, preventing hypothermia, stopping external sources of hemorrhage and immobilizing fractures. First responders with minimal pediatric training should not spend extra time attempting intravenous access in small children and infants. However, if experience permits, intravenous access should be obtained before hypothermia and/or blood loss prevent easy access. The optimal approach to the severely injured child who is close to a pediatric trauma center may be to "scoop and run." The initial assessment in the hospital environment continues the resuscitation begun in the pre-hospital phase. The initial assessment includes repeating the primary survey, continuing the ongoing resuscitation, performing a secondary survey and making plans for definitive care. The

unique aspects of pediatric airway management, fluid and electrolyte management and response to shock and injury will be discussed as they pertain to the initial assessment of the seriously injured child.

The primary survey serves to simultaneously identify and correct life-threatening injuries, establish the means to resuscitate the child, and define treatment priorities. This sequence is outlined by the following initials: A-B-C-D-E. Where A is airway control, B is breathing, C is circulation and control of hemorrhage, D is disability or neurologic assessment, and E is exposure of all potential injuries.

Establishing and maintaining an open airway is the most important initial action of the evaluating trauma team. Airway patency must be continuously assessed as airway compromise may occur at any time. Maintenance of the airway has particular importance for the treatment of the child with a head injury. Maintenance of a patent airway prevents hypoxia, which can be responsible for secondary brain injury. The adverse effect of inappropriate airway management has been demonstrated by Gentleman and Jennett who found a higher mortality rate among those head injured patients who suffered hypoxia. Control of the airway also provides an element of safety during transport to and performance of radiologic studies. Evaluation of airway patency begins with observation of the respiratory efforts of the child. The child with a normal respiratory rate (Table 2), Glasgow coma scale >10 and no stridor can be observed while the remainder of the primary and secondary survey is conducted. An impaired respiratory effort is often due to upper airway obstruction. The large tongue, tonsils and adenoids in the small child put them at increased risk for airway obstruction. Simple maneuvers such as the chin lift or jaw thrust often correct the anatomic obstruction. Apneic children and children requiring immediate operative intervention should be intubated.

Once the need for intubation is established, an intravenous line is placed and preoxygenation is begun with bag-mask ventilation with 100% oxygen. All equipment necessary should be available and in good working order. At a minimum this includes several laryngoscopes, uncuffed and cuffed endotracheal of various sizes, oral and tracheal suction, a saturation monitor and Magill forceps. Paralysis and sedation facilitate the process of intubation. Muscle relaxants such as succinylcholine or vecuronium are given as a slow intravenous push after the patient is adequately preoxygenated. Sedation is concomitantly achieved with either fentanyl and or midazolam. All patients with potential cervical spine instability should have manual in-line cervical stabilization performed during intubation. Prior to intubation an appropriate endotracheal tube is selected. Most infants and children should have an uncuffed endotracheal tube placed. Several rules of thumb exist to aid in selection of the appropriate size endotracheal tube. The size of the child's fifth finger or nares can be used to approximate the endotracheal tube size. The formula: $\text{age}(\text{years}) + 16/4$ can

also be used to estimate tube size. Regardless of the size chosen, endotracheal tubes of one size smaller and one size larger should be immediately available. Most infants and children can be intubated with a Miller type blade. A curved laryngoscope blade (Macintosh) is more effective when intubating larger children and adolescents. After passage of the endotracheal tube, its location is confirmed by auscultation and chest x-ray. When a patient arrives with an endotracheal tube placed in the field, no assumptions should be made that it is in good position. Its position should be confirmed upon arrival before any further assessment steps are taken. Occasionally facial trauma, laryngeal trauma or preexisting anatomic abnormalities prevent placement of an oral-tracheal airway and a surgical airway becomes necessary. A surgical cricothyrotomy is the preferred approach in the child older than 3 to 4 years. In younger children, the cricothyroid membrane is smaller and a formal tracheostomy may be necessary.

After the airway is established ventilation is evaluated (B=breathing). Both the spontaneously breathing child and the intubated child are evaluated for the presence of cyanosis, symmetric chest excursions, equal breath sounds and normal oxygen saturation determined by pulse oximetry. In-line capnography is also useful if available. Intubated children should have an arterial blood gas sent as soon as possible to assess arterial CO₂, O₂ and pH. Unstable patients with decreased ipsilateral breath sounds should have the position of their endotracheal tube immediately checked. If the position of the endotracheal tube is satisfactory, a needle thoracentesis should be performed to rule out a potential tension pneumothorax. Stable patients with decreased ipsilateral breath sounds should have an AP chest x-ray performed. Life-threatening chest injuries that may require urgent relief include a hemothorax, pneumothorax, open chest wound, flail chest, pulmonary contusion, and cardiac tamponade. The trauma patient with a hemothorax or pneumothorax should have a chest tube placed. Most thoracostomy tubes should be placed in the 6th intercostal space in the midaxillary line. Initial management of a pulmonary contusion and cardiac tamponade will be discussed later.

After the airway is secure, ventilation is established and life threatening chest conditions are recognized, the adequacy of the circulation is evaluated. Appropriate evaluation of the pediatric circulation requires a knowledge of age appropriate vital signs. It is helpful to have normal vital signs for the infant, toddler, and adolescent displayed in a prominent position in the trauma room. Assessment of an infant's vital signs is made more difficult by the inherent variability in normal blood pressure values. Moss et al noted a 40-point variability in systolic blood pressure readings in both premature and full-term infants. xii This finding emphasizes the importance of observing trends in vital signs during the ongoing resuscitation of the child. Decisions regarding the need for fluid

resuscitation should be made using both clinical observation and objective data from hemodynamic monitoring. The patient should be disrobed and examined for pallor, capillary refill, strength of distal pulses and external blood loss. Pulse oximetry, automated blood pressure measurement and continuous EKG monitoring should be established as soon as possible. Patients who are hemodynamically unstable or hypoxic should have an arterial line placed. Patient who fulfill these criteria should also have a urinary catheter placed to monitor hourly urinary output. Placement of the urethral catheter should be done only after inspecting the meatus for blood and the pelvis for instability.

Recommendations for resuscitation of the pediatric patient conform to established ATLS guidelines. xiii The patient suspected of having circulatory compromise is given an intravenous bolus of 20 ml/kg of lactated Ringer's solution. The patient is observed for a response to this intervention. If no clinical improvement occurs, a second bolus of 20 ml/kg is given. Inadequate response to these interventions suggests major blood loss and 10 ml/kg of type specific blood is transfused. If type specific blood is not available then low titer O negative packed red blood cells are administered. Cardiac tamponade should be considered in the patient with poor response to volume resuscitation and no obvious source of hemorrhage. If time permits, echocardiography is a useful diagnostic and therapeutic adjunct. If the patient is hemodynamically unstable, a subxiphoid pericardiocentesis is performed.

Adequate treatment of hemorrhagic shock is dependent upon the ability of caretakers to obtain venous access. This is often more difficult in children than in adults. Children's veins are smaller and often are underneath significant subcutaneous fat. Nevertheless, virtually all children can have intravenous access established. Like adults, children should have the largest bore catheters placed in the upper extremities if possible. Locations for intravenous access in the upper extremities include the cephalic vein at the wrist and the cephalic and basilic vein in the antecubital fossa. If these options are not available then a cutdown on the saphenous vein at the ankle should be done. Subclavian vein catheterization is useful when done by experienced personnel. Many authors would suggest the placement of an intraosseous infusion line immediately if peripheral access is not easily obtained. Intraosseous infusion takes advantage of delivery of solutions through the medullary cavity and emissary veins of both long and flat bones. Common locations for placement of these lines in children are the proximal tibia, distal femur, lateral malleolus, and iliac crest. Intraosseous infusion has not historically been thought to be optimal for volume resuscitation due to limitations on flow rates through the medullary cavity. However large and small animal studies using hemorrhagic shock models have demonstrated the efficacy of crystalloid resuscitation using

intraosseous infusion. xiv These findings suggest expanding the application of this technique to include prehospital use.

Once the A-B-Cs of the primary survey are performed attention is directed to estimation of the neurologic status of the patient (D=disability). This is done by evaluating the level of consciousness, looking for focal neurologic deficits, and gauging pupillary responsiveness. Consciousness is immediately assessed by observing whether the patient is alert, responsive to verbal stimulation, responsive to painful stimulation or is unconscious. A more rigorous evaluation can be performed using the Glasgow Coma Scale for Children. The first exam is used as a baseline for comparison. If muscle relaxants are used to assist intubation then a baseline exam must be documented prior to their use.

After a rapid neurologic assessment, the patient is disrobed to facilitate detection of all injuries (E = exposure). Pressure is applied to all sources of hemorrhage. Special consideration should be made for the maintenance of body temperature in the infant and child. Small children have less body fat, a higher surface area, and higher metabolic rate. These factors all potentiate the loss of body heat to the environment. Children that need to be extricated from automobiles are at particular risk for hypothermia due to prolonged exposure. Efforts to prevent hypothermia include the use of warming blankets, heating lamps, warmed iv fluids, and heated trauma rooms.

The secondary survey completes the initial assessment of the patient and begins the transition to definitive care of the identified injuries. A more complete history of the accident and medical history of the patient can often be obtained at this time. During the secondary survey the A-B-Cs should be continuously reassessed due to their dynamic nature. Neurologic injuries may evolve. An airway may be lost due to depressed consciousness. Ventilation may become difficult. Each problem requires a systematic reassessment of the A-B-Cs. The secondary survey is a thorough head-to-toe examination of the patient. The head and neck are examined for palpable skull fractures, scalp lacerations, cervical spine tenderness, CSF otorrhea and rhinorrhea, facial deformities and pupillary response. The chest should be examined for symmetric breath sounds, rib tenderness or deformity and abnormal heart sounds.

The abdomen is then examined for the presence of injury. Most children swallow air or develop gastric distension during bag - valve -mask ventilation. This may lead to the false impression of abdominal tenderness or distension. To prevent this, a nasogastric tube may need to be passed and it's position confirmed. Examination of the abdomen begins with inspection for bruising which may suggest a blow or force to the abdomen. Examples include the bruising due to handlebar injuries or seat belt injuries. The abdomen is then gently palpated for tenderness. The abdominal exam is often useless due to neurologic injury. Patients at increased risk for

abdominal injury should undergo a CT scan of the abdomen and pelvis. A rectal exam is performed to evaluate rectal tone and to identify rectal bleeding. Compression of both iliac wings is performed to identify pelvic instability. The secondary survey is completed with examination of the extremities. Symmetry of the extremities and gross deformities are noted. Distal pulses and capillary refill should be documented.

The secondary survey is completed by performing radiologic studies. A chest x-ray, abdominal x-ray, and cervical spine series should be performed on all seriously injured children. Children who are unconscious or who have an altered mental status should have a CT scan of the head performed. Stable children with abdominal tenderness should undergo a CT scan of the abdomen and pelvis. The resuscitation phase is completed by determining the disposition of the patient and the need for evaluation by consultant.

IV. INITIAL INJURY MANAGEMENT

A. HEAD AND NECK

Cranioerebral injuries:

Cranioerebral injury is the most common cause of death in the pediatric trauma patient. Non-lethal injuries result in serious morbidity and long term functional disability for thousands of children every year. It is estimated that over 100,000 children a year are admitted to the hospital for the evaluation of head trauma. xv Approximately 5% of these admissions are for severe head trauma (GCS < 8). xvi Child abuse is the most common cause of death in children between the ages of six months and one year and head injury is the most common reason for this. The mechanism of head injury relates to the age of the child and the severity of the injury. If all head injuries are analyzed, then falls (35%) are the most common reason for injury, followed by motor vehicle accidents (25%). xvii If only severe head injuries are analyzed, then motor vehicle accidents result in the majority of injuries. The younger the child the greater the chance that the head injury resulted from a fall. Other common causes of head injury include bicycle accidents, sports accidents, and assaults.

Data from the National Pediatric Trauma Registry revealed a 6% mortality rate from head injury. When this data was compared to that of the predominantly adult Major Trauma Outcome Study, children were demonstrated to have significantly better outcome. 17 This relationship was also maintained when increasingly severe injuries were compared by the Abbreviated Injury Scale. This observation was independently confirmed by Luerssen et al during their analysis of over 8,000 patients in three metropolitan hospitals xviii This data suggests that the pediatric patient has a greater ability to withstand neurologic injury. Whether this is due to greater plasticity of the nervous system, greater physiologic reserve or other factors is unknown. Several unique physiologic and anatomic

characteristics of children influence the management of their head injuries. As mentioned previously, the young child has a relatively large head and weak neck muscles, placing them at greater risk of deceleration injuries. Therefore the infant is a greater risk of cerebral injury during forceful shaking. The young child has a more compliant skull due to the thinner bones and nonfused cranial sutures. As a result, force applied to a single area of the skull may result in that area deforming and absorbing all of the force and not producing a coup-contracoup type injury. One also must realize that pediatric brain injury not only disrupts the present state of central nervous system function but also may disrupt the capacity for further development.

