



FOR  
Health Professionals

# Is this bleeding normal?

## Evaluating a child with a suspected bleeding disorder

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# Disclosures

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# Objectives

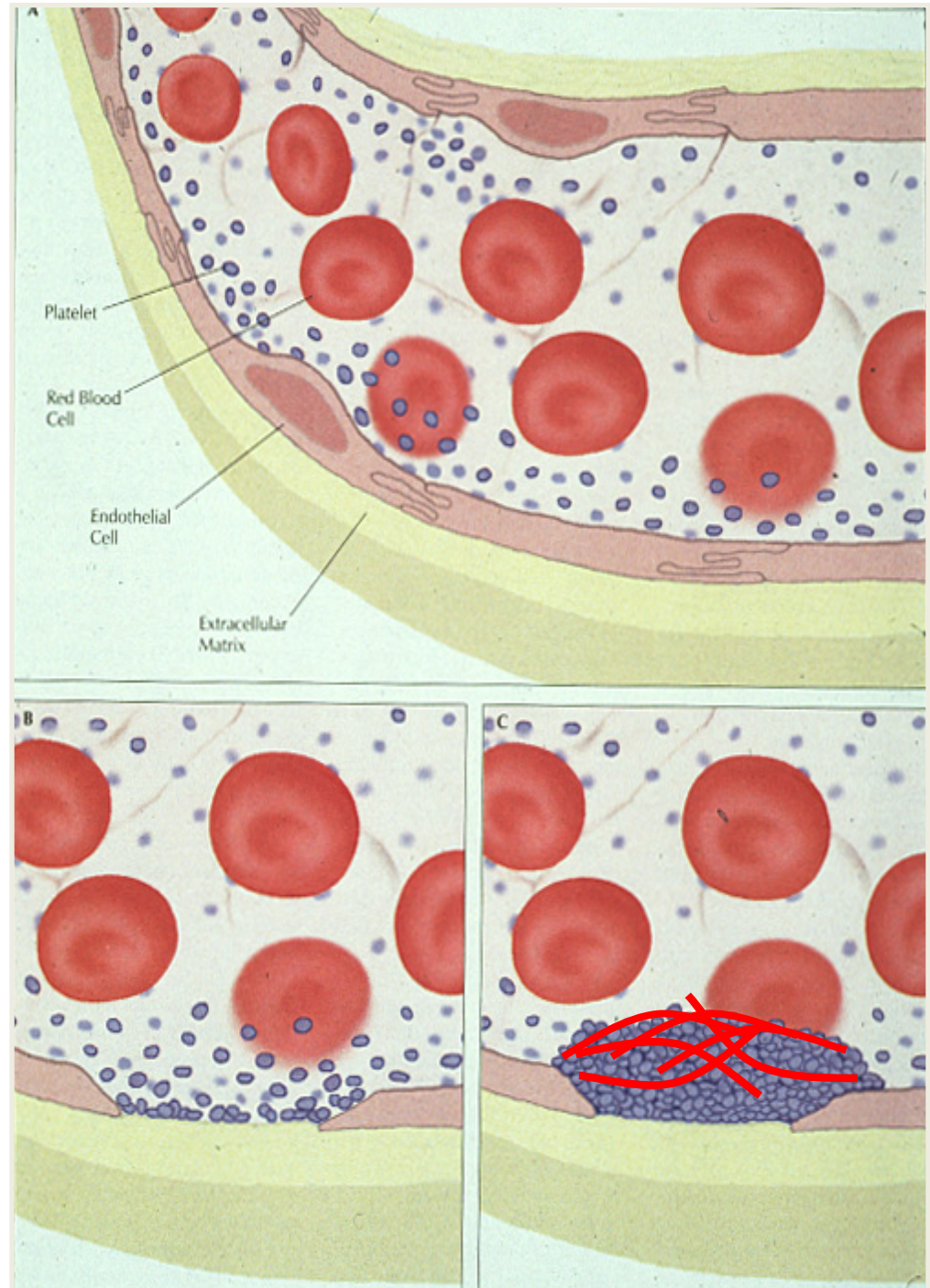
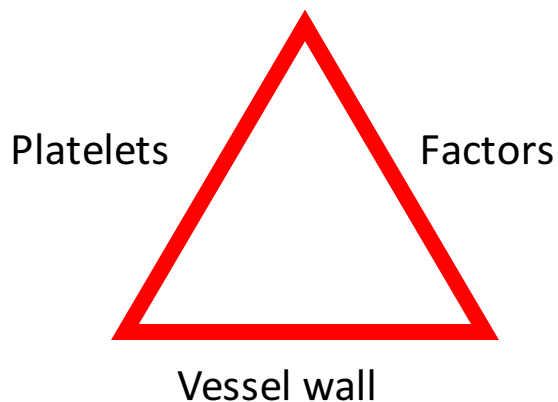
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After this session participants will:

1. Have an approach to evaluating a child who presents with bleeding
2. Be familiar with common bleeding disorders in children as well as other common causes of bleeding
3. Recognize presentations of bleeding in children requiring urgent referral to a hematologist

What do you need to form a blood clot?

