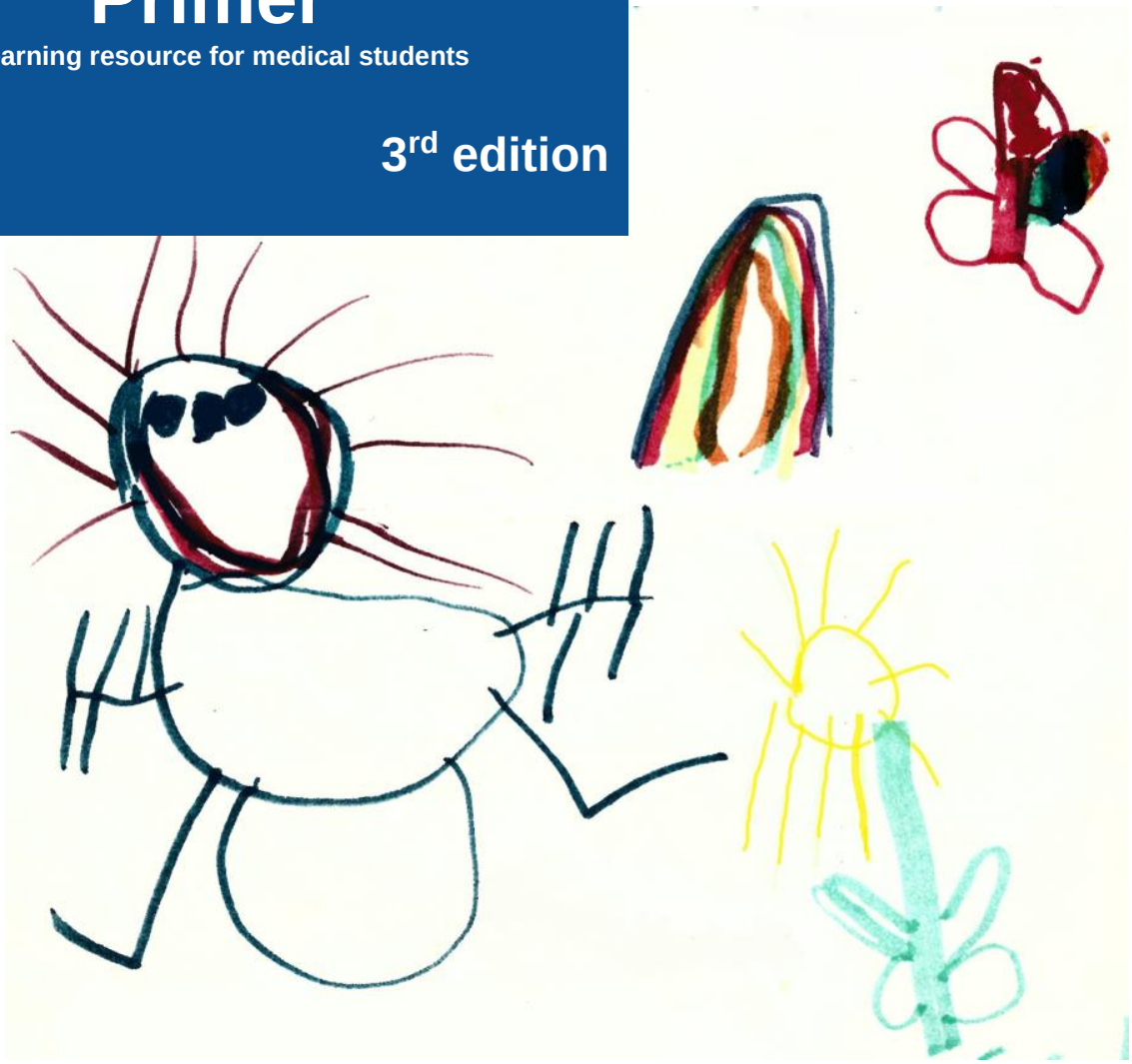
# A Pediatric Surgery Primer

A learning resource for medical students

3rd edition



Geoffrey K. Blair, MD, FRCSC

# A PEDIATRIC SURGERY PRIMER

## A LEARNING RESOURCE FOR MEDICAL STUDENTS

## 3RD EDITION

Geoffrey K. Blair, MD, FRCSC

Clinical Professor of Surgery

Department of Surgery

University of British Columbia

Dedicated to children everywhere.

Think about this:
If we all strived to do only that which
helps and does not harm children,
then all would be right in our world.

# Preface to the 3rd edition

Pediatric Surgery is a broad and interesting field. Within that "species" we know of as babies and children (ages 0-16 and beyond) Pediatric Surgery ranges through the diagnosis and management of congenital anomalies, infective disorders, cancer, thoracic disease, trauma, head and neck disorders, etc. Of all the surgical sub-specialties, it is in many ways the most "general". In some ways, it is a hybrid specialty combining aspects of Pediatrics with Surgery. We deal with young patients who, by virtue of their age, often cannot give testimony. Worried parents are also part of the package. Professionalism and a caring, compassionate attitude are vital. Our rewards as Pediatric Surgeons are to see our patients have a better start to a long life before them. Children, by and large, are pure and simple in their motives, unsullied by motives of secondary gain, or getting time off work or compensation for their disease. They just want to get back to being kids. They are innocent victims of their troubles and we have many opportunities—indeed a responsibility—to serve as their rescuers wherever possible. It is a heady duty, but, for the most part, it is truly a joy and tremendously interesting.

The field of Surgery is progressing rapidly and as I look back on my decades as a pediatric surgeon I am amazed at the changes which I have seen. Some of what was considered 'best practice' or 'contraindicated' or even 'incurable' when I was in medical school are now the opposite. Keep an open mind, and continuously look at and carefully consider the evidence which supports or refutes the status quo. This message applies to any field on Medicine, not just Surgery. Medical education has also changed a great deal in the past few decades. Learning how to THINK about clinical problems is the key. Of course, in order to properly think your way through any problem you must have a certain fund of knowledge, together with a cognitive scheme or framework to put all the facts together.

However, I maintain that there are two vitally important elements which must bookend the thinking process in order to be a good doctor. Firstly, you must CARE, truly care about your patient. If you don't care about your patient and what may happen to them then your thinking will be incomplete, haphazard or even misdirected. While caring for your patient and thinking your way to a best solution you must then ACT. You could be the smartest diagnostician in the world, but your patient, or in the case of young children, their parents, will ask, "What can you now do to help, doctor?" Action on your part may be advising an operation, prescribing a medication, doing some further investigations, willfully taking a 'wait and see' approach and following up, referring the patient to another health care provider, or a number of different action options. And sometimes the appropriate action is simply to admit that no solution is available but that you continue to care and will be there with them in the midst of the uncertainty.

Surgeons are generally good at dealing with uncertainty. There are many times in a surgeon's career when uncertainty abounds but action is needed. It may be an 'acute abdomen' where the underlying diagnosis is uncertain but because of the state of the patient an operation must be done. Most medical students have done Sciences before they begin their medical school

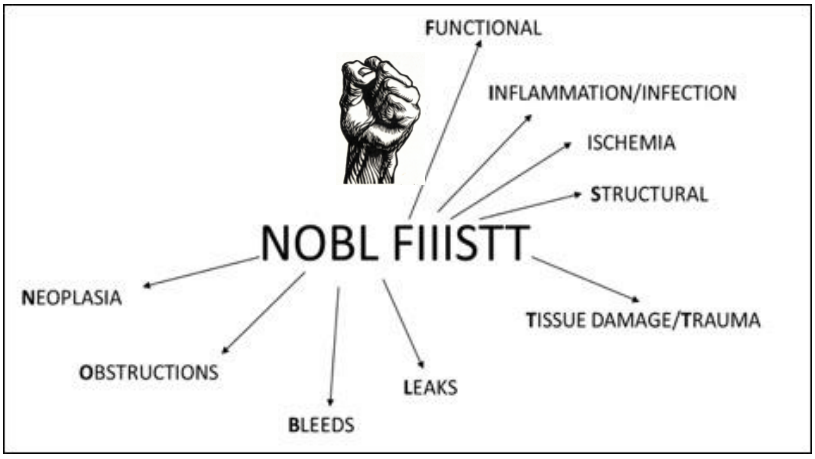
training. As such, they have been learning in what I call the 'Temples of Accuracy'. It may have been the Organic Chemistry lab where the exact weight of a product of a chemical reaction is weighed out on one of those scales so accurate and sensitive that they need tiny doors closed on the weighing chamber in a case an errant puff of air passing by throws off the reading. You may recall the Physics lab where a spectroscope reading could tell you precisely what elements were glowing in the flame or the Mathematics class where the correct answer was 56.2457, and not 56.2689.

Students coming into medical school from these 'Temples of Accuracy' may believe that the practice of Medicine has the same exactitude of the science lab. Instead of accurate scales or spectroscopes providing precise readings, modern medical instruments like CT and MR scanners and blood tests will unfailingly provide similar accurate answers to what ails the patient. A good doctor these days need never face significant uncertainty before taking action.

Think again… Unfortunately, it just isn't so. Get comfortable with some uncertainty.

My instruction for you is simply to read this Pediatric Surgery Primer. It is not a textbook in the usual sense—there are many fine textbooks out there. Rather, it is a means whereby together we'll think our way through some important Pediatric Surgery problems. In medical education lingo we speak of these representative cases as 'worked examples'. Yes, I shall be inevitably tossing some facts at you. After all, you do need facts to build your thinking on; but as its title implies, this book is meant to 'prime' your thinking.

In the same way that you'll need cognitive frameworks to think about clinical problems, I too need a framework to help educate your thinking process. I use NOBL FIIISTT in this Primer. 'NOBL FIIISTT' is simply an acronym which represents a framework to aid surgical teaching. It is one framework, amongst others, which can perhaps help you learn and think about the spectrum of disorders surgeons deal with. Think of each element of NOBL FIIISTT representing a pathologic process and they are not mutually exclusive. Take a look at this diagram:




By the way, there's a story behind the fist in this picture. I'll tell you about it at the end of the book if you're interested.

It's in the situations of cancerous tumours, bowel obstructions, hemorrhage and the many, many other disease processes in the NOBL FIIISTT spectrum where surgeons can help. Before you jump to pinning diagnostic labels ponder the important issue of, "What is going on inside this patient that is causing their problem?" Simply put, by understanding what's wrong you can proceed to ameliorate the situation.

G.K Blair, 2023

# INTRODUCTION

As a medical student, there are some childhood diseases, conditions, clinical presentations and concepts that you will be expected to understand and logically think through. Some of these are quite different than their adult counterparts and some are similar. Where I speak of children, or childhood, unless otherwise stated I am referring to infants, children and adolescents- ages 0 through 16+. Here's how this book is laid out:

The Canadian Association of Paediatric Surgeons in 2022 ratified this latest version of Learning Objectives for Undergraduate Medical Students:

## CAPS National Learning Objectives for Graduates of Canadian Medical Schools

**The following represents what the Canadian Association of Paediatric Surgeons (CAPS) believes are the necessary and reasonable Learning Objectives for <u>all</u> medical students to achieve by the time of their graduation as pertaining to the surgical problems of infants, children and youth:**

**Pediatric Surgical Clinical Considerations:**

Bear in mind the following when gathering histories, doing physical examinations and undertaking investigations in babies and children with potential surgical problems.

- **History**: Most often the history is obtained from the parent(s) / responsible caregiver but even young children can and should be addressed in history-gathering as well. Also explore the psycho-social context of the child and family as it can be of critical importance in the care of both.

- **Physical examination:** Children often can be fearful of doctors (and the parents may show their fear as well). Respect the child and use a gentle approach, assuaging their fears through distractions, games, and gaining their trust through friendliness. Never be dishonest (e.g. telling the child, "this isn't going to hurt" when you're about to do something that you know will be uncomfortable for the youngster). Take it slow and involve a parent in the examination process. Be appropriately thorough and do not neglect to do what's necessary (e.g. DREs when indicated).

- **Investigations:** Ask only for tests which will be truly helpful and avoid testing where results will give little guidance (e.g. CRP in abdominal pain). Understand the operational characteristics of any test you plan to do (e.g. potential harm from false positive/negative result). Consider the potential harmful effect of the test (e.g. CT scanning and radiation exposure to a child increasing future risks of malignancy). Ensure that requested tests will be done properly (e.g. AXR alone is only a supine view and of little use if considering a bowel obstruction; a supine and upright film will give far more information).

- **Physiology & Anatomy:** Babies and children, because of their particular stage of growth and development often manifest and react to disease in ways very different than the adult. Remember, "Children are not just small adults."

**Specific Learning Objectives:**

Given a patient with a typical history, signs and investigation results who has any disease/injury on the following list, the student will be able to:

a) Identify the disease/injury as the most likely diagnosis;
b) Appropriately include other less likely, but still possible diagnoses in the differential diagnosis and justify their inclusion using knowledge about their presenting features (not expected to be comprehensive)
c) Explain to the patient/parent the cause (if known), generally how common the disorder is and the typical natural history;
d) Denote potential life/limb/organ-threatening pathology and possible need for timely consultation/referral
e) Discuss with the patient/parent the options and recommendations for management;
f) Provide the patient/parent with a general prognosis;
g) Outline a general management plan (need not include the specifics of surgical or intensive care management)

1. **Inguino-scrotal/Abdominal wall pathology:**
   - Testicular torsion
   - Inguinal hernia (asymptomatic or symptomatic, including incarcerated)
   - Hydroceles

- Cryptorchidism
- Umbilical hernia

2. **The 'Acute Abdomen'—specifically the following:**
    - Intestinal volvulus/malrotation
    - Appendicitis (both early and ruptured)
    - Intussusception
    - Meckel's Diverticulum
    - Bowel obstructions (general diagnosis and management) *

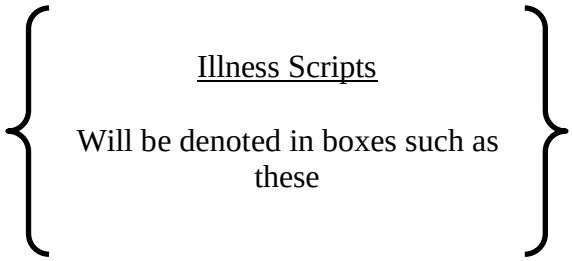
3. **Infantile hypertrophic pyloric stenosis**
4. **Biliary atresia**
5. **Pediatric malignancies:**
    - Neuroblastoma
    - Wilms' tumour
    - Lymphoma
6. **Acute congenital surgical disease:**
    - Esophageal atresia (& potential accompanying VACTERL syndrome elements)
    - Imperforate anus
    - Intestinal atresia
    - Hirschsprung's disease
    - Congenital diaphragmatic hernia
    - Gastroschisis & Omphalocele
7. **Pediatric trauma:**
    - Pediatric Poly-trauma
    - Occult trauma & Non-accidental Injury

* 'Bowel obstructions' include some of the specific items listed, such as intestinal atresia, but because the general approach to bowel obstructions in children, whatever the cause, is so important it is listed under the 'acute abdomen'.

---

In order that your learning remains an active rather than passive process but also realizing your clinical inexperience in Pediatric Surgery we will together 'walk through' some **representative cases** which will help you start to think through some common Pediatric Surgical problems—how they might present clinically, how you can build a differential diagnosis based on that clinical information, and how you can logically and sequentially refine your differential diagnosis by gathering more information through appropriate testing. *The cases are presented in simple point-form and in blue italics.*

Along with the representative cases to think through I'll tell you of some features of other Pediatric Surgical conditions through **"Illness Script"** boxes. Simply put, an illness script is a short snapshot of how a disease typically—but NOT always— presents.




I shall also refer to some aspects of **treatment**, but don't worry; this is not a course on how to become a Pediatric Surgeon. Some basic surgical treatments will be presented. You should know some surgical treatment basics after all.

"**Checkpoints**" are placed at intervals. These are meant to help you package your learning points and will expand on what the representative cases have illustrated.

I'll also throw you some "Checkpoint Challenges" related to those conditions. You'll have to draw on that new knowledge, probably refer to other educational resources and THINK—and I'll give you Answer Hints in the Appendix. Only hints, though. You'll retain more if you have to do some 'digging' for the answers. The Checkpoint Challenges also provide you with some topics you can ask your surgical teachers about, as I have tried to formulate some of them around 'debateable' issues. You'll also see in the Answer Hints that I wax on about some of my opinions and I cannot guarantee that my pediatric surgical colleagues will agree with everything I say. That's ok, as you should know that surgery is replete with its controversial views.

Here and there I've added a few **historical facts and other whimsies** that you may find interesting but you'll not ever be tested on. Sometimes knowing some historical trivia can be a memory aid.

You are encouraged—indeed it will be necessary—to use this Primer in conjunction with other learning resources such as standard surgical textbooks. Some important surgical conditions in children I just mention in passing but you'll learn and remember more if you look them up yourself.

I am hoping that it feels like we are just having a chat about some important points of Pediatric Surgery. This isn't a textbook. For you, a novice in medicine, it is a "Primer" to help you get started, indeed, to "prime" your thinking about childhood surgical disease.

Let's proceed.

LET'S BEGIN WITH THE N OF NOBL FIIISTT:

# NEOPLASIA

## NOBL FIIISTT

**Case NW**
*2-year-old boy, previously healthy*
*CC- Lump in abdomen*
*HPI- Healthy, no pain, no associated symptoms, no urinary symptoms*
*P/E- Large left abdominal mass*

**Consider:**

This is how most extracranial solid malignant tumours commonly present in childhood- as a visible or palpable "lump"- with no pain or other manifestations. Often the parents will report that they first noticed this at bath time, the child being otherwise totally asymptomatic. Take every new lump appearing in childhood seriously.

Brain and central nervous system tumours are the most common solid malignancies of childhood. However, the 3 most prevalent childhood solid malignancies, usually arising from the abdomen or pelvis are:

- **Neuroblastoma**
- **Wilms' tumour (nephroblastoma)**
- **Lymphoma**
- Other possibilities include:
    - Rhabdomyosarcoma
    - Ewing's sarcoma (a type of bone cancer)

- Hepatoblastoma & Hepatocellular carcinoma (liver cancers)
- Other less common cancers

**What are you going to do?**

- A thorough **history and physical exam** is **<u>always</u>** expected.
- Was there exposure to radiation? Is there a family history? Does the child have a known genetic syndrome that predisposes to cancer development- Down, Beckwith-Wiedemann, Li-Fraumeni, Fanconi Anemia, etc.?

*Further history is non-contributory in this child.*
*P/E reveals a large, non-tender, smooth mass occupying the left flank, easily visible.*

You should now have serious suspicions that this mass is a malignancy.

You realize you must act promptly. Yes, it is possible that a left colon full of stool or simply a toddler's normal protuberant abdomen could be all that's going on here, but you felt a definite abnormal mass lesion.

**What investigations do you arrange?**

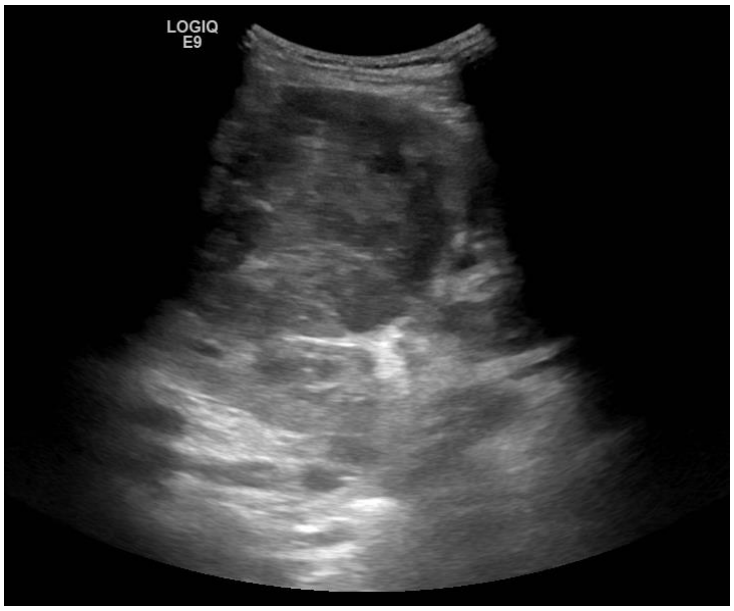
- **Urinalysis**- Simple- you can do this in your office or clinic, results are within minutes and it costs almost nothing. Should be done.
- **Basic blood work**- Think "tumour markers', CBC and LFTs
- **Imaging**- Think carefully. This is required ASAP- because it could be a fast-growing cancer, and because the child's parents are beside themselves with anxiety. (Understandably so!)
  - <u>X-ray</u>: a simple <u>a</u>bdominal x-ray is not going to tell you much. A chest x-ray might reveal lung metastases though.
  - <u>CT scan:</u> more difficult to arrange, involves significant radiation, and may need special techniques and contrast enhancement. CT scans have an important role, but not generally as the first line of imaging in children.
  - <u>MR scans</u>: may be difficult to arrange, involves no radiation, often needs special radiology expertise, and demands serious sedation or general anesthesia in a 2-year-old such as because of the scan time.
  - <u>Ultrasound</u>: Yes, you got it! Ultrasound! No radiation, but still needs special expertise, but easier to arrange, the child needn't be sedated and can give very important information about the characteristics of this boy's mass.

It is **NOT** appropriate to just fill out an imaging requisition and send it in. and it's not enough to simply mark "urgent" on the requisition. To properly arrange the next imaging steps, you should **personally contact a radiologist** and discuss the problem. That discussion is important for a number of reasons:

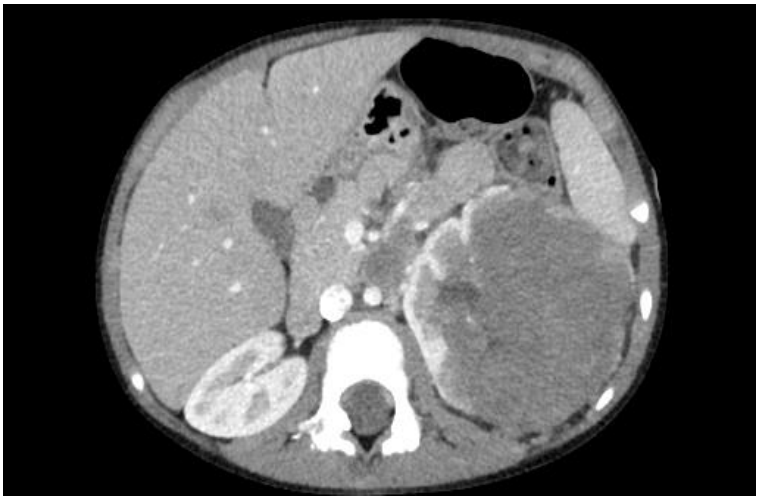
- It draws in the radiologist as your colleague helping you do "the right thing" here. Radiologists are your important partners!
- It ensures that if special imaging techniques are required they are anticipated. The more the radiologist knows about the case the better the imaging will be.
- It expedites the imaging. It is not right that this 2-year-old wait any significant length of time for these studies to be done and reported.