The initial management of the child with head injury begins with assessment of the severity of injury. Assessment begins with an evaluation of the level of consciousness. This is best performed by using the Glasgow Coma Scale or the modified Glasgow Coma Scale for infants. Patients are assigned numerical values for their motor response, verbal response, and eye opening. A normal patient would have a score of 15 and a brain dead patient would have a score of 3. Next pupillary and brain stem function are assessed. The pupils are assessed for direct and consensual response to light. Brain stem function is evaluated with either the dolls eye response or by the cold caloric test. Both of these maneuvers evaluate the function of the vestibular pathway, which is carried via the eighth nerve into the vestibular nuclei of the midbrain. Finally, motor function is evaluated by observing the ability of the patient to move their limbs. During the assessment, a history of head trauma and its sequela is elicited. Caretakers and rescue personnel are asked to describe any loss of consciousness, post traumatic seizures, vomiting, or focal motor deficits. The initial evaluation is completed with examination of laboratory studies. Blood alcohol levels and urine toxicology screens should be performed in at risk patients to estimate the effect of these drugs on the neurologic exam.

The severity of the injury determines the need for admission and the need for further diagnostic and therapeutic steps. In the absence of other injuries, a child should be admitted to the hospital if they have a history of head trauma and a seizure, skull fracture, loss of consciousness, altered mental state or significant headache. Children with head trauma suspected to be due to child abuse should be admitted regardless of associated symptoms. All children with severe head injuries (GCS < 8) or focal neurologic deficits should have a CT scan of the head performed. The CT scan is obtained to identify mass lesions, midline shift, contusions, fractures and secondary signs of elevated intracranial pressure (ICP). If operable mass lesions are identified, the patient is immediately transferred to the operating room for craniotomy and decompression. Patients with evidence of increased ICP and nonoperable lesions are treated with hyperventilation, elevation of the head, and diuresis with Mannitol. Patients with a GCS of < 8 should have

ICP monitoring. If possible this is done with a ventriculostomy catheter, which allows both pressure monitoring and removal of cerebral spinal fluid. Patients with severe injuries or a history of post-traumatic seizures are treated with phenytoin. Specific craniocerebral injuries will now be discussed.

Skull fractures: Approximately one fourth of pediatric patients with a history of head trauma will have a skull fracture detected.^{xix}The majority of skull fractures in children are linear fractures. These fractures are significant only in relation to the underlying brain pathology. Therefore, the child with a linear skull fracture and no abnormal neurologic signs can be observed with the anticipation that no brain injury will likely manifest itself later. Other less common skull fractures that children may get include diastatic fractures, basilar fractures, and depressed fractures. Diastatic skull fractures are unique to children as they are a skull fracture through a cranial suture. This most commonly occurs through the lambdoid suture. Diastatic skull fractures can be complicated by leptomeningeal cysts, which are protrusions of the meninges through the fracture site. These may demonstrate progressive growth and require operative closure. The rare occurrence of leptomeningeal cysts justifies follow-up skull films in the patient under three years of age who sustains a skull fracture. Basilar skull fractures occur rarely in children and manifest themselves as they do in adults with CSF rhinorrhea, otorrhea, and mastoid bruising. Depressed skull fractures occur when both tables of the skull are disrupted and pushed below the surrounding skull by a distance greater than the thickness of the skull. Depressed skull fractures are treated operatively if they are open fractures, if there is a CSF leak, and if there is a mass lesion below it.

Epidural hematoma: An epidural hematoma results from hemorrhage between the skull and dura. It often results from a laceration of the middle meningeal artery, but may also result from disruption of a subcortical vein. It is often associated with an overlying skull fracture. An epidural hematoma may present in several ways. The classic presentation is a child who sustains head trauma and has a brief loss of consciousness followed by recovery and a "lucid interval." After a variable period of time there is a rapid loss of consciousness due to expansion of the hematoma.

Patients with severe head trauma may present with an epidural hematoma and underlying brain injury, resulting in coma. A minority of patients will have a small epidural hematoma and will be neurologically normal. These patients can be closely observed in an intensive care setting. Patients with an epidural hematoma and neurologic dysfunction undergo craniotomy and evacuation of the hematoma.

Subdural hematoma An acute subdural hematoma occurs from traumatic disruption of bridging veins, resulting a blood collection between the dura and the surface of the brain. The clinical outcome is often worse than an

epidural hematoma due to the associated underlying brain injury. Evacuation of the hematoma is performed if its presence is thought to contribute significantly to neurologic status. An infant with an acute subdural hematoma should be considered to be a victim of child abuse until proven otherwise.

Cervical spine injuries:

Children injure their cervical spines less than adults. Despite this fact, the need for cervical immobilization and suspicion of cervical injury is paramount when managing the pediatric trauma patient. The personal and societal cost of the spinal cord injured patient is enormous. Anatomic differences between adults and children result in a reduced incidence of cervical spine injury in children. It also results in a different distribution of injuries in children when they do occur. The pediatric cervical spine does not attain adult attributes until 8 years of age. The cervical vertebrae exist as nonfused ossification centers prior to this age. The cartilaginous nature of these elements make them less susceptible to fracture. The flexibility of the cervical spine in the infant and young child is also likely to be a protective element. This flexibility is due to laxity of the intraspinal ligaments, the horizontal nature of the facet joints, and the relative weakness of the muscles of the neck. Unlike adults, the infant and young child has a proportionally greater chance of injuring the upper cervical vertebrae because of their large heads and flexible necks. Nitecki and Moir found that in patients under 8 years of age 87% had an injury at the third cervical vertebrae or above. In this study, higher fatality rates were associated with higher levels of cervical injury, with a C-1 fracture having a 17% fatality rate. xx Older children and adolescents usually sustain cervical spine injuries as a result of motor vehicle accidents and sporting activities. Injuries in younger children usually occur during falls. Child abuse may be the cause of cervical spine injury in the infant and young child.

All seriously injured pediatric patients should have cervical immobilization with an appropriate size Philadelphia collar placed at the scene of the accident. A history of neck pain, numbness, or weakness should be sought from the conscious patient. Physical exam determines the presence of extremity motor function, deep tendon reflexes, and sensation. The bulbocavernosus reflex should be performed in any patient suspected of spinal cord injury to assess for cord "sparing". A lateral cervical spine film should be performed as part of the initial assessment. Complete assessment mandates that all seven of the cervical vertebrae are seen to the junction with the first thoracic vertebrae. In order to see the C-7,T-1 junction, the shoulders may need to be pulled down or a "swimmers view" performed. The cervical spine film should be examined for alignment of vertebral bodies, height of the vertebral bodies, and enlargement of the prevertebral stripe. Many children have either a straight or kyphotic appearance to the cervical vertebrae. This may confuse the inexperienced examiner into

suspecting an injury. If necessary, flexion-extension views of the stable patient are required to resolve this question. After the secondary survey, the cervical spine is completely assessed by performing an anterior-posterior (AP) view and an odontoid view. Considerable experience is necessary for the interpretation of these views in the young child as several developmental anomalies may mimic fractures. CT scanning is used for the assessment of children with persistent pain and no fracture on plain films. CT scanning is also helpful for the evaluation of fracture areas, the assessment of subluxation injuries, and visualization of the spinal cord in injured patients.

The specific injuries children sustain are similar to those occurring in adults. These are atlantoaxial injuries, rotary subluxations, odontoid fractures, and arch fractures. The principles of treatment are also similar to those applied to adults. Skeletal traction is used to reduce the vertebral column and achieve alignment when fracture or spinal cord compression is present. Gardner-Wells tongs are used for older children and adolescents and halo traction is used for younger children. Although the use of steroids has not been rigorously tested in children, the results in adults suggest its application to children.

A unique injury of the spinal cord in children merits a separate discussion. Due to the pliable nature of a child's vertebral column, injury to the spinal cord can occur in the absence of a fracture. This entity, spinal cord injury without radiographic abnormality (SCIWORA) can occur at any spinal cord level but occurs at higher levels in younger children. ^{xxi} This syndrome often presents hours to days after the initial injury. Most lesions are complete and recovery is rare.

Facial injuries: Several case series document the relatively uncommon occurrence of maxillofacial injuries in children. The majority of injuries are nasal fractures, dental fractures, mandibular fractures, and external ear injuries. ^{xxii,xxiii,xxiv} When maxillofacial injuries are noted, there must be a strong suspicion of associated cranial and orthopedic injuries. Successful management of oral and maxillofacial injuries requires a multispecialty approach including dentists, oral surgeons, plastic surgeons, pediatric general surgeons, otolaryngologists and occupational and physical therapists. The anatomic deformity and ensuing hemorrhage from these injuries often complicate airway management. The patient with depressed consciousness or poor respiratory effort should have an orotracheal airway established if intubation allows visualization of the larynx. Patients with severe maxillofacial injuries or suspected laryngeal injury should have a tracheostomy or cricothyrotomy performed. Patients with suspected injury to the paranasal sinuses or nasal fractures should not have nasotracheal intubation performed. These patients should also have an orogastric tube placed to avoid passage of the nasogastric tube through the cribriform plate into the cranium. Specific injuries and their management will be discussed.

Dental injures Dental injury accompanies both serious and minor facial trauma. Minor facial trauma often results in soft tissue injury to the mouth and face and loss of dentition. Loss of deciduous teeth in the absence of mandibular injury does not result in morbidity or deformity. It is unusual for injury of the deciduous tooth to result in injury to the underlying permanent teeth. Injury to the permanent teeth must be handled in an expeditious manner. Once recognized, the injured tooth should be placed back in the socket. If the child is unconscious or too young to do this, the tooth can be placed in milk or saline. The tooth should not be debrided if it is clean. All attempts should be made to replace the tooth by one hour. Once replaced in the socket, it is sprinted in place.

Mandibular injuries Children have an equal tendency to sustain mandibular injuries as adults. Mandibular injuries are defined by their location. These include symphysis, angle, dentoalveolar, condylar, and body fractures. Like adults, these injuries can be missed and all children should have their occlusion checked prior to discharge. Minor injuries without displacement of bone can be treated conservatively. Many fractures are treated with intermaxillary fixation. This is achieved by either stainless steel dental wiring, placement of Erich arch bars, or by using acrylic cement to join adjacent teeth. Deciduous and new permanent teeth offer a less stable source of fixation, potentially compromising fracture healing. Other treatment options include external fixation with the "Joe Hall Morris" device, and internal fixation.

Midfacial fractures Owing to the lack of prominence and cartilaginous nature of the midface in the child, injuries are rare. When they occur, they are classified and managed according to adult principles. Classification of pediatric injuries follows the LeFort I, II, and III system. Injuries are stabilized by buttressing them to the stable skeletal structure above the plane of injury.

Nasal injuries Most facial fractures in children are nasal fractures. They result from a variety of injuries including motor vehicle accidents, sporting activities, assaults, falls, and child abuse. Most nasal fractures can be diagnosed clinically. All trauma patients should have a nasal speculum exam to assess for fractures and septal hematoma. Untreated hematomas of the nasal septum can result in infection and septal necrosis. If a hematoma is identified it is incised in a dependent fashion and the nose is packed to prevent recurrence. Radiographs are often not helpful. CT scanning may be useful in planning the repair of complex nasal injuries involving the nasoethmoid area. Simple nasal fractures are either reduced immediately or treated 3-4 days later.

Sinus injuries Sinus injuries in young children are uncommon because of the delayed formation of the maxillary and frontal sinuses. The most common injury is to the maxillary sinus, which is the first to pneumatize.

Maxillary sinus injuries should be suspected in children with evidence of significant facial trauma. Physical findings suggestive of maxillary injury are unilateral midface depression and periorbital bruising. Opacification of the maxillary sinus is a clue to the diagnosis. CT scanning provides the anatomic detail necessary for definitive treatment. Treatment options include observation, intranasal antrostomy, or anterior maxillary antrostomy.

Ear injuries Injury to the ear is largely unrecognized as problem in pediatric trauma. The ear is the most commonly injured organ of sensation in motor vehicle accidents. The ear can be injured in one of three locations: external ear, middle ear, and inner ear. During the secondary assessment the external ear should be observed and the ear canal inspected with an otoscope. The examiner should note the presence of blood or clear fluid in the ear canal. The tympanic membrane should be observed for disruption. Injuries to the external ear include simple and complex lacerations, hematomas, burns, and avulsion. Like a hematoma of the nasal septum, a hematoma of the ear must be immediately drained to prevent necrosis of the cartilage and overlying perichondrium. Injuries to the middle ear and inner ear may result in tympanic membrane perforation, perilymphatic fistula, ossicle injury, and facial nerve injury. All trauma patients should be assessed for conductive and sensorineural hearing loss, dizziness, tinnitus, and vertigo.