1. A normal blood vessel
2. Platelets:  
Normal number  
Normal function
3. Coagulation factors:  
I to XIII



# Referral to Pediatric Hematology

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“Dear Pediatric Hematologist:

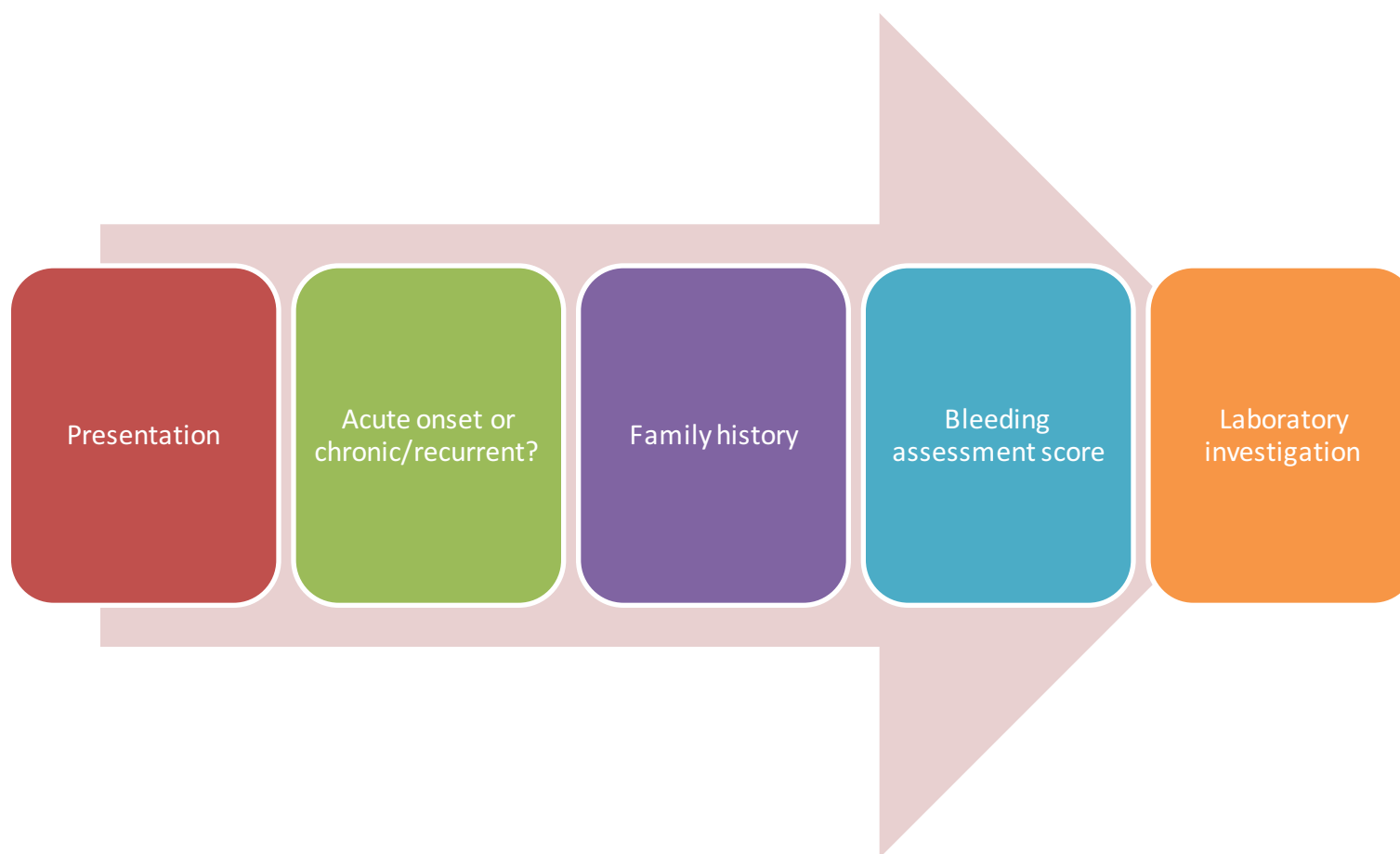
Please see this seven year old boy with recurrent epistaxis for investigation of a bleeding disorder.”