*Results:*

- *Urinalysis- Normal*
- *Blood work- Normal*
- *CXR- Normal*
- *Abdominal ultrasound- Large left flank solid mass, probably arising from the superior aspect of the left kidney. Interrupted left renal vein blood flow and possible tumour thrombus extending into IVC. Consistent with Wilms' tumour, but possibly neuroblastoma. Right kidney and other organs are normal.*
- *A CT scan was also done soon after the US*




The radiation of a CT scan in this case is now entirely justified by the US findings. So now we know it is serious and probably malignant.



**What are you going to do now?**

You figure it is probably a Wilms' tumour, and you should **contact the parents**, discuss the findings and outline to them what you think should be next. They will ask you about Wilms' tumours. You don't know all the details yet and cannot say for certain if this actually is a Wilms' tumour, but they should be told that it is one of the commonest solid tumours of childhood.

Modern treatment protocols are in place and the outlook can be very good. Of course, you cannot guarantee anything- but you are with them on this journey.

> **This needs expertise**. Call a pediatric cancer specialist - an oncologist, a pediatrician, or a pediatric surgeon.

**What's next?**

We are not going to discuss the treatment protocols for childhood malignancies except for these important points:

- Further investigations: specialized imaging will be done- such as that CT scan of the chest- that can give important anatomic information. It helps us **stage** the tumour.

- Staging: We do not expect you to memorize cancer staging for different cancers. You should know the basic paradigm of the **TNM staging system**, which varies depending on the tumour type and denotes:

  **T**- the **tumour** size and possible local invasion
  **N**- the **nodal** status (e.g. local spread vs more extensive spread to lymph nodes)
  **M**- the presence or absence of **metastases**.

  Staging tables then convert the TNM status into specific stages. Staging the tumour materially helps plan the therapy.

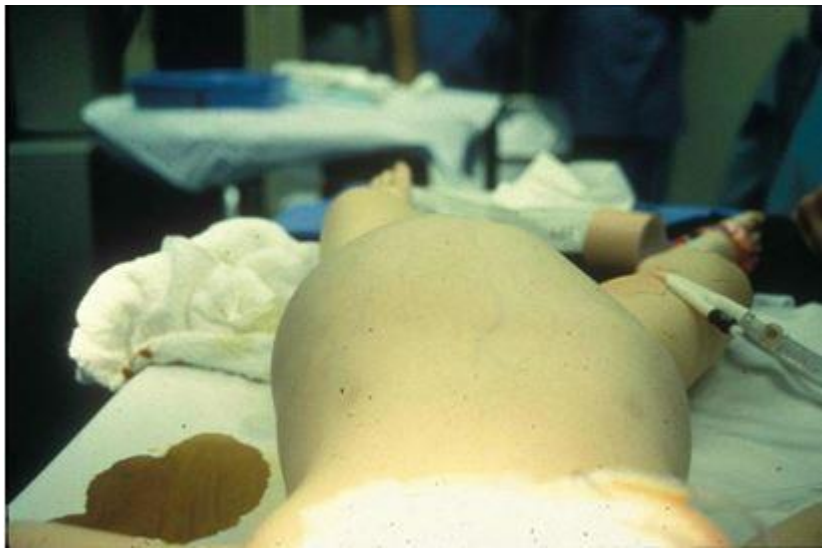
- Therapy choice: modes used in particular sequences are usually dictated by the tumour factors, the Stage and Grade of the tumour—but also by patient factors. In general, therapies in childhood solid tumours may involve:
  - Chemotherapy
  - Surgery
  - Radiation

- Biopsy?
  - It's decided that a tissue diagnosis <u>is</u> required in this case (although not necessarily in all Wilms' tumours. Suffice it to say that 'it depends…"). This means a biopsy. In general, for children's cancer we **do not** do needle biopsies. Needle biopsies provide cells for cytology assessment and are good for carcinomas, but most childhood tumours are <u>not</u> carcinomas. (Sometimes we can rely on 'core needle biopsies' which gives the pathologist core samples of tissue and not just cells.)

*This boy underwent an **incisional biopsy**- namely a piece of the tumour was removed surgically. The other option would be an **excisional biopsy**, wherein the entire tumour (which in this case would necessitate a left nephrectomy as well) is removed. An excisional biopsy is used if the tumour is resectable and not invading nearby structures. Remember this boy's renal vein and IVC had tumour within them (shown on abdominal U/S).*

*Biopsy pathology report: Wilms' tumour, favourable histology. ("Favourable" in this case refers to certain histologic features of differentiation.)*

This report on the histology of this tumour helps us 'grade' the tumour. Although it may seem logical to have the stage and grade of the tumour known before initiating any treatment, other factors come into play. For instance, if the tumour does not appear to have spread locally on imaging, an upfront nephrectomy may be considered appropriate treatment, not knowing the histologic grade until the whole kidney is out. Bottom line—there are different evidence-based protocols for different tumour situations.



You can see the massive size of this tumor as this boy lies on the OR table. It's amazing that with tumours this size, children are most often asymptomatic. Amazing also that tumours in children can grow to such a large size without anyone noticing. We should routinely palpate children's abdomens when they visit the doctor, even if they're there because of an earache! Generally, the earlier they're detected, the better the prognosis!

*Therapy:*

- *Insertion of a long-term central venous catheter for chemotherapy.*
- *Neoadjuvant chemotherapy followed by left nephrectomy and complete tumour excision. (The neoadjuvant chemo caused the renal vein/IVC tumour thrombus to shrink allowing complete tumour excision.)*
- *Post-operative radiation therapy to the tumour bed.*

Wilms' tumour survival overall ~85%- but depends on stage and grade

*Long-term follow-up:*

*Now 5 years post treatment he is followed by ultrasound and CXR and is cancer free. He is a normal 7-year-old in all respects except for the large transverse surgical scar on his abdomen. He will require follow-up for decades, for the long-term effects of his cancer and its treatment. What long-term risk would his radiation therapy incur?*

***A brief history of Wilms' Tumour***

*A German doctor, Max Wilms, described this tumour of childhood in 1899 and its unique "triphasic" histology. The tumour was certainly known before 1899. Indeed, the first nephrectomy for this tumor was performed in 1877. However, since Wilms had described its microscopic makeup and proposed an embryonic origin, the eponym, "Wilms' tumour" was popularized by the year 1900. You'll be smart to remember it's Wilms' and not Wilm's, although these days the apostrophe may be left off, i.e. Wilms tumour. Max Wilms died in 1918 from sepsis incurred while performing a tracheostomy.*

## CHECKPOINT: CHILDHOOD NEOPLASIA

- Children are not just small adults!
- Abdominal masses in kids - think possible cancer (although some solid tumours are benign)
- Often large masses are asymptomatic.
- Promptness!
- Take a history and do a proper physical exam!
- Basic testing (Blood testing for tumour "markers", urinalysis—but normal results cannot be reassurance.)
- Ultrasound is often a good first imaging choice
- Needle biopsies usually are not appropriate in childhood tumours
- Get help- bring in a specialist

CHECKPOINT CHALLENGE #1: A MOTHER BRINGS HER 1-YEAR-OLD GIRL TO YOUR CLINIC AND SAYS THAT HER BABY HAS BEEN STRAINING AT STOOL A LOT RECENTLY. YOU GATHER A COMPLETE HISTORY AND DO A THOROUGH PHYSICAL EXAMINATION—WHICH INCLUDES A DIGITAL RECTAL EXAM. (YES—APPROPRIATE EVEN IN BABIES!) YOU FEEL A HARD, IMMOBILE 'LUMP' ABOUT 3 CM. IN DIAMETER POSTERIOR TO THE ANORECTUM IN THE REGION OF THE TIP OF THE COCCYX. YOUR THOUGHTS? (YOU CAN LOOK AT THE ANSWER HINTS TO THIS AND THE OTHER CHECKPOINT CHALLENGES IN THE APPENDIX.)

**OK- Let's pick up the pace here…**
**This is SURGERY. SURGEONS DO THINGS. Diagnosis is good and necessary—but only because it helps you plan what to DO. It is vitally important to try to understand what's wrong. Think PATHOPHYSIOLOGY.**

# OBSTRUCTIONS

## NOBL FIIISTT

***Case OD***
*Newborn male, 37 weeks gestation*
*5 hours old*
*CC-Bilious, GREEN vomit*
*HPI- polyhydramnios detected in late pregnancy*
*Vaginal delivery*
*Good APGARs*
*Vomited 60 ml. "bile"*

**In general terms what is the suspected disease process here?**

True bilious emesis is GREEN, so if someone reports a child vomiting bile, ask if it was GREEN. Yellow doesn't count!
Now we are talking the forest here- not the trees. So, based on this history thus far, this child has a **bowel obstruction**, until proven otherwise. The exact type of bowel obstruction is not important yet. Pathophysiology first—you'll get to the diagnosis eventually.

In children, **GREEN** vomitus is a bowel obstruction until proven otherwise.

**"Beware the child who vomits GREEN"**

**Based on that, establish a plan for acute, immediate care.**

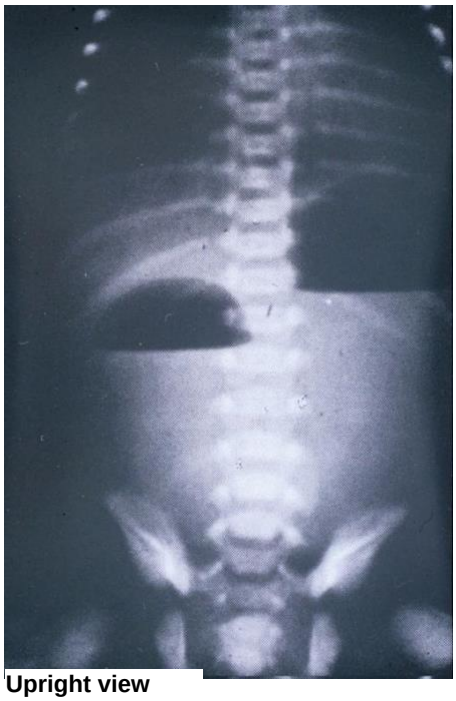
I REALLY want you to remember that so, I often stand on the furniture when I say it!

Before you get fancy, realize that you've got a tiny baby who is vomiting bile, and probably is obstructed. A baby who has a GI obstruction is fluid deficient with both poor intake and lack of absorption but is also '3rd spacing'. The GI tract is dilated proximal to wherever the obstruction is. Vomiting loses both fluid and electrolytes and also places the child at risk of aspiration.

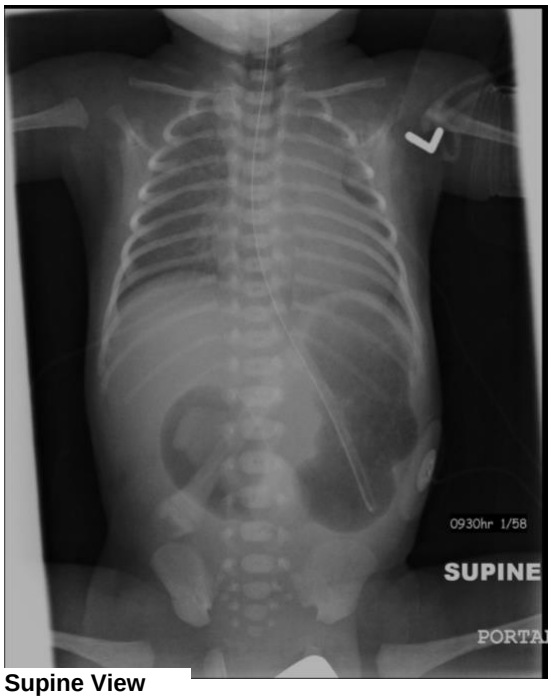
Decompress the GI tract, and support the baby. We talk about "drip & suck"; NG tube and an IV. (We won't dwell on the details of fluid and electrolyte replacement in babies. Remember the 4-2-1 rule for maintenance IV therapy though.)

Your mind is awash with all the various causes of newborn bowel obstructions. While you think about that you might want some simple tests:

- Bloodwork: You may order blood work, but remember this baby just got off the best dialysis machine there is—the placenta—so, chances are the blood work will be normal. Normal bloodwork at this stage is no reassurance.
- Imaging: You could ask for some simple imaging
    - Abdominal ultrasound: **No**! Not the right answer! Ultrasound is confounded by gas and with a GI obstruction there probably will be a lot of bowel gas. Even if there wasn't, unless there is some sort of mass lesion causing the bowel obstruction, an ultrasound won't be able to delineate its cause.
    - X-ray: **Yes!** (There is radiation, but simple radiographs are low dose- unlike a CT.)



**Upright view**




**Supine View**

Essentially, you should never order just a 'flat plate' of one view of the abdomen—and that's often what you'll get if you simply ask for an abdominal x-ray. Stipulate you want two views. You want a supine view, and you need a view where gravity can allow fluid and air to form a visible interface—i.e. air-fluid levels. Usually that gravity view is an upright film, but often in babies we get lateral views or lateral-decubitus views.

*Abdominal x-ray report indicates— **"Double-bubble**" – namely there is evidence that the obstruction is high in the GI tract with no gas distally. This is classic for **Duodenal Atresia.***

(The word "atresia" refers to any luminal structure in the body that doesn't form correctly and is thereby obstructed. One can have bowel atresia, esophageal atresia, biliary atresia, vascular atresia, airway atresia, etc.)

**Duodenal atresia:**

- A congenital condition—**Where there is one congenital anomaly, look for others. e.g. ⅓ of duodenal atresia cases have Trisomy 21**
- Believed to be a failure of 'vacuolization'—the duodenal lumen opens up early in fetal life when the solid core of duodenal tissue forms vacuoles which then coalesce
- Associated with congenital heart disease
- Polyhydramnios is often seen prenatally
- Classic 'double-bubble' on AXR
- Primary repair—duodeno-duodenostomy- We find the obstruction at laparotomy and do a short bypass of it, sewing proximal duodenum to the distal duodenum

How would esophageal atresia present in a newborn? (We'll consider this later…)

*No further images required. The baby was taken to the operating room on Day 2 of life and underwent a laparotomy and a duodeno-duodenostomy. He did well. Chromosomal testing for Trisomy 21 (Down syndrome) was negative. Echocardiography was also normal.*

**Remember VACTERL:** an acronym for these malformations that often occur together:

**V**ertebral defects
**A**nal atresia
**C**ardiac defects
**T**racheo-**E**sophageal fistula/atresia
**R**enal anomalies
**L**imb abnormalities

If a baby has three or more of these features, we say they have VACTERL syndrome

**Newborn Bowel Obstructions- Differential Diagnoses:**

- Duodenal atresia
- Small bowel atresia
- Colonic atresia
- Imperforate anus—surprisingly the **most common** cause of newborn bowel obstructions!
- Hirschsprung's disease- aganglionosis of distal bowel
- Meconium ileus- GI obstruction $2^0$ to thick, tenacious meconium (e.g. with cystic fibrosis)
- Malrotation with Ladd's bands- a sort of congenital adhesion
- Malrotation with volvulus- a twisting of the bowel leading to obstruction and vascular compromise
- Extrinsic obstructions- Meckel's bands, duplication cysts, etc.

**But OD's story is not over. Eight years later…**

***OD: 8-year-old male***
*CC- Abdominal pain x 3-4 days*
*HPI- Intermittently vomiting bile (green) x 3 days. Hasn't passed gas or stool for 2 days. Years of post-prandial discomfort.*
*HPH- Duodenal atresia repair day 2 of life*
*P/E- VS- HR 100, RR 13, BP 100/73, T $37^O$ Thin. Mild abdominal distension. Mild abdominal tenderness generally. Old, well-healed surgical scar.*

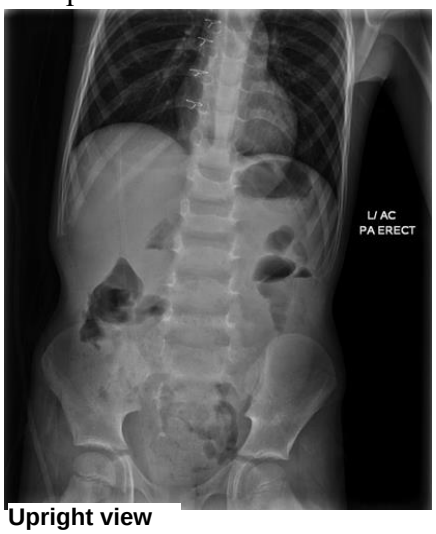
- Pathophysiologically, what do you think is going on here?—That's right! A bowel obstruction— the triad of bilious (green) emesis, abdominal distension, & obstipation
- What should our working diagnosis be?
- Early management?
- Tests?

With previous abdominal surgery and a clinical picture of a bowel obstruction, it would be usual to assume a **working diagnosis** of an **adhesive bowel obstruction**- an acquired condition. Always ask about, and look for, indications of previous abdominal surgery. Amazingly people can forget. In this case a partial, intermittent or incomplete bowel obstruction secondary to adhesions would certainly account for his years of post-prandial discomfort. What was formerly an incomplete obstruction is now worse.

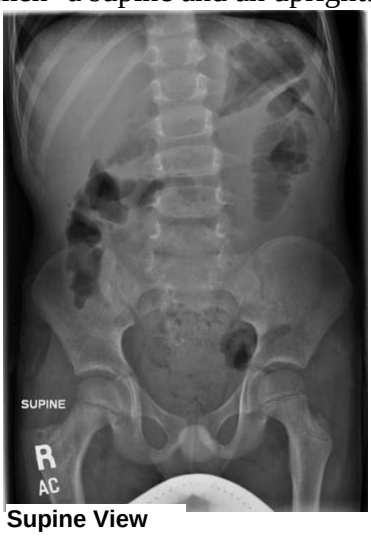
Sounds like he needs:

- 'Drip and Suck'- NG tube and IV maintenance
- + volume **resuscitation**. After days of vomiting he, of course, is volume-depleted. Don't hold back. He would need a lot of volume.
- His BUN and creatinine would be high, and his serum potassium and chloride may be low. Perhaps his sodium too. All of that should be checked and corrected.

And you would ask for…an AXR. (not an ultrasound and not a CT scan). But remember- not just a "flat plate". Get two views of his abdomen- a supine and an upright.



**Upright view**



**Supine View**

*The clinical picture and the x-ray evidence of a few air-fluid levels and some distal gas indicate a probable adhesive bowel obstruction.*
*CBC: WNL—Hgb- 145 g/L , WBC- 8 x $10^9$/L , Platelets- 250 x $10^9$/L*

**<u>Adhesive Bowel Obstructions</u>**

- Generally, 2˚to previous surgery/inflammation
- Resuscitation! They are volume and electrolyte depleted
- 'Drip and suck'- IV and NG—Generously restore volume (they often are more volume-depleted than you think) and electrolyte balance
- Monitor & repeated P/E- persistent pain, fever, high WBC are bad
- Some resolve with just 'drip & suck' treatment and close, clinical monitoring but fever,

persistent abdominal tenderness and an elevated WBC are danger signs!

- CT scan usually is not required in children, but sometimes is indicated.
- Laparotomy or laparoscopy and to cut/divide the obstructing adhesions, i.e. a 'lysis of adhesions' if no resolution

There is no absolute way to prevent adhesions. Even after a lysis of adhesions they reform to variable extents. Meticulous surgical technique, minimal bowel handling and maybe laparoscopic surgery are some ways we can perhaps prevent troublesome adhesions after operation.

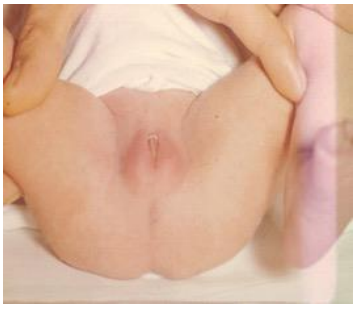
*He was followed closely and frequently examined and after 48 hours of 'drip & suck' he was clinically much better. (A fever, worsening or non-resolution of symptoms, or an elevated WBC could've indicated an operation.) His pain had resolved shortly after NG insertion. He started to pass gas. His NG drainage diminished. Repeat X-rays normalized. His NG tube was removed, and he began to drink clear fluids. Within a day he was eating and was able to go home. It could happen again, but so far, he's been OK.*

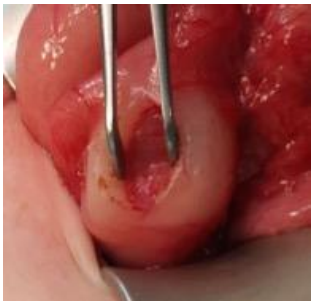
His father wondered if he should have an operation anyways, to 'cut the scar tissue' so that he wouldn't have any more bowel obstructions. How would you answer his dad's question?

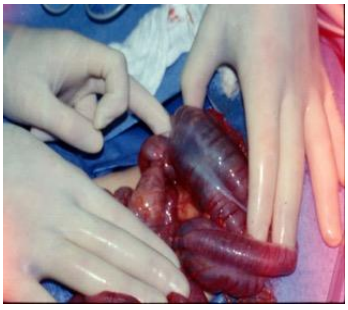
Age is important when considering a DDx for pediatric obstructions!

## Pediatric abdominal obstructions

| Newborn: | Infant: | Child: |
|---|---|---|
| -GI atresias | -Pyloric stenosis | -Intussusception |
| -Malrotation | -Intussusception | -Appendicitis |
| -Meconium ileus | -Malrotation | - Adhesions |
| -Hirschsprung's | -Hirschsprung's | -Malrotation |
| -Imperforate anus | | |
| -Biliary atresia | | |
| -Urinary obstruction | | |


Imperforate anus


Pyloric stenosis


Intussusception

## PYLORIC STENOSIS AND INTUSSUSCEPTION (YOU MUST KNOW THESE TWO CONDITIONS!)

**Intussusception**

The usual 'Illness script" (although many won't display the entire "script"):

- Occurs 3 months to 3 years- rarely outside this age range
- 10 minutes cramping intermittent 'knees up' abdominal pain, with apparent wellness in between episodes
- "Red currant jelly" stool (stool mixed with bloody mucous)
- Palpable 'sausage-shaped' abdominal mass

Testing & Rx: RESUSCITATE FIRST! (they can be quite sick in later stages), IV, Radiology- detectable on ultrasound, radiologically-mediated enema reduction, operation if necessary.

