Faculté de Médecine, Ulg

Aide aux ARCS

ALGORITHMES OU ARBRES DÉCISIONNELS EN PÉDIATRIE

Aide aux arcs de médecine

Algorithmes, arbres décisionnels, Images spécifiques

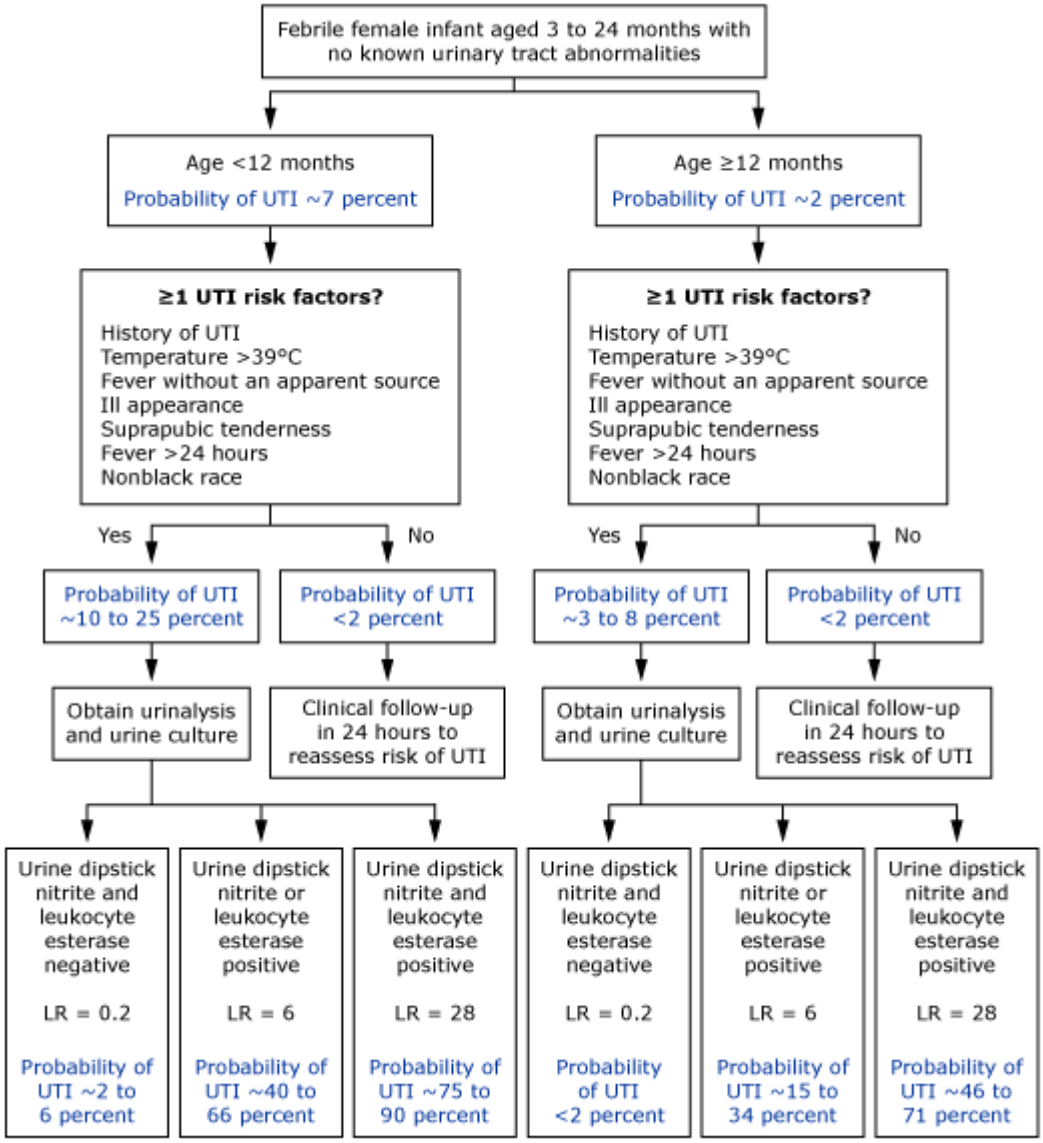
Faculté de Médecine
Université de Liège

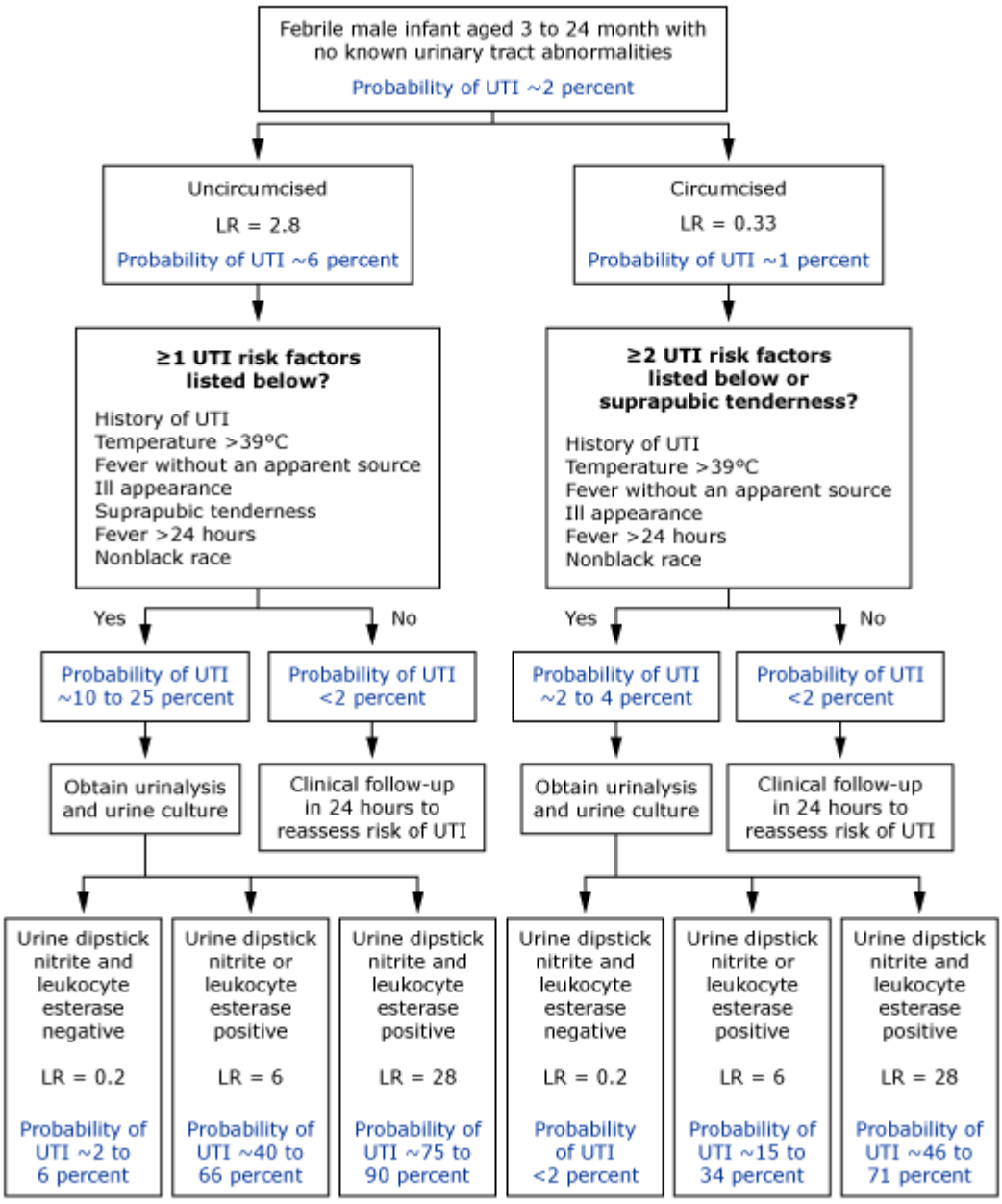