B. CHEST INJURIES

Chest injuries in children are uncommon, yet when they occur result in significant morbidity and mortality. Approximately 5% of all pediatric patients admitted for trauma will have a chest injury. However, this minority of patients experiences a mortality rate approximating 25%.²⁵ Most children sustain chest injuries as a result of blunt trauma. Chest injuries in older children and adolescents result from pedestrian and passenger motor vehicle accidents. Injuries in infants and younger children occur due to falls, motor vehicle accidents and child abuse. Mortality in children with thoracic injuries occurs predominantly from associated injuries. When data from the National Pediatric Trauma Registry (NPTR) was analyzed by Cooper et al, only 14% of those patients dying with thoracic injuries died from their thoracic injuries alone. Most children with thoracic injuries die from closed head injuries. Of 1,553 thoracic injuries registered with the NPTR only 230 (15%) were due to penetrating trauma. Overall mortality from penetrating chest trauma was 14%, which is essentially equal to that of blunt chest trauma (15%).^{xxv} Anatomic differences between children and adults and different mechanisms of thoracic injury results in a different distribution of injuries within age groups. Because of their pliable thoracic cavities, children sustain less rib

fractures than adults, therefore children may sustain significant intrathoracic trauma without external signs. The pliable nature of the child's thoracic cage also prevents it from being a significant protective mechanism. This may be the reason why children sustain pulmonary contusions more frequently. Most children have no preexisting pulmonary disease and this may partly explain why children with pulmonary contusions are intubated for shorter periods of time than adults. Despite the rare occurrence of thoracic injuries in children they must be suspected, diagnosed, and treated rapidly because of their immediate threat to life. Pediatric trauma teams, accustomed to nonoperative approaches, must be ready to quickly intervene to treat one of these life threatening injuries.

Intrathoracic and chest wall injuries

Rib fractures: Approximately half of children sustaining thoracic trauma have rib fractures. Although the frequency of rib fractures in children in each clinical series varies, (30% to 50%) it is widely held that rib fractures occur less frequently in children than in adults. xxvi,xxvii As was mentioned previously, the pliable chest wall of the child may sustain no injuries while significant intrathoracic injury occurs. In Nakayama's report, 52% of the children sustained serious intrathoracic injuries in the absence of rib fractures. 27 However, when rib fractures do occur, the risk of mortality increases. Children with increasing numbers of ribs fractured have a proportional increase in mortality. 26 Fracture of the first rib in children does not indicate the same risk of great vessel injury that it does in adults. 27

The identification of rib fractures in a child should heighten the suspicion for serious intrathoracic injury such as pneumothorax, hemothorax, and pulmonary contusion. Rib fractures should be treated symptomatically with oral or intravenous analgesics, intercostal nerve blocks, and occasionally with epidural analgesia. The impaired respiratory effort associated with painful rib fractures results in hypoventilation and atelectasis. Early ambulation and chest physiotherapy help avoid pulmonary complications.

Pulmonary contusion: Pulmonary contusion is a common sequela of chest trauma in children. In most series it predominates as the most common intrathoracic injury. xxviii Pulmonary contusions in children have a more favorable outcome than in adults. Less children require mechanical ventilation and for shorter durations. Unlike pulmonary contusions in adults, those in children present early. Many children with a pulmonary contusion have an associated intrathoracic injury such as a hemothorax or pneumothorax. Children identified as having a pulmonary contusion should have their oxygenation monitored frequently in the first 48 hours. Most children will only require supplemental oxygen for a brief period of time. Chest x-rays should be repeated during the first 48 hours to assess the status of the pulmonary injury and to identify the late presentation of associated

injuries. Most pulmonary contusions will resolve in 7-10 days. The occasional pediatric patient will develop respiratory failure as a result of a pulmonary contusion. Conventional ventilation, which minimizes barotrauma, should be used initially. The rare pediatric patient may develop fulminant respiratory failure, which requires extracorporeal life support.

Pneumothorax and hemothorax When considered together, pneumothorax and hemothorax are the second most common pediatric chest injury. Both injuries can result in immediate cardiorespiratory collapse. Unfortunately, these injuries may be either self-limited or indicate serious intrathoracic pathology. Nakayama noted the presence of pneumothorax in 37.1 % and hemothorax in 13.3% of the patients he reviewed. Predictably, both injuries occur at a higher frequency when penetrating thoracic trauma is considered. Approximately 2/3rds of the patients with penetrating thoracic trauma in the NPTR database had either a hemothorax or pneumothorax.²⁷ The flexibility of the child's mediastinum is thought to predispose them to tension pneumothorax, a potential complication of a simple pneumothorax. Most pneumothoraces can be diagnosed with the initial AP chest x-ray. Unfortunately, some pneumothoraces are only visible with an upright chest x-ray. In addition, some pneumothoraces are only manifest after the initiation of positive pressure ventilation.

Some small pneumothoraces in spontaneously breathing patients can be observed. All large pneumothoraces and those in mechanically ventilated patients should be treated with tube thoracostomy. An appropriate size chest tube should be placed in the fourth to fifth intercostal space in the mid-axillary line on the affected side. A chest x-ray should be performed to assess the placement of the tube and to confirm evacuation of the pneumothorax. Most pneumothoraces resolve with 2-3 days of suction. A persistent or large air leak should immediately raise the suspicion of a tracheobronchial injury. These are unusual injuries but they are responsible for many of the operative procedures performed on children with thoracic trauma. If a tracheobronchial injury is suspected, flexible bronchoscopy is performed. Occasionally, late injuries are diagnosed by CT scanning.^{xxix} Small tears in the membranous trachea can often be managed nonoperatively. Larger injuries are approached through either a thoracotomy or median sternotomy. Injured tissue is debrided and an end-to-end anastomosis is performed. These injuries are optimally repaired early to avoid pulmonary infection and loss of pulmonary parenchyma. The initial form of treatment for an identified hemothorax is placement of a chest tube. Ideally, the largest possible tube is placed to aid in the evacuation of blood. The initial and subsequent blood loss through the tube should be noted. Sources of blood loss within the chest cavity include intercostal muscles and vessels, pulmonary parenchyma and great vessels. Low pressure vessels in the lung usually stop bleeding when the lung is expanded. Indications for thoracotomy are a chest tube output of 2-3

ml/kg/hr, an initial output of greater than 20-30% of estimated blood volume, or rebleeding after initial cessation of blood loss.

Cardiac injuries

Cardiac injuries after thoracic trauma in children are rare. Blunt injuries to the pericardium and heart occur in all age children. Injuries most commonly result from motor vehicle accidents, falls, and crushing forces. Penetrating injuries most commonly occur in adolescents as a result of stabbings or gunfire. A common sequelae of cardiac and pericardial injury is pericardial tamponade.

Pericardial tamponade occurs when blood fills the pericardial space and impairs cardiac function. The myocardium, coronary vessels, pericardial vessels and great vessels can all be sources of bleeding that cause pericardial tamponade. The young child is theoretically at higher risk because of the smaller volume of the pericardial space. Blood in the pericardial space impairs end-diastolic filling, coronary artery flow, and ventricular wall motion. This results in progressive myocardial dysfunction, hypotension, and eventual cardiac arrest. The diagnosis of pericardial tamponade proceeds from a high index of suspicion. It should be considered in all hypotensive patients who have sustained thoracic trauma. Clinical findings such as muffled heart sounds, cyanosis, and elevated central venous pressure, and electrocardiograms changes are unreliable. The unstable patient who is suspected of having pericardial tamponade should undergo needle pericardiocentesis. Drainage of small amounts of blood will often restore the hemodynamic status of the patient. An echocardiogram can be performed in the stable patient. Patients with penetrating wounds of the thorax and upper abdomen should be considered to be at high risk for this injury. Suspicious injuries in the appropriate setting are managed immediately with a pericardiocentesis. Unstable patients with penetrating injuries of the thorax should undergo emergency thoracotomy for stabilization prior to definitive management of other intrathoracic injuries. Patients who undergo successful pericardiocentesis and require operative intervention for other reasons should have the induction of anesthesia preceded by subxiphoid pericardiotomy under local anesthesia. This is done to prevent potential cardiovascular collapse due to anesthetic depression of an already impaired myocardium.

Myocardial Contusion: It is unclear what the true incidence of myocardial contusion is in children. The incidence of myocardial contusion depends on the diagnostic studies used. Two pediatric studies using clinical criteria found the incidence of myocardial contusion to be 6 and 7.7%. 25,xxx Ildstad, looking at myocardial contusion in children prospectively with MUGA scans, found the incidence to be 43%. xxxi Much of the difficulty lies in defining what the "gold standard" for diagnosis. Langer et al found poor correlation between ECG, CPK with isoenzymes, echocardiography

and nuclear pyrophosphate scan in the diagnosis of myocardial contusion in 41 patients with blunt thoracic trauma. xxxii It is clear that children at high risk are adolescents with adult type "steering wheel" injuries and young children with crush injuries. These children and those with a history of thoracic trauma and chest pain or arrhythmias and those with sternal fractures should be evaluated for the presence of a myocardial contusion. Auscultation should be performed to rule out any murmurs suggestive of intracardiac injury. Patients should be placed on continuous EKG monitoring to observe for arrhythmias. Measurement of serum CPK isoenzymes seems to add little to the evaluation. xxxiii At risk patients should undergo echocardiography to assess for wall motion abnormalities and valvular dysfunction. The presence of a myocardial contusion does not seem to increase the anesthetic risk of urgent operative procedures.

Cardiac Concussion: Cardiac concussion refers to abnormalities of cardiac function which result from a sudden, high velocity force to the chest. These may result in sudden arrhythmias such as ventricular fibrillation and asystole. These injuries are rare but usually occur during sporting events in which the child is hit in the chest with a ball. Other rare cardiac injuries include valvular disruption, acquired ventricular septal defects, coronary artery lacerations and atrial or ventricular rupture. If possible, atrial or ventricular injuries are repaired without cardiac bypass. This avoids anticoagulation, which could complicate the management of associated injuries of the brain or abdomen. It is appropriate to discuss emergency room thoracotomy for pediatric trauma patients

within the context of intrathoracic injuries. Emergency room thoracotomy is rarely indicated in children. Children who present without vital signs from blunt trauma do not benefit from an emergent thoracotomy. The indications for thoracotomy are similar to those for adults with one exception. Valid indications include:

- 1 . Witnessed loss of vital signs in the emergency room
2. Cardiac arrest due to penetrating thoracic trauma
3. Rapid hemodynamic instability due to massive chest tube blood loss
4. Failure of closed chest massage.

Thoracotomy to obtain aortic control for intrabdominal hemorrhage does not seem to be a valid indication for emergency room thoracotomy as enough collaterals exist in children to prevent this maneuver from being effective. The child with intrabdominal injuries and hemodynamic instability is best treated in the operating room with intrabdominal vascular control.

Diaphragmatic Injuries

Most injuries to the diaphragm result from blunt trauma. The majority of patients injure their left hemidiaphragm. Diaphragmatic injuries from penetrating injuries occur less frequently and occur equally on the right and left. Significant force is required to injure the diaphragm and as a result the incidence of associated injuries is high. xxxiv Rib fractures are associated with diaphragmatic injuries on both sides. Diaphragmatic injury on the right side is associated with liver injury and injuries on the left are predictably associated with splenic injury. The patient with diaphragmatic rupture may either be asymptomatic or have rapid cardiovascular collapse. A number of findings seen on chest x-ray suggest the diagnosis of diaphragmatic rupture. These are:

1. Displacement of the tip of the nasogastric tube into the chest.
2. Haziness or an indistinct appearance to the diaphragm
3. Air- fluid levels in the chest.

If doubt remains, water soluble contrast in either the colon or stomach can often establish the diagnosis. Several options exist for repair of these injuries. A transabdominal approach permits easier reduction of hollow and solid viscera from the chest and allows inspection for associated intrabdominal injury. A thoracic approach, either through an extension of the incision or separate thoracotomy may be necessary to treat any intrathoracic injuries. The diaphragm is closed with interrupted, nonabsorbable sutures. Many surgeons reinforce their closure with pledgeted sutures. Occasionally a penetrating injury causes enough tissue loss to the diaphragm that prosthetic material is required for closure.

Esophageal injuries

Blunt esophageal injury is very rare. This is due to the distensible nature of the esophagus and its location in the posterior mediastinum. Most esophageal injuries are iatrogenic. Most esophageal injuries due to trauma result from penetrating trauma, usually in the neck in older children. Management of esophageal injuries in children follows the same principles for adults.

C. ABDOMEN

Initial diagnostic approach

The majority of intraabdominal injuries that children sustain are due to blunt trauma. Therefore, most injuries are not directly observable. An appropriate diagnostic approach to these injuries should identify those children in need of operative therapy early. In addition, the application of appropriate diagnostic tests should be based on the ability to predict the frequency of injury. Clinical evaluation of the child is often hampered by lack of cooperation due to fear and anxiety. The high frequency of head injuries in seriously injured children often mandates a more liberal use of diagnostic studies.

Approximately 8% of children reported to the National Pediatric Trauma Registry had an intraabdominal injury. The solid organs are most commonly involved (spleen 30%, liver 28%, kidneys 28%). Injury to hollow viscera are recorded 14% of the time. It is unusual for a child to die of an isolated intraabdominal injury. Only 9% of the children with isolated abdominal injury died. 25

The diagnostic approach to a stable child suspected of having an abdominal injury should evaluate for solid organ injury and rule out hollow visceral injury. As most children with abdominal injuries are treated nonoperatively, it is imperative for solid organ injury to be imaged accurately. Fortunately, the risk of hollow visceral injury in blunt trauma is sufficiently low to support a nonoperative approach.