1. Does this child have a bleeding disorder?
2. What kind of investigation does he need?



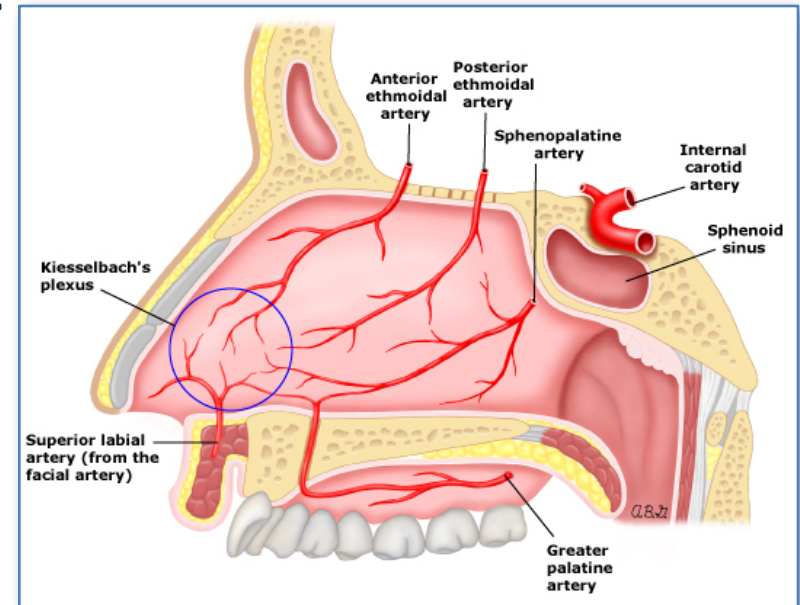
# Evaluation of the bleeding patient

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## Tip #1: Most children with epistaxis do not have a bleeding disorder

- 30% of children < 5 years and 56% of children aged 6 - 10 years have had at least one nosebleed.
- Why so common?
  - **Anatomy**
- Local causes
  - ✓ Trauma: nose-picking\*; foreign bodies
  - ✓ Mucosal irritation: dry air\*; allergic rhinitis; inhaled irritants, URIs or other local infection
  - ✓ Anatomic abnormalities, deviated septum, benign tumors
- Systemic causes



(UpToDate 2016)

## Bleeding assessment tools (BAT)

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- Standardized questionnaires that can help quantify clinical bleeding symptoms/signs
  - Consist of a series of questions and a scoring system
  - BATs have been created for different purposes and with different scoring systems; several have been validated
- BATs can be:
  - Age specific: adults or children
  - Disease specific: VWD, platelet function disorders
  - Symptom specific: epistaxis and menorrhagia

**ISTH-BAT can be used for all ages, all symptoms.**

It has been evaluated in >1000 adults and >300 children to develop normal ranges.



# ISTH-BAT scoring sheet

SYMPTOMS (up to the time of diagnosis)	SCORE				
	0 <sup>s</sup>	1 <sup>s</sup>	2	3	4
Epistaxis	No/trivial	- > 5/year or - more than 10 minutes	Consultation only*	Packing or cauterization or antifibrinolytic	Blood transfusion or replacement therapy (use of hemostatic blood components and rFVIIa) or desmopressin
Cutaneous	No/trivial	For bruises 5 or more (> 1cm) in exposed areas	Consultation only*	Extensive	Spontaneous hematoma requiring blood transfusion
Bleeding from minor wounds	No/trivial	- > 5/year or - more than 10 minutes	Consultation only*	Surgical hemostasis	Blood transfusion, replacement therapy, or desmopressin
Oral cavity	No/trivial	Present	Consultation only*	Surgical hemostasis or antifibrinolytic	Blood transfusion, replacement therapy or desmopressin
GI bleeding	No/trivial	Present (not associated with ulcer, portal hypertension, hemorrhoids, angiodysplasia)	Consultation only*	Surgical hemostasis, antifibrinolytic	Blood transfusion, replacement therapy or desmopressin

14 categories: hematuria, tooth extraction, surgery, menorrhagia, postpartum hemorrhage, muscle hematoma, hemarthrosis, CNS bleeding, other.

([www.isth.org/resource/resmgr/ssc/isth-ssc\\_bleeding\\_assessment.pdf](http://www.isth.org/resource/resmgr/ssc/isth-ssc_bleeding_assessment.pdf))

## Value of BATs

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### ISTH-BAT cut-off scores:

- $\geq 4$  for men
- $\geq 6$  for women
- $\geq 3$  for children

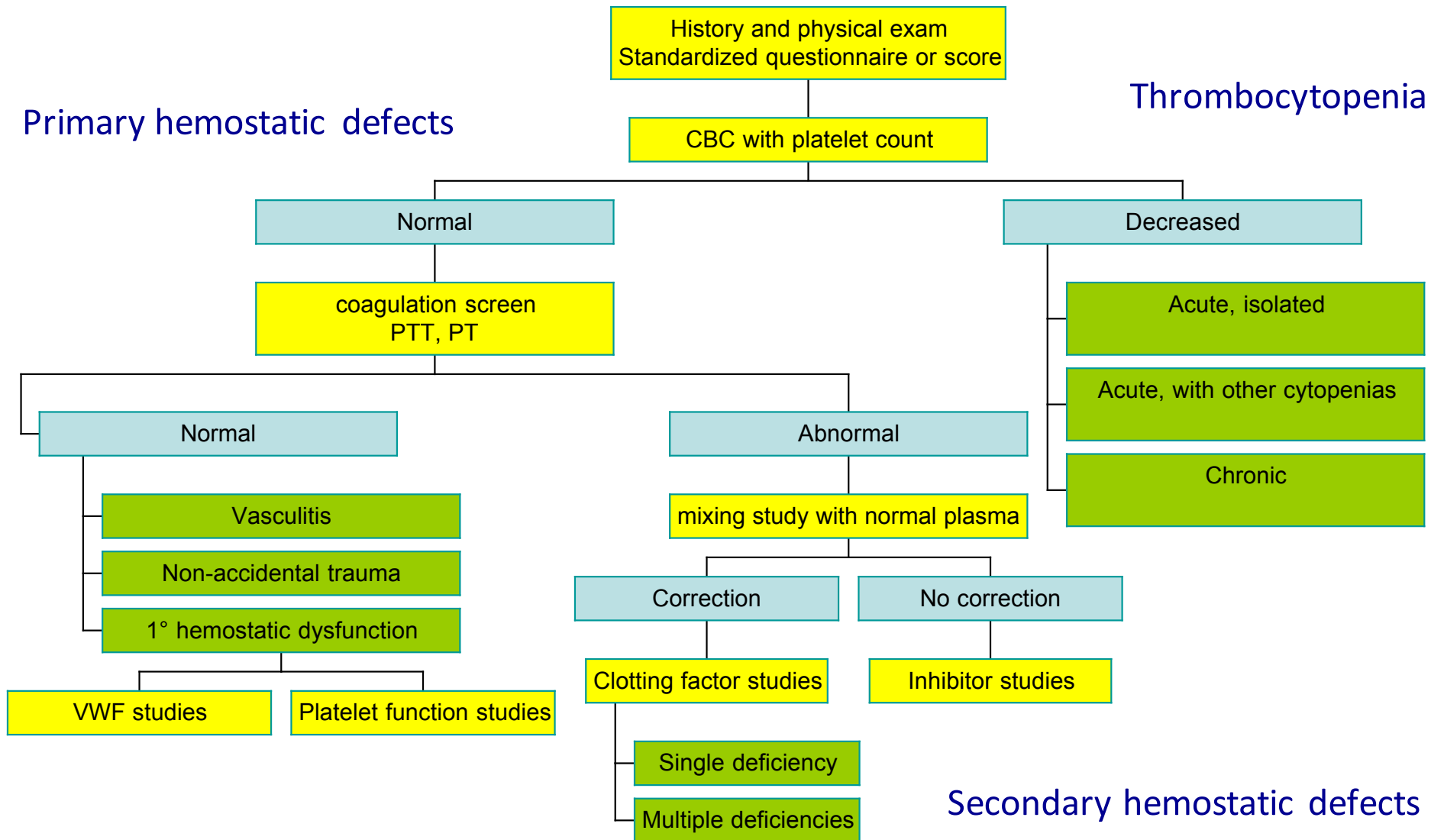
### Utility of ISTH-BAT:

- Sensitivity 97%
- Specificity 50%
- NPV 99%

### Tip #2:

A validated BAT score does not provide a diagnosis, but it can help to identify which patients do not need further investigation.