Pédiatrie et néonatalogie universitaire

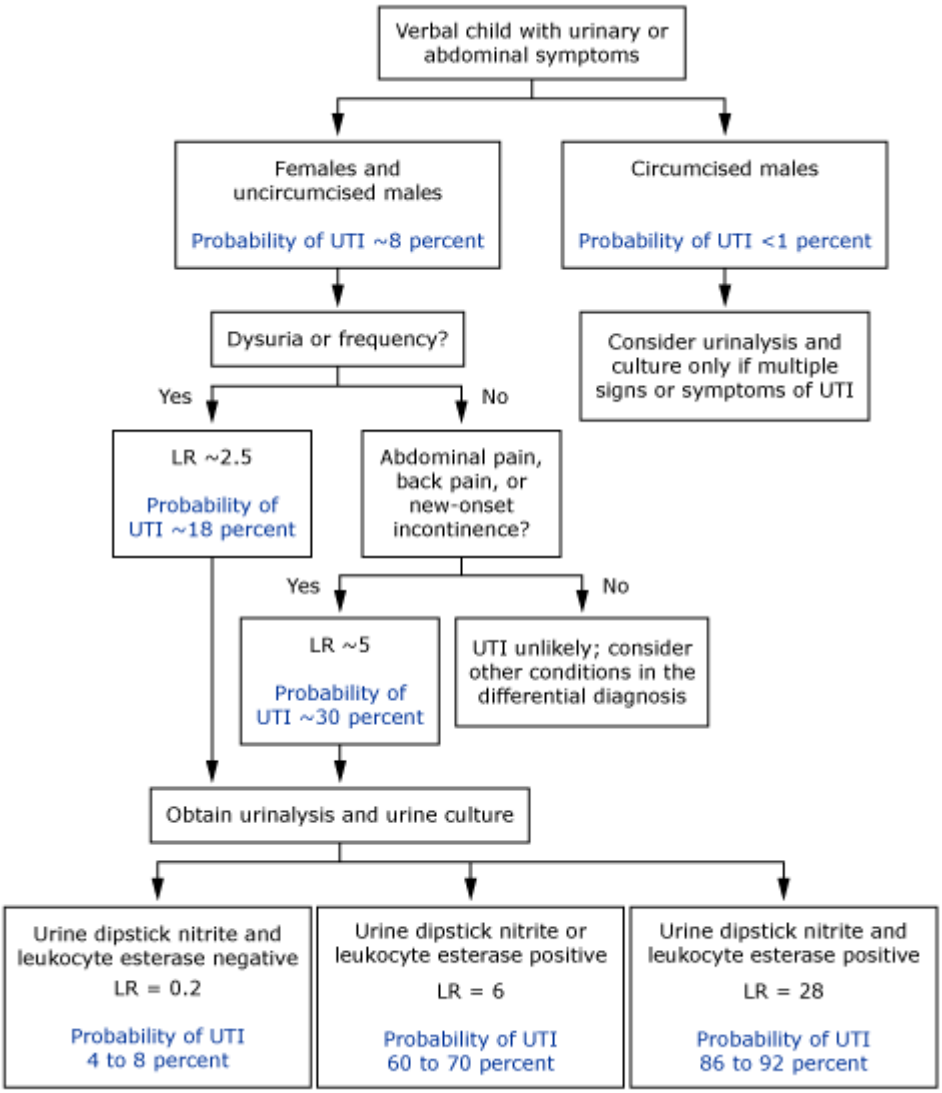
Professeur Oreste Battisti

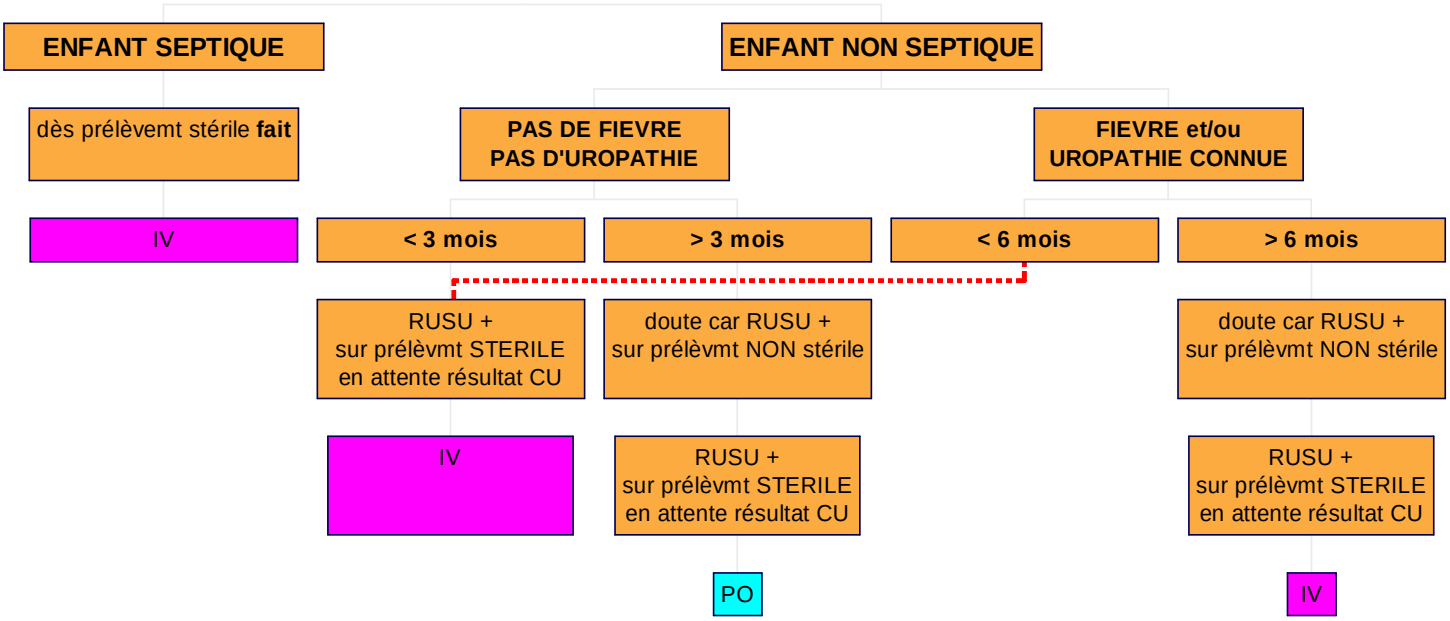
➔ L'arbre urinaire

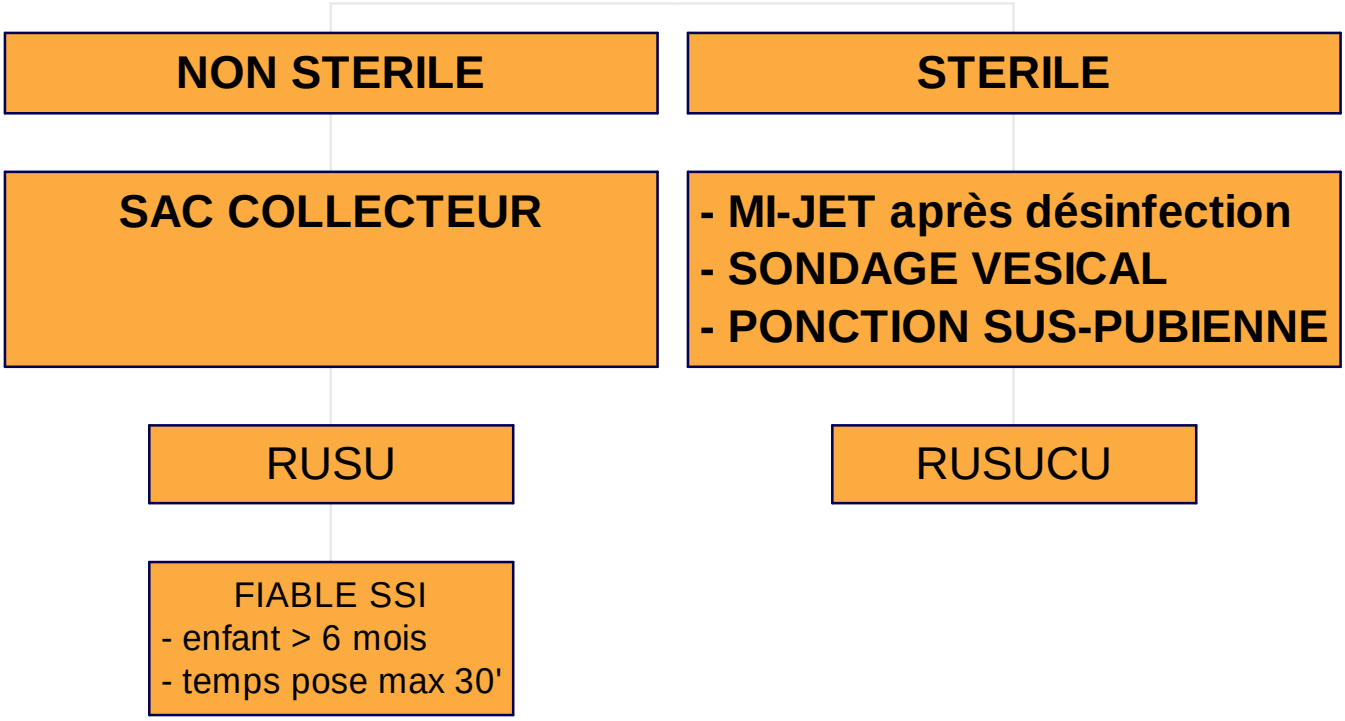
L'infection urinaire

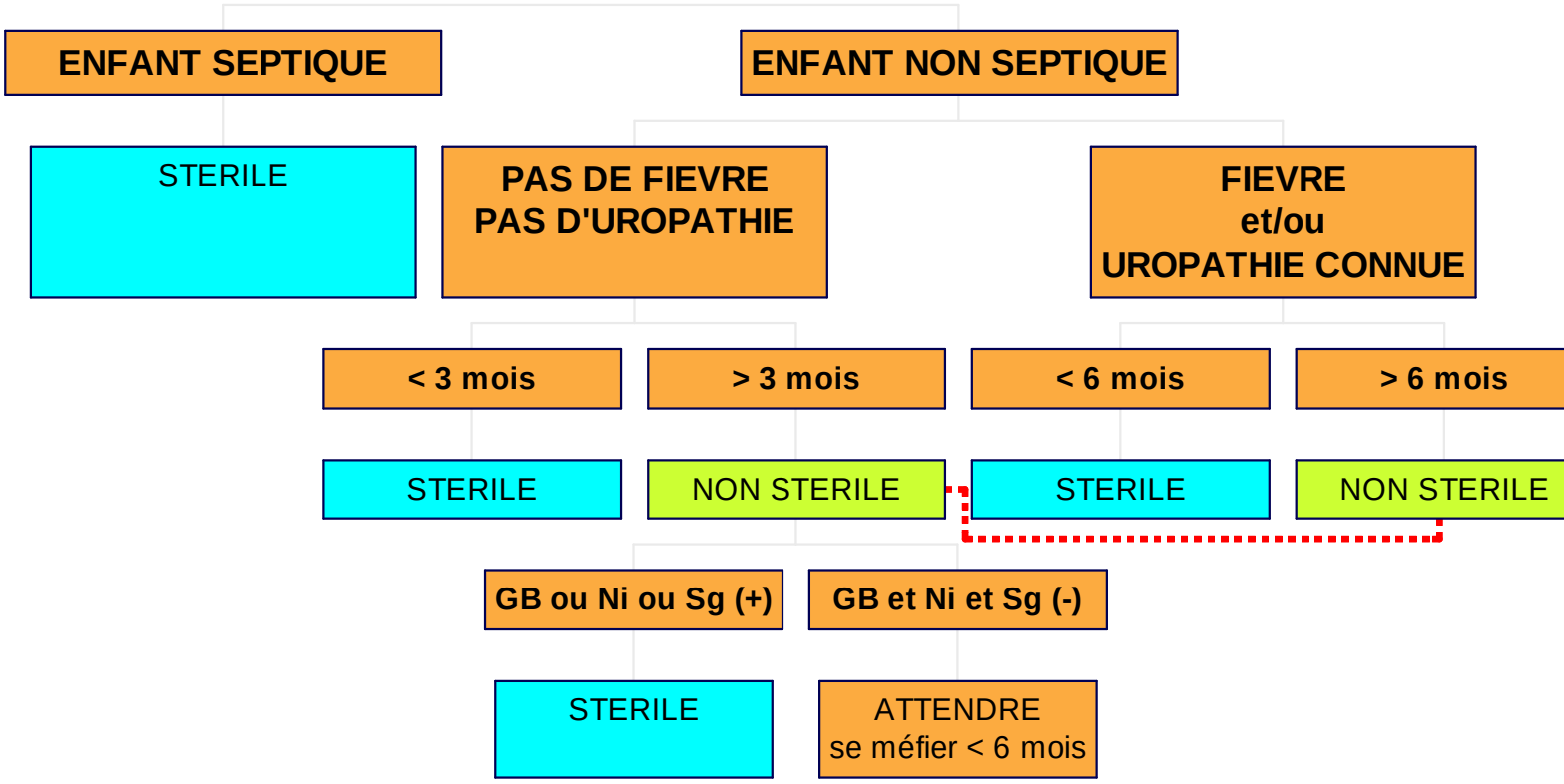