**Infantile Hypertrophic Pyloric Stenosis**

The usual 'Illness script":

- Remember "**PPP**": **P**ersistent, **P**rojectile, **P**rogressive emesis
- Occurs at 2-8 weeks of age- NOT in the very early newborn period
- The hypertrophied pylorus feels like 'Olive' on palpation of the epigastrium
- Hypochloraemic, hypokalaemic metabolic alkalosis

Testing: 'lytes, venous blood gas, abdominal ultrasound

Rx: RESUSCITATE FIRST!- volume and 'lytes, then a pyloromyotomy

*I suggest you read up on infantile hypertrophic pyloric stenosis and intussusception.*

## CHECKPOINT: PEDIATRIC GI OBSTRUCTIONS

Re: GI obstructions

- **BEWARE THE CHILD WHO VOMITS GREEN**
- Life-threatening/ Resuscitation required
- Think pathophysiologically—what's wrong?
- Congenital vs Acquired?

Re: Any obstruction

- History & Physical exam!
- Consider the child's age then consider what types of obstructions occur at that age
- Sensible testing- e.g. ask a radiologist
- Specialized treatments in some specialized conditions
- Look for associated conditions. e.g. VACTERL
- It's not just the bowels that get obstructed—in babies consider biliary atresia, urinary obstructions, and CSF obstructions…

***Have you noticed?***
*Surgeons seem to be obsessed with food!*
*Look how often we refer to food when we speak of disease—*
*"olives", "red currant jelly", "sausage-shaped masses"…*

CHECKPOINT CHALLENGE #2: A 7-YEAR-OLD BOY PRESENTS TO THE ER WITH AN 18-HOUR HISTORY OF INTERMITTENT CRAMPY ABDOMINAL PAIN AND PASSAGE OF DARK RED STOOL. A THOROUGH HISTORY AND PHYSICAL EXAM COUPLED WITH TWO-VIEWS OF HIS ABDOMEN AND AN ABDOMINAL ULTRASOUND POINT TO AN INTUSSUSCEPTION AS BEING THE UNDERLYING CAUSE!

THIS IS CERTAINLY OUT OF THE USUAL AGE RANGE FOR INTUSSUSCEPTION. WHAT IS GOING ON? (A HINT IS IN THE APPENDIX, BUT NOT IN THIS BOY'S APPENDIX.)

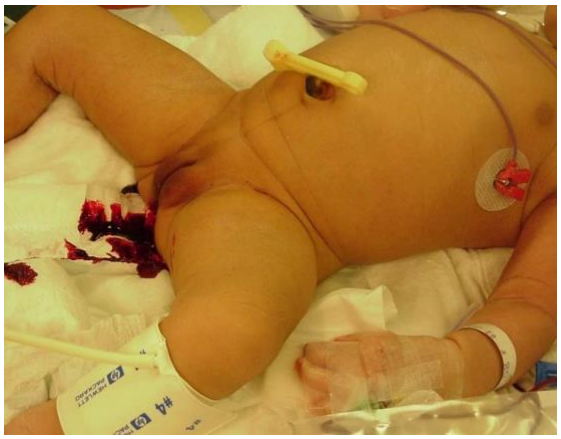
# BLEEDING

## NOBL FIIISTT

One way to organize our thoughts around bleeding is to think about "medical" bleeding vs "surgical" bleeding. Simply put, surgical bleeding may be defined as bleeding where surgery plays a significant role in stopping the bleeding. This does not mean that we can ignore "medical" aspects in surgical bleeds, for instance, a concomitant coagulopathy. That would be an error, but the categorization allows us to focus our learning on those bleeding conditions where a surgeon should be involved in the care of the child. Let's pick a bleeding problem to work through:

***Case BV***

*1-day old newborn female,*
*39-week gestation.*
*CC—passing significant blood PR*
*HPI- non-contributory*
*P/E- Tachycardic*
*Tender abdomen*



This is significant bleeding and demands your urgent attention. **So-what now?**

We have to **think**. We have to reason "INDUCTIVELY". Inductive reasoning takes us from this particular patient's situation to a broader category: the **diagnosis**. [Reference: Brush JE, The Science of the Art of Medicine, 2015 John E. Brush, Jr publisher, eBook available through iBooks]

First, it would help to know a short list of pathologies which may cause significant rectal bleeding in a newborn baby. Let's consider the following possibilities and rule them in or out with what we know:

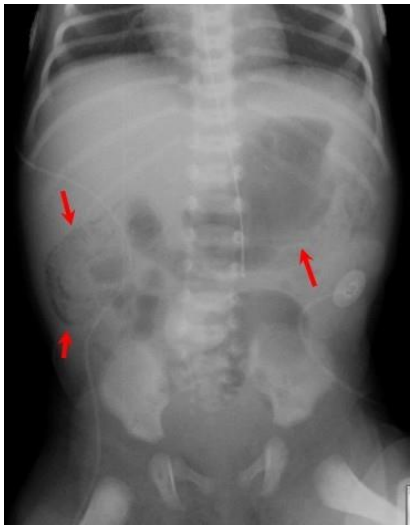
**Differential Diagnosis:**

- Swallowed blood?- Babies when they pass through the birth canal can rarely swallow significant quantities of mother's blood. Or they may swallow blood from mother's nipple while breastfeeding. However, when the blood passes through their GI tract the baby is well and will not have a tender abdomen with tachycardia. You could waste valuable time in this case by testing the defecated blood with an Apt Test (look it up!) which will indicate that it is mother's blood and not baby's if it had been swallowed.

- Baby has a coagulopathy? Possible, and bear in mind that "medical" and "surgical" bleeding are not discrete categories. Always a good idea with any significant GI bleeding to check the patient's coagulation status. Did the baby get the routine newborn shot of IM Vitamin K? But again, the baby's abdomen shouldn't be tender in that situation.

- An intussusception?- Intussusceptions are exceedingly rare—really rare—in very young babies. Remember, intussusceptions generally occur between the ages of 3 months to 3 years. This is a one-day old babe. You are probably way off the mark if you think the baby has an intussusception!

- Necrotizing Enterocolitis?- Babies with "NEC", as we call it, certainly can pass significant blood per rectum and they are tender and tachycardic and sick. However, they are almost always 3 days old or older, usually have been fed, and are usually premature and/or have had a perinatal asphyxiation event or are on NEC-inducing medications. Certainly, we can keep it in mind, but it doesn't fit well with this clinical picture. An AXR in NEC often will show bubbles of air in the bowel wall, a condition called 'pneumatosis intestinalis'. (Red arrows on this AXR)



- Vascular malformation of the GI tract that is now bleeding? – For some reason people often include this high on their differential list, but it is actually very, very rare.

- Colonic polyp perhaps—Polyps are acquired lesions and do not occur in one-day-old babes.

- Peptic Ulcer?- Newborn babies are actually achlorhydric for the first few days; and remember: "no acid, no ulcer".

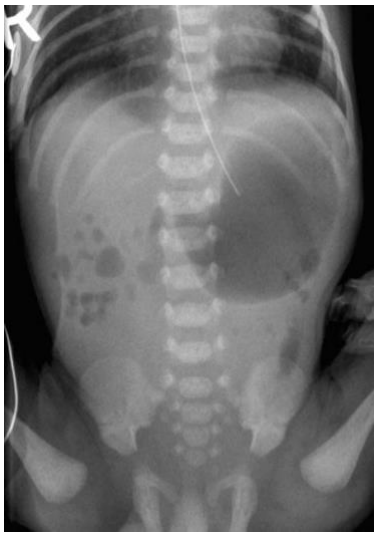
- Intestinal Malrotation with Acute Volvulus?- If the baby is born with a GI malrotation, an acute volvulus- or twist- of the intestines may occur at any time. The true incidence of intestinal malrotation is not known, occurring perhaps as frequently as 1/100 in autopsy studies to a less frequent 1/4000 in clinical studies, but it's not rare. The chances of an intestinal malrotation turning into a life-threatening intestinal volvulus is also unknown but when it happens it is often in the newborn period. It occurs suddenly causing an acute vascular compromise of the mesentery which in turn leads to intestinal ischemic necrosis and congestive bleeding and sloughing of intestinal mucosa. The baby might be hemodynamically compromised, and the baby's abdomen would seem to be tender. There may also be abdominal distension and bilious (green) emesis because the duodenum is often obstructed by the twist and by abnormal congenital "Ladd's bands" that are often present in intestinal malrotation which sweep from the right lateral abdominal wall across

the duodenum to the ascending colon, which is abnormally located in the upper mid-abdomen.

Therefore, using inductive reasoning, which is not infallible, we must assume this baby has **Intestinal Malrotation with an Acute Volvulus**—a very dangerous and life-threatening condition. We must also include NEC as a secondary possibility. Even if the baby wasn't too sick—and this baby is—assuming the worst is usually the best option.

*An IV is started and fluids are administered quickly. An 8 Fr nasogastric tube is inserted and placed on suction. There is no blood noted in the NG aspirate, but the baby just vomited green.*

**What does "8Fr" mean?**
8Fr is pronounced "8 French" and the "French" size denotes the outside circumference of the tube which, basically for the purposes of calculation, is in millimetres. By the way, if you know the circumference of a tube, how can you calculate its diameter? (refer to Grade 6 math!)
For purposes of decompressing the GI tract, an 8Fr NG would be the smallest tube you would use on a baby. Using anything smaller is useless.



*However, this baby worsens quickly with more blood passed per rectum, more apparent abdominal tenderness and her hemodynamic status rapidly worsens as well.*

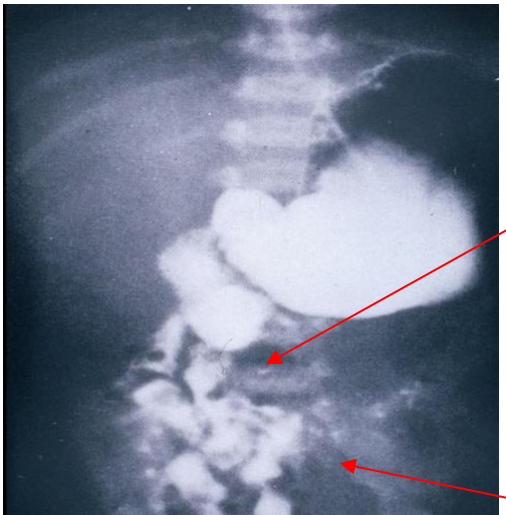
*A quick crib-side AXR is done.* *(supine only—-someone thought that would be adequate, which is a common mistake. If possible, you should always get 2 views— a supine view and a dependent view which would show any free air or air-fluid levels if present.)*

*The AXR is abnormal but non-specific. No apparent pneumatosis intestinalis, for instance. The stomach is big and filled with gas. The NG should be pushed in further.*

Wouldn't it be great to have a test that could confirm malrotation and volvulus here? Shouldn't we always strive for diagnostic certainty?

An Upper GI Contrast x-ray is an excellent way to accurately denote intestinal malrotation.

In intestinal malrotation the duodeno-jejunal (DJ) junction does not cross the midline.

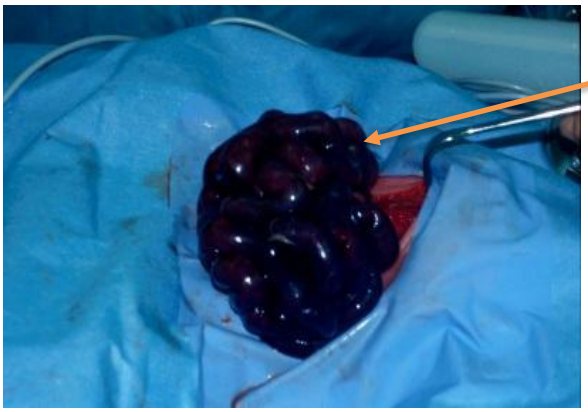


This UGI series shows intestinal malrotation with the small bowel coursing, in a sort of spiral, down the right side of the abdomen. The duodeno-jejunal flexure is to the right of the midline. This means the bowel and its mesentery is not fixed to the retroperitoneum but is floating freely and able to twist into a volvulus. The duodenum here is somewhat large, possibly because of Ladd's bands partially obstructing it. (I should add that a radiologist needs to interpret studies like these.)

**So- should we just order an UGI series?**

Unfortunately, this baby is so very sick and unstable any specialized x-ray study would result in her death. You just don't have time! Many times, we have to act in the face of uncertainty!

*The baby underwent an emergency laparotomy. An acute volvulus was found with a 720$^{O}$ twist of the mesentery. The intestine was black and markedly ischemic. The bowel was untwisted, and a Ladd's procedure was performed (Bowel untwisted, and placed in 'non-rotation', i.e. colon primarily on the left, small bowel primarily on the right, appendectomy done). The bowel 'pinked up', mesenteric blood flow restored, and no bowel was resected. The baby recovered smoothly.*



You can see here how black the bowel was at laparotomy. This baby needed surgery now! Any delay would have been deadly.

Let's take a look at another, different case of bleeding.

***Case BW:***
*3 y.o. male*
*CC- bleeding PR*
*HPI- Painless dark red and black bleeding PR x 2 days. O/W healthy.*
*P/E- Pallor. Tachycardic. Normal abdomen.*
*DRE- dark blood/melena on finger.*

This doesn't sound like the last case, does it? The abdomen isn't tender, and the child isn't complaining of any pain whatsoever. But, like the last case, he is tachycardic.

This is a good example of how you can learn to recognize "**Illness Scripts."** Perhaps you'd call it a "**Spot Diagnosis**." I showed you some Illness Scripts for Pyloric Stenosis and Intussusception.

The recognition of illness scripts is different than the inductive approach that we took with this last case of intestinal malrotation with volvulus. This is not to imply that either is better, or more

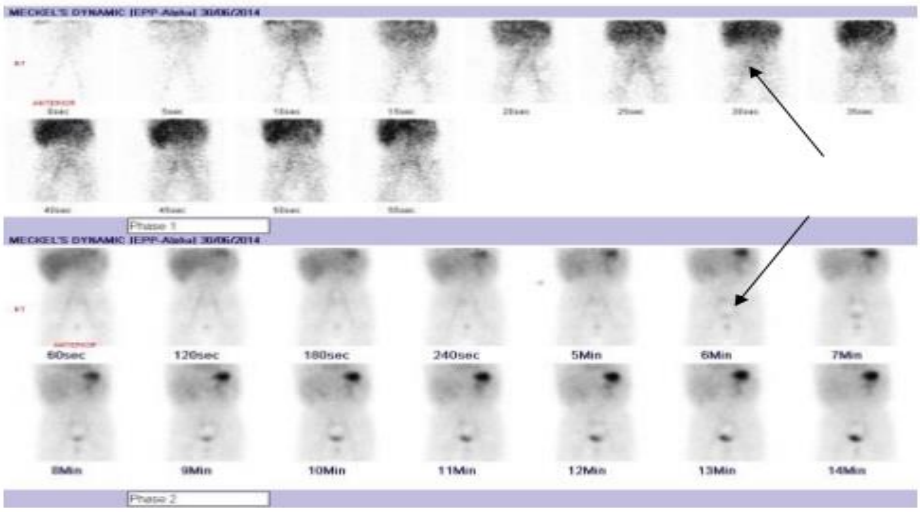
accurate, or that elements of induction cannot be used with illness scripts or vice versa. Both are good and can lead you down the correct path—and sometimes the wrong path.

Simply put, the more experience you gain, the more easily you'll recognize **patterns of disease presentation**. When you are just starting out you are forced to work more slowly and methodically factoring in each separate fact presented.

So, when I hear this story from my medical student who has just taken a history and examined the lad I recognize this illness script as typical of **a bleeding Meckel's diverticulum.** Could there be another diagnosis here? Of course! So, we have to be careful. Let's ask ourselves some questions:

- Does this boy need an immediate operation? **No.**
- Does this boy need systemic support? **Yes.** He's bleeding so a good size IV needs to be started, a cross-match done, plus a CBC and coagulation profile.
    - CBC shows hemoglobin at 60 g/dl (really low!), Coag studies are normal.
    - We initiate a transfusion—reasonable with this level of symptomatic anemia.
- What other tests would help?
    - Other blood tests? **No real need.**
    - AXRs? **Won't show anything specific.** P/E reveals his abdomen to be normal.
    - Abdominal ultrasound? Now here, you have to ask, what are you looking for? What could an abdominal ultrasound possibly show us here? A tumour? **No- an ultrasound here is a waste of time.**
    - A Meckel scan? **Now you're talking!**

*An urgent Meckel scan is done. – it is positive*



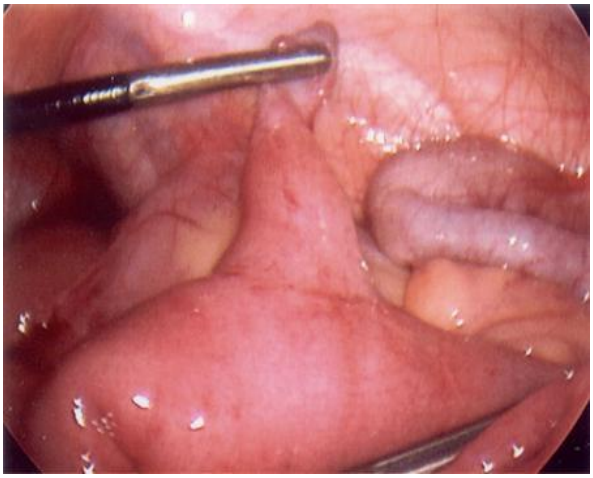

A Meckel scan utilizes the injection of Technetium-99m (99m Tc) which gathers in areas of gastric mucosa. We see in these images the radionuclide gathering in the stomach and in the bladder as it is secreted. The arrow indicates an area of abnormal update- in this case a Meckel's

diverticulum. This nuclear medicine scintiscan is quite accurate in children, but as you can see in the images here, they can be very difficult for the non-radiologist to read.

A Meckel's diverticulum with ectopic gastric mucosa that secretes acid easily causes the adjacent small bowel mucosa to ulcerate and bleed—profusely. The bleeding is usually enough to cause a significant acute anemia. Because the blood has been in the presence of acid, like upper GI bleeding it is often melena or at least dark purple.

We now know the source. Best to surgically deal with this urgently.

*The boy undergoes a laparoscopic resection of the Meckel's diverticulum. He recovers quickly and is discharged in 2 days.*



Laparoscopic view of Meckel's Diverticulum being held up at its tip by the instrument. (photo courtesy of Dr. KuoJen Tsao)

**Meckel's Diverticulum**
Usual illness script:

- Bleeding PR, painless, dark red/purple or black melena
- Pallor
- Tachycardic
- Normal abdomen
- DRE: Dark blood/Melena on finger

Testing: Meckel scan, CBC, X-match
Rx: Resuscitation, Surgical resection

***Who was Professor Johann Meckel?***

*Johann Friedrich Meckel was an anatomist born in 1781, in Halle, Germany. Professor Meckel specialized in defects and abnormalities that occur during embryonic development. The Meckel's diverticulum was first described by Wilhelm "the Father of German Surgery" Fabry in 1598, but remained unnamed for 200 years! Much like our friend Max Wilms, Meckel's name has been attached to this previously discovered abnormality as a result of his research on its embryology and anatomy in 1809. As a matter of fact, Meckel is a prominent eponym, as the Meckel family spawned a long line of Professors of Anatomy, who described a number of anatomic features and anomalies.*

## CHECKPOINT: BLEEDING

*(any kind of bleeding- not only GI bleeding)*

- Bleeding is a sign/symptom of the underlying pathology. Determine if it is significant bleeding vs insignificant bleeding- i.e. is the patient anemic or showing signs of hypovolemia?
- Differentiate 'Surgical' bleeding vs 'Medical' bleeding- i.e. surgical bleeding is amenable to surgery; ligating vessels, Interventional Radiology (IR) embolization, etc. Medical bleeding, such as a coagulopathy, needs medical treatment; fresh frozen plasma, etc. Often bleeding patients have both medical and surgical bleeding, however.
- Support the patient, resuscitate/transfuse as necessary.
- Prevent shock! (Children descend rapidly into shock and that's big trouble!)
- Find the cause. Treat the cause.

CHECKPOINT CHALLENGE #3: LET'S SAY THE RADIOLOGIST LOOKING AT THE MECKEL SCAN ON PATIENT BW SAYS IT IS "EQUIVOCAL"—WHAT THEN?

CHECKPOINT CHALLENGE #4: A 12-YEAR-OLD GIRL PRESENTS WITH A HISTORY OVER THE PAST YEAR OF A NUMBER OF EPISODES OF INTERMITTENT ABDOMINAL PAIN, NAUSEA, AND ON TWO OCCASIONS SHE HAS HAD GREEN EMESIS. ON EACH OCCASION THE SYMPTOMS HAVE SUBSIDED SPONTANEOUSLY. THERE IS NO OTHER PERTINENT HISTORY. PHYSICAL EXAMINATION, INCLUDING HER ABDOMEN, IS ENTIRELY NORMAL. SHE UNDERGOES INVESTIGATIONS WHICH INCLUDE A NORMAL ENDOSCOPY BUT AN UPPER GI SERIES SHOWING INTESTINAL MALROTATION WITH NO OBSTRUCTION. WHAT TO DO?

# LEAKS

## NOBL FIIISTT

'Leaks'—an odd and actually broad category of surgical woes. Let's consider what happens when various bodily compartments leak.

***Case LP:***
*15-year-old female, previously healthy, walks into your ER*
*CC- Shortness of breath*
*HPI- Acute onset while watching television. No history of trauma. Never had this complaint before.*
*P/E- decreased A/E on right*

What is your immediate management?

Remember the ABCs!
**A- Airway***: She is phonating easily and seems not to have any airway issues. You assess her trachea and it is midline.* Why is that important?

**B-Breathing:** *RR 30/min. She is not cyanotic on room air. An oxygen saturation probe on her finger reads $SaO_2$=95%. But you provide her with nasal prongs anyway- $O_2$ at 2 l/min providing approximately 28% $O_2$. You confirm decreased breath sounds on the right.*

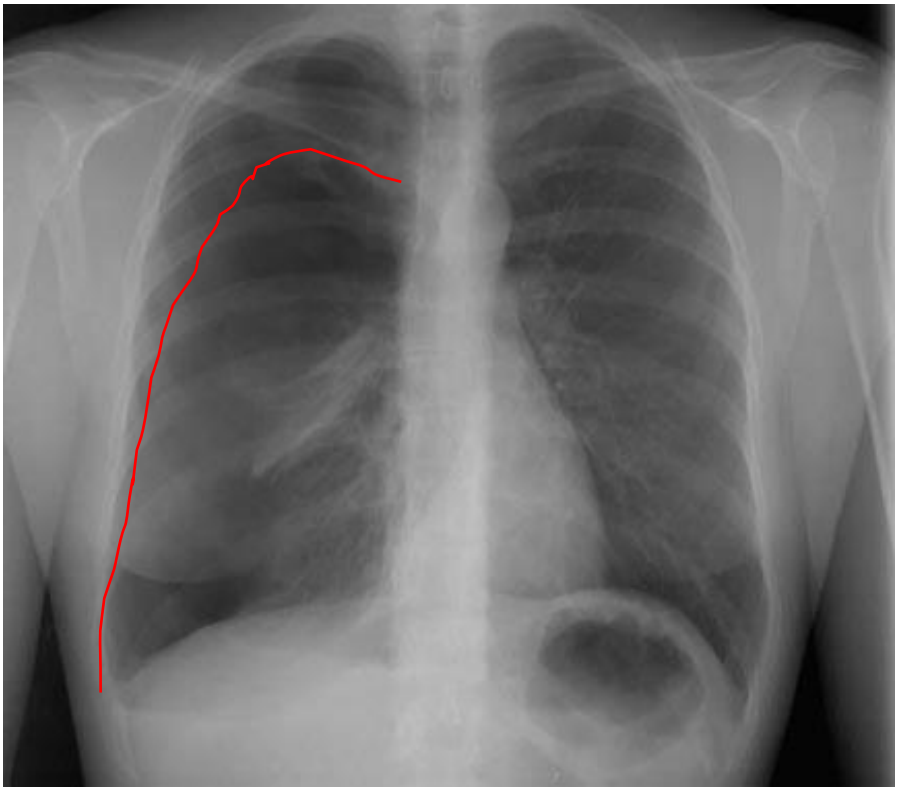
**C-Circulation*n*:** *HR 75/min, BP 120/80. You are considering starting an IV, but she is so stable and in a monitored section of the ER that you decide to hold off for now.*

Let's say it's the middle of the night.