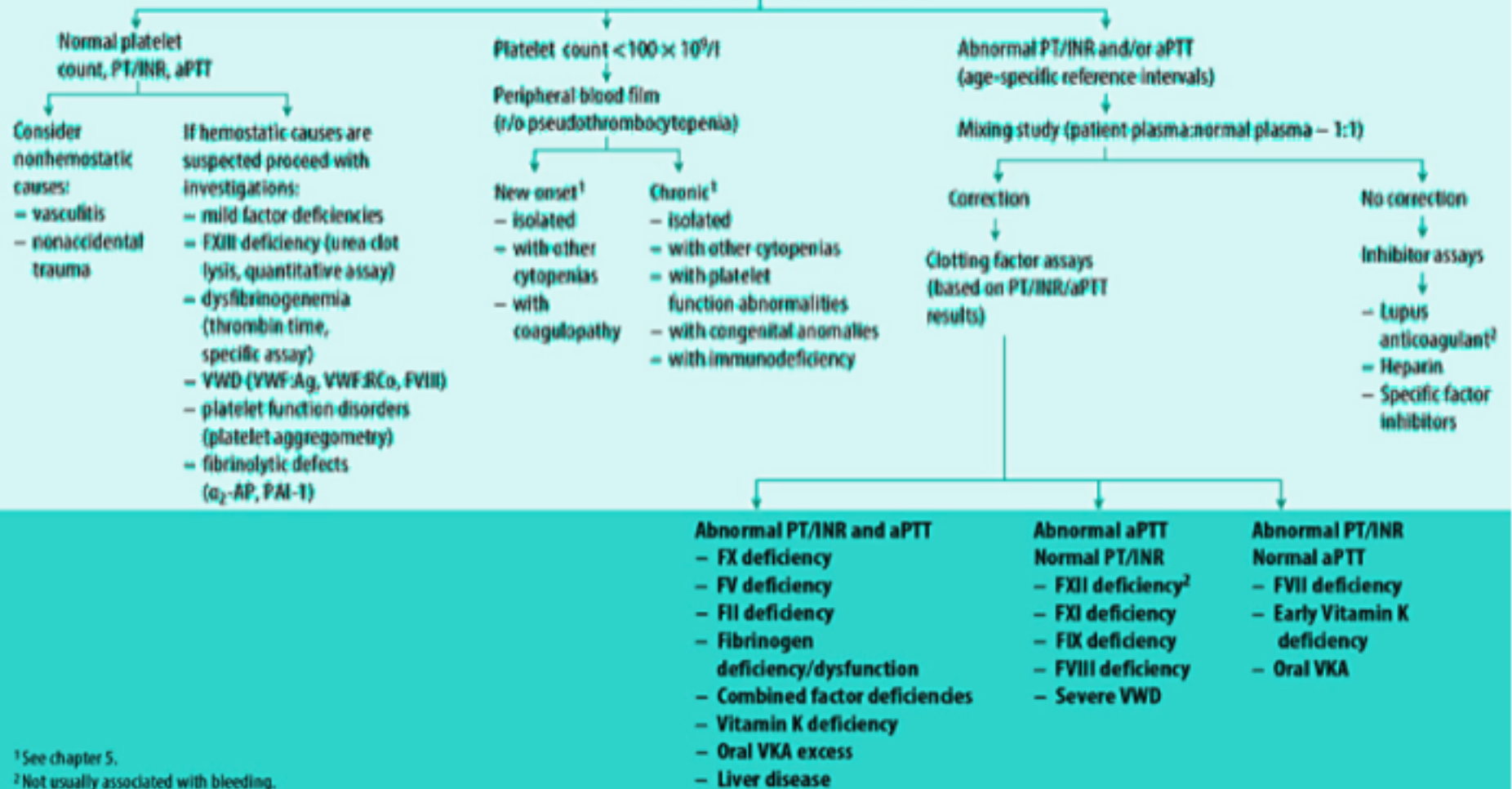
# Approach to the bleeding child



### Child with bleeding symptoms

Medical history: age, sex, past medical history, use of medications  
 Bleeding history: standardized bleeding questionnaire  
 Family bleeding history: standardized bleeding questionnaire, ethnicity  
 Physical examination: hemodynamic status, pattern of bleeding, other findings

Initial laboratory tests: CBC, PT/INR, aPTT



<sup>1</sup> See chapter 5.

<sup>2</sup> Not usually associated with bleeding.

(From: Revel-Vilk S, Rand ML, Israels SJ. Approach to the Bleeding Child. In: SickKids Handbook of Pediatric Thrombosis and Hemostasis. Karger 2013)

## What laboratory tests should I order?

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Most algorithms are based on basic testing followed by specialized testing:

1. CBC with platelet count

2. PT/INR, PTT →

3. Specialized testing

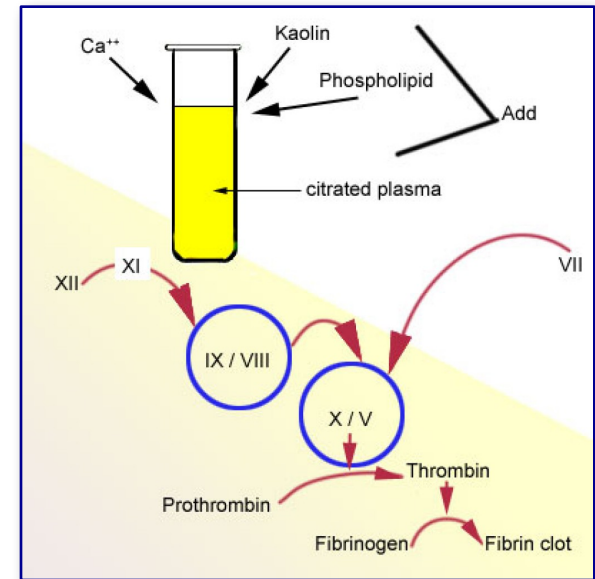
*(requires consultation with a Hematologist or Hematopathologist)*

It depends where you practice:

- Recent changes to DSM guidelines have removed the **PTT** from general requests except for UFH monitoring or as part of evaluation for DIC
- Other laboratory services still provide PTT testing
- **PTTs** can always be requested via Hematopathologist

# Why the **PTT** is a poor test for bleeding

- **Inappropriately sensitive:**
  - Lupus anticoagulants
  - Clotting factor deficiencies that do not cause bleeding (FXII)
- **Inadequately sensitive:**
  - Mild clotting factor deficiencies
- **Not built to measure:**
  - Von Willebrand Disease
  - Factor XIII deficiency
  - Some fibrinogen abnormalities
  - Fibrinolytic defects
  - Platelet disorders