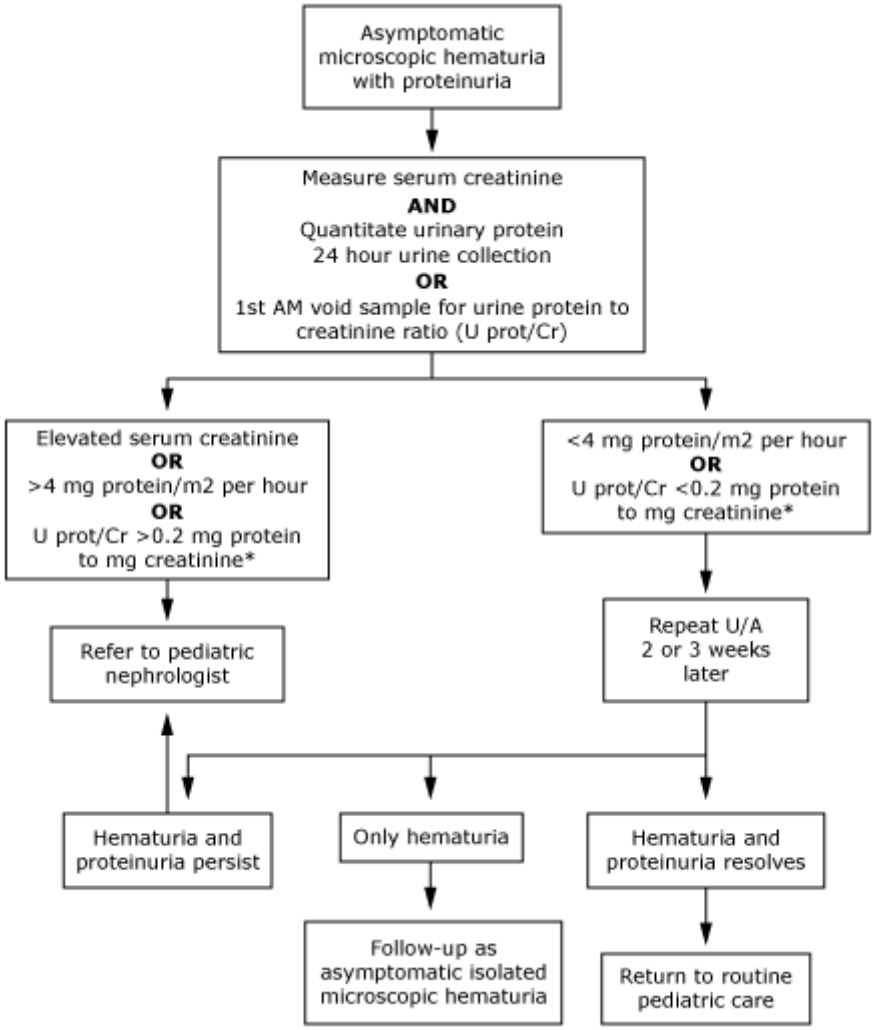

Febrile male infant aged 3 to 24 month with no known urinary tract abnormalities
Probability of UTI ~2 percent
Uncircumcised
LR = 2.8
Probability of UTI ~6 percent
Circumcised
LR = 0.33
Probability of UTI ~1 percent
≥1 UTI risk factors listed below?
History of UTI
Temperature >39°C
Fever without an apparent source
Ill appearance
Suprapubic tenderness
Fever >24 hours
Nonblack race
≥2 UTI risk factors listed below or suprapubic tenderness?
History of UTI
Temperature >39°C
Fever without an apparent source
Ill appearance
Fever >24 hours
Nonblack race
Yes
No
Yes
No
Probability of UTI ~10 to 25 percent
Probability of UTI <2 percent
Probability of UTI ~2 to 4 percent
Probability of UTI <2 percent
Obtain urinalysis and urine culture
Clinical follow-up in 24 hours to reassess risk of UTI
Obtain urinalysis and urine culture
Clinical follow-up in 24 hours to reassess risk of UTI
Urine dipstick nitrite and leukocyte esterase negative
LR = 0.2
Probability of UTI ~2 to 6 percent
Urine dipstick nitrite or leukocyte esterase positive
LR = 6
Probability of UTI ~40 to 66 percent
Urine dipstick nitrite