Evaluation of the abdomen is a component of the secondary survey. Specific mechanisms of injury raise suspicion for intraabdominal injury. The child restrained by only a lap belt is at risk for hollow visceral injury and lumbar fracture. The child falling onto a bicycle handle bar is at greater risk for duodenal and pancreatic trauma. If a child is cooperative, the abdomen should be palpated and signs of peritoneal irritation sought. Unfortunately, most young children are not able to be easily examined. It may be necessary to place a nasogastric tube to relieve abdominal distension due to air swallowing and gastric dilatation. A rectal examination should be performed to detect blood suggestive of intestinal injury. The unstable children with abdominal distension should be taken to the operating room for laparotomy. If it is unclear whether the abdominal findings are due to hemoperitoneum, then diagnostic peritoneal lavage may rarely be indicated. Ultrasound may also be helpful if this is immediately available. If the child is stable, a decision must be made regarding further diagnostic evaluation. It is unusual for plain abdominal films to be useful. They contribute meaningfully if free air is detected or a pelvic or spine fracture is noted. Frequently, the most important decision is whether to proceed with CT imaging of the abdomen and pelvis. CT imaging of pediatric abdominal injuries is the standard of care. CT imaging of the abdomen and pelvis in

children reliably identifies intraperitoneal and retroperitoneal injury. Identification of solid organ injury is aided by injection of intravenous contrast. This is of particular importance in evaluating renal injury. The use of oral contrast has not been shown to aid in the diagnosis of hollow organ injury and therefore its use is not recommended.

While evaluation of solid organ injury is well established by CT scanning, evaluation of bowel injury is more difficult. Fortunately, this is an infrequent result of blunt trauma in children. Unequivocal evidence of bowel perforation is usually not provided by CT scanning. In most clinical series contrast extravasation, free air or bowel disruption is not detected. However, virtually all patients with bowel perforation have indirect signs of bowel injury. These include free intraperitoneal fluid in the absence of solid organ injury, bowel wall thickening, bowel wall enhancement and bowel wall dilatation. xxxv

The most common indication for abdominal CT scanning in children is an inability to evaluate the abdomen because of a head injury or inability of the child to cooperate. The mechanism of injury is also an important indication for CT scanning. Mechanisms of injury, which suggest the need for CT evaluation, are lap belt injuries, handlebar injuries, child abuse and falls. It is unusual for lab values to solely justify CT scanning. One exception is elevation of the SGOT > 200 and the SGPT >100. These are very sensitive indicators of parenchymal liver injury. xxxvi However in our experience laboratory values were not sufficiently predictive to identify organ injury and, furthermore that patients had returned from CT scan prior to the return of their laboratory values

The role of diagnostic laparoscopy in children is not well defined. It has had preliminary application in adult trauma situations. It seems to be most useful for examining the abdomen in stable patients with wounds that have penetrated the abdominal wall and peritoneum. Experimental evidence suggests an elevation in ICP results from pneumoperitoneum. xxxvii As many children with significant abdominal trauma have head injuries this is a serious concern for the application of trauma laparoscopy to children with blunt abdominal trauma.

Injuries to solid viscera

Spleen Trauma is the most serious threat to the well being of the child. The most common mechanism of injury to the child is blunt trauma and the most commonly injured organ is the spleen. Successful nonoperative management of splenic injuries has resulted from improved diagnostic methods, appreciation for the immunologic role of the spleen and creation of critical care units for children. The spleen was formerly thought to have no useful function and this bias was reflected in the practice of immediate

removal following injury. Although laboratory work had indicated important immune functions of the spleen in 1910, recognition of its importance followed the report of King and Schumaker over forty years later. This report detailed the occurrence of fatal infections in two of five patients who had undergone splenectomy for hematologic disease. xxxviii A body of literature soon developed which corroborated these findings and gave credence to the syndrome of "19 overwhelming post splenectomy infection". (OPSI). A long held bias suggested that the spleen was not repairable. However, discovery of the segmental nature of the blood supply of the spleen led to successful attempts at splenorrhaphy. Finally, the development of modern CT scanning allowed the early detection of splenic injury, thus obviating the need for peritoneal lavage and/or diagnostic laparotomy.

The spleen is the most commonly injured intraabdominal organ following blunt trauma. It is unusual for it to be injured following penetrating trauma in children. Iatrogenic injury occurs less commonly than in adults and when it does usually occurs during performance of an antireflux procedure. The spleen develops in the dorsal mesogastrium, placing it in direct contact with the pancreas and in proximity with the stomach. Its blood supply, the splenic artery, is a branch of the celiac axis. The splenic artery usually passes above and behind the body and tail of the pancreas. It then divides into segmental branches at the hilum. Michels analyzed the splenic artery in 100 spleens and found an average of 17 branches at the hilum. xxxix Two to six venous trunks exit the spleen and form the splenic vein, which joins the superior mesenteric vein and forms the portal vein. Anatomic studies indicate that venous drainage within the spleen also follows a segmental pattern. The presence of this segmental blood supply and the absence of intersegmental anastomoses facilitate conservative splenic surgery and nonoperative management.

The realization that the absence of a spleen predisposed to infection led to investigation of the normal function of the spleen. Information regarding splenic function is derived from clinical observation of splenectomized patients and laboratory data from experiments involving splenectomized mammals. It is apparent that the spleen contributes to immunoglobulin production.

Splenectomized animals have deficient IgM levels and IgM responses to intravenous antigen challenge. xl This deficient response seems specific to intravenous stimuli, as appropriate responses to subcutaneous antigens are mounted. This deficiency seems to result from a decrease in the number of cells producing immunoglobulin. Other important immunoglobulins (IgA, IgG) do not seem to be decreased as a result of splenectomy. xli The spleen also produces tuftsin and properdin. Splenectomized animals have deficiencies in both these peptides. Tuftsin is a tetrapeptide that is thought to act on the neutrophil to promote phagocytic activity. The spleen is also

known to filter and destroy effete red blood cells. The effect of the absence of this activity in the splenectomized patient is unknown.

Splenic injuries in children usually result from blunt mechanisms. In most series, motor vehicle accidents are the most frequent cause of splenic injury. Falls are the next most common etiology. xlii,xliii The spleen is particularly vulnerable in the child because of the lack of protection afforded by the flexible rib cage. The majority of children are hemodynamically stable upon presentation. Unstable children usually have associated intraabdominal and/or extraabdominal injuries.

The majority of children with blunt injuries to the spleen can be managed nonoperatively. The initial assessment defines the patient at risk for splenic injury on the basis of physical findings or mechanism of injury. The diagnostic procedure of choice is an abdominal CT scan. This defines disruption of splenic architecture and identifies coexisting intraabdominal and retroperitoneal injuries. Bolus intravenous contrast should be administered to define the splenic injury.

The hemodynamically stable child with a splenic injury is initially admitted to the hospital. Serial hematocrits are performed during the first 48 hours. Recently a recommendation on the management of splenic trauma that was created by “averaging” the management patterns of a group of pediatric surgeons has been proposed. These guidelines apply to the patient with isolated spleen or liver trauma. Any patient with a grade 4 or higher injury is admitted to the ICU, whereas all the other grades are admitted to the floor. The length of the hospital stay is formulated by adding one to the grade (i.e. grade 2, stays 3 days) and for length of time free from contact activities grade plus 2 (weeks). Most pediatric surgeons allow early ambulation. There does not seem to be a need for the routine performance of post-injury scans as it seldom influences clinical behavior. xliv

The patient with instability and a splenic injury, or the patient who becomes unstable or needs continued transfusion is taken to the operating room. The initial focus of laparotomy should be splenic conservation. The transverse orientation of the arteries and veins supplying the spleen theoretically predispose to transverse fractures. The absence of significant collateral arterial circulation limits bleeding and facilitates splenorraphy. If splenorraphy is contemplated, the spleen should be mobilized. Surgical options include wrapping the spleen in hemostatic agents, partial splenectomy with or without parenchymal suturing and direct suture of bleeding vessels with omental placement on the raw splenic surface. The patient with ongoing hemorrhage from other injuries, a disintegrated spleen or a totally devascularized spleen should undergo prompt splenectomy. Drainage of the left upper quadrant is not necessary in the absence of a pancreatic injury.

All patients undergoing splenectomy are at risk for overwhelming post-splenectomy infection (OPSI). First described in 1952 by King and Schumaker, it represents an overwhelming infection with a case fatality rate of approximately 50%. The majority of infections are due to encapsulated organisms such as H.influenza, S. pneumonia, and N. meningitides. Most infections occur within 2 years of splenectomy but they can occur at any time in the future. Splenectomized patients should receive vaccines against S. pneumoniae, H. influenza, and N. meningitidis. Most surgeons place their patients on oral penicillin for 1-2 years. Patients should be counseled on the hazards of OPSI and the need to seek immediate medical attention in the event of fever.

Liver Modern management of pediatric liver injuries emphasizes selective nonoperative management. 36,xlv Mortality from pediatric liver injuries results from associated injuries (head, IVC) or exsanguination from diffuse parenchymal injury or major hepatic venous injuries. Biliary tract injuries will be considered after a discussion of liver injuries. Children with liver injuries present with a wide spectrum of severity of injury to the liver. Most children have small parenchymal lacerations that do not require operative therapy. At the other end of the spectrum is the child with fatal juxtahepatic venous injury. Cooper noted that of children sustaining serious intraabdominal injury, 27% had a liver injury. 25 Liver injury is also an indicator of the presence of associated injuries. Children with liver injuries rarely die of these injuries. Rather, mortality results from concomitant head injury.

Most liver injuries result from either motor vehicle related accidents or to a lesser degree, falls. Liver injury should be suspected in any seriously injured child. Admission liver function tests are a fairly sensitive indicator of parenchymal injury when the SGOT is greater than 200 and the SGPT is greater than 100. 36 CT scanning is the best imaging method for the evaluation of liver injuries. It allows definition of parenchymal lacerations, intrahepatic hematomas and other solid organ injuries. Most children have injuries to the posterior segment of the right lobe. Hemodynamically unstable patients suspected of having a liver injury are taken immediately to the operating room. Most patients are hemodynamically stable when observed to have a liver laceration on CT scan. These patients are managed in a similar fashion to those patients with a splenic injury. See above section for protocol for ICU, hospital stay and activity restrictions. Serial Hematocrits should be checked on these patients until a stable pattern has been established.

Liver injuries requiring laparotomy are managed with increasing levels of intervention. Most injuries will stop bleeding with direct pressure. Hemostatic agents may help with this. If this is unsuccessful, suture ligation of bleeding points is attempted. Should bleeding continue, occlusion of the hepatic artery and portal vein (Pringle maneuver) should be performed.

Formal hepatic resections should be a last resort, as most injuries can be managed by resection debridement. An occasional patient will have a juxtahepatic venous injury. This should be suspected in the patient with an abdominal injury that is rapidly unstable and in the patient who has persistent liver bleeding despite inflow occlusion. If this is suspected preoperatively, one approach is to perform a median sternotomy first. This allows control of the suprahepatic vena cava prior to releasing the tamponading effect of the abdominal wall when the abdomen is opened. The best approach in many patients may be to pack the right upper quadrant and prevent the inevitable occurrence of intraoperative hemorrhage, coagulopathy, hypothermia and death. Then, when the patient is stable they are brought back to the operating room with shunting or venovenous bypass available and the venous injury is approached in a deliberate fashion under optimal conditions.^{xlvi}

Nonoperatively managed liver injuries uniformly heal within 3-6 weeks. Complications that have been noted include bile leaks and hemobilia. Biliary tract injuries in children are infrequent occurrences. Blunt injury most frequently occurs to the gallbladder. This should be treated with cholecystectomy. Because most injuries to the extrahepatic bile ducts result from penetrating trauma it is rare in children. When it occurs, treatment principles, follow those for adults. Minor injuries to the bile duct may be treated with closure and t-tube drainage. Anything more than a mild injury should be treated with a biliary enteric bypass to avoid the high stricture rate that occurs with reanastomosis of the injured bile duct.

Pancreatic and duodenal trauma

Injuries to the pancreas and duodenum will be discussed together because of their anatomic relationship and frequency of coexisting injury. When all ages are considered, a minority of injuries occur to the pancreas and duodenum in children. Only 2% of duodenal injuries occurred in children in the review by Corley et al. ^{xlvii} Pancreatic injuries result from blunt trauma in 1 to 10% of all children sustaining blunt abdominal trauma. Due to the shared blood supply and proximity, any injury to either organ mandates assessment of both.

Duodenal injury

The duodenum's location and relationship to other organs has important implications for injury detection and management. The duodenum is fixed in a retroperitoneal position in its second and third parts. Its posterior location protects the duodenum from anterior blows, however, forces directly on the vertebral column may disrupt the duodenum. Penetrating injuries to the duodenum are potentially lethal due to the proximity of the

portal vein, hepatic artery and superior mesenteric artery and vein. Any injury to the duodenum should arouse suspicion of a pancreatic injury.