- What sort of things may be going on inside this girl causing this clinical picture?
- You are developing a differential diagnosis…
- What is your **working diagnosis**?
    - The working diagnosis is either the one you are betting on or the one you **must absolutely rule** out for the patient's immediate welfare.

With a working diagnosis in mind, what further information do you want? What testing would help? It's usually best to keep it simple and easy at first. (e.g. an MR scan right now to rule out a mediastinal tumour is not a good idea!)

CXR you say? Yes- easy, quick and readily accessible.



The red line marks the approximate edge of the right collapsed lung. Approximately 40% of the total right pleural cavity is now filled with air outside the right lung.

*The CXR reveals a 40% pneumothorax on the right. There is some associated right lower lobe atelectasis. The mediastinum is not shifted. The left lung and other structures are within normal limits.*

How are we going to help this girl? Air has somehow 'leaked' into her right pleural space.

First, let's consider how this pneumothorax occurred.

Remember, it all happened while she was sitting watching TV, so it wasn't a traumatic pneumothorax. A broken rib hasn't punctured her lung. Some individuals, especially lanky teenagers, have 'bullae' on the apices of their lungs. These air-filled blisters can spontaneously burst. Air then escapes the lung and begins to fill the pleural space, resulting in partial collapse of the lung. This is perceived in the early stages as dyspnea, often associated with ipsilateral chest pain. If the leak is large, then more air leaks into the pleural space. This results in a profound collapse of the lung.
With every inspiration more, air escapes into the pleural cavity. This pleural air has no escape, and eventually a **tension pneumothorax** may be the result which can **deviate** the mediastinum (remember I asked why it was important that her trachea is midline?), causing not only severe

respiratory compromise, but also may impair venous return to the heart, which in turn may decrease the cardiac output. A quick death may be the result of a tension 'pneumo' if it is not treated. Tension pneumothoraces are happily rare in cases spontaneous pneumothorax—but not in traumatic pneumothoraces. Review your trauma ABCs to remind yourself on how, in extreme cases, you can treat a tension 'pneumothorax'.

*Under local anesthesia a chest tube is placed in the right pleural cavity. This is attached to an 'underwater seal' (which is like a one-way valve for air- air can go out, but not back in) and a collection chamber which allows for escape of the pleural air, re-expansion of the right lung.*

You'll see different types of chest tubes used. Some are large and clear (e.g. 24 Fr), while others are small and opaque (e.g. 10 Fr), with a "pigtail" curl inside the chest. Ask clinicians why they chose the one they used. In most patients, the leaking bulla will seal itself over a few days. If not, then video-assisted thoracoscopic surgery (VATS) may be done to surgical seal the leak. The bullae are sealed surgically and the parietal pleura is either scraped and irritated, or actually stripped away so that the re-expanded lung will 'stick' to the chest wall and not collapse again.

LP had a 40% pneumothorax. 'Pneumos' of 25% or less may be treated by watchful waiting without a chest tube hoping for spontaneous resolution.

*After 3 days, her chest tube stopped bubbling, her CXR showed complete re-expansion of the lung. Her chest tube was removed and she was discharged home.*

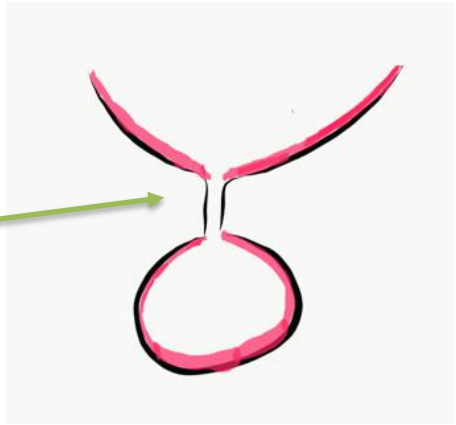
## FISTULAE

A **fistula** is a 'leak' of sorts.

What is a **fistula**?

We often use terms without regard to their exact meaning.

- Definition: A fistula is an abnormal connection between two epithelialized (or endothelialized) surfaces.



Fistula tract (Pink is epithelium)

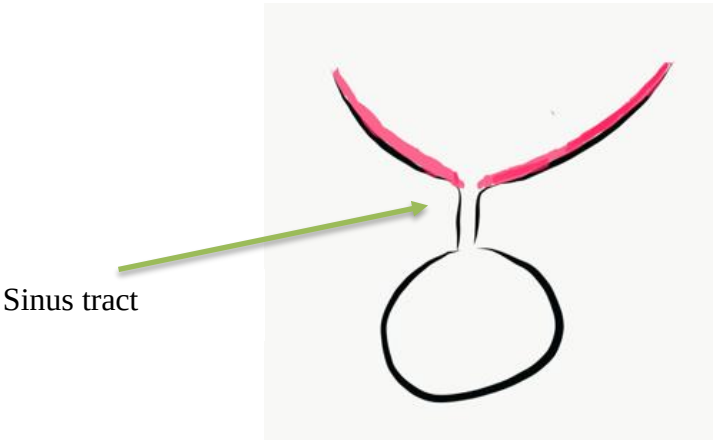
Lots of examples:

- A gastro-colic fistula
- A tracheo-cutaneous fistula
- A recto-urethral fistula
- A bilio-cutaneous fistula

- A brancho-cutaneous fistula
- An arterio-venous fistula (two endothelialized surfaces)

While we are at it, let us define **a sinus tract**. We are not speaking of anatomic sinuses like the maxillary sinus, but pathologic sinus tracts are sometimes simply referred to as a 'sinus'. Its definition is subtly different from that of a fistula.

- Definition: A sinus tract is an abnormal connection between two surfaces only one of which is epithelialized.




Example:

- A draining sinus- Sometimes a deep pathologic collection of fluid or pus will drain to an epithelialized surface like the skin or intestinal mucosa. The abscess is not lined by epithelium, but the skin or bowel is—thus it is a sinus..

Very often a fistula is mistakenly referred to as a sinus, and vice versa. Think about the surfaces connected and then use the proper term. Chances are, as a medical student, you'll encounter fistulae and sinus tracts. There are many different reasons why they form and many ways they can close.

When you encounter a fistula, or a sinus tract ask the questions:

- Is it a fistula or a sinus?
- What caused this?
- What prevents it from closing or healing?
- What can we do to help it close or heal?
- Or… do we want it to close?

We'll return to fistulae and pathological sinuses later… but in the meantime, think (or look up) what sorts of disease processes lead to pathological fistulae and sinus tracts.

## CHECKPOINT: FISTULAE, SINUS TRACTS AND LEAKS

- What's leaking?

- Where is it leaking?
- Why is it leaking?
- Can I fix the leak?
- If I can't fix the leak, can I control it?

CHECKPOINT CHALLENGE #5: CHECKPOINT CHALLENGE #5: THE 15 YEAR-OLD GIRL, LP, WHO WAS TREATED FOR HER SPONTANEOUS PNEUMOTHORAX, *THE DAY AFTER HOSPITAL DISCHARGE FLEW TO FLORIDA TO VISIT HER AUNT. SHE ALMOST DIED ON ROUTE WHEN AT 38,000 FEET IN THE JETLINER SHE SUFFERED SEVERE RESPIRATORY EMBARRASSMENT. THE PLANE MADE AN EMERGENCY LANDING IN TULSA, OKLAHOMA, WHERE PARAMEDICS DECOMPRESSED A RECURRENT RIGHT PNEUMOTHORAX.* WHY DID ALL THAT HAPPEN? IMAGINE YOU ARE ON THAT PLANE AND THE CAPTAIN CALLS FOR MEDICAL HELP—WHAT WOULD YOU DO?

CHECKPOINT CHALLENGE #6: OFTEN, PATIENTS WITH A UNILATERAL SPONTANEOUS PNEUMOTHORAX WILL HAVE A CT SCAN OF THEIR CHEST. LET'S SAY LP HAS A CT AND IT SHOWS BULLAE ON THE RIGHT SIDE, AS EXPECTED, BUT ALSO BULLAE ON HER LEFT SIDE, EVEN THOUGH SHE HAS NO PNEUMOTHORAX ON THE LEFT. WHAT IS THE ADVICE?

CHECKPOINT CHALLENGE #7: LP'S BULLAE AT THE TOP OF HER RIGHT LUNG IS LEAKING INTO HER RIGHT PLEURAL CAVITY. IS THIS A FISTULA? IS IT A SINUS? DOES IT MATTER?

# FUNCTIONAL DISORDERS

## NOBL FIIISTT

The word "functional" above refers simply to how the body or an organ functions and is not pertaining to the "functional" problems in psychiatry.

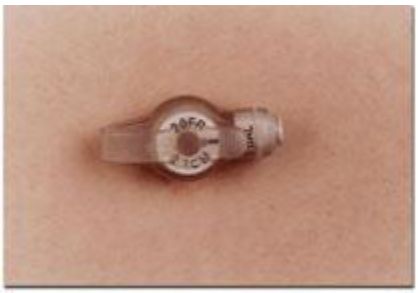
***Case FG:***
*1 month old female*
*CC- Feeding difficulties*
*HPI- Preterm. Microcephaly.*
*Feeding study indicates 'aspiration'*
*Currently fed via NG*

How can a surgeon help this little patient?

Up to now we have been discussing disease wherein the surgeon can intervene and treat that disease for resolution. In this little one there is no surgeon who can 'fix' this child's profound neurologic disability. But it doesn't mean that a surgeon cannot help.

This is an example of a disorder of **function**. The child cannot swallow properly and thus cannot eat normally. An NG tube may be used to feed her, but a chronic NG feeding tube is uncomfortable and difficult to maintain. In this situation, a surgically placed **gastrostomy tube** for feeding may be appropriate. It doesn't cure the child, but it affords an easy to maintain direct route into the stomach for feeding and avoids the discomfort and hassle of a NG feeding tube. One benefit of a gastrostomy rather than a feeding NG tube in children who need long term tube feeding is actually a social benefit. Children who have NG tubes are immediately labelled as "sick" and a hidden gastrostomy tube can promote more normal and healthier social interactions with other children and adults.

Gastrostomy tubes, or G-tubes as they are known, come in many different forms and types. This is one type of 'low-profile' G-tube.



By the way-Is a gastrostomy a fistula?

(Yep-a therapeutic fistula)

We have talked about GI Obstructions and in that section I mentioned **Hirschsprung's disease**. Hirschsprung's disease presents as a **GI obstruction**, most often in early infancy. However, it too can be considered as a **disorder of Function**. (NOBL FIIISTT categories are not mutually exclusive—e.g. Hirschsprung's is in both the Obstruction and Functional categories.)

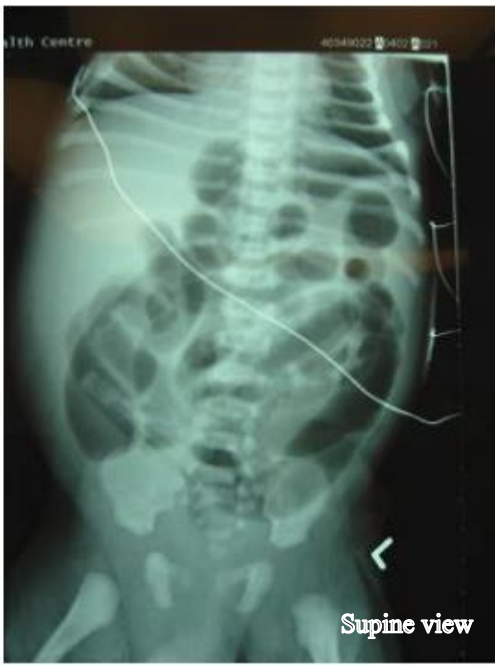
Consider this case, which illustrates many of the classic features of Hirschsprung's:

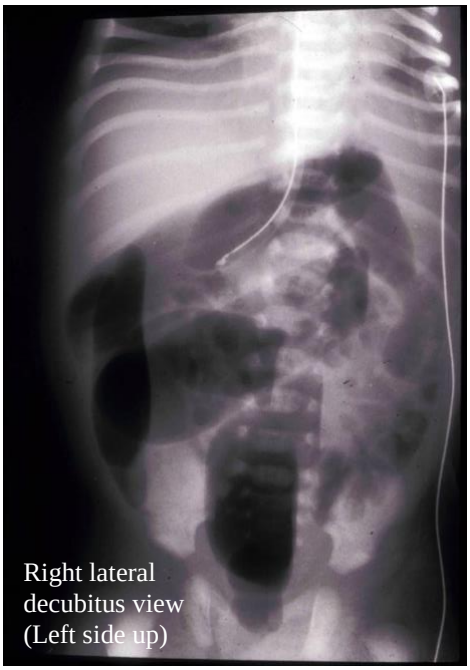
***Case FH:***
*Full-term male. .*
*CC- Failure to pass meconium in the first 24 hours*
*HPI- Unremarkable pregnancy. Vaginal birth Good Apgar scores.*
*At 36 hours- intolerant of feeds. No meconium passed.*
*O/E- abdominal distension, non-tender abdomen, anus patent, DRE with little finger results in a 'gush' of loose meconium out the anus.*

Most normal babies pass meconium within the first 24 hours of life. **Don't ever miss a case of imperforate anus on a newborn exam, which is the commonest cause of bowel obstruction in the newborn.** But, if they have an anus and haven't passed meconium in the first 24 hours, think Hirschsprung's disease.

Imaging?

- In Case FH it would be reasonable to obtain 2 views of the abdomen. Two AXRs – one with the patient supine and another view that shows gravity at work. By that I mean it would usually show air-fluid levels in the bowels if there is an obstruction.



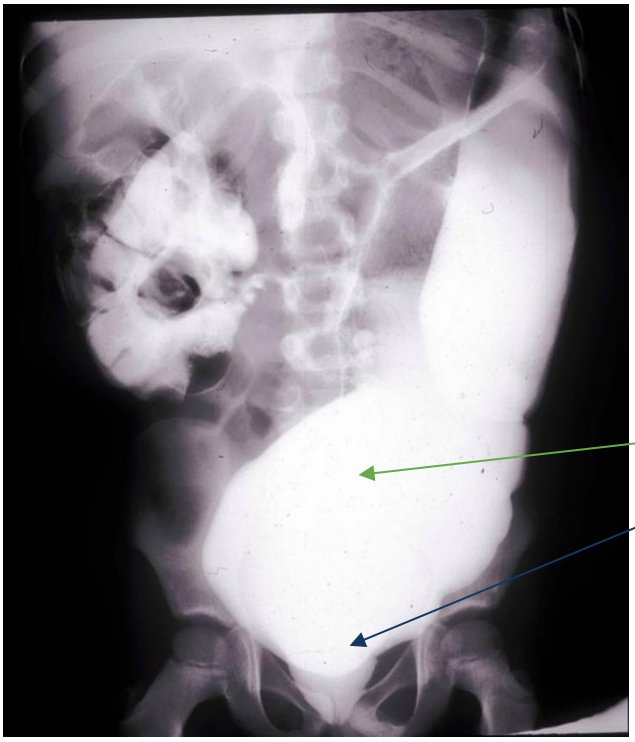

*The AXRs reveal evidence of bowel distension, some air-fluid levels on the right lateral decubitus view on the right and no air in the rectum, all suggestive of Hirshsprung's disease.*

**What is going on here?**

Hirschsprung's disease is pathologically characterized by a lack of ganglion cells in the rectal wall. This nerve cell lack leads to a sort of chronic unrelenting spasm of the rectum, and a functional obstruction of the lower GI tract. The lack of ganglia is always a feature of the rectum in Hirschsprung's with the absence of ganglia extending a variable distance proximal up the colon and rarely even into the small intestine. A digital rectal exam can sometimes break the rectal spasm for a moment and one then witnesses a sudden rush of meconium passing through as we saw in patient FH.

**Investigations?**

**A colon contrast enema** (sometimes termed a barium enema but often other contrast agents are used instead of barium) can most often (but not always) reveal the narrowed spasmodic rectum, with distension of the normally innervated colon above.

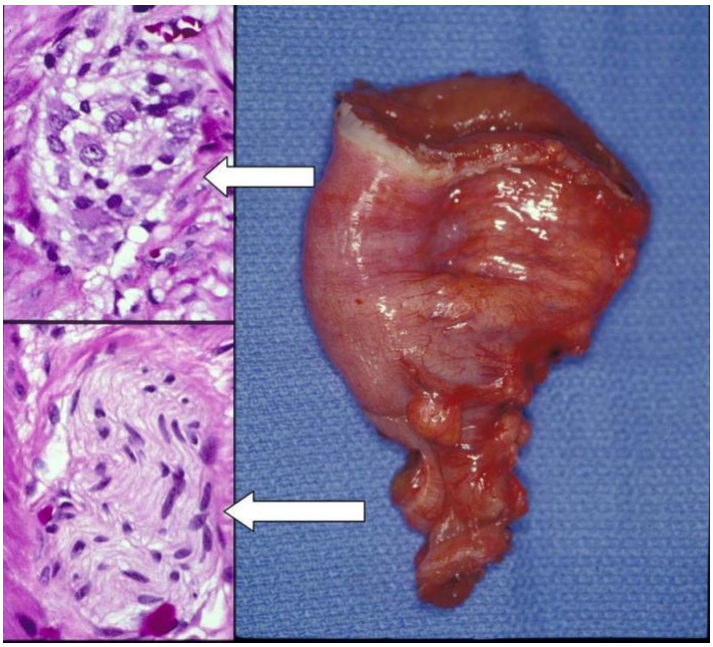


*A colon contrast study reveals a narrow rectum and a dilated **sigmoid colon** indicating the probable **transition zone** (from aganglionic to ganglionated bowel above) at the recto-sigmoid. The narrowed rectum is difficult to appreciate here.*

A colon contrast study like this can help us plan for surgery. But we still need more evidence that this is Hirschsprung's disease. The clinical picture and the radiologic studies on patient FH are classical, but there are conditions that may mimic Hirschsprung's and with major surgery as the Hirschsprung cure, we had better look to see if ganglion cells are actually absent—**with a rectal biopsy**. Three or four small samples of the inner wall of the rectum may be taken with a small endoluminal biopsy instrument at various levels. The pathologic diagnosis of Hirschsprung's disease is made when the pathologist reports:

*Mucosal-submucosal rectal biopsies of patient FH reveal a complete absence of ganglia. (See figure below)*

The right half of this composite figure shows a surgically excised portion of rectosigmoid. On the left you can see histologic evidence of ganglia in the dilated portion above an apparent transition zone. But below the transition zone, approximately where the initial rectal biopsies were taken, the ganglia are absent. Also the nerve trunks are hypertrophied—another histologic indicator of Hirschsprung's disease. You can see in this pathology specimen why the colon contrast enema looked the way it did.



**Treatment?**

This disease, a disease of disordered neurologic function causing bowel obstruction may be successfully treated with surgery. Essentially the aganglionic portion of colon is removed and the ganglionated colon is 'pulled through' and sewn to the anal canal. The operation, which has technical variations, is called a **"pull-through**". Prior to the conception of the pull-through operation, the only help for a patient with Hirschsprung's disease was a colostomy. If a Hirschsprung baby's obstruction isn't promptly relieved, the disease can be fatal, often very rapidly fatal.

*Baby FH underwent a pull-through operation and did well, stooling normally.*

***Case FT:***
*14-year-old girl presents with a 2-year history of significant hyperthyroid symptoms— tachycardia, weight loss, anxiety— and a large goiter.*
*Investigations indicate Graves' disease.*
*Hyperthyroid symptoms generally controlled with antithyroid medication*

*A pediatric endocrinologist refers FT to a pediatric surgeon.*

Two things of note here:

1. Firstly, you may think that Graves' disease is an 'adult' disorder. Although there are many pediatric-specific diseases (like Infantile Hypertrophic Pyloric Stenosis) that adults don't have to contend with, there are definitely some adult-predominant illnesses from which children are not necessarily spared. Dealing with a child with an adult-predominant disease has special challenges, such as in this case of FT. FT

has a long-life ahead of her and what are the long term effects and burden of taking anti-thyroid medication the rest of her life?

2. Certainly a thyroidectomy can rid her of the hyperthyroidism and her need for antithyroid drugs, but post-thyroidectomy she'll be rendered HYPOthyroid—life-long—and need life-long thyroid hormone replacement. What are the long term effects of that?

FT's case will need some good thought and discussion, not only between the endocrinologist and the surgeon, but, of course, with FT and her parents. It's one of many disorders of Function wherein the surgeon may play a specific role.

## CHECKPOINT: FUNCTIONAL DISORDERS

- Sometimes we can fix things, sometimes we can't.
- Disorders of GI motility is a good example of a disorder of FUNCTION.
- If we cannot fix the FUNCTION is there a way to work around the problem?

### CHECKPOINT CHALLENGE #8: BABY FH AS DESCRIBED ABOVE HAD A DEFINITE BOWEL OBSTRUCTION WITH HIS HIRSCHSPRUNG'S DISEASE AND ALTHOUGH HE WAS INTOLERANT OF FEEDS AND HAD SOME ABDOMINAL DISTENSION, HE NEVER VOMITED "GREEN". WHY?

### CHECKPOINT CHALLENGE #9: WHAT NEUROLOGIC DISEASE WHICH CAN AFFECT BOTH CHILDREN AND ADULTS SOMETIMES BENEFITS FROM A THYMECTOMY?