## What the **PTT** is good for:

- Severe clotting factor deficiencies
- Multiple clotting factor deficiencies
- Unfractionated heparin monitoring

## Tip #3: Laboratory screening tests may not help you

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- The exception is the CBC
- Critically ill children often have abnormalities if they have sepsis or consumptive coagulopathy
- Other children:
  - Rarely have abnormalities of PTs unless they have Vitamin K deficiency
  - Abnormal PTTs are more likely to be caused by infection-associated LA than a factor deficiency
  - More common bleeding disorders require specialized testing

# Referral to Pediatric Hematology

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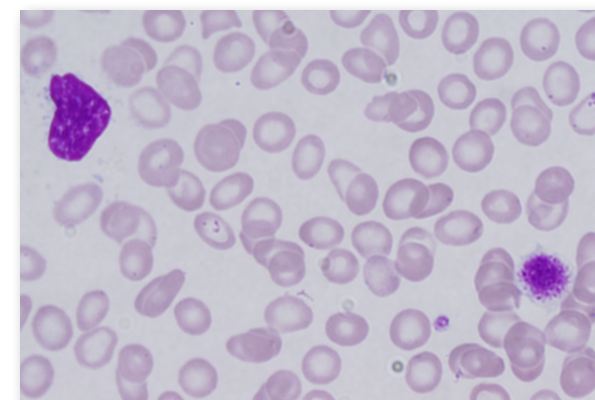
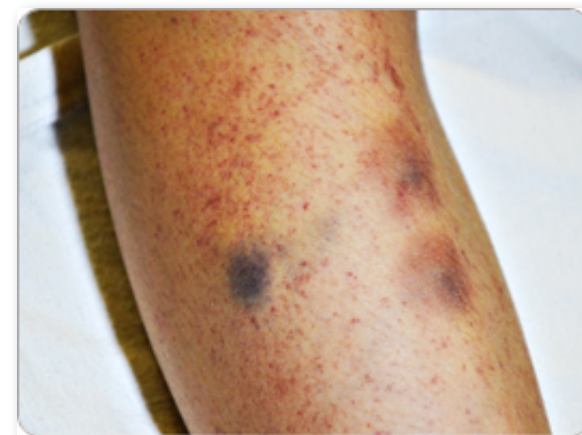
“Dear Pediatric Hematologist:

Please see this 5 year old girl, previously well, with sudden onset of bruising, a petechial rash and epistaxis. She previously had an adenoidectomy without bleeding complications.”

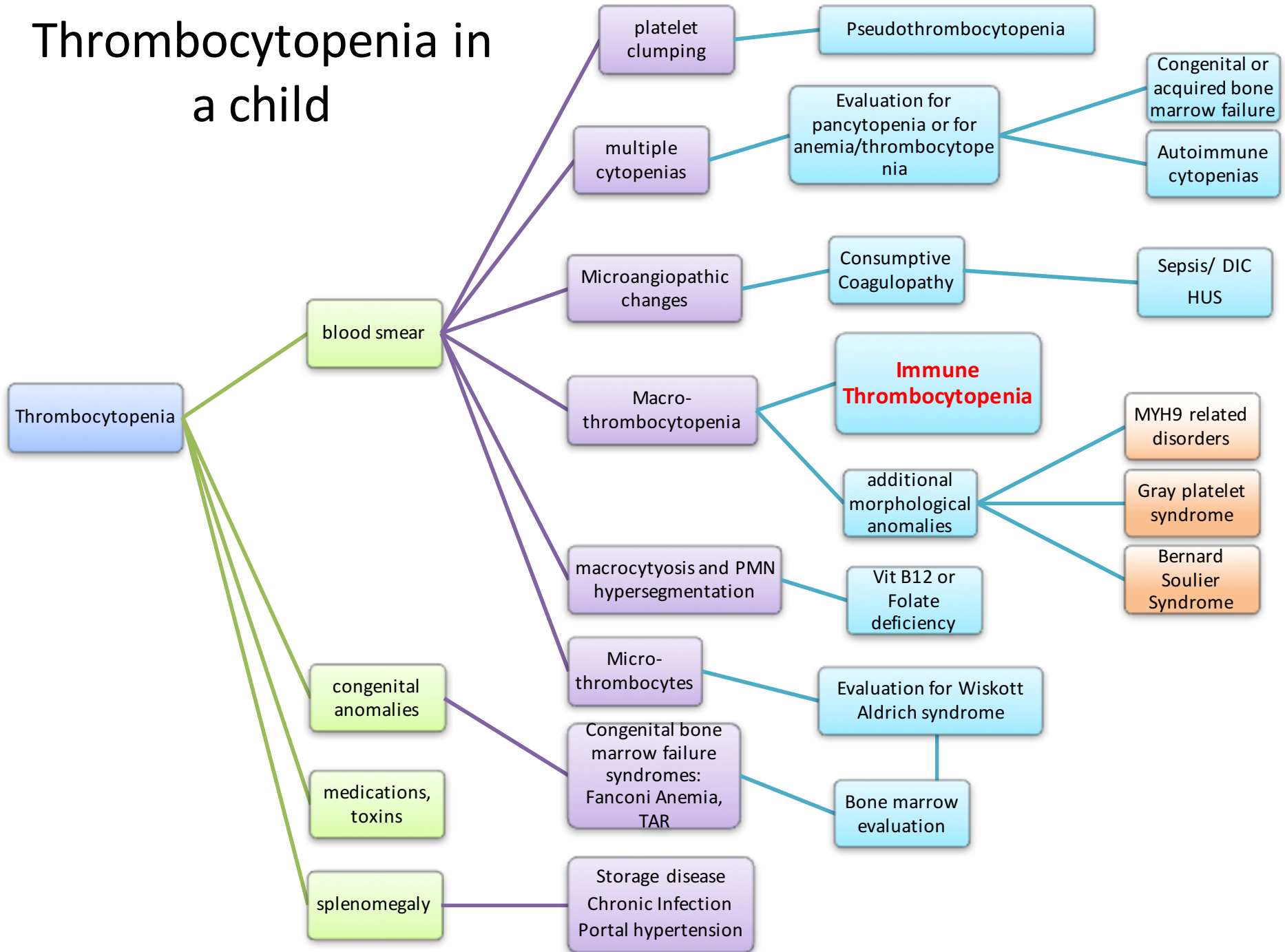
1. Does this child have a bleeding disorder?
2. What kind of investigation does she need?