Most injuries to the duodenum in children result from blunt trauma. Most occur from isolated blows to the epigastrium resulting from falls and bicycle accidents. Recognition of these types of injuries is often delayed days to weeks. This results from the absence of peritonitis from contained rupture of the retroperitoneal duodenum. Patients with minor injuries often have minimal right upper quadrant or epigastric tenderness. An occasional child will present with right lower quadrant tenderness that mimics appendicitis.

The diagnosis of duodenal injury is aided by a high index of suspicion in certain clinical situations, in particular those patients who have sustained falls or blows to the epigastrium. The pathognomonic sign of duodenal rupture on abdominal radiographs is retroperitoneal air bubbles. However, this is not a consistent sign in early injury. Other less specific signs include scoliosis toward the right, obliteration of the right psoas and ileus. If no other injuries are suspected, a hypaque swallow defines duodenal anatomy and will usually demonstrate extravasation. In most cases however, associated abdominal injury will be suspected and the diagnostic procedure of choice will be a CT scan. Every attempt should be made to fill the stomach adequately with contrast material so that the duodenum can be opacified. Minor duodenal injuries can often be observed. Small duodenal injuries are closed primarily and periduodenal drains placed. Duodenal closure can be buttressed with either an omental or jejunal patch. Significant injuries of the duodenal wall should be repaired and accompanied by proximal diversion of gastric contents. A rational choice is the Vaughn pyloric exclusion in which the pylorus is closed with either absorbable or nonabsorbable suture material and a gastrojejunostomy is fashioned. Blunt duodenal trauma may result in an intramural hematoma. If large enough this may obstruct the duodenum and result in a gastric outlet obstruction. Initial therapy includes nasogastric decompression and parenteral nutrition. If conservative therapy is prolonged then extramucosal drainage is performed.

Pancreatic injuries

The pancreas occupies a retroperitoneal position with intimate contact with both the c-loop of the duodenum and the hilum of the spleen. Like the duodenum, it is draped over the vertebral column and is similarly vulnerable to forces which compress it on the spine. Most pancreatic injuries in children result from either bicycle or motor vehicle accidents. They often present in a delayed fashion. Early manifestations of injury include bleeding and abdominal pain. Pancreatic ascites, peritonitis and pseudocysts are all potential late presentations of pancreatic injuries. Computed tomography is the most useful diagnostic procedure. It allows

assessment of the integrity of the parenchyma and defines lesser space fluid collections. ERCP has been shown to be useful to assess for pancreatic duct disruption in children. ^{xlviii} Minor pancreatic injuries should be treated with external drainage. Pancreatic transaction with duct disruption should be managed by distal resection with splenic preservation. The management of post traumatic pancreatic pseudocysts is controversial. Small pseudocysts will often resolve spontaneously. Immature pseudocysts causing local complications should be drained externally. It is unclear whether "mature" pseudocysts should be drained externally or internally. Both approaches have been used with success.

Combined pancreaticoduodenal trauma

High velocity blunt trauma or penetrating trauma results in serious combined injuries of the pancreas and duodenum. Early mortality results from exsanguination due to injury to visceral vessels around the pancreas and duodenum. After hemorrhage is controlled, the pancreas and duodenum are assessed. Serious injury to both organs may require pancreaticoduodenectomy. If the patient is unstable it is inappropriate to proceed with an operation of this magnitude. The safest approach is to drain the abdomen, pack off bleeding sites and stabilize the patient in the intensive care unit. Once the patient is resuscitated, the definitive operation can be performed.

Injuries to the kidney, ureter, bladder, and urethra

Urinary tract injury is a common sequela of blunt abdominal trauma in children. Most injuries involve the kidney and they are frequently associated with other intraabdominal injuries. Essentially all children with severe renal trauma will have associated injuries. ^{xlix} Any of several signs or symptoms may alert the clinician to the presence of urinary tract injury during the initial assessment. Patients may complain of abdominal pain, flank pain, pelvic pain or inability to void. Physical findings suggestive of urinary tract injury are blood at the urethral meatus, cephalad displacement of the prostate, pelvic instability and the presence of swelling or hematomas of the flank.

The presence of any of these signs or symptoms warrants diagnostic evaluation. Patients with microscopic hematuria and abdominal findings also need radiologic studies. The study of choice is a CT scan of the abdomen and pelvis with IV contrast administration. This is the study of choice because it can evaluate the function of both kidneys, can identify associated intraabdominal injuries and can detect renal parenchymal injury. Children with inability to void, pelvic fracture or blood at the meatus should have a retrograde urethrogram performed to look for urethral injury. Specific injuries to the kidney, ureters, bladder, and urethra will be discussed.

The kidney is the most commonly injured component of the urinary tract in children and adults. Children are thought to have more frequent renal injuries than adults. This is believed to be due to the greater vulnerability of the child's kidney due to the flexibility of the rib cage, paucity of retroperitoneal fat and larger relative size of the kidneys. Abnormal renal development may predispose to renal injury following trauma. The reported incidents of preexisting renal anomalies in selected series of renal trauma ranges from 1 to 23%.¹ The most common anomaly is hydronephrosis. Wilm's tumor may present with rupture following minor trauma. Most renal injuries are detected following CT scanning for symptomatic hematuria or looking for other intraabdominal injuries. Several classifications schemes exist to categorize injuries to the kidney. li,lii The most practical scheme divides injuries into minor and major ones. Minor injuries include contusions, superficial cortical lacerations and isolated disruption of the fornix. Major injuries are pedicle injuries, deep lacerations with disruption of the collecting system and the "shattered" kidney. The majority (85%) of renal injuries are minor and can be observed. Patients with pedicle injuries suspected on the basis of nonvisualization on CT scan should undergo angiography. Transabdominal exploration should be performed and control of the renal artery and vein obtained. Thrombosed arterial segments should be debrided. Occasionally, interposition saphenous vein grafting is necessary. Most serious renal injuries without pedicle injury can be managed nonoperatively with operative intervention reserved for continuing blood loss or urinary sepsis.

Occasionally the surgeon identifies a perinephric hematoma during urgent laparotomy for hemodynamic instability. If the hematoma is not expanding it should be left undisturbed. If exploration of the hematoma is necessary, then an on the table IVP should be performed to confirm the presence and function of the contralateral kidney. Bladder injury in children results from blunt, penetrating and iatrogenic trauma. In infants and small children, the pelvis is proportionately smaller than adults. Therefore the urinary bladder is intraabdominal and at greater risk for injury. Bladder injuries can be divided into extraperitoneal and intraperitoneal types. Bladder rupture should be suspected in patients with pelvic fractures, blood at the urethral meatus and hematuria. The diagnosis can usually be made by observing contrast passage into the bladder during the pelvic component of the CT scan. Visualization of the bladder is improved by clamping the urinary catheter prior to scanning. Occasionally patients with intraperitoneal bladder rupture present late. Manifestations of this result from the effect of uroascites. These include peritonitis, azotemia and abdominal distension.

Extraperitoneal bladder rupture is best managed by prolonged catheter drainage. Most of these injuries are associated with pelvic fractures and hematomas. Catheter drainage is usually successful and avoids the need to disturb a stable pelvic hematoma. If operative intervention is necessary for

other injuries, intravesical closure of extraperitoneal bladder injuries should be entertained. Intraperitoneal bladder injuries should be managed with operative exploration, debridement and multilayer closure of the bladder. The colon and ureter should be inspected for injury. Bladder drainage with either suprapubic cystostomy or a urethral catheter should continue over 7 to 10 days postoperatively.

Ureteral injuries are rare in children. Younger children usually sustain these injuries as a result of blunt trauma. Adolescents are more likely to have a ureteral injury because of penetrating trauma. The mechanism of blunt injury to the ureter and collecting systems is speculative. One theory suggests that sudden violent upward movement of the kidney during truncal flexion results in distraction and separation of the upper ureter. Other authors have recognized the association of fractures of the vertebral column and have suggested that direct injury to the ureters results from bony fragments or direct compression. ^{liii} It is difficult to diagnose ureteral injuries. Only a minority of patients will have hematuria. Few will have flank pain or swelling. Most patients with serious abdominal trauma will undergo CT scanning. Attention should be paid to the collecting system and ureter of both kidneys. Perirenal or periureteral fluid collection should raise the suspicion of injury. Failure to observe contrast in one ureter should also raise concern for ureteral disruption. Patients with penetrating injury to the retroperitoneum should have the entire ureter inspected. Most blunt injuries to the ureter in children are located at the ureteropelvic junction. Most penetrating injuries occur in the middle of the ureter. If the injury is recognized early, treatment should include debridement of the devitalized ureter, performance of an ureteroureterostomy and placement of a double-J stent. If there is loss of ureteral length, other approaches such as a transureteroureterostomy, ureteroneocystostomy with a psoas hitch or a Boari flap may need to be performed.

Urethral injuries in children are rare and usually occur in males. Frequent mechanisms of injury in males are pelvic fractures or straddle injuries. Most authors recommend initial placement of a suprapubic cystostomy and late urethral reconstruction in the male with a disruption of the prostatic urethra.

Injuries of the stomach and small bowel

Injuries to the small bowel and stomach from blunt abdominal trauma are unusual and can be difficult to diagnose. Injury to the small intestine is likely if penetrating injury to the abdomen has occurred. Blunt injury to the intestine results most commonly from motor vehicle accidents and child abuse. Lap belt injuries are associated in particular with hollow visceral injuries. ^{liv} Both intraabdominal and extraabdominal injuries coexist and make diagnosis of hollow visceral injuries more difficult. The mechanism of blunt intestinal injuries is speculative. Proposed mechanisms include

rupture of "closed loops", shearing at points of fixation (adhesions, ligament of Treitz) and compression against the vertebral column. Injuries to the small intestine and stomach may present immediately, within days or months later. Patients present immediately when the injury results in disruption of the bowel wall. Pneumoperitoneum and peritonitis are early indications of full thickness injury to the bowel. If the injury to the bowel wall is not complete, a contusion can form. Subsequent necrosis of the bowel wall at that location may present as peritonitis 3 to 4 days post-injury. If enough of the bowel wall is contused, a stenosis may result in bowel obstruction weeks to months later. lv

As most blunt abdominal trauma is managed nonoperatively in children, a high index of suspicion for hollow visceral injury must be maintained. A history of direct injury to the abdomen should be sought. Use of a lap belt, in the absence of a shoulder restraint, increases the concern for intestinal injury. The abdomen should be examined for contusions or lap belt ecchymoses, tenderness, distension and absence of bowel sounds. Laboratory studies are unlikely to be helpful in the early evaluation of these injuries. A nasogastric tube should be passed and aspirated to assess for the presence of bright red blood. An x-ray of the abdomen and chest should be performed to assess for free air. The position of the nasogastric tube within the stomach area should be identified. The absence of free air in the abdomen does not rule out intestinal perforation as the majority of these injuries do not result in pneumoperitoneum. As most children with significant blunt abdominal trauma undergo CT scanning, its ability to diagnosis blunt intestinal injury is essential.

Separate studies by Sherck in adults and Wang in children have confirmed the sensitivity (92% and 93% respectively) and specificity (94% and 95% respectively) of CT evaluation of intestinal injury. lvi,lvii These data suggest the utility of CT scanning to screen for intestinal injury. Those children without suggestive findings have a very low chance of having a missed injury. However, positive CT findings may "overcall" intestinal injuries and may lead to nontherapeutic laparotomies. CT findings suggestive of bowel injury include unexplained peritoneal fluid, bowel wall enhancement, bowel discontinuity, interloop fluid, pneumoperitoneum, bowel wall thickening and contrast extravasation. The most sensitive of these findings is the presence of peritoneal fluid in the absence of solid visceral and pelvic injury.

Despite its potential, diagnostic peritoneal lavage (DPL) has very little place in the diagnosis of blunt intestinal injury in children. Its most sensitive result, hemoperitoneum, is not helpful as most intraperitoneal bleeding is due to solid visceral injuries, the vast majority of which are managed nonoperatively in children. Unless food particles are identified, findings such as bacteria, alkaline phosphatase or elevation of the peritoneal white blood cell count are often equivocal or not elevated early in these

injuries. In order to minimize morbidity and recognize intestinal injuries early, CT scan findings must be correlated with clinical impressions and frequent examinations. The following evaluation and management plan is suggested. Patients with diffuse peritonitis and/or pneumoperitoneum should undergo laparotomy. Children with head injuries or equivocal exams should undergo CT scanning. Those with several findings suggestive of bowel injury or those undergoing brain surgery should undergo laparotomy. Children with minimal findings of bowel injury should be observed and reexamined frequently. Equivocal cases may undergo laparoscopy if the examiner is comfortable with examination of the small bowel by this method.

The child with suspected bowel injury should be given broad spectrum antibiotics preoperatively. A vertical midline incision should be performed and the entire small bowel and colon examined. It is extremely important to open the lesser space and inspect the posterior surface of the stomach for injuries. The abundant blood supply of the stomach and small bowel allows repair of most injuries without functional loss. Injuries to the stomach have included perforation of the greater and lesser curvatures and body of the stomach. Injuries to the stomach should be debrided and closed in two layers. Injuries to the small intestine can be debrided and oversewn. If a significant part of the circumference of the small bowel is injured, a resection and anastomosis should be performed. After the injuries are managed, the peritoneal cavity is irrigated. Most children have uncomplicated postoperative courses.