*Robert James Graves (1796-1853)*

*Graves' disease, otherwise known as primary thyrotoxicosis, is named after an Irish physician. He described this condition in 1835 and although we name it after him because of that, the disease was in fact first described by an English physician, Caleb Hillier Parry, ten years earlier in 1825! Not only was poor Parry cheated out of being eponymically attached to primary thyrotoxicosis, Parry also described what we call Hirschsprung's disease way before Hirschsprung! Is there no justice in this world?*

*However, there is another reason to be grateful for Dr. Graves, for he was the first to realize that medical students should be involved in clinical ward work for the sake of their education. Prior to Graves, medical students simply attended lectures, anatomy labs and read books.*

*Lastly, you've maybe heard the old saying "Feed a cold, starve a fever." Well, Dr Graves typified the attitude we should all have towards "established medical doctrine" in that he habitually and continually would question its rationale. His dying wish for his gravestone epitaph is a lesson to us all:*

*Robert James Graves (1796-1853)*
*"He fed fevers"*

# INFLAMMATION/INFECTION
## NOBL FIIISTT

Inflammation does not necessarily mean Infection

***Case IA:***
*11-year-old boy*
*CC- Abdo pain X 24 hours. Denies any trauma.*
*HPI- Previously healthy.*
*Started as mid-abdominal pain, now localized in RLQ*
*P/E Localized tenderness and guarding at McBurney's point*

As we work our way through this very common clinical scenario let us ponder the following in sequence:

- What's going on inside this boy to cause these symptoms & signs?
- Establish a differential diagnosis
- Establish a working diagnosis
- What tests do you want to do?
- What tests are unnecessary?

**Inside:**

It seems likely that something inflammatory and quite probably infective in this boy's belly is the trouble, although we can't right now dismiss obstructive problems. Neoplasia, structural, functional, ischemia, leaks, and trauma are much less likely.

**Differential Diagnosis:**

The list of possible differential diagnoses in the inflammatory/infective category in a case like this is very long and includes appendicitis, of course. Less likely are diagnoses such as mesenteric adenitis, gastroenteritis, or rare entities like a Meckel's diverticulitis, etc. It is odd but true that a disease as common as appendicitis can continue to perturb and deceive us. It does so on a regular basis, even when you've dealt with thousands of appendicitis patients over the years.

I shall tell you that this lad's clinical picture is absolutely 'classic' for appendicitis, thus you would be correct if appendicitis was your **working diagnosis**. I ask you now:

**What tests would you like to do?**

This boy has classic findings of appendicitis. But you, having recently been steeped in the ways of the 'Temples of Accuracy' want confirmation! Maybe you are thinking that you cannot rely on the story you hear, or what your palpating fingers are telling you. What you wish for is some way which can tell you with 100% certainty that he has appendicitis. After all, the cure for appendicitis demands the child be cut open! If you're looking for "certainty" in your medical practice or in life, you will be disappointed. You must, as I say, get comfortable with uncertainty.

It doesn't mean that you're reckless or negligent, but rather that you must search for the solutions which have "the weight of evidence" supporting them. So, in terms of possible tests let's consider:

- A **CBC**? This is reasonable and inexpensive and can give you important evidence. *In this boy his white blood cell count (WBC) is elevated at 14 x $10^9$/l (normal=3.5–10.5 × $10^9$/L) and he has a noted 'left shift' on blood smear indicating high numbers of immature white blood cells.* This is the usual sort of WBC seen in appendicitis. However, easily 10% of people with acute appendicitis may have a normal WBC. A blood count is certainly <u>not</u> confirmatory.
- A **Urinalysis**? A 'dipstick' urinalysis costs less than one cent and can give you important clues about the state of hydration and the possibility of a urinary tract infection. It should be done routinely. *In this lad it is normal.*
- A **C-reactive protein**? CRP is a very non-specific test, gives little or no guidance and may easily be normal in cases of appendicitis—and, at current pricing costs about $20. Compare to a $5 CBC cost. This boy does not need his CRP measured.
- An **AXR?** So, let's say you think about doing an AXR— 2 views. No use there, unless you're lucky enough to see a calcified fecalith in the appendix, you cannot tell much about the appendix on a plain AXR. What about a CT scan? No good either because the amount of radiation a CT scan imparts on a child's abdomen has been shown to run a 0.1% risk of future cancer development. One tenth of one percent sounds like a small chance, and I suppose it is, but tell that to the boy's father. He won't want even that small chance of a future cancer for his son. We generally avoid, if possible, doing abdominal CT scans on children in cases of possible appendicitis, for that reason.
- Ahhh! **An abdominal ultrasound!** No radiation, and your radiologist is pretty good at it. You've seen how ultrasound can diagnose gallstones, hydronephrosis and fetal anomalies. You can get accurate confirmation of your working diagnosis of appendicitis that way, without any bad radiation.

**There's that word, "confirmation" again…**

It's important in situations like this to think a few steps ahead and consider some 'what if' scenarios. We know that ultrasound varies in accuracy from patient to patient and radiologist to radiologist in the detection of appendicitis. A reasonable estimate might be a 95% accuracy rate for a good radiologist to detect appendicitis. It's reasonable to think that 5% of the time the ultrasound is wrong. In this case then there may be a 5% chance the ultrasound will not detect appendicitis when appendicitis actually is the diagnosis (false negative).

Clinically, you believe this lad has appendicitis and needs an appendectomy. So, what if you get that ultrasound and it doesn't show appendicitis, will you be happy **NOT** operating on this boy? Appendicitis quickly goes on to perforation and sepsis in children and can actually cause death. In this case, a false negative ultrasound could be harmful, leading you away from the proper course of treatment for this boy.

Therefore, in this **<u>classic</u>** case the proper course of action is actually to proceed to operation <u>without</u> any imaging. Learn that ALL tests have a **finite accuracy rate**. ALL tests can have

false-negatives and false-positives. Learn, as you go, the finite, operational accuracy characteristics of the tests you are ordering and **think** of the 'what ifs'.

I am NOT suggesting that ultrasound has no place in the diagnosis of appendicitis. It is very useful in situations where the clinical picture is not clear. A properly performed and interpreted abdominal ultrasound can give you valuable evidence when you need it pointing towards or away from appendicitis. I just do not want you to think that ultrasound can "confirm" the diagnosis.

**Think**. That is all I ask. Whether you consider asking for an ultrasound, a C-reactive protein, a CBC, or whatever—**Think**. Tests can be invaluable guides and can help us immeasurably in clinical puzzles. Many children present with vague clinical findings only suggestive of appendicitis, and ultrasound can be tremendously helpful in those cases.

Tests should be ordered after careful thought. This case illustrates what I call…

**Blair's Four Rules of Testing:**

1. Do not order a test unless the results of that test will truly change your management.
2. Do not order a test if you do not know the tests inherent accuracies in the context of your patient, otherwise you will not know what the test result actually means for your patient.
3. Consider the risk of the test and whether the risk of testing is appropriate in the context of your patient's present and future health.
4. If you are going to order a test, order it properly.

Furthermore, as responsible medical doctors working in a health care system that has finite resources, it is vital that we understand our role in **Resource Stewardship**.

In this regard you should acquaint yourself with **Choosing Wisely Canada**

Check out this website:
http://www.choosingwiselycanada.org/

**Back to our patient…**

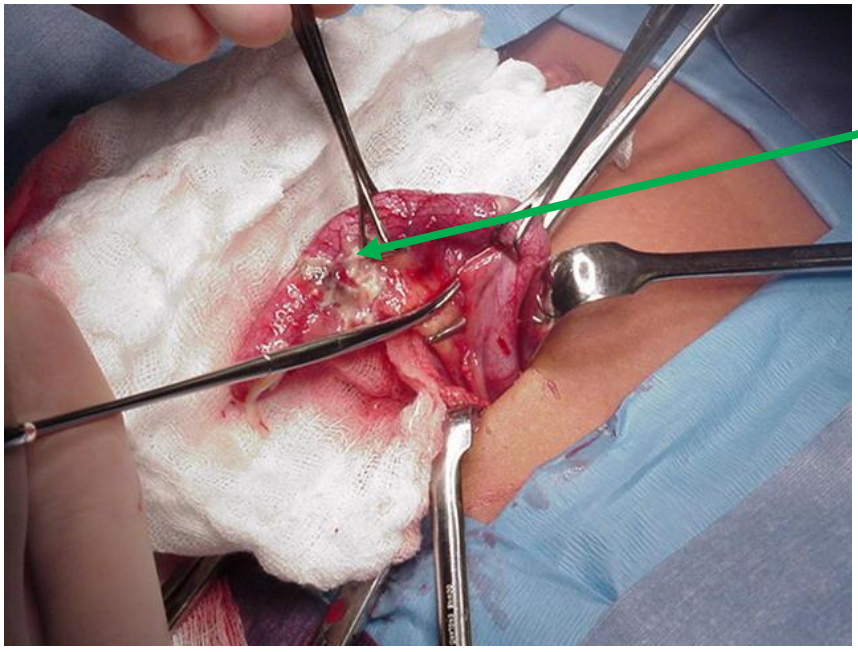
- *A urinalysis and CBC were done and the results were as noted.*
- *No imaging was done.*
- *An IV was started and because he was slightly volume depleted. A bolus of normal saline was administered. He was kept NPO.*

*Based on the history, the clinical findings, and with the added evidence of the elevated WBC, the parents were informed of the probable diagnosis of acute appendicitis; the possibilities of other diagnoses (e.g. Meckel's diverticulitis, Crohn's, etc.) including finding a normal appendix; the*

*advisability of an urgent laparoscopic appendectomy under a general anesthetic; the option (in this case not advised) of not operating and treating with IV antibiotics; and the possibility of complications such as post-operative wound or intra-abdominal infections, or intra-operative damage to other organs, bleeding, the need for conversion to an 'open' non-laparoscopic operation, anesthetic complications and other misadventures—all generally uncommon and not expected. Questions were invited. They gave their consent.*

*A prophylactic broad-spectrum antibiotic was administered intravenously within one hour of starting the operation and a laparoscopic appendectomy was performed under a standard general anesthetic. An inflamed, but unruptured appendix was found. No further antibiotics were administered. The child was given intermittent IV morphine for post-operative pain. The next day, he was well, eating and drinking, and was discharged home on acetaminophen for pain prn. He returned to school a few days later and was playing soccer with his friends one week later.*

Not all children have such a standard, classic case of appendicitis. The variants, the problems and the challenges of this common disease will continue to interest us for decades to come. Your expertise in diagnosing and managing this disease and all others will only come with experience and hours of study.



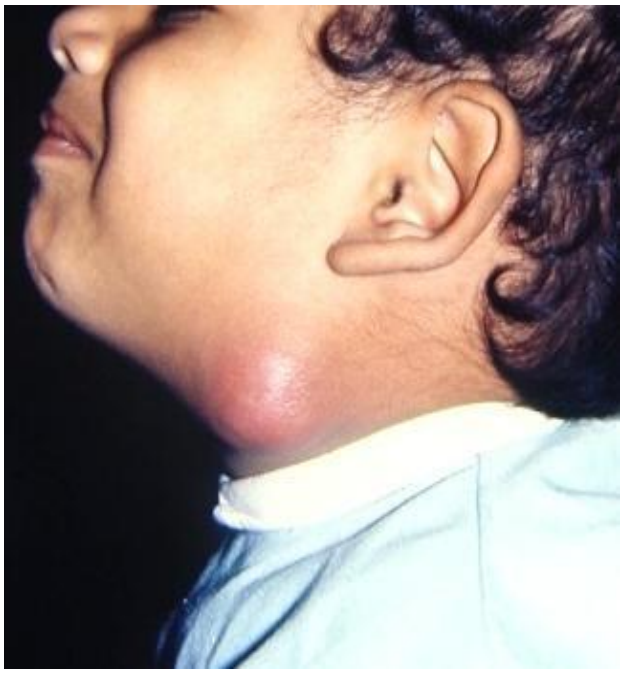
This photograph was taken during an open (i.e. not laparoscopic) appendectomy. The inflamed appendix here shows a small perforation in its mid-portion. These days, most appendectomies are done laparoscopically.

***Dr. Abraham Groves (1847-1935)—A Canadian surgical pioneer.***

*It was 1883 and the entity we know as appendicitis was not realized. At that time it was thought to be an inflammation of the caecum, and the prescribed treatment was to simply drain a RLQ abscess— if the patient was lucky enough to live that long. In that year, a country doctor/surgeon in rural Ontario, Dr. Abraham Groves, was about to drain a RLQ abscess in a very sick 12-year-old boy. He wrote, "On making the opening an inflamed appendix was found. This was removed by first ligating the organ at its origin and also the appendiceal mesentery, which was then cut through… On the third day, when I went to see the boy, he was doing well…the patient recovered." Dr. Groves modest account of this operation is now acknowledged to be the first intentional appendectomy, at least in North America, for the disease we now know as appendicitis. A Canadian surgical hero!*
*(My grandmother actually worked with him as one of his scrub nurses! I just had to tell you that!)*

It is important to be very familiar with inflammation and the many ways it may manifest.

Take a look at this toddler's neck. The swelling is obvious. What else can you say about the **lump**?



- It's red.
- The boy isn't very happy. It's painful.

I can tell you 2 other things that the picture cannot tell you-

The lump is palpably **warm**.
And the boy **isn't moving his neck well**.

This lesion demonstrates nicely the **5 basic attributes of inflammation** (in Latin!):

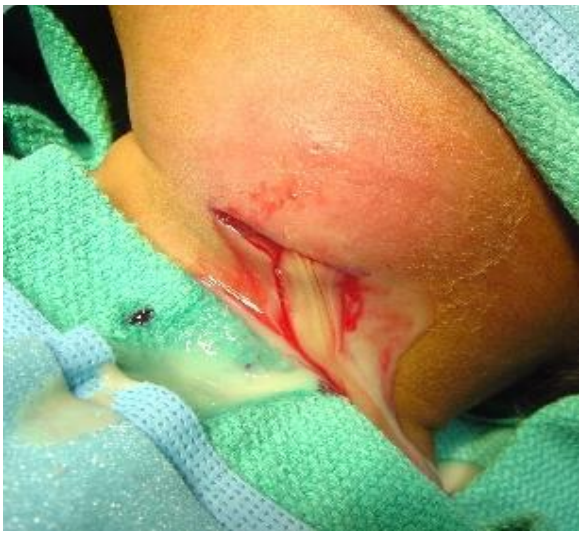
1. **Rubor**- redness
2. **Calor**- warmth
3. **Dolor**- pain
4. **Tumor**- swelling
5. **Functio laesa**- disabled function (his neck movement in this case)

What do think this lump is? Cellulitis? Lymphadenitis? An abscess?

Imagine you are in the produce section of the grocery store. You want to buy a ripe tomato to have in a salad that evening. You don't want one that is too ripe and mushy, nor do you want one that is hard, unripe and tasteless. You pick up a nice red tomato, and gently squeeze it. It's perfect.

You know what a good ripe tomato feels like. That's what a superficial abscess feels like.

That's what this lump feels like. Don't order an ultrasound. An ultrasound done for a superficial abscess often over-calls lumps as abscesses- before the abscess is 'ripe'. If you can pick a nice ripe tomato in the grocery store, you can diagnose a superficial abscess without the need for a 'test'.



**An abscess is a collection of pus**.
We can help infection resolve faster by draining the pus—an Incision & Drainage or "I & D". This basic surgical procedure has been around for literally thousands of years. Look for opportunities to do one! It's a bit gross, but very satisfying and helpful.
But you must wait till the abscess is 'ripe'. Doing an I & D too soon isn't helpful.
This is a photo of the pus drained from the boy's neck abscess.

Cervical abscesses like this are common in toddlers, usually caused by Staph or Strep infections of the cervical lymph nodes.

**Of course, there are many other inflammatory conditions of childhood that can come to the attention of a surgeon. Some have no apparent infective aspects, such as Crohn's disease, ulcerative colitis, thyroiditis, etc. In most cases the surgical management is similar to the adult management paradigms.**

## CHECKPOINT: INFLAMMATION/INFECTION

- Inflammation is usually our friend—a vital part of the immune system
- How does inflammation help the patient?
- Infection is the enemy.
- What do surgeons do to prevent infection?

CHECKPOINT CHALLENGE #10: LET'S SUPPOSE THAT OUR 11-YEAR-OLD PATIENT, IA, WAS INITIALLY MISDIAGNOSED AND TWO WEEKS LATER PRESENTED WITH A LARGE PELVIC ABSCESS SECONDARY TO THE RUPTURE OF HIS ACUTE APPENDICITIS MANY DAYS PREVIOUSLY. WHICH OF THE FOLLOWING THREE SYMPTOMS/SIGNS IS MOST COMMONLY SEEN IN PELVIC ABSCESSES:
☐FEVER ☐DIARRHEA ☐ PAIN

Checkpoint Challenge #11: The toddler with the neck abscess shown above presented without a fever or any other systemic signs. He undergoes an I&D procedure as in the photo shown. Does he require a course of antibiotics?

Checkpoint Challenge #12: Another toddler presents with a lump in the anterior midline of his neck. His parent's just noticed it and it seems slightly red and tender. Consider the possibilities.

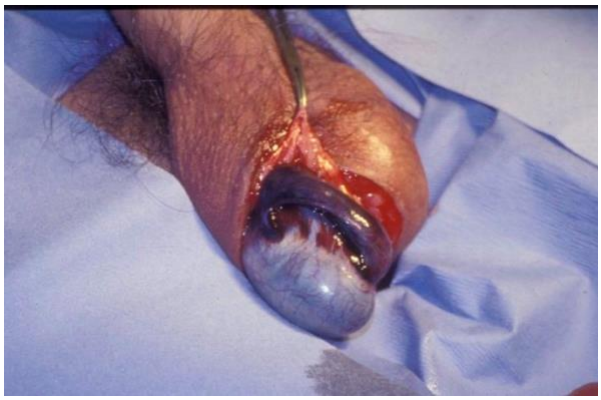
# ISCHEMIA
## NOBL FIIISTT

***Case IT:***
*13-year-old boy*
*CC- Acute scrotal pain X 3 hours*
*Started suddenly while watching TV*
*HPI- Previously healthy.*
*P/E Swollen and extremely tender right scrotum*

The **working diagnosis** for what we call the 'acute scrotum' must be **testicular torsion**. You don't have much time here. The testicle is suffering acute **ischemia**.

I was taught that when faced with an "acute scrotum" doing any tests like ultrasounds, Doppler studies looking for blood flow, etc. are only "things to do until the doctor arrives." You've got a **maximum of 6 hours** to save a testicle that is twisted. Three hours have passed since the pain started; another hour to check him into the ER; another hour to see him and examine him. Now you're up to 5 hours. He has got to go to the OR **now**!

This is another situation where you have to cope with uncertainty. Sometimes clinically you can determine that it is not testicular torsion, but rather epididymo-orchitis, or an acute hydrocele or hernia, or a torsion of the appendix testes—all of which are on a differential diagnosis list for the "acute scrotum". But unless you can be confident in establishing one those diagnoses, a trip to the operating room for a scrotal exploration is the safest thing to do.



Most cases of testicular torsion result from a lack of fixation of the testicle within the tunica vaginalis. This is called the 'bell clapper deformity' and allows the testicle to twist on its vascular pedicle. The result is as you see here.

*An urgent scrotal exploration was done and an acute torsion of the testicle was found and untwisted. 10 minutes later the testicle was nicely perfused. A typical bell-clapper deformity was found. A bilateral fixation orchidopexy was done to prevent either testicle from twisting in the future. He was discharged home a few hours later and did well.*

Remember case BV who presented with rectal bleeding and turned out to be suffering from an acute intestinal volvulus? I could've used BV as a case of ischemia too! The NOBL FIIISTT items overlap and are not necessarily mutually exclusive.

## CHECKPOINT: ISCHEMIA

- Time is of the essence when there is ischemia.
- How does venous occlusion lead to ischemia?
- How long after complete arterial occlusion do irreversible changes occur?

CHECKPOINT CHALLENGE #13: WHEREAS ADULTS OFTEN SUFFER ISCHEMIC HEARTS, BRAINS AND INTESTINES BECAUSE OF ATHEROSCLEROSIS, CHILDREN ARE NOT AFFLICTED BY THAT GENRE OF VASCULAR DISEASE. HOWEVER, A 6-YEAR-OLD LAD OF MIDDLE EASTERN DESCENT IS KNOWN TO HAVE SICKLE CELL ANEMIA AND PRESENTS WITH ACUTE ABDOMINAL PAIN AND A SEVERE DROP IN HIS HEMOGLOBIN LEVEL. IN WHAT WAY IS HE SUFFERING FROM ISCHEMIA?

# STRUCTURAL

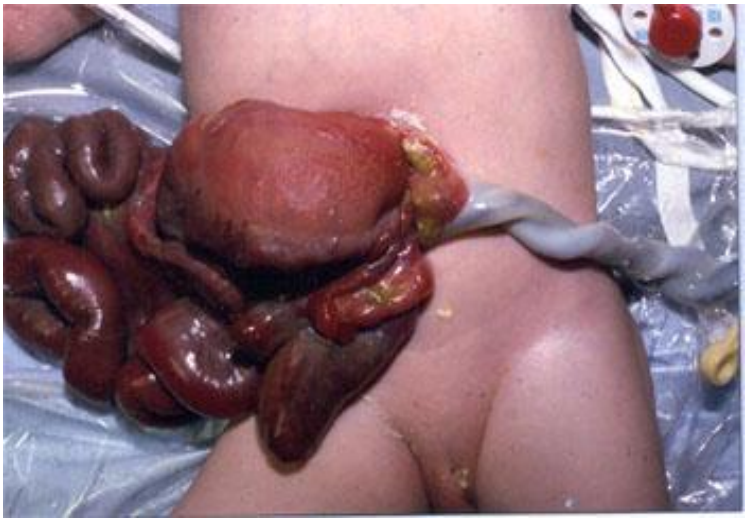
## NOBL FIIISTT

We are going to spend some time discussing "structural" problems. Much of Pediatric Surgery is dealing with the structural effects of embryology gone wrong. These structural pathologies may be common, as inguinal hernias are, or uncommon, as are the 1 in 5000 infants who are born with esophageal atresia.

## ABDOMINAL WALL HERNIAS

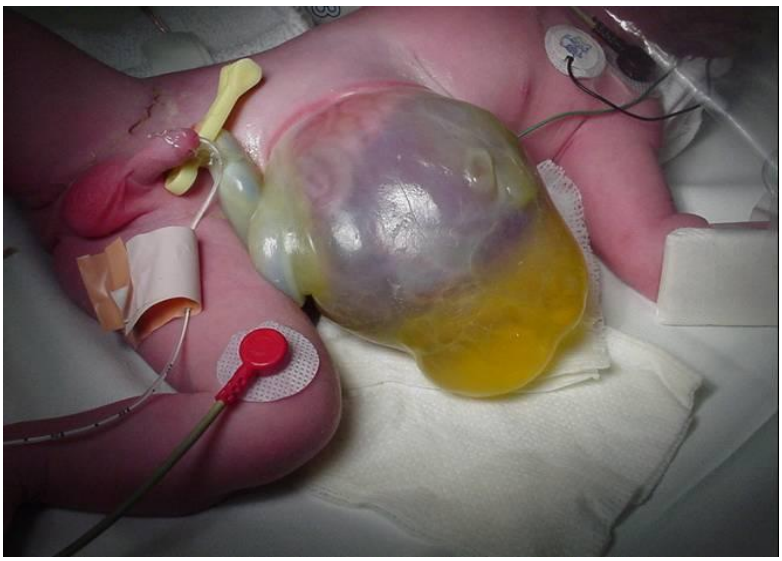
If we consider the definition of a "hernia" as "the protrusion of an organ or part of an organ or other structure through the wall of the cavity normally containing it", then it is reasonable to categorize hernias (or herniae) as "structural issues".