	Patient	Ref. Range
WBC x 10 <sup>9</sup> /L	11.2	5.0-15
HGB (g/L)	115	115-125
HCT	0.34	0.34-0.4
RBC x 10 <sup>12</sup> /L	3.9	3.9-5.4
MCV (fL)	79.8	75-87
MCH (pg)	27.2	24-34
RDW (%)	13.1	11.4-14.4
Retic. x 10 <sup>9</sup> /L	65	25-75
Platelets x 10 <sup>9</sup> /L	<b>4</b>	150-400
Blood film	Lymphocytosis Occ. Large platelets	



# Thrombocytopenia in a child



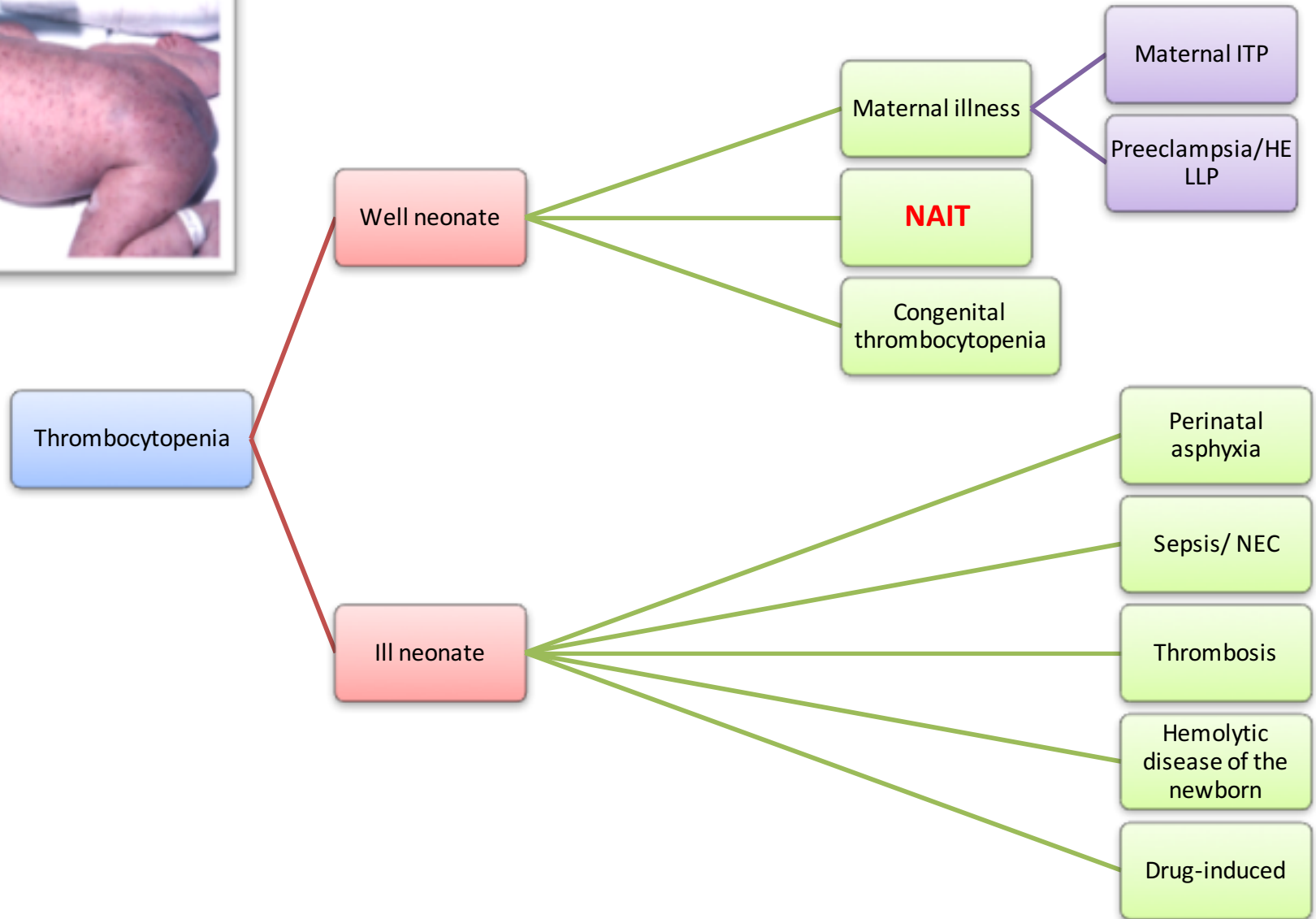
## Primary ITP in children

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- Incidence: 4/100,000 children
- Mean age: 5.7 years
- Seasonal variation
- More common in boys than girls
- Previously healthy children
  - >60% have a history of antecedent infectious illness or immunization in the preceding 4 weeks
- Typical abrupt onset of bleeding
- Isolated severe thrombocytopenia
  - >75% have platelet counts  $<20 \times 10^9/L$

# The special case of thrombocytopenia in a neonate

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## Tip #4: Thrombocytopenia has many causes in children

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- Acute onset of isolated thrombocytopenia in otherwise well children is most likely to be ITP
- Thrombocytopenia associated with other cytopenias raises concerns for bone marrow disorders
- Persistent thrombocytopenia raises the possibility of congenital causes; look for signs of associated anomalies

# Referral to Pediatric Hematology

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“Dear Pediatric Hematologist:

Please see this 3 month old boy who has developed extensive bruising and swelling at site of venipuncture. No family history of a bleeding diathesis.”

1. Does this child have a bleeding disorder?
2. What kind of investigation does he need?

	Patient	Ref. Range
WBC x 10 <sup>9</sup> /L	12.8	5.0-15
RBC x 10 <sup>12</sup> /L	3.8	2.7-4.9
HCT (L/L)	0.34	0.28-0.44
HGB (g/L)	120	90-140
MCV (fL)	85	74-115
RDW (%)	13.5	11.4-14.4
Retic. x 10 <sup>9</sup> /L	65	25-75
Platelets x 10 <sup>9</sup> /L	320	150-400
Blood film	Typical for age	
PT (sec)/ INR	12/ 1.1	10-12.5/0.9-1.1
PTT (sec)	<b>87</b>	26-38



Factor VIII 1% (50-150)  
Factor IX 62% (50-150)