Colon injuries

Traumatic colon injuries in children occur infrequently. Most colon "injuries" result from iatrogenic causes. Like other hollow visceral injuries in children, a high index of suspicion must be present to make the diagnosis of an intraabdominal colon injury. Early detection of injury is important due to the bacterial exposure that results from disruption of the unprepared colon. Principles of repair are the same in children as for adults. Multiple mechanisms result in intraabdominal colon injuries. These include lap belt compression, gun shot wounds, stab wounds and direct blows. Iatrogenic causes of colon injury include perforation during radiologic procedures such as enema reduction by air or barium, perforation during colonoscopy and perforation due to laparoscopic cautery injury. Rectal injuries result from child abuse, foreign bodies, endoscopy and pelvic fractures.

The child with a potential colon injury is resuscitated and managed like any other child with significant trauma. Unfortunately, the diagnosis is difficult to make unless a penetrating injury obviously disrupts the colon. Signs suggestive of injury are peritonitis, pneumoperitoneum and rectal blood. The CT scan may show intraperitoneal fluid, pneumoperitoneum or edema of the colon wall. Patients with a history of rectal trauma and

pneumoperitoneum or peritonitis may benefit from a water soluble contrast enema to identify the site of perforation.

There are many surgical options to treat the intraabdominal colon injury. The most important decision for the surgeon is whether to perform a colostomy. Small colon wounds in patients with minimal contamination can be closed without diversion. Clinical conditions that suggest need for colostomy in the adult are:

1. shock
2. extensive fecal contamination
3. extensive hemoperitoneum
4. two or more intraabdominal injuries
5. more than 8 hours between the injury and repair

It is unclear whether all of these criteria apply to children. Despite the dissimilarities between the right and left colon, primary closure of injuries of both sides of the colon is safe in selected patients. Extensive wounds mandate colon resection and either primary anastomosis or proximal colostomy (or ileostomy) and Hartman's pouch procedure. Significant rectal injuries should be primarily repaired, irrigated and proximally diverted. Presacral drains should be placed and may exit transperineally or transabdominally. All patients should have extensive peritoneal irrigation, and be placed on 5 to 7 days of broad spectrum antibiotics that includes anaerobic coverage. The most frequent postoperative complications are infections. These include intraabdominal abscesses, pneumonia, sepsis and wound infections. Colostomy malfunction, small bowel obstruction and osteomyelitis of the pelvis also occur to a lesser degree.

Special issues

Pelvic fractures Pelvic fractures in children are unusual occurrences. They are important to consider in any seriously injured child because of their association with other injuries and because bleeding from a pelvic hematoma may be life threatening. Pelvic fractures in younger children result from pedestrian-automobile accidents. Pelvic fractures in adolescents result from being automobile occupants and from falls. The mortality rate in children due to pelvic fractures is 1.4 to 25 %. This appears to be lower than in adults.

The forces required to fracture the pelvis are substantial. As such, associated injuries are to be expected. Injuries associated with pelvic fractures can be divided into those with proximity to the pelvis and those distant to the pelvis. Proximity injuries include urethral, bladder, rectal, vaginal, perineal and vascular injuries. Despite a relatively high incidence of hematuria in pelvic fractures in children, urethral and bladder injuries are

uncommon. Reichard et al reported an incidence of bladder and urethral injury of 4 and 0% respectively. 59 These numbers are less than the range reported in adults. Injuries to the rectum, vagina and perineum occur less frequently, but can result in serious short term and long term morbidity. The most frequent distant injuries are craniocerebral and abdominal. The association with head injuries is consistent with the mechanism of pedestrian-automobile collision in which the victim is struck and thrown. Children frequently injure their heads when they hit the ground. It is more likely for children to die of an associated head injury than from hemorrhage into the retroperitoneum.

A fractured pelvis should be suspected in any child with significant trauma. Pedestrian victims of accidents are more likely to sustain these injuries. During the secondary survey, potential pelvic injury is evaluated. Significant pelvic injury can result in obvious deformity or leg length discrepancy. Hematomas of the lower abdomen, flank and perineum are a clue to pelvic fracture. All patients should undergo rectal examination to assess for blood, rectal tone and location of the prostate. An elevated prostate suggests potential urethral injury. Females with pelvic fractures should undergo vaginal examination to evaluate for lacerations secondary to bone fragments.

The urethra should be inspected for the presence of blood, and if present, a urethrogram should be performed. An AP x-ray of the lower abdomen and pelvis is routine in the evaluation of all children with significant trauma. This should be performed without a gonad shield so as not to miss sacroiliac injuries. Further delineation of complicated pelvic fractures requires a CT scan.

Most children with pelvic fractures develop a pelvic hematoma. Most of these have self-limited bleeding. Children with unstable pelvic fractures and serious bleeding should undergo immediate placement of external fixation to "close" the pelvis and tamponade the bleeding. Continued bleeding should be treated with angiographic embolization of coils or gelfoam. A stable pelvic hematoma encountered during a laparotomy for blunt trauma should be left alone.

Vascular injuries

Vascular injuries to children were once predominantly the result of diagnostic procedures. Technical advances with these procedures have led to a decline in these injuries. Coincident with this decline has been an increase in the occurrence of these injuries as a result of blunt and penetrating trauma. Vascular injuries to children result from diverse mechanisms. Penetrating injuries result from bullets, knives, and broken glass. Injuries to the effected vessels include transactions, intimal flaps and

pseudoaneurysms. Blunt trauma may cause joint dislocation, fractures and soft tissue injury. Specific injuries to effected blood vessels include thrombosis, contusion or laceration.

Appropriate management of vascular injuries in children require an appreciate of the unique physiologic characteristics of children. The smaller size of children's blood vessels predispose them to vasospasm. Thus, vessels in spasm may cause ischemia in the absence of a surgically repairable injury. The small size of children's blood vessels also makes their repair technically more difficult. The implications of vascular injury on extremity growth in children must also be considered. Chronic ischemia of both the lower and upper extremity is known to impair longitudinal bone growth. The functional significance of this observation seems more important for lower extremity growth. lx,lxi

Children with vascular injuries are managed within the context of standard resuscitation and assessment protocols. Major vascular injuries to the chest or abdomen should be immediately suspected in the unstable patient. Many vascular injuries of the extremity will be identified during the secondary survey. Significant extremity injuries are best managed by multidisciplinary teams owing to the frequent association of fractures, nerve injury and soft tissue loss. Early coordination of care results in the best functional result for the patient. Blunt and penetrating injuries will be discussed by anatomic areas.

Cervical vascular injuries are very rare in children and usually result from penetrating injuries. Principles of management are identical to those for adults. Children with injuries between the sternal notch and jaw and active bleeding or obvious injury to the trachea or esophagus undergo surgical exploration. A number of options exist for repair of the carotid artery including lateral arteriorrhaphy, saphenous vein interposition, resection and anastomosis and transposition of the external carotid to the internal carotid artery. Stable children with penetration of the platysma or equivocal signs of injury should undergo a four vessel angiogram, esophagoscopy, bronchoscopy and an esophagogram. Injuries to the major arteries of the thorax in children are very unusual, usually fatal and most often result from blunt trauma. Left sided hemothoraces, apical capping and sternal fractures should raise suspicion for these injuries. Upper rib fractures do not seem to correlate with aortic injury in children as they do in adults. Repair of these injuries is facilitated by cardiopulmonary bypass and often requires interposition grafting. Most intrathoracic vascular injuries occur in the setting of severe multisystem trauma and contribute to the poor outcome in these patients.

The most common site of vascular injury in children is the extremities. The initial assessment of the injured extremity includes examination for deformities, palpation of pulses and assessment of neurologic function. The

neurologic exam is often compromised by the age of the child or by diminished consciousness. Active bleeding should be stopped with direct pressure. The use of tourniquets or "blind" clamping is to be discouraged. Associated fractures should be splinted and soft tissue injuries covered with sterile dressings. Major vascular injuries that are associated with fractures and dislocations are uncommon in children. However, failure to detect these injuries early may result in motor and sensory deficits, growth failure and amputation. As such, certain orthopedic injuries need to be recognized as high risk situations for vascular injury. These injuries are distal femur fractures, supracondylar humerus fractures and knee dislocations. Vascular injuries result from traction on the vessels and laceration from fracture fragments. Injury to adjacent soft tissues may damage the collateral circulation and worsen ischemia. Patients with diminished pulses and femoral fractures should undergo arteriography. Patients with absent pulses after reduction of a supracondylar fracture should also undergo arteriography or immediate exploration. Arteriography should be performed in all cases of posterior knee dislocations. After identification and location of the arterial injury, the patient should be taken to the operating room. Fasciotomy should be considered early if ischemia has been prolonged. All members of the operative team should be present during the entire operation. An unaffected lower extremity should be prepped in the event that saphenous vein harvesting is necessary. Fracture reduction should be performed after isolation and identification of the affected vessels. Isolated intimal flaps or contusions may be resected and an end-to-end anastomosis performed. More extensive injuries may require a reversed saphenous vein graft. Fractures associated with significant soft tissue loss may need to be treated with external fixation. After satisfactory fracture reduction is achieved, the vascular repair is performed. Penetrating extremity injury in children results from glass fragments, knives, and bullets. Physical findings mandating operative exploration include pulse deficits, poor perfusion, active bleeding and nerve injury. Arteriography does not often add significant information in these instances. The use of arteriography for "proximity" injuries is controversial. The conservative approach is to perform angiography for all of these types of injuries. This seems most appropriate for the management of these injuries in children, which occur infrequently and thus, limit the clinician's exposure, ability and experience with this problem. Penetrating extremity injuries are managed in the standard fashion. A leg is prepped to make the saphenous vein available. Proximal and distal control are obtained. Major venous injuries are repaired when possible. Nerve injury should be sought and identified. The artery is either primarily repaired or debrided and an end-to-end anastomosis performed. If the anastomosis is under tension, an interposition vein graft should be placed. Distal single vessel injuries such as those to the radial or ulnar arteries, or branches of the tibio-peroneal trunk are not necessarily

repaired. Fasciotomy should be again considered at the end of the operation if significant time to revascularization has occurred.

Abdominal vascular injuries in children occur infrequently. They result in significant morbidity and mortality because they are usually unsuspected and result from high energy injuries, which cause associated problems. Blunt trauma is more likely to cause major venous injuries. The majority of these patients will be hemodynamically unstable and have associated injuries. Fayiga et al evaluated 19 children with abdominal venous injury and found 11 (58%) with liver injuries, 6 (32%) with splenic injuries and 2 (11%) with kidney injuries. Injuries to the retrohepatic veins and adjacent inferior vena cava are particularly difficult to manage. Size limitations in children restrict the application of adult techniques such as atriocaval shunting. Total vascular isolation of the liver with veno-venous bypass should be considered for serious injuries of this type. Visceral arterial and aortic injuries result from both penetrating and blunt trauma. Injuries to the superior mesenteric artery can result in rapid exsanguination. This injury should be repaired by either arteriography or interposition grafting. Mesenteric venous injuries should be repaired prior to arterial repair to prevent venous congestion. Second look laparotomy should be considered in all instances of SMA or SMV injury. Injuries to the inferior mesenteric artery can result in large retroperitoneal hematomas and can be confused with infrarenal aortic injury. Proximal and distal control of the aorta should be obtained. If repair is not possible, ligation of the IMA is well tolerated. Renal artery injury usually results from deceleration injury. Attempts should be made to re-vascularize the kidney within 4 hours.

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Appendix A: VITAL SIGNS BY AGE

Age	Wt (kg)	HR Avg/min	RR Avg/Min	BP (sys) (mm Hg)	ET tube ID Mm	Laryn - Goscope Blade	Chest tube (Fr)	NG/ Foley (Fr)
NB	1	145	<40	42±10	2.5	0	8-10	5 feeding
NB	2-3	125		60±10	3.0	1st	10-12	5-8 feeding
1m	4	120	24-35	80±16	3.5			
6m	7	130		89±29	3.5			
1y	10	130	20-30	96±30	4.0	1	16-29	8
2-3y	12-14	120		99±25	4.5			
4-5y	16-18	100		99±20	5.0-6.0	2	20-28	10
6-8y	20-26	100	12-25	105±13	6.0-6.5			
10-12y	32-42	75		112±19	7.0	2-3	28-32	10-12
>14y	>50	70	12-18	120±20	7.5-8.5	3	32-42	>12

APPENDIX B: PEDIATRIC HEMATOLOGY VALUES

CBC:

	Birth	1m	2m	6m	7m	2y	6y	10y	12y
WBC	5-20	5-19.5	6-17.5			5-14.5		4.5-13.5	
RBC	3.6-4.9	2.7-3.8	3.1-3.8	3.7-4.5	3.9-4.6		4.0-4.6		4.1-4.6
Hgb	13.5-16.5	10-14		9.5-11.5	10.5-12	11.5-12.5	11.5-13.5		12-14
Hct	39-51	31-43	28-35		33-36	34-37	35-40		36-41
MCV	86-105	85-104	77-106	74-91	70-78	75-81	77-86		78-90
MCH	28-34		26-30		23-27	24-27	25-29		25-30
MCHC	28-33	29-33		30-33		31-34			
RDW	11.5-14.5								
Plt	150-400								
MPV	7.4-10.4								

DIFFERENTIAL:

	Birth	2w	6m	1y	4y	6y	10y	16y
Meta	0-4							
Band	0-4							
Seg	53-70	20-48	17-54	25-50	27-55	30-55	40-59	40-62
Eos	0-4							
Baso	0-1							
Lymph	26-40	40-85	67-77	60-67	37-52	30-48	34-48	27-40
Atyp	0-0							
Mono	4-12	5-20	4-10	3-10	3-9	3-8		3-9

Appendix C: Latex Allergy Protocol and Precautions

Supportive Data:

As the use of latex has increased over the years as a protection against bloodborne pathogens, latex allergies are increasing among those at highest risk including health-care workers, chronically ill patients (especially persons with spina bifida or urogenital anomalies) and latex industry workers. Sensitivities can range from dermatitis, hives and nasal congestion, to asthma, food cross-reactions and anaphylaxis. The following protocol is intended to provide patients with as safe an environment as possible.