Let's begin with the dramatic:

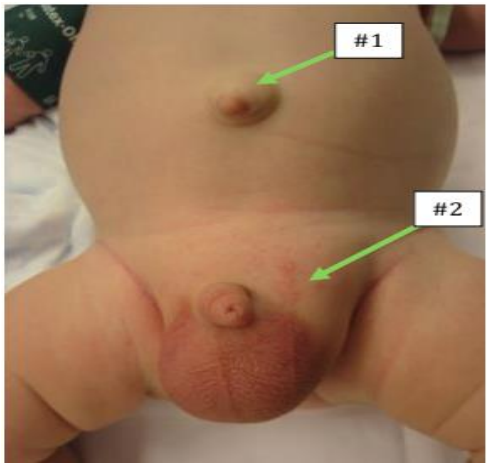


This is **gastroschisis**. This congenital problem can be diagnosed prenatally by fetal ultrasound. Characteristically the stomach, small bowel and colon are herniated through a defect that is to the right of the insertion of the umbilical cord. Although there are exceptions, most often it is only the GI tract which is herniated. Embryologically it is difficult to fully explain. Gastroschisis usually is an isolated anomaly, that is, the babies usually are anatomically normal otherwise. However, approximately 20% 0f gastroschisis babies will have an intestinal atresia.

This is an **omphalocele**. The abdominal organs, primarily bowel and liver herniate through a variably-sized central abdominal defect at the umbilicus. Unlike in gastroschisis, the guts in an omphalocele are covered by a thin membrane. Omphaloceles can also be reliably detected prenatally by ultrasound. 50% of omphalocele babies will have other major congenital defects- e.g. neurologic, chromosomal, cardiac.



In both gastroschisis and omphalocele, the baby needs to have the abdominal organs placed into their proper domain. In both situations, the abdominal cavity is congenitally small thus making it difficult to 'pack' the guts in! Most often we stage the reduction. In gastroschisis we place the bowels in a silastic 'silo' which is then squeezed daily for up to a week to gradually place the intestines back in the abdomen. In omphalocele, we must attend to any other congenital anomalies such as congenital heart disease that may compromise the child's health, but we take advantage of the fact that the defect is covered by a membrane. We 'paint' the membrane with an anti-infective ointment like silver sulfadiazine and dress the membrane protectively and gently compress the omphalocele. Over months, the organs fit into the abdomen, and eventually the fascia is surgically closed. Nutrition is tricky in both types of babies and often Total Parenteral Nutrition (TPN) must be given for weeks or months before the bowel is ready to do its job.



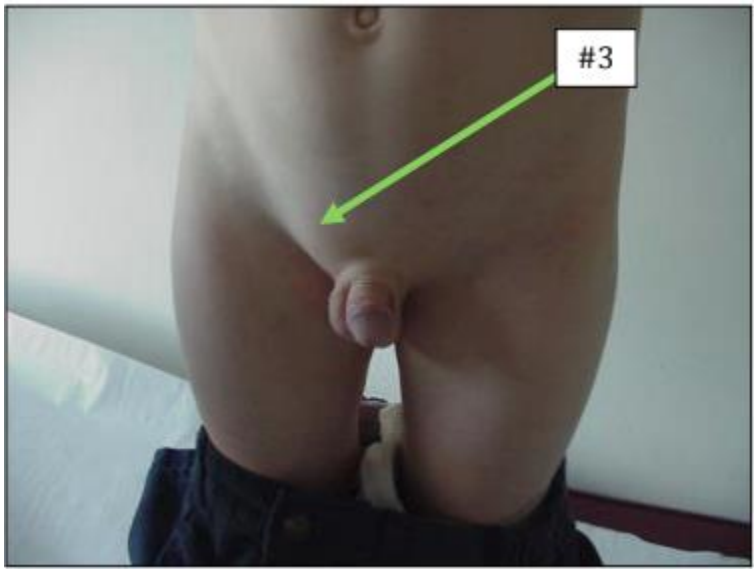

Now consider the following pictures. I have placed numbered arrows at areas I want you to consider.

**#1**- This is an **umbilical hernia**, which in the infant should really be regarded as a normal variant. The umbilical fascia comes together as a circular ring, but often children have a benign fascial opening here. When they cry or strain the umbilical skin will bulge- and this can alarm the parents.

It is non-pathologic and holds no risk of incarceration (wherein the bowel gets entrapped and ischemic). It does not cause pain, but when babies cry with infantile colic the resultant umbilical bulge is often blamed. Taping or binding them or taping coins over them are all traditional methods of treatment—all are ineffective and contraindicated. Usually these umbilical hernias only start to close at age 1 year.

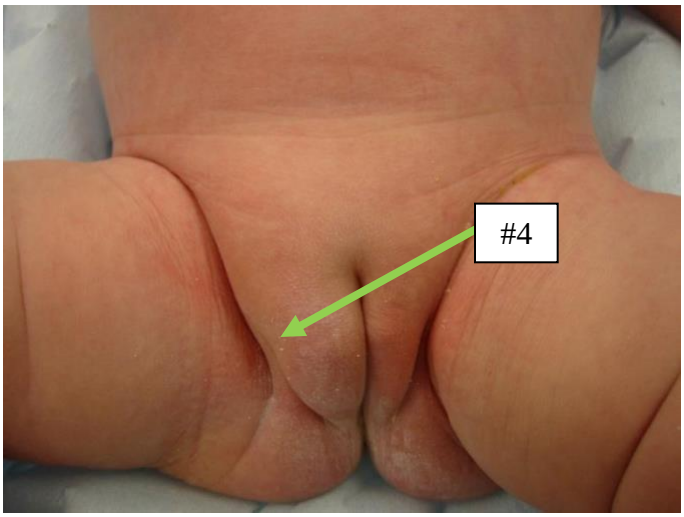
Most close on their own and parental reassurance is the key. A few persist beyond age 5 years. We tend to repair these in school-age children, and generally not before.

**#2-** This is an **indirect inguinal hernia**. They are common, occurring in about 1-2% of boys and 0.5% of girls. They can appear in early infancy and are especially common in premature babies. It represents a congenital patent processus vaginalis, which is supposed to close prior to birth. The protrusion is at the internal inguinal ring. (Now's a good time to review inguinal anatomy!) A hernia 'sac' follows down the inguinal canal towards the scrotum. Parents typically are the first to notice these bulges which tend to come and go over the day. Twenty percent present as bilateral hernias. Usually hernias contain bowel and are reducible, i.e. can be pushed back in place. However, some may **incarcerate,** which is characterized by an irreducible bulge, pain, tenderness and eventually signs of bowel obstruction. A **strangulated** hernia is when the incarcerated bowel within the hernia becomes ischemic, as well as the testicle, since bowel incarceration in an indirect inguinal hernia also occludes the adjacent testicular vasculature. Repair should be undertaken shortly (i.e. ideally within a few weeks) following a hernia's detection to prevent possible incarceration. Incarceration of an inguinal hernia is a serious emergency and repair is technically difficult.




**#3** This too is an **indirect inguinal hernia**. Even though they are congenital, often the patent processus vaginalis is very small in infancy and hidden from clinical view. As the child ages the patency enlarges until it manifests as an inguinal bulge. Again, it's very often the parents who notice it first. Early on the hernia is asymptomatic, but with time will cause pain. Often in its early stages of enlargement it is only visible when the child is standing as in this picture.

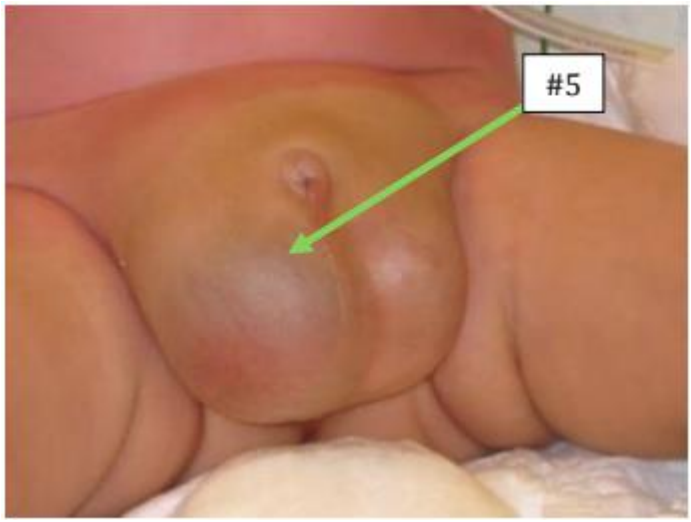
Ultrasound examination is surprisingly inaccurate and unreliable, and we actively **discourage** its use in hernia detection and diagnosis. The diagnosis is purely clinical. Surgical repair, which is a tying off of the patent processus vaginalis, should be booked shortly after clinical diagnosis. Parents often assume that we have to use mesh in the repair because they have heard of, or experienced, a mesh repair of hernias in adults. Mesh is not required in children as the posterior inguinal wall, where adults have a direct inguinal hernia, is generally intact and strong.




**<u>#4</u>** Girls can get **indirect inguinal hernias** as well.

They are less common than in boys but have the added danger of not only having bowel incarcerate in them but also an ovary. Both may strangulate. Early repair is indicated as it is in boys reasonably soon after the clinical diagnosis is made.

**<u>#5</u>** Is this an **indirect inguinal hernia** or a **hydrocele**? If it's a hydrocele is it a communicating one or a non-communicating hydrocele?

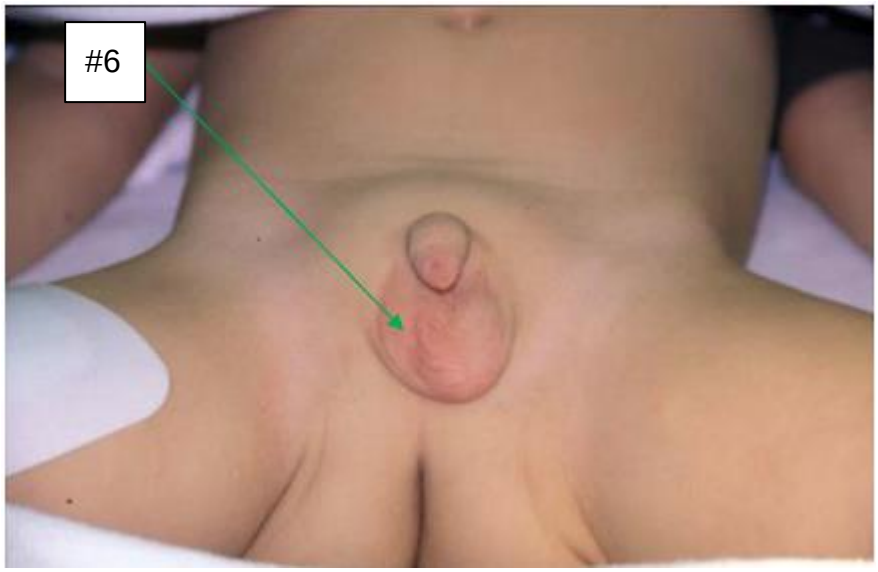



Clinically, hydroceles are generally painless in babies and children. A hernia will bulge in and out. Communicating hydroceles also can often vary in size dramatically over the period of the day. Non-communicating hydroceles tend to be static in size. Palpating a hernia usually reveals bowel that can be appreciated and reduced with a palpable 'gurgle'. Hydroceles, even the communicating ones, won't manually reduce. Most often you can palpate above the hydrocele and appreciate normal cord structures. With some practice in hernias, you can often feel the hernia sac, as a thick portion of the cord. Hydroceles easily **transilluminate**, but hernias do not- provided that you don't use a very powerful light which can shine through bowel in a darkened room and mislead you. Once again, ultrasounds are essentially <u>useless</u> as a means of assessment of hernias and childhood hydroceles.

Case #5, by the way, is **a communicating hydrocele**. Its fluid tends to create a bluish colouration to the scrotal sac (refer to optical physics:'Rayleigh scattering'). By and large, because they are so benign, we tend to simply 'watch' hydroceles in infants, both communicating and non-communicating. However, we must realize that a communicating hydrocele is potentially an indirect inguinal hernia in evolution, because it represents a narrow patent processus vaginalis. Older children who may suddenly have a hydrocele appear usually

will prove to have a small patency to their processus vaginalis. Elective repair is usually indicated.

**#6 Where's his right testicle?**
The best term for this condition is cryptorchidism, in this case, right-sided. Cryptorchidism means "hidden testicle". The following are possibilities (**1 and 5 are most common**).




1. The testicle is **undescended**. The testicles descend in early fetal life from their site of formation in the abdomen. Most often it will stall in the inguinal canal, but sometimes high in the abdomen.
2. His right testicle was descended but **atrophied** because of a newborn or fetal testicular torsion, which from time to time we see, or for reasons of supposedly inadequate vasculature.
3. **Agenesis** of his right testicle.
4. **Ectopic** right testicle. The testicle descended but veered off to lodge in the upper thigh or lower abdominal wall.
5. It's a **retractile** right testicle. It's in the scrotum when he's calm and warm, but when he is upset or in cooler air the cremasteric muscle pulls it up into the inguinal canal.

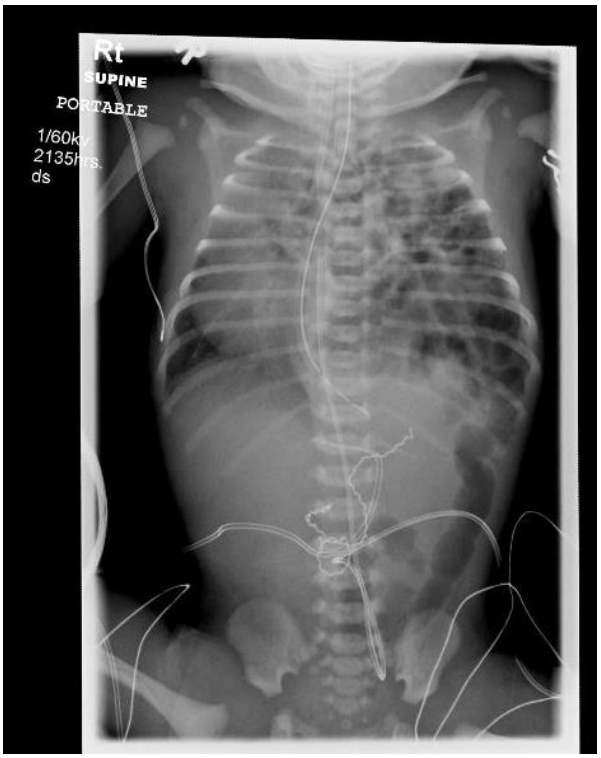
A retractile testicle can be manually brought down into the scrotum and when released should stay there for at least 10 seconds. Think of it as an overactive cremasteric reflex. Retractile testicles are absolutely benign. Reassurance is all that is needed—as long as you are sure. Follow-up at least once in 6-12 months is a good idea.

An undescended testicle should be brought down. If the testicles have not descended by 6 months of age they aren't going to descend and an **orchidopexy** to fix the testicle in the scrotum should be done- ideally by age 1 year.

What do you think my advice regarding the use of ultrasound in cryptorchidism is?
That's right- it's generally useless! It seems that ultrasound should be able to find a hidden testicle, but studies have shown **significant inaccuracies**—revealing testicles when surgery has shown no testicle. Or where ultrasound has failed to find a testicle and surgery then finds it, simply undescended, in the inguinal canal. Do not order ultrasound scans for undescended testicles—you'd be wasting health care funds and quite possibly it may lead to a misdiagnosis.

Back to some less common, but very serious structural diseases we deal with in babies and children. By the way, both of the following entities have an incidence of about 1 in 5000 live births.

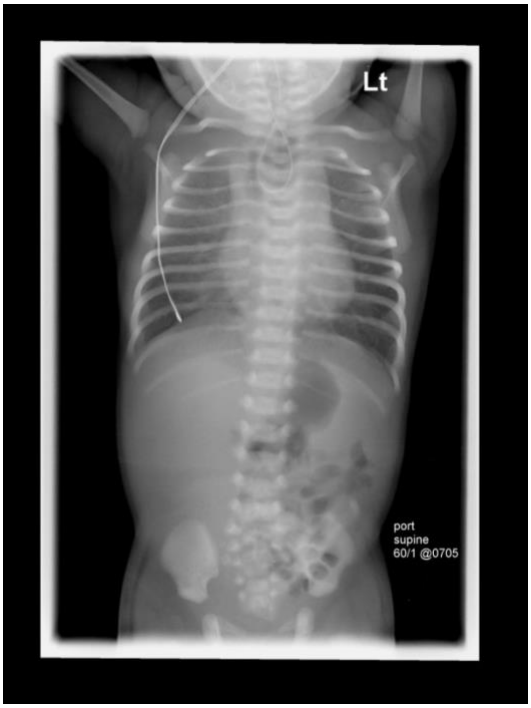
## CONGENITAL DIAPHRAGMATIC HERNIA



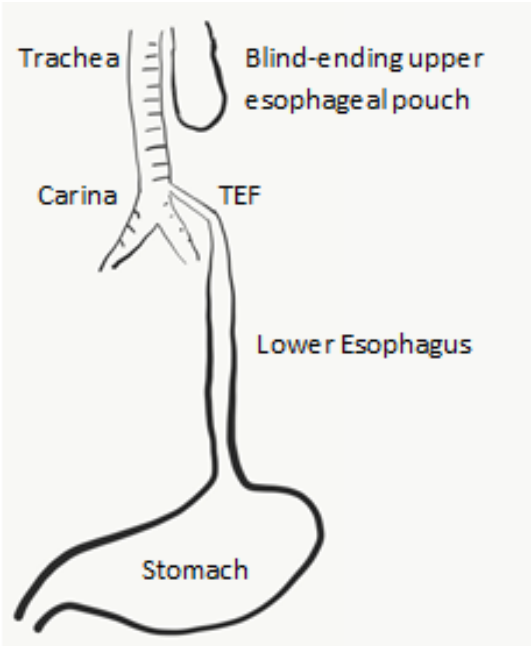

If the diaphragm doesn't form correctly, then as the fetus develops, the developing lungs have to compete for space with the developing bowels. (Check out this newborn's CXR- all those gas-filled bowel loops in the left chest crowding his little lungs!) The lungs lose, and children who are born with diaphragmatic hernias have small lungs, especially on the side where the diaphragmatic hole or 'defect' is. This is most often on the left side in the posterior-lateral aspect- the so-called Foramen of Bochdalek (hence you may hear the term "Bochdalek hernia"). These infants tend to have significant respiratory distress in the first minutes of life, with cyanosis. There'll be little, or no breath sounds on the herniated side, and the abdomen will be scaphoid, rather than the chubby abdomen that babies usually have. In the old days, we would operate immediately to open the abdomen and pull the guts out of the chest. We thought that would allow the lung to expand and alleviate the baby's respiratory distress. It took us decades to realize that didn't work! The lungs, especially on the side of the hernia, are congenitally small, and so is the baby's pulmonary vascular bed. Remember how a normal baby's pulmonary vascular pressure drops precipitously upon the child's first breath? Because of the small pulmonary vascular bed (and other things) this pressure drop cannot happen in these babies. No wonder so many of these babies died in the old days.

Now, we are smarter. We delay operating on these babies and support them for a few days in the ICU until their **persistent fetal circulation** and persistent **pulmonary hypertension** is somewhat improved. Take time to review the remarkable physiologic changes that happen at birth. This disease is an excellent example of why you need to know all that anatomy and physiology you learned earlier in medical school.

## ESOPHAGEAL ATRESIA




Recall duodenal atresia? Embryologically, atresia may occur in any hollow organ and the esophagus is no exception. So, think of a baby with a blocked esophagus. They cannot swallow, so they appear like they are frothing at the mouth minutes after birth, and suctioning doesn't seem to resolve it. We say these babies are "mucousy". If you try to insert an NG tube you can't. It will stop about 10 or 11 centimeters from the nares or curl in the upper 'pouch' of the atretic esophagus as it does in this x-ray. You have to continue to suction them while you get an urgent CXR.
Sometimes the CXR will show a 'gasless' abdomen, which makes sense, because if they can't swallow their saliva, then they can't swallow air either, and it's swallowed air that we see as gas in the bowel. However, in most cases of esophageal atresia, the CXR will show gas in the bowel as it shows in the CXR here. Why is that?




This diagram shows the usual disordered anatomy in most cases of esophageal atresia. You can see there is a distal trachea-esophageal fistula (TEF*). This TEF allows gas from the trachea to get into the GI tract with every breath.

So, if the child can't swallow and there is a TEF, think about all the bad things that can happen.

We Pediatric Surgeons love to fix esophageal atresia which is done surgically through the chest by means of a thoracotomy or using a minimally invasive approach, thoracoscopically. The results usually are good. Remember though, when there is one congenital anomaly, look for others.

**In Britain the acronym is TOF —trachea-oesophageal fistula*

**Remember the acronym: VACTERL –** Each letter stands for anomalies that tend to cluster with the others:

- **V**ertebral defects
- **A**nal atresia
- **C**ardiac defects
- **T**racheo-**E**sophageal fistula/atresia
- **R**enal anomalies
- **L**imb abnormalities

Three or more of these together is called "VACTERL syndrome".

## CHECKPOINT: STRUCTURAL DISEASE:

- Much of Pediatric Surgery treats congenital structural disease (abdominal wall defects, hernias, atresias, etc.)
- An understanding of embryology, embryopathology and pathophysiology is helpful.
- If you detect one congenital anomaly, then look for others!

CHECKPOINT CHALLENGE #14: A 2-YEAR-OLD BOY IS NOTED TO HAVE A RIGHT INGUINAL HERNIA. ON FURTHER EXAMINATION YOU NOTE A RIGHT UNDESCENDED TESTICLE AS WELL. THE LEFT TESTICLE AND GROIN IS NORMAL. BRIEFLY OUTLINE A PLAN FOR THIS LAD.

CHECKPOINT CHALLENGE #15: ONE WEEKEND A NUMBER OF YEARS AGO, I SAW 4 NEWBORNS WITH ESOPHAGEAL ATRESIA. (SO MUCH FOR THE SUPPOSED 1 IN 5000 INCIDENCE!). ONE OF THE BABIES HAD A LARGE OMPHALOCELE IN ADDITION TO HIS ESOPHAGEAL ATRESIA. IN THAT BABY WHAT WOULD BE THE PRIORITIES OF OPERATIVE MANAGEMENT?

# TRAUMA/TISSUE DAMAGE

## NOBL FIIISTT

**Children are not just small adults!**

That is a familiar refrain from Pediatric Surgeons, imploring our colleagues not to treat kids in the same way adults are treated. The set of pathologies and disease that afflict children, as you have learned, are very different from the inflictions of adulthood. Children physiologically, emotionally and anatomically can react very differently to disease processes as compared to adults. Yet we have important principles of management that are the same no matter what the age of the patient. One of these is the initial approach to the traumatized patient. The ABC's…

1. **A**-AIRWAY
2. **B**-BREATHING
3. **C**-CIRCULATION
4. **D**-DISABILITY
5. **E**-EXPOSE

Let's consider some cautionary cases that highlight the risks of hidden injury in children.

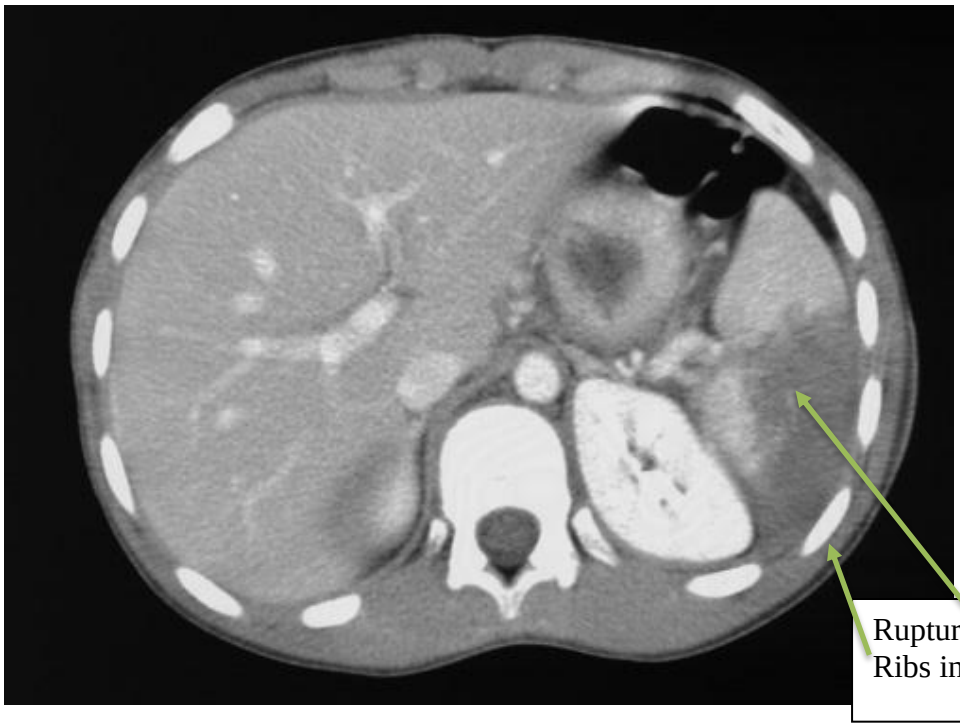
***Case TS:***
*10-year-old girl, previously well, tripped while walking across a parking lot falling across a cement curb and landing on her left side. Four hours later she complains of abdominal pain, and left shoulder pain. She was unable to sleep that evening and because of the continued pain was brought into the local ER- 8 hours after the injury.*
*P/E: HR-135 BP-100/60 RR-24 T-37.3*
*Airway & Breathing- normal*
*Abdo tenderness in LUQ and epigastrium with guarding.*
*Shoulders and limbs assessed as normal*
*No visible bruising*
*CXR- Normal. No rib fractures*

It's hard to imagine that a simple trip with no resultant bruising or fractures could be anything serious.

*It was felt that the slight tachycardia, the abdominal tenderness/guarding coupled with Kehr's sign* indicated the need for a CT scan.*

***CT Scan results:***

*Kehr's sign perhaps should be called Kehr's symptom. It is a classic **referred pain** situation wherein blood (or free air) irritating the underside of the diaphragm is interpreted as C3,4,5 dermatome pain.




*The CT scan indicated a ruptured spleen with minimal intraperitoneal blood. No rib fractures. No other injuries apparent.*

The CT scan was a good call. A FAST ultrasound probably would have been **falsely negative**. FAST has not shown to be as reliable in children as in adults. Even a detailed ultrasound scan could have missed this ruptured spleen!

Children do not have the muscle bulk and rib stiffness that protects the spleen and liver in adults. A relatively low kinetic energy hit can result in solid organ damage in a child without concomitant rib fractures. Be suspicious of 'occult' **solid organ injuries** such as spleen, liver and kidney damage in children with relatively minimal injury. We don't like irradiating children, but in pediatric trauma, a CT scan is often necessary.

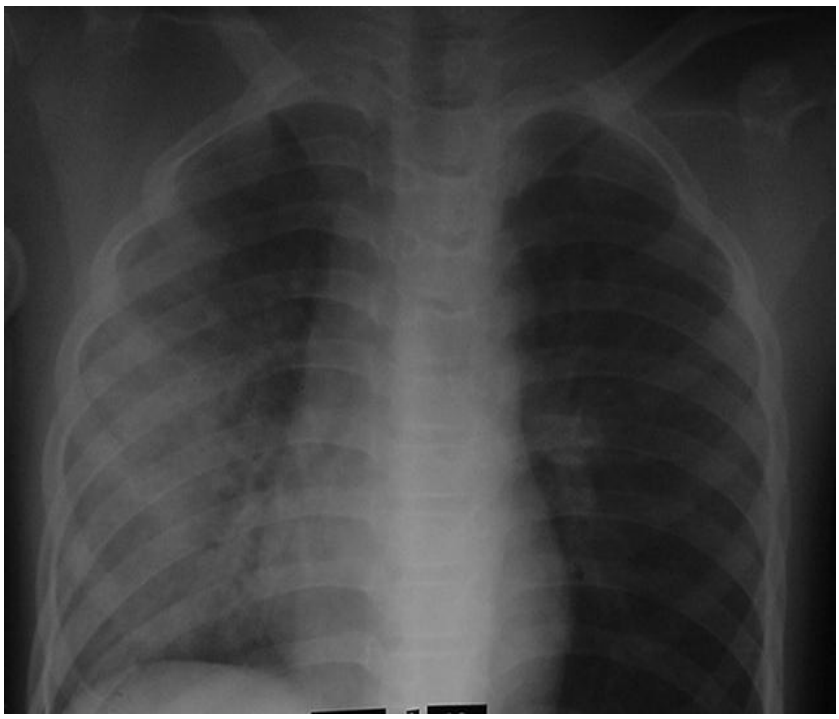
***Case TC:***
*6-year-old boy, previously healthy, was playing on the driveway when he was inadvertently run over by his father backing up his van.*
*He was extracted immediately and rushed to the nearest ER.*
*P/E vital signs all within normal limits. $SaO_2$ 97% on room air (normal)*
*Airway normal. Breathing normal, chest clear but a tire mark is evident across his chest.*
*Face and conjunctiva revealed prominent multiple petechiae.*

*You decide to do a CXR*

***CXR report-*** *No evident lung parenchymal changes. No rib fractures. Mediastinum normal. Normal CXR.*
How upsetting it was for the father! But how lucky for the child that he apparently suffered no injuries!

Should he be sent home?
**No way!**

*4 hours later while still under observation, his $SaO_2$ dropped to 75% (low). A repeat CXR was done after placing him on $O_2$.*



***CXR report-*** *Significant lung parenchymal changes consistent with traumatic lung contusion.*

*He required intubation and ventilation with high $FiO_2$. He was extubated 3 days later and was discharged home fully recovered, a week later. No sequelae.*

In the context of this sort of injury, the multiple facial petechiae is termed **traumatic asphyxia.** This occurs when the chest is crushed under a weight and the glottis is closed causing a sudden rise in thoracic pressure and CVP which in turn causes capillary bursting in the head and neck region.

This case, like the ruptured spleen case, illustrates that significant internal organ damage may occur in children despite no, or little, outward evidence of significant injury. You must learn to be **suspicious**. Consider the mechanism of injury. Consider the 'soft shell' of a child's body. Look for subtle signs indicating deeper trouble. Realize that a significant injury in child may be delayed in presentation. For instance, a child often will maintain a normal blood pressure despite significant hypovolemia/blood loss until just prior to their catastrophic plunge into hypotension, shock and death. A child's apparent stable physiology can be deceptive. Often, it's best to err on the side of admitting and monitoring, 'just in case'.

**"Much of the time my job as a Pediatric Surgeon is watching and worrying…"**

We have discussed how children can harbour significant injuries. Sadly, probably more often than we realize, even the mechanism of childhood injury can be obscure to us. **Child abuse and non-accidental injury** is something we must always be wary of. The perpetrators of these crimes may act out of anger, impulse, frustration or mental illness. They try to hide their crime. If they are the parents, they will sometimes offer plausible stories of how their child got injured. Very young children, of course, cannot give their side of the tale and older children might be fearful of telling the truth.

I have found in many instances that any of the following '**3 suspicions'** may be the hint that the child was the victim of non-accidental trauma:

- Suspicious History
- Suspicious Parents
- Suspicious Findings

It may be a story of injury that somehow just doesn't make sense. Or perhaps your suspicions may arise when parents have inconsistencies in their story. Strangely some abusive parents will appear almost too loving and concerned, as though they are trying to over-compensate for their regretful abuse of the child. There may be a strange pattern of burn injuries or bruising to a child's body.

If you even slightly suspect non-accidental trauma then you are ethically and legally bound to call in a Child Abuse team. These individuals are trained to detect and deal with the situation in a sensitive manner. Do not feel badly if you have sounded an alarm that turns out to be false—it's better than being suspicious and not blowing the whistle. Next time the child may be… you know…

"So, what about **Tissue damage**?", you may ask. Trauma is often defined as injury caused by a kinetic force, like a car hitting a child. However, children can suffer, just as adults do, burns or electrical injuries which are the result of thermal or electrical forces acting on tissue. Granted, we perhaps could lump it all under 'Trauma' but since burns and electrical injuries have a different set of problems I added the '/Tissue damage' moniker. Just as with trauma, remember in burns and electrical injuries children are not just small adults.

## CHECKPOINT: TRAUMA/TISSUE DAMAGE

- Why are children more vulnerable to traumatic injury?
- Children are not just small adults!
- Think child abuse when you wonder about:
    - Suspicious History
    - Suspicious Parents
    - Suspicious Findings

CHECKPOINT CHALLENGE #16: A YOUNG GIRL AND HER MOTHER WERE VICTIMS AS PEDESTRIANS OF AN APPARENT HIT & RUN ACCIDENT ON A RESIDENTIAL SIDE STREET. THEY WERE BOTH FOUND UNCONSCIOUS ON THE ROAD BY A PASSER-BY WHO QUICKLY CALLED 911. THEY WERE BOTH BROUGHT INTO THE ER IN ONE AMBULANCE. THE MOTHER'S TEMPERATURE ON ADMISSION WAS 37$^{O}$ BUT THE CHILD'S TEMPERATURE WAS ONLY 35.9$^{O}$. WHY?

CHECKPOINT CHALLENGE #17: A 1-YEAR-OLD BOY HAS A SECOND-DEGREE SCALDING INJURY TO HIS ENTIRE RIGHT LEG. WHAT PERCENTAGE OF HIS BODY SURFACE AREA IS BURNT?

**Well done! We are finished. (Unless you still are going back and forth from the Checkpoint Challenges and the Answer Hints in the appendix, in which case, carry on!)**

We've gone over a lot of material. I hope it's made you think. I hope it's prompted a curiosity in you to learn more. I hope you're keen to get out there, to help some children and be the best doctor you can be.

Good luck!

# APPENDIX (BUT NOT THE KIND THAT YOU REMOVE!)

## ANSWER HINTS FOR THE CHECKPOINT CHALLENGES WITH SOME "FYI" REFERENCES.

*(THE CHALLENGES IN THE PRIMER BODY ARE REPEATED HERE FOR YOUR CONVENIENCE)*

***CHECKPOINT CHALLENGE #1**: A MOTHER BRINGS HER 1-YEAR-OLD GIRL TO YOUR CLINIC AND SAYS THAT HER BABY HAS BEEN STRAINING AT STOOL A LOT RECENTLY. YOU GATHER A COMPLETE HISTORY AND DO A THOROUGH PHYSICAL EXAMINATION—WHICH INCLUDES A DIGITAL RECTAL EXAM. (YES- APPROPRIATE EVEN IN BABIES!) YOU FEEL A HARD, IMMOBILE 'LUMP' ABOUT 3 CM. IN DIAMETER POSTERIOR TO THE ANORECTUM IN THE REGION OF THE TIP OF THE COCCYX. YOUR THOUGHTS?*

—YOU MIGHT INITIALLY THINK THAT THE HARD LUMP YOUR DIGIT IS FEELING IS SIMPLY HARD STOOL, BUT IN THIS CASE IT IS AN IMMOBILE MASS. FECAL MATTER, NO MATTER HOW HARD, IS ALWAYS SOMEWHAT MOBILE. NOW, YOU MIGHT HAVE HEARD OF "SACROCOCCYGEAL TERATOMAS" WHICH, MOST OF THE TIME, ARE BIG (USUALLY HUGE!) TUMOURS EASILY SEEN ARISING FROM THE COCCYX IN NEWBORNS. THEY HAVE REAL MALIGNANT POTENTIAL. SAY IF A CHILD IS BORN WITH A VERY SMALL TERATOMA ARISING FROM THE SAME AREA THAT DOESN'T MANIFEST UNTIL, SAY, ONE YEAR OF AGE?
IMPORTANT MESSAGES: A DRE IS INDICATED, AND NOT TO BE AVOIDED WHEN CHILDREN HAVE STOOLING ISSUES & ALTHOUGH "COMMON THINGS ARE COMMON" THE UNCOMMON DOES COME ALONG!

FYI—PHI JH. SACROCOCCYGEAL TERATOMA : A TUMOR AT THE CENTER OF EMBRYOGENESIS. J KOREAN NEUROSURG SOC. 2021 MAY;64(3):406-413. DOI: 10.3340/JKNS.2021.0015. EPUB 2021 APR 29. PMID: 33906346; PMCID: PMC8128526.

***CHECKPOINT CHALLENGE #2**: A 7-YEAR-OLD BOY PRESENTS TO THE ER WITH AN 18-HOUR HISTORY OF INTERMITTENT CRAMPY ABDOMINAL PAIN AND PASSAGE OF DARK RED STOOL. A THOROUGH HISTORY AND PHYSICAL EXAM COUPLED WITH TWO-VIEWS OF HIS ABDOMEN AND AN ABDOMINAL ULTRASOUND POINT TO AN INTUSSUSCEPTION AS BEING THE UNDERLYING CAUSE! THIS IS CERTAINLY OUT OF THE USUAL AGE RANGE FOR INTUSSUSCEPTION. WHAT IS GOING ON?*

—REMEMBER THE USUAL AGE RANGE FOR INTUSSUSCEPTION IS AGE 3 MONTHS TO 3 YEARS. IN THAT AGE RANGE THE INTUSSUSCEPTIONS ARE USUALLY WHAT WE SOMETIMES CALL "IDIOPATHIC" EVEN THOUGH WE BELIEVE THAT THEY PROBABLY OCCUR BECAUSE A "PEYER'S PATCH" ACTING AS A LEAD POINT. MAYBE "LEAD POINTS" IS A KEY TO THIS 7-YEAR-OLD'S INTUSSUSCEPTION?

FYI— HTTPS://WWW.MERCKMANUALS.COM/EN-CA/PROFESSIONAL/PEDIATRICS/GASTROINTESTINAL-DISORDERS-IN-NEONATES-AND-INFANTS/INTUSSUSCEPTION

***CHECKPOINT CHALLENGE #3****: LET'S SAY THE RADIOLOGIST LOOKING AT THE MECKEL'S SCAN ON PATIENT BW SAYS IT IS "EQUIVOCAL"—WHAT THEN?*

—OOOOH… TOUGH ONE! SO, YOU THINK THERE ARE PROBABLY OTHER WAYS TO DETECT A BLEEDING MECKEL'S DIVERTICULUM, IF THAT IS, INDEED, WHAT THE PROBLEM IS HERE. BUT DO YOU KEEP ON TESTING. TESTING AND TESTING? OR AT SOME POINT DO YOU SAY, "I REASONABLY BELIEVE THAT THE CAUSE OF THE GI BLEED HERE IS A MECKEL'S DIVERTICULUM AND THEREFORE IT'S REASONABLE TO PROCEED TO…"

FYI—SHARMA RK, JAIN VK. EMERGENCY SURGERY FOR MECKEL'S DIVERTICULUM. WORLD J EMERG SURG. 2008 AUG 13;3:27. DOI: 10.1186/1749-7922-3-27. PMID: 18700974; PMCID: PMC2533303.

***CHECKPOINT CHALLENGE #4****: A 12-YEAR-OLD GIRL PRESENTS WITH A HISTORY OVER THE PAST YEAR OF A NUMBER OF EPISODES OF INTERMITTENT ABDOMINAL PAIN, NAUSEA, AND ON TWO OCCASIONS SHE HAS HAD GREEN EMESIS. ON EACH OCCASION THE SYMPTOMS HAVE SUBSIDED SPONTANEOUSLY. THERE IS NO OTHER PERTINENT HISTORY. PHYSICAL EXAMINATION, INCLUDING HER ABDOMEN, IS ENTIRELY NORMAL. SHE UNDERGOES INVESTIGATIONS WHICH INCLUDE A NORMAL ENDOSCOPY BUT AN UPPER GI SERIES SHOWING INTESTINAL MALROTATION WITH NO OBSTRUCTION.*

— REMEMBER, "BEWARE THE CHILD WHO VOMITS GREEN!". IT'S GOOD THAT SHE HAD THAT UPPER GI CONTRAST STUDY. COULD IT BE THAT WITH HER NOW KNOWN INTESTINAL MALROTATION SHE AS AN 'INTERMITTENT' VOLVULUS? THE PROBLEM WITH INTESTINAL MALROTATIONS IS THAT THE BOWEL MESENTERY DOES NOT HAVE ITS USUAL 'TWO-POINT FIXATION' TO THE RETROPERITONEUM (AT THE DUODENO-JEJUNAL JUNCTION, A.K.A. THE "LIGAMENT OF TREITZ" AND THE RETROPERITONEAL ATTACHMENT OF THE CECUM). THIS LACK OF FIXATION ALLOWS THE INTESTINES TO, AT TIMES ROTATE, I.E. A VOLVULUS.

FYI—CHECK THIS OUT! THE GOVERNOR OF NORTH CAROLINA HAS A MESSAGE FOR YOU: HTTPS://FILES.NC.GOV/GOVERNOR/DOCUMENTS/FILES/INTESTINAL-MALROTATION-AND-VOLVULUS-AWARENESS-DAY.PDF

***CHECKPOINT CHALLENGE #5****: THE 15 YEAR-OLD GIRL, LP, WHO WAS TREATED FOR HER SPONTANEOUS PNEUMOTHORAX, THE DAY AFTER HOSPITAL DISCHARGE FLEW TO FLORIDA TO VISIT HER AUNT. SHE ALMOST DIED ON ROUTE WHEN AT 38,000 FEET IN THE JETLINER SHE SUFFERED SEVERE RESPIRATORY EMBARRASSMENT. THE PLANE MADE AN EMERGENCY LANDING IN TULSA, OKLAHOMA, WHERE PARAMEDICS DECOMPRESSED A RECURRENT RIGHT PNEUMOTHORAX. WHY DID ALL THAT HAPPEN? IMAGINE YOU ARE ON THAT PLANE AND THE CAPTAIN CALLS FOR MEDICAL HELP—WHAT WOULD YOU DO?*

THINK ABOUT, OR LOOK UP, BOYLE'S LAW (OR WHAT HAPPENS TO YOUR BAG OF POTATO CHIPS ON AN AIRPLANE?). WHAT SHOULD SHE HAVE BEEN TOLD WHEN DISCHARGED FROM HOSPITAL? INSOFAR AS WHAT YOU SHOULD DO IF YOU WERE CALLED TO HELP HER ON THAT PLANE, TAKE A LOOK AT THE ARTICLE CITED HERE. BY THE WAY, THERE IS A GOOD CHANCE THAT SOMETIME IN THE FUTURE YOU'LL BE CALLED TO ADDRESS AN INFLIGHT MEDICAL EMERGENCY.

FYI—FLATT AE. A FLIGHT EMERGENCY. PROC (BAYL UNIV MED CENT). 2009 JAN;22(1):24-5. DOI: 10.1080/08998280.2009.11928464. PMID: 19169395; PMCID: PMC2626355.
&— HTTPS://THORAX.BMJ.COM/CONTENT/THORAXJNL/57/4/289.FULL.PDF

***CHECKPOINT CHALLENGE #6**: OFTEN, PATIENTS WITH A UNILATERAL SPONTANEOUS PNEUMOTHORAX WILL HAVE A CT SCAN OF THEIR CHEST. LET'S SAY LP HAS A CT AND IT SHOWS BULLAE ON THE RIGHT SIDE, AS EXPECTED, BUT ALSO BULLAE ON HER LEFT SIDE, EVEN THOUGH SHE HAS NO PNEUMOTHORAX ON THE LEFT. WHAT IS THE 'BEST PRACTICE' ADVICE?*

—THE PROTOCOLS FOR 'BEST PRACTICE' IN THE CARE OF PATIENTS WITH SPONTANEOUS PNEUMOTHORAX ARE STILL A MATTER OF DEBATE. I WANTED TO OFFER YOU THIS CHALLENGE SO THAT YOU ARE REMINDED THAT MUCH OF WHAT WE DO IN SURGERY, AND IN MEDICINE IN GENERAL DEMANDS CONSTANT QUESTIONING. THE FOLLOWING ARTICLE REFERENCES SUCH DEBATES IN THEIR WORDING OF "SHOULD BE CONSIDERED", "MIGHT BE BENEFICIAL", AND "FURTHER STUDIES ARE REQUIRED".

FYI— NOH D, KEUM DY, PARK CK. OUTCOMES OF CONTRALATERAL BULLAE IN PRIMARY SPONTANEOUS PNEUMOTHORAX. KOREAN J THORAC CARDIOVASC SURG. 2015 DEC;48(6):393-7. DOI: 10.5090/KJTCS.2015.48.6.393. EPUB 2015 DEC 5. PMID: 26665105; PMCID: PMC4672973.

***CHECKPOINT CHALLENGE #7**: LP'S BULLAE AT THE TOP OF HER RIGHT LUNG IS LEAKING INTO HER RIGHT PLEURAL CAVITY. IS THIS A FISTULA? IS IT A SINUS? DOES IT MATTER?*

—WE SHOULD NOT GET HUNG UP ON THE TERMS, BUT SINCE I AM ASKING, IS THE PLEURAL CAVITY LINED WITH EPITHELIUM? ARE THE ALVEOLI LINED WITH EPITHELIUM? IF THERE'S AN ABNORMAL COMMUNICATION BETWEEN THEM THEN WE COULD RIGHTLY CALL THAT COMMUNICATION A…(I'LL LET YOU ANSWER.)

***CHECKPOINT CHALLENGE #8**: BABY FH HAD A DEFINITE BOWEL OBSTRUCTION AND ALTHOUGH HE WAS INTOLERANT OF FEEDS AND HAD SOME ABDOMINAL DISTENSION, HE NEVER VOMITED "GREEN". WHY?*

—THINK ABOUT THE LEVEL OF THIS BABY'S GI OBSTRUCTION IN THIS CASE. IT IS VERY IMPORTANT FOR YOU TO REALIZE THAT THE LACK OF ONE "CLASSIC" SYMPTOM OR SIGN, SUCH AS GREEN EMESIS, DOES NOT MEAN THE CHILD DOESN'T HAVE THE DISEASE. HIRSCHSPRUNG'S DISEASE MAY PRESENT IN A MYRIAD OF WAYS AND GREEN BILIOUS VOMITING, ABDOMINAL DISTENSION AND LACK OF STOOL PASSAGE IS ONE, BUT BROWN EMESIS CAN OCCUR, AND THE

CHILD MAY PRESENT WITH "HIRSCHSPRUNG'S ENTEROCOLITIS" WHICH MAY MANIFEST AS FULMINANT DIARRHEA, AND, BY THE WAY, MAY BE QUICKLY FATAL! THE MESSAGE HERE IS THAT NO MATTER WHAT THE DISEASE AND ITS ASSOCIATED "ILLNESS SCRIPT" IS, CHILDREN WILL NOT UNCOMMONLY "GO OFF SCRIPT" TO CHALLENGE YOU.

FYI—THERE ARE LOTS OF GOOD REVIEW ARTICLES OUT THERE WHICH CAN GIVE YOU AN OVERVIEW OF HIRSCHSPRUNG'S DISEASE. HOWEVER, I FIND THE TEXTBOOKS SOMEWHAT DEFICIENT AS THEY OFTEN SAY LITTLE OR NOTHING ABOUT HIRSCHSPRUNG'S ENTEROCOLITIS. CHECK OUT: HTTPS://WWW.NCBI.NLM.NIH.GOV/PMC/ARTICLES/PMC3462485/#:~:TEXT=HIRSCHSPRUNG%2DASSOCIATED%20ENTEROCOLITIS%20(HAEC)%20WAS%20FIRST%20RECOGNIZED%20IN%20THE,ABDOMINAL%20DISTENTION%2C%20DIARRHEA%20AND%20SEPSIS.

***CHECKPOINT CHALLENGE #9**: WHAT NEUROLOGIC DISEASE WHICH CAN AFFECT CHILDREN AND ADULTS SOMETIMES BENEFITS FROM A THYMECTOMY?*

—IF YOU KNOW THIS ONE THEN GOOD FOR YOU. CHECK THE FYI ARTICLE BELOW WHICH HAS THE ANSWER IN THE TITLE! THERE IS SO MUCH WE DON'T KNOW ABOUT THIS PARTICULAR "DISEASE OF FUNCTION" AND SO MANY OTHER DISEASES. EXACTLY HOW A SURGICAL REMOVAL OF A THYMUS COULD HELP SUCH A PATIENT, ESPECIALLY, IT SEEMS PEDIATRIC PATIENTS, IS A MYSTERY. NOT SURPRISINGLY THE EXACT ROLE OF THYMECTOMY IS STILL DEBATED.

FYI— SIEB JP. MYASTHENIA GRAVIS: AN UPDATE FOR THE CLINICIAN. CLIN EXP IMMUNOL. 2014 MAR;175(3):408-18. DOI: 10.1111/CEI.12217. PMID: 24117026; PMCID: PMC3927901.

***CHECKPOINT CHALLENGE #10**: LET'S SUPPOSE THAT OUR 11-YEAR-OLD PATIENT, IA, WAS INITIALLY MISDIAGNOSED AND TWO WEEKS LATER PRESENTED WITH A LARGE PELVIC ABSCESS SECONDARY TO THE RUPTURE OF HIS ACUTE APPENDICITIS MANY DAYS PREVIOUSLY. WHICH OF THE FOLLOWING THREE SYMPTOMS/SIGNS IS MOST COMMONLY SEEN IN PELVIC ABSCESSES:*
*☐FEVER ☐DIARRHEA ☐PAIN*

—DOES IT REALLY MATTER? "IN MY EXPERIENCE" (AND YOU'LL PROBABLY START USING THAT PHRASE YOURSELF ONE DAY) DIARRHEA IS SURPRISINGLY MORE COMMON IN CASES LIKE THIS RATHER THAN PAIN OR FEVER, BUT I HAVEN'T DONE A RETROSPECTIVE ANALYSIS. IT'S JUST MY IMPRESSION. THE POINT IS THAT, WHEREAS YOU MAY INTUITIVELY THINK IT WOULD BE PAIN OR FEVER, PATIENTS WITH A MISSED APPENDICITIS MAY OFTEN PRESENT WITH A PICTURE SEEMINGLY MORE IN KEEPING WITH A GASTROENTERITIS OR INFLAMMATORY BOWEL DISEASE. THE DIARRHEA OCCURS BECAUSE THE PELVIC COLLECTION OF PUS IRRITATES THE RECTUM. A DRE CAN OFTEN REVEAL AN EASILY PALPABLE PELVIC MASS, THE ABSCESS, AS THE CULPRIT WITH A SUBSEQUENT ULTRASOUND TO CHARACTERIZE IT. KNOWING THIS SORT OF DECEPTIVE PRESENTATION OF "MISSED APPENDICITIS" CAN PREVENT NEEDLESS DIAGNOSTIC WORKUPS FOR BOWEL INFECTIONS AND IBD. IT CAN BE THE KNOWLEDGE OF VARIABLE PRESENTATIONS OF

DISEASE WHICH CAN SEPARATE THE AVERAGE CLINICIAN FROM THE GREAT CLINICIAN. STRIVE TO BE GREAT THROUGH SEEING LOTS OF PATIENTS, READING AND THINKING.

***CHECKPOINT CHALLENGE #11:*** *THE TODDLER WITH THE NECK ABSCESS SHOWN ABOVE PRESENTED WITHOUT A FEVER OR ANY OTHER SYSTEMIC SIGNS. HE UNDERGOES AN I&D PROCEDURE AS IN THE PHOTO SHOWN. DOES HE REQUIRE A COURSE OF ANTIBIOTICS?*

—WE COULD DEBATE THIS UNTIL THE COWS COME HOME. ONE DOES NOT WANT TO BE CAVALIER IN THE TREATMENT OF CHILDREN WITH INFECTIONS. HOWEVER, THE SCENARIO I HAVE SET UP HERE IS NOT UNCOMMON—A CHILD WITH A FAIRLY SUPERFICIAL CERVICAL ABSCESS, AND NO SYSTEMIC SYMPTOMS OR SIGNS OTHERWISE, NEEDING THE DEFINITIVE TREATMENT OF AN INCISION & DRAINAGE. IN THIS SITUATION ANTIBIOTICS ARE REALLY RATHER SECONDARY. IT CAN BE JUSTIFIABLE TO ADMINISTER A SHORT COURSE OF ANTIBIOTICS, FOR INSTANCE, SAY A PRE-OP IV DOSE OF A BROAD-SPECTRUM CEPHALOSPORIN AND A VERY SHORT COURSE, OF ONLY A DAY OR SO AFTERWARDS. PUS SHOULD BE SENT FOR CULTURE AND SENSITIVITY, BUT IT ALMOST UNDOUBTEDLY WILL BE A COMMON STREP OR STAPH SPECIES. IT IS THE CLINICAL STATE OF THE CHILD WHICH SHOULD DICTATE TREATMENT, NOT, GENERALLY SPEAKING, THE MICROBIOLOGY REPORT (UNLESS IT SHOWS SOMETHING WEIRD LIKE ACTINOMYCOSIS). SUBJECTING A CHILD TO A FULL COURSE OF ANTIBIOTICS IN THIS SORT OF SITUATION IS NEEDLESS AND IS PROBABLY TREATING YOUR ANXIETIES MORE THAN DOING ANY GOOD FOR THE CHILD. WE KNOW THAT ANTIBIOTICS ARE PROBABLY USED TOO MUCH, TOO LONG AND TOO FREQUENTLY AND CAN HAVE UNWANTED EFFECTS. FOLLOWING THE CHILD CLINICALLY IS THE BEST GUIDE. AND DON'T EXPECT THE AREA OF THE ABSCESS TO SUDDENLY LOOK AND FEEL NORMAL EITHER. A DRAINED ABSCESS CAN EASILY TAKE 6 WEEKS FOR THE VESTIGES OF SWELLING TO ALMOST DISAPPEAR. A PERSISTENT SMALL—BUT NOT INFLAMED— 'LUMP' IN THE AREA FOR WEEKS AFTERWARDS NEED NOT CAUSE CONCERN.

FYI—IT IS NOT MY INTENT TO CONFUSE YOU ABOUT THE PROPER EVALUATION AND TREATMENT OF NECK ABSCESSES, BUT THERE ARE DIFFERENT OPINIONS OUT HERE. ALSO, REALIZE (AGAIN) THAT CHILDREN ARE NOT JUST SMALL ADULTS. HERE'S A PAPER WHICH TALKS ABOUT NECK ABSCESSES IN GENERAL BUT MIGHT LEAD YOU TO BELIEVE THAT EVERY NECK ABSCESS NEEDS A CT SCAN AND A FULL GAMUT OF BLOOD WORK. I OFFER YOU THIS REFERENCE SO THAT YOU CAN APPRECIATE HOW IN MANY AREAS OF MEDICINE AND SURGERY YOU NEED TO LEARN PERSPECTIVE AND HOW TO APPLY WHAT YOU KNOW TO THE PARTICULAR PATIENT YOU ARE DEALING WITH AT THAT TIME AND IN THAT CONTEXT.

MCDOWELL RH, HYSER MJ. NECK ABSCESS. [UPDATED 2022 SEP 19]. IN: STATPEARLS [INTERNET]. TREASURE ISLAND (FL): STATPEARLS PUBLISHING; 2023 JAN-. AVAILABLE FROM: HTTPS://WWW.NCBI.NLM.NIH.GOV/BOOKS/NBK459170/

***CHECKPOINT CHALLENGE #12****: ANOTHER TODDLER PRESENTS WITH A LUMP IN THE ANTERIOR MIDLINE OF HIS NECK. HIS PARENT'S JUST NOTICED IT AND IT SEEMS SLIGHTLY RED AND TENDER. CONSIDER THE POSSIBILITIES.*

—One way to classify neck masses in children is "lateral vs midline" or perhaps even better "Anterior cervical triangle vs posterior cervical triangle". The 'lump' in question is related to the embryology of the thyroid gland. These entities are certainly not rare, and infection is sometimes the way they first present. Be aware that there is a great deal of anxiety-provoking literature out there that might have you believe that absolutely every lump a child has in their neck needs a CT scan (vide supra) and a needle biopsy. Not so. (Needle biopsies of childhood neck nodes is not a recommended practice, but needle biopsies—FNA— of thyroid nodules is). But getting to the point in practice where you can say to a parent, with confidence, "There's nothing to worry about here" takes years of experience. You don't want to take chances. Often the best "test" is calling a colleague to assess. That colleague may be another doctor in your clinic, a pediatric surgeon or an ENT surgeon. Whatever course of action you take regarding a neck mass in a child probably the most important is to follow-up in days or weeks depending on the situation. What you don't want is for the parent to have the child re-assessed in six months when the mass is much larger, saying, "but you said six months ago it was nothing to worry about, so even when I saw that it was getting worse I figured you didn't want to see him again!" I do not want fear to dictate your medical practice, but we are paid to worry. A simple follow-up is often the ticket!

FYI—This article, although not perfect, at least affords good suggestions for thoughtful workups in cases of neck masses in children. However, take note that many of the recommendations are of a "C" category. Sometimes the best approach is to pick up the phone and ask a specialist what their advice would be. (Woe unto us and our health system if we get to the point where we can't simply ask each other about proper patient management!)

Meier JD, Grimmer JF. Evaluation and management of neck masses in children. Am Fam Physician. 2014 Mar 1;89(5):353-8. PMID: 24695506.

***Checkpoint Challenge #13**: Whereas adults often suffer ischemic hearts, brains and intestines because of atherosclerosis, children are not afflicted by that genre of vascular disease. However, a 6-year-old lad of Middle Eastern descent is known to have Sickle Cell Anemia and presents with acute abdominal pain and a severe drop in his hemoglobin level. In what way is he suffering from ISCHEMIA?*

—We speak of a "sickle cell crisis", but what's another name for the condition wherein a crisis causes abdominal pain? That name implies the "ischemic" aspect of the disease

FYI—Borhade MB, Kondamudi NP. Sickle Cell Crisis. [Updated 2022 Aug 29]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK526064/

***CHECKPOINT CHALLENGE #14:*** *A 2-YEAR-OLD BOY IS NOTED TO HAVE A RIGHT INGUINAL HERNIA. ON FURTHER EXAMINATION YOU NOTE A RIGHT UNDESCENDED TESTICLE AS WELL. THE LEFT TESTICLE AND GROIN ARE NORMAL. BRIEFLY OUTLINE A PLAN FOR THIS LAD.*

—IN MOST CASES OF UNDESCENDED TESTICLE THERE CAN BE FOUND A CONCOMITANT INDIRECT INGUINAL HERNIA, EVEN THOUGH IT MAY NOT HAVE CLINICALLY MADE ITSELF APPARENT. INDEED, THE PRESENCE OF THE HERNIA SAC, WHICH IS THE FAILURE OF CLOSURE OF THE PATENT PROCESSUS VAGINALIS IS THOUGHT TO BE PERHAPS CAUSALLY LINKED TO THE TESTICLE NOT DESCENDING. IN THE COURSE OF AN OPERATION FOR AN UNDESCENDED TESTICLE—AN ORCHIDOPEXY—THIS INDIRECT INGUINAL HERNIA SAC IS LIGATED. SO, IN A 2-YEAR-OLD BOY LIKE THIS WHO PRESENTS WITH A RIGHT INGUINAL HERNIA AND SHOULD HAVE IT REPAIRED, DO YOU NOT THINK WE WOULD DO AN ORCHIDOPEXY AT THE SAME TIME WHEN THE RECOMMENDED AGE FOR ORCHIDOPEXY IS ABOUT 1 YEAR OF AGE? SOUNDS LIKE A GOOD PLAN.

FYI— LESLIE SW, SAJJAD H, VILLANUEVA CA. CRYPTORCHIDISM. [UPDATED 2022 NOV 28]. IN: STATPEARLS [INTERNET]. TREASURE ISLAND (FL): STATPEARLS PUBLISHING; 2022 JAN-. AVAILABLE FROM: HTTPS://WWW.NCBI.NLM.NIH.GOV/BOOKS/NBK470270/

***CHECKPOINT CHALLENGE #15:*** *ONE WEEKEND A NUMBER OF YEARS AGO, I SAW 4 NEWBORNS WITH ESOPHAGEAL ATRESIA. (SO MUCH FOR THE SUPPOSED 1 IN 5000 INCIDENCE!). ONE OF THE BABIES HAD A LARGE OMPHALOCELE IN ADDITION TO HIS ESOPHAGEAL ATRESIA. IN THAT BABY WHAT WOULD BE THE PRIORITIES OF OPERATIVE MANAGEMENT?*

—YOU HAVE TO THINK, "WHAT PATHOLOGY IN THIS BABY IS THE GREATEST IMMEDIATE THREAT?" REMEMBER, IN OMPHALOCELES THERE IS A MEMBRANE WHICH COVERS THE HERNIATED GUTS. IN ESOPHAGEAL ATRESIA THE BABY CANNOT SWALLOW AND EMERGES INTO THE WORLD FROTHING SALIVA AND MUCOUS AT THE MOUTH. FURTHERMORE, THERE MAY BE A FISTULOUS CONNECTION BETWEEN THE ESOPHAGUS AND WHAT PART OF THE BABY'S ANATOMY? WHAT RISK IS THAT? AND NOTE THAT I ASKED WHAT WOULD BE THE PRIORITIES OF **OPERATIVE** MANAGEMENT. REMIND YOURSELF OF THE TREATMENTS OF OMPHALOCELE AND ESOPHAGEAL ATRESIA.

***CHECKPOINT CHALLENGE #16****: IT WAS EARLY DECEMBER WHEN A YOUNG GIRL, ABOUT 2-YEARS OLD AND HER MOTHER WERE PEDESTRIAN VICTIMS OF AN APPARENT HIT & RUN ACCIDENT ON A RESIDENTIAL SIDE STREET. THEY WERE BOTH FOUND UNCONSCIOUS ON THE ROAD BY A PASSER-BY WHO QUICKLY CALLED 911. THEY WERE BOTH BROUGHT INTO THE ER IN ONE AMBULANCE. THE MOTHER'S TEMPERATURE ON ADMISSION WAS 37$^{O}$ BUT THE CHILD'S TEMPERATURE WAS ONLY 35.9$^{O}$. WHY?*

—WE ARE NOT TOLD WHAT THEY ARE WEARING OR HOW LONG THEY WERE LYING THERE BEFORE BEING TRANSPORTED INTO THE HOSPITAL, BUT WE'RE TOLD IT WAS IN DECEMBER, SO IF THIS HAPPENED IN CANADA, IT WAS A CHILLY TIME OF YEAR. SO, HERE'S A TINY BIT OF MATH TO HELP YOU. IT TURNS OUT THAT THE RATIO (S/V) OF THE SURFACE AREA (S) TO THE VOLUME (V) OF A SPHERE= 3/R, WHERE R=THE RADIUS OF THE SPHERE. SO, THE SMALLER THE SPHERE, PROPORTIONATELY THE LARGER THE S/V RATION IS, AND THIS GENERAL RULE OF INVERSE PROPORTIONALITY APPLIES TO ANY 3-DIMENSIONAL SHAPE—SUCH AS A HUMAN BODY

(ALTHOUGH THE S/V RATIO WILL BE DIFFERENT DEPENDING ON THE SHAPE). THE INJURED UNCONSCIOUS BODIES OF THE SMALL CHILD AND THE LARGER ADULT ARE LYING THERE ON A COLD DECEMBER DAY, RADIATING THEIR BODY HEAT. YOU TOK ALL THAT MATH AND PHYSICS IN HIGHSCHOOL AND PROBABLY IN UNIVERSITY TOO. FEEL FREE TO USE IT—IT'S STILL IN YOUR BRAIN.

FYI— HTTPS://VAN.PHYSICS.ILLINOIS.EDU/ASK/LISTING/791

***CHECKPOINT CHALLENGE #17****: A 1-YEAR-OLD BOY HAS A SECOND-DEGREE SCALDING INJURY TO HIS ENTIRE RIGHT LEG. WHAT PERCENTAGE OF HIS BODY SURFACE AREA IS BURNT?*

—CHILDREN ARE NOT JUST SMALL ADULTS. REMEMBER THE "RULE OF NINES", AND CHECK OUT THE FYI AND OTHER REFERENCES

FYI— HTTP://WWW.EMTRESOURCE.COM/EMERGENCIES/BURNS/RULE-OF-NINES/

AND WHAT ABOUT THAT FIST?


THE IMAGE OF THAT UPRAISED FIST YOU HAVE PROBABLY SEEN BEFORE, NOT IN THE CONTEXT OF SURGERY, BUT REPRESENTATIVE OF REVOLUTION. THE IMAGE, ENTITLED THE "HAND" WAS FIRST CREATED IN THE HEYDAY OF THE TURBULENT '60S AS A WOODCUT BY A BERKELEY, CALIFORNIA ARTIST AND REVOLUTIONARY, FRANK CIECIORKA. ONLY A FEW PRINTS WERE MADE OF THE ORIGINAL WOODCUT AND A FRIEND OF MINE, LINCOLN CUSHING, AN ARCHIVIST OF REVOLUTIONARY ART AND IMAGERY, HAS ONE OF THEM, GIVEN TO HIM BY THE ARTIST HIMSELF. LINCOLN ENCOURAGED ME TO USE THE IMAGE. FRANK CIECIORKA, WHO DIED IN 2008, TOOK PRIDE IN THE WIDESPREAD ADOPTION OF HIS FIST IMAGE BY MANY GROUPS LOOKING FOR MEANINGFUL SOCIETAL CHANGE. SINCE THE 1960S THE 'FIST' HAS BEEN A RALLY CRY TO QUESTION THE STATUS QUO AND STAND STRONG FOR JUSTICE FOR ALL. A GOOD MESSAGE!

REF: CUSHING, LINCOLN. ALL OF US OR NONE: SOCIAL JUSTICE POSTERS OF THE SAN FRANCISCO BAY AREA. HEYDAY, BERKELEY, 2012


And what's with the cover? One of my little patients drew this picture for me. He told me the big bulge below his tummy was his hernia. Pretty smart!

ACKNOWLEDGEMENTS

This primer started years ago as a conversation with Aoife O'Leary, a medical student curious to learn more about Pediatric Surgery. Aoife expertly and seemingly effortlessly took my original hodge-podge of documents, pictures, and diagrams, and formatted them all into the first edition of this little book. Thank you, Aoife!

I also want to thank the many UBC Medical Students, who have offered up their encouragement and cogent suggestions for each of this Primer's editions, including this third one. I especially acknowledge with gratitude Tisha Dasgupta and Cyrus Bhiladvala, for their hard work at creating the 2nd edition's complex layout and their patient tolerance of my many requests. Now for this 3rd edition, I want to express my gratitude to Damian Duffy, the Executive Director of UBC's Office of Pediatric Surgical Evaluation and Innovation and one of OPSEI's cooperative students, Josh Zhang, who worked on the formatting of this 3rd edition.

My colleagues and friends within the UBC Department of Surgery have given me invaluable support in this educational endeavor. In particular, Ms. Charlene Gafur, our UBC Department of Surgery Senior Program Assistant for Undergraduate Years 3 & 4, continues to work tirelessly to help bring this and so many other educational resources and opportunities to our medical students. Your work is valued and very appreciated by me and many others. Thank you.

It is wonderful that my pediatric surgical colleagues, both faculty and residents, enjoy teaching and are so good at it. Of course, what makes teaching particularly enjoyable is when our students enjoy learning.

Remember always to CARE, to THINK, to ACT…

About the author:

Dr. Geoffrey K. Blair, MD, FRCSC is a retired Pediatric Surgeon and a Clinical Professor of Surgery in the Department of Surgery at the University of British Columbia, and the past Director of the Undergraduate Surgical Education Program at UBC.