# What does hemophilia look like in young children?





## Tip #5: Hemophilia in young boys looks different

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- An unusual bruising pattern or soft tissue bleeding
  - ✓ Always be alert to the possibility of non-accidental trauma
- Joint bleeds are rare in children before they are walking
- There is not always a family history:
  - ✓ 30% of cases are the result of new mutations

An aPTT is helpful if the question is hemophilia

## When to consider referral to Pediatric Hematology

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1. Thrombocytopenia (platelet count  $<100 \times 10^9/L$ ), particularly if accompanied by additional cytopenias
2. A severe or unusual pattern of bleeding in a young child\*
3. A child with bleeding symptoms and a significant family history of bleeding [with or without a diagnosis]; sooner if surgery is planned\*
4. Menorrhagia in adolescent girls, severe enough to cause iron-deficiency anemia or impact ADL \*

\* Likely to have a positive BAT score

## Take Home Messages

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1. Most mucocutaneous bleeding in children is not the result of a hemostatic disorder.
2. A careful history and a BAT may be more helpful than laboratory screening tests (except CBC) in determining the need specialized testing.
3. Remember that your pediatric hematologist cannot always give you a definitive diagnosis for mucocutaneous bleeding.
4. Be alert to severe bleeding conditions that require urgent attention.

# ISTH Bleeding Assessment Tool:

[https://www.isth.org/resource/resmgr/ssc/isth-ssc\\_bleeding\\_assessment.pdf](https://www.isth.org/resource/resmgr/ssc/isth-ssc_bleeding_assessment.pdf)

Questions?

*[sisraels@cancercare.mb.ca](mailto:sisraels@cancercare.mb.ca)*

## Question:

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Frequent epistaxis in a child is more likely to be a sign of a bleeding disorder if any of the following are true, except:

- A. He has seasonal allergies.
- B. He previously had nasal cautery to manage the epistaxis.
- C. His brother also has epistaxis.
- D. He has a positive history for bleeding at sites other than the nose.

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