I. Assessment

A. Assess:

1. all patients for latex allergy at the point of entry in registering for or admission to any inpatient or outpatient service, utilizing the “Latex Allergy Screening Tool”.
2. room or patient care area for latex equipment or products prior to bringing the patient into area. (E.g. latex gloves, ambu bag etc).
3. for signs and symptoms of latex allergy associated with latex exposure (nasal drip/congestion, sneezing, eye irritation, tearing, itching, skin rash, reddened edematous skin, hives or welts on body, edema of any part of body, chest pains/tightness, difficulty breathing, wheezing (bronchospasm), anaphylaxis-hypotension.

II. Care and Management

A. Place “Latex Precaution” alert signage as follows:

- a. latex alert wrist band
- b. on door and above bed of inpatient.
- c. on outside of chart.
- d. on medication administration record

B. Notify other departments when sending patients with latex precautions for services.

C. Use only latex-free products and equipment. If latex gloves are required for barrier precautions in certain circumstances, low protein- powder free latex gloves shall be utilized. Cloth may be placed between the patient’s skin and latex-containing equipment if latex free alternative is not available.

-
- D. Wash hands after latex glove removal prior to entering patient room/area. Avoid getting glove powder on uniform when caring for other patients.
 - E. Do not allow latex balloons in hospital. Mylar balloons are allowed.
 - F. All injectable medication in vial form will be drawn up into latex free syringes in the same aseptic manner used for non-latex allergic patients. Use stopcocks for the administration of IV medications (bolus and secondary). **DO NOT USE LATEX PORTS.** Cover latex ports with latex free tape.
 - G. Use latex free oxygen masks, CPR face masks and ambu bags. Replace elastic bands on facemasks with cloth.
 - H. Consult with physician regarding:
 - 1. the need for pre-medication prior to surgery to reduce the incidence of latex allergy reactions.
Keypoint: Pre-medication is not a substitute for latex avoidance. Anaphylaxis has been reported in spite of pre-medication.
Keypoint: See attachment for suggested protocols. Pre-medication requires an MD order.
 - 2. need for patient to be the first scheduled for daily activity in a department (e.g. x-ray, surgery).
Keypoint: Aerosolized glove powder containing latex can be exposed to mucous membranes triggering anaphylaxis.
 - I. For severe anaphylaxis due to latex allergy, implement usual emergency procedures with additional measures:
 - a. immediately remove irritating agent
 - b. use non-latex equipment including gloves, oxygen equipment and ambubag.

II. Complications/Reportable Symptoms

- A. Notify MD:
 - 1. if latex allergy screening tool reveals signs and symptoms or risks of latex allergy in previously undiagnosed patient.
 - 2. signs and symptoms of latex sensitivity reactions. (See Assessment 1.c)

III. Patient/Family Education

- A. Teach patient/so:
1. significance of latex (rubber) allergies
 2. signs and symptoms of latex allergy
 3. importance of reporting to physician for assessment and further recommendations.
 4. importance of recognition and removal of latex products in the environment.
 5. importance of carrying latex allergy identification alert and the necessity to
 6. notify all healthcare, school and daycare professionals about allergy.
 7. how to use emergency epinephrine (EPI-Pen) if ordered by MD.

IV. Documentation

Keypoint: Documentation of exact events in patient progress notes is essential for confirmation of latex as the causative agent when patient signs and symptoms are present.

- A. Document:
1. signs and symptoms of latex allergy in progress notes
 2. notification of physician
 3. initiation of latex allergy precautions
 4. medical and nursing interventions
 5. patient responses and tolerance to interventions.
 6. patient/family teaching

Pre-Medication List for Latex Allergic Patients

Note: Pre-medication prior to surgery to reduce the incidence of reaction is **NOT** a substitute for latex avoidance. Anaphylaxis has been reported in spite of pre-medication.

Children:

1. **Prednisone (PO) or its equivalent:** 1 mg/kg/dose (50 mg maximum) at 13 hours, 7 hours and within one-hour pre-procedure/pre-operatively. Then every 6 hours post-operatively for 24 hours.

OR

- Methylprednisolone (Medrol) IV:** 0.5 mg to 1 mg/kg/dose (125 mg maximum) at 13 hours, 7 hours and within one-hour pre-procedure/pre-operatively. Then every 6 hours post-operatively for 24 hours.
2. **Diphenhydramine (Benadryl) IV/IM:** 1 mg/kg/dose (50 mg maximum) within 45 minutes of procedure/surgery, then every 6 hours for 24 hours.

-
3. **Ranitidine (Zantac) IV:** 0.5 mg/kg/dose (50 mg maximum) within 30 minutes of procedure/surgery, then every 8 hours for 24 hours.

Adults:

1. **Prednisone (PO) or its equivalent:** 20 mg at 13 hours, 7 hours and within one-hour pre-procedure/pre-operatively. Then every 6 hours post-operatively for 24 hours.
OR
Methylprednisolone (Medrol) IV: 40 mg at 13 hours, 7 hours and within one-hour pre-procedure/pre-operatively. Then every 6 hours post-operatively for 24 hours.
2. **Diphenhydramine (Benadryl) IV/IM:** 50 mg within 45 minutes of procedure/surgery, then every 6 hours for 24 hours.
3. **Ranitidine (Zantac) IV:** 50 mg within 30 minutes of procedure/surgery, then every 8 hours for 24 hours.
OR
Famotidine (Pepcid) IV: 20 mg every 12 hours for 2 doses.

APPENDIX D: PICC AND MIDLINE OCCLUSIONS

Types of Occlusions

Mechanical Obstruction

External causes such as:

- Empty IV bag
- Infusion pump turned off
- Kinked tubing
- Closed clamp
- Occluded injection port
- Occluded IV filter
- Sutures are too tight around catheter

Other causes:

- Patient position kinks off the catheter internally
- Catheter migration

Thrombotic Obstruction

Intraluminal Thrombus

- Internal lumen of catheter is obstructed by clotted blood or the accumulation of fibrin
- Results from blood remaining in the catheter following flushing or from coughing or valsalva increased venous pressure causing retrograde blood flow into catheter
- Inadequate flushing after blood draws allows layers of fibrin to accumulate and will over time narrow or obstruct the lumen

Fibrin Tail

- The accumulation of fibrin can result in a tail extending off the tip of the catheter
- Usually does not interfere with infusion but could interfere with aspiration
- Known as withdrawal occlusion, when the tail occludes the tip upon aspiration

Venous Thrombosis

- Fibrin accumulates when damage occurs to the endothelial layer of the vein wall
- It is a venous thrombosis if it completely occludes the vessel
- May involve the catheter causing obstruction

Non-Thrombic Occlusions

- Lipid deposits and drug precipitates can also cause obstruction in the lumen of catheters

-
- Factors affecting the formation of precipitates include the pH of drugs and solutions, inappropriate diluent, drug-drug incompatibility, heparin lock without saline flush and inadequate flushing/heparinization procedures
 - Common drugs causing mineral precipitates are diazepam, calcium gluconate, and phenytoin
 - Lipid deposits (waxy buildup) are more commonly seen with the “3 in one” or “all in one” TPN solutions

Dealing with Obstructions

Mechanical Obstructions

- When an obstruction is suspected, first check all the external factors
- Start at the fluid container (Empty? Air in line?) working down to the site (fluid leaking at site?) checking clamps, looking for kinks, sutures too tight, etc.
- Change patient’s position to see if that corrects the problem
- Chest radiograph to rule out migration may be an option

Thrombotic Occlusions

- Rule out mechanical obstructions and that catheter is in proper position
- Rule out a precipitate obstruction
- Using a 10ml syringe attempt to aspirate the clot gently from within the lumen
- Do not use force, guidewires, or flushing to clear the catheter. This could damage/rupture the catheter or cause a clot to embolize
- To use a thrombolytic agent, such as Urokinase, make sure policies and procedures are followed and there is a physician’s order
- BARD recommends the 3 way stopcock method as the safest way to declot a PICC/midline catheter (see declotting section)
- Urokinase may not be effective against extraluminal clots or fibrin sheath
- Urokinase may not be effective for occlusions older than 7 days
- Urokinase works by activating plasminogen to plasmin, which leads to lysis of clot (Heparin does not lyse clots-it only inhibits coagulation)

Non-Thrombotic Occlusions

- Thrombolytic agents (i.e., Urokinase) are ineffective against drug precipitates or lipid deposits
- Use sodium bicarbonate or hydrochloric acid for drug precipitates. These drugs return the pH to normal and which allows the drug to return to its soluble state.
- Use 70% ethanol (ethyl alcohol) or sodium hydroxide for lipid deposits
- Make sure to follow policy and procedures or have a physician's order

Two Ways to Solubilize Medications/Precipitates

Agent	Hydrochloric Acid (HCl)	Sodium bicarbonate (NaHCO ₃)
Dose	1 mL of 0.1 N HCl (prepared and filtered by a pharmacist)	volume of catheter of 8.4% solution (1 mEq/mL is commercially available)
Dwell time	20 minutes and repeat 1 or 2 more times	precipitate usually clears in 1 to 2 minutes
Mechanism	decrease pH to solubilize acidic drugs or precipitates	increases pH to solubilize basic drugs or precipitates
Cautions	minimal risk of metabolic acidosis febrile reaction when infused rather than aspirated	none

Two Ways to Solubilize Lipit Deposits

Agent	70% Ethanol (ethyl alcohol)	Sodium hydroxide (doda 1ye, NaOH)
Dose	volume to fill catheter (prepared and filtered by a pharmacist)	Infuse 10mL of 0.1 NHCI at 1 mL one hour, followed by a 2-hour lock, and then infuse 0.9% NaCl at 1 mL per hour. Quick flush with 20 mL 0.9% NaCl. Repeat from start one time if necessary
Dwell time	1 to 2 minutes	above
Mechanism	solvent for lipid	solvent for both protein and lipid
Cautions	unpleasant taste when ethanol is flushed through the catheter	

*Macklin, Denise. How to manage PICCs. AJN 97(9): 26-32, September 1997

Drugs Incompatible with Heparin

Alteplase (Activase)
Amikacin (Amikin)
Amiodarone (Cordarone)
Ciprofloxacin (Cipro)
Dacarbazine (DTIC-Dome)
Diazepam (Valium)
Doxorubicin (Adriamycin)
Droperidol (Inapsine)
Erythromycin
Gentamicin
Haloperidol (Haldol)
Idarubicin (Idamycin)
Kanamycin
Methotrimeprazine (Levoprome)
Netilmicin
Phenytoin (Dilantin)
Tobramycin (Nebcin)
Triflupromazine (Vesprin)
Vancomycin

3 Way Stopcock Method for Declotting a PICC or MidLine

The following procedure describes step by step how to utilize the 3 way Stopcock Method to declot a PICC, Midclavicular, or Midline.

BARD ACCESS SYSTEMS recommends this method for declotting for the following reasons:

It eliminates the problem of catheter rupture

It allows declotting without using excessive force to push or pull by removing the excess pressure from the catheter

By applying negative pressure, a vacuum is created within the catheter

By use of stopcock, the declotting agent is drawn into the catheter by the vacuum to the location of the clot

With this method, only the amount of declotting agent that is needed is administered due to the fact that only the dead space created by the aspiration is filled

Luer-lock syringes must be used with this method

Extreme care must be taken to avoid excess negative pressure in order to prevent a sudden forceful flow of the thrombolytic agent in the catheter that could dislodge the clot resulting in embolism

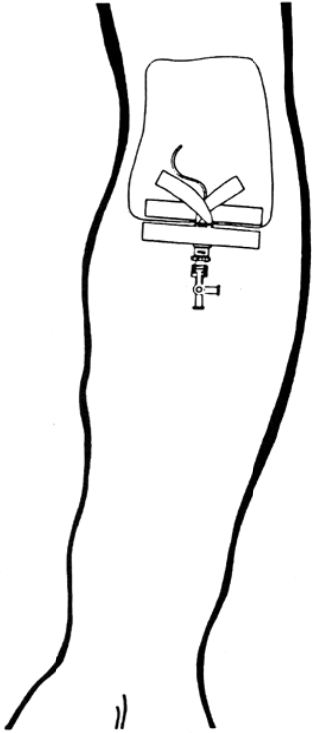
DECLOTTING PROCEDURE **STEP 1**

STERILE SUPPLIES NEEDED:

- 1 10cc Luer Lock Syringe
- 1 5cc Luer Lock Syringe
- 1 Three-way stopcock

Declotting agent
Sterile gloves
Sterile mask

Using sterile technique:
Apply a three-way stopcock directly to the hub with the stopcock off to the patient (turn key should be in the 12 o'clock position as shown).



NOTE: Always attach stopcock directly to catheter hub

DECLOTTING PROCEDURE **STEP 2**

Visualize a clock with the catheter at 12 o'clock



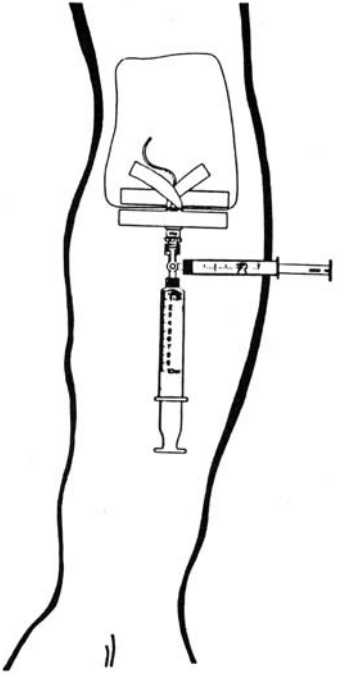
Attach the empty 10cc syringe at 6 o'clock



Attach the syringe with the clotting agent at 3 o'clock



NOTE: Stopcock, syringe, and catheter connections must be airtight.



DECLOTTING PROCEDURE **STEP 3**

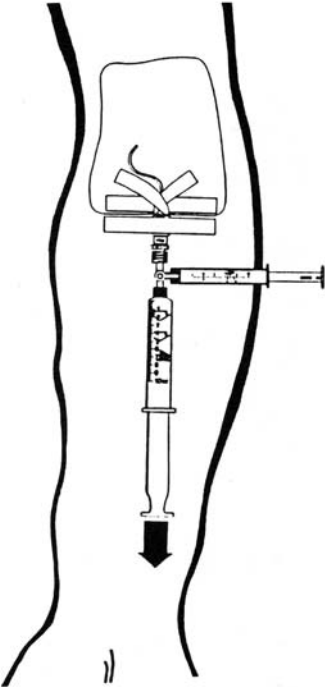
Point the turn key toward the declotting agent (3 o'clock).



Pull the plunger in the 10cc syringe (6 o'clock) back as far as possible causing a negative pressure in the catheter.



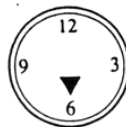
NOTE: Stopcock, syringe and catheter connections must be airtight.



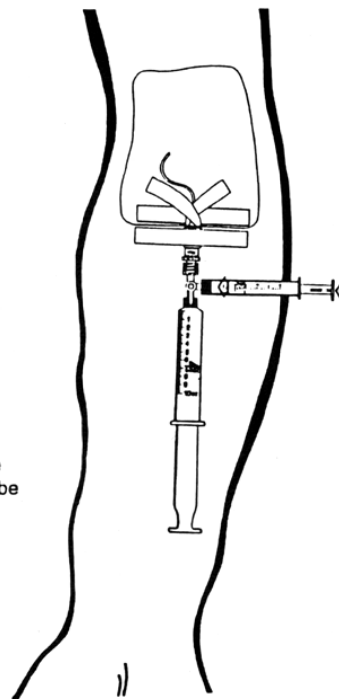
DECLOTTING PROCEDURE

STEP 4

While maintaining the negative pressure on the 5cc syringe, point the turn key toward the 10cc syringe (6 o'clock).



As the negative pressure resolves, the required amount of de clotting agent necessary to reach the clot will be pulled automatically into the catheter. Do not manually push the de clotting agent into the catheter. This could rupture the catheter. Allow the negative pressure to pull the necessary amount into the catheter. This may be a very small amount.

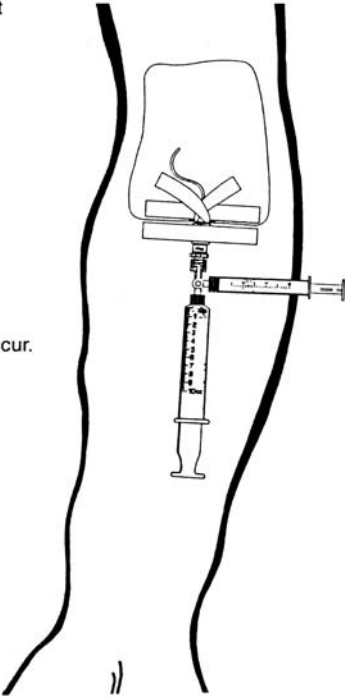


DECLOTTING PROCEDURE	STEP 5
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At this time point the turn key toward the patient
(12 o'clock).



Wait the recommended time for declotting to occur.



DECLOTTING PROCEDURE **STEP 6**

When ready to aspirate point the turn key to the 5cc syringe (3 o'clock).

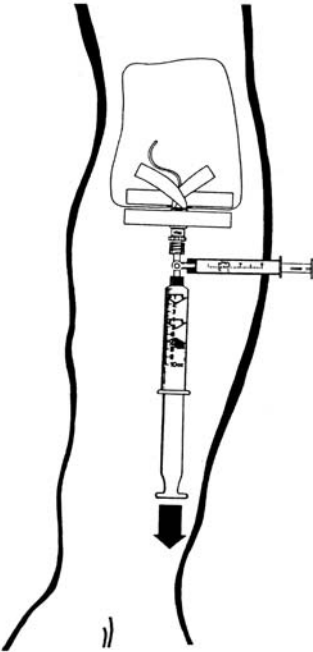


Attempt to aspirate by pulling back on the plunger of the 10cc syringe (6 o'clock).



Withdraw 6cc of blood to remove the declothing agent.

Flush with 20cc saline followed by heparin lock.



NOTE: More than one attempt may be required to achieve patency.

Adapted from "Declothing Peripherally Inserted Central Catheters," Ron Bonstell and Joseph Brown, Gesco International.

**APPENDIX E: DICTATION PROCEDURES:
Shriners Hospital for Children**

USING ANY TOUCH-TONE PHONE, FOLLOW THESE STEPS:

(for STATs, notify medical transcription at ext. 3680, 5294, 5295, 5293)

If you have questions or need assistance, call the Medical Transcription Office @ extension 3680. The office is open Monday -Thursday from 6am – 4 pm, and on Friday from 6am to 3 pm.

1. Dial **6000** (or **503- 412-6891** if outside Shriner's)
2. After recorded message, enter your surgeon User ID# :
Zallen: 300259; Silen 300092; Harrison 300025; Bliss 300187
3. Enter the **two digit work type number:**
 - 06 – Letters
 - 70 – Inpatient H&P
 - 71 – Operative Report
 - 72 – Discharge Summary
 - 73 – Consultation
 - 74 – Inpatient Progress Notes
 - 76 – Outpatient Progress Notes
 - 78 – Peri-Operative Note (PARQ)
 - 79 – Short-Stay Discharge Summary
 - 80 – Short Stay H&P
 - 82 – Initial H&P (New Patient)
4. Enter the 5-digit medical record number
5. Enter 5 to end one report and begin another without hanging up.
6. Enter 9 on the keypad to end dictation.

DICTATION PROCEDURES: Shriners

<u>FUNCTION CONTROLS</u>	<u>KEYPAD</u>
Listen	1
Record	2
Short Rewind	3
Pause	4
End Report	5
Fast Forward to end	6
Fast Forward (short)	7
Rewind (to beginning)	8
Disconnect	9
Clear IDs	*

OPERATIVE NOTE FORMAT

Pt. Name and MRN
Date of Operation
Name of Dictator
Pre-op Dx.
Post op Dx.
Procedure(s) Performed
Surgeon(s)
Assistant(s)
Anesthesia
Specimen(s) Removed
Implants
EBL
IV Fluids
Tourniquet Time
Drains
Complications
Indications for Procedure
Findings
Description of Procedure

H&P FORMAT

Pt. Name and MRN
Admission Date
Name of Dictator & Attending
History:
Chief Complaint
History of Present Illness
Past Med/Surg History
Family Hx
Social Hx
Developmental Hx
Allergies and Medications
Immunization Status
Review of Systems
Pain Assessment
Physical Exam:
General, HEENT, Neck ,
Chest, Heart, Abdomen,
Neurological and Orthopedic
Xrays
Impression & Diagnosis
Plan

APPENDIX F: PEDIATRIC PAIN MANAGEMENT GUIDELINES:

Typical Initial Pediatric Pain Med Dosages By Age

*** = patient requires continuous SaO2 monitoring**

Neonate (0-30days)

Outpatient

- Acetaminophen 10-15mg/kg PO Q6H or 30-40mg/kg PR x1 then 20mg/kg PR Q6H

Inpatient (in addition to above)

- Morphine 0.05-0.1mg/kg IV Q4H*
- Morphine IV cont infusion 0.01-0.02mg/kg/hr*
(NICU/PICU only)

Infant (1month-1year)

Outpatient

- Acetaminophen 10-15mg/kg PO Q4H or 30-40mg/kg PR x1 then 20mg/kg PR Q6H
Concentration: 10mg/0.1mL infant Tylenol
- Ibuprofen (must be >6m of age) 4-10mg/kg PO Q6H
Concentration: 40mg/mL infant ibuprofen
- Can consider low dose Oxycodone if pain not managed with Tylenol or Ibuprofen:
Oxycodone 0.05mg/kg PO Q4-6H
Concentration: 5mg/5mL

Inpatient (in addition to above)

- Morphine 0.05-0.1mg/kg IV Q2-4H*
- Morphine IV cont infusion 0.01-0.04 mg/kg/hr* (NICU/PICU only)
- Fentanyl 1-4mcg/kg IV Q2-4H* (NICU/PICU only)
- Fentanyl 0.5-1mcg/kg/hr IV & titrate up PRN* (NICU/PICU only)

Child

Outpatient

- Acetaminophen 10-15 mg/kg PO Q4H; 10-20mg/kg PR Q4H
Concentration: 32mg/mL children's tylenol
- Ibuprofen 4-10mg/kg PO Q6H
Concentration: 20mg/mL children's ibuprofen
- Oxycodone 0.05-0.15mg/kg PO Q4-6H
Concentration: 5mg tab or 5mg/5mL
- Vicodin/Lortab 0.1-0.2mg/kg (hydrocodone component) Q3-4H (up to 10mg)
Concentration: 7.5mg hydrocodone;500mg acetaminophen/15mL

Inpatient (in addition to above)

- Ketorolac 0.5mg/kg IV Q6H (max 5d use)
- Oxycodone 0.05-0.15mg/kg PO Q4-6H
 - *Concentration: 5mg tab or 5mg/5mL*
- Morphine 0.05-0.1mg/kg IV Q2-4H*
- Morphine IV cont infusion 0.01-0.02 mg/kg/hr* (NICU/PICU only)
- Morphine PCA- see PCA order sheet for dosing*
- Dilaudid 0.03-0.08mg/kg PO Q3-4H
 - *Concentration: 1mg/mL*
- Fentanyl 1-2mcg/kg IV q30-60min* (NICU/PICU only)
- Dilaudid 0.015mg/kg IV Q3-6H*
- Dilaudid PCA- see PCA order sheet for dosing*

Remember to consider a bowel regimen for children on narcotics.

Consider Pain Service consultation if infant/child is not palliated with these medications.

OREGON AREA HOMECARE COMPANIES

<u>Company</u>	<u>Phone</u>	<u>Fax</u>
Apria	503-258-2200	503-253-4954
Byram Medical	503-233-2201	503-233-1984 866-447-4426 (enterals)
Coram	503-684-3046	503-684-6627
Good Samaritan	541-768-5360	541-768-5383
Homecare IV of Bend	541-382-0287	541-385-6260
Home Parenteral Care (Eugene)	541-683-3700	541-683-3415
Kaiser infusion	503-499-5219	
Optioncare	866-347-8660	866-347-8662
Optioncare (WA)	360-690-4125	1-877-885-3981
Pacific Home Care (Eugene)	541-751-9868	541-683-3415
Parkway (Roseburg)	541-677-2438	541-677-6587
Providence Home Svs	503-215-4646	503-215-4985
Sacred Heart in Eugene	541-461-7650	541-461-7686
Samaritan Health Svcs. (Corvallis)	541-757-5254	
Shapes (Salem)	503-370-5719	503-561-2522
Shapes	503-561-2536	503-561-2537 (enterals)

NOTES:

Please submit any corrections/suggested additions to this manual to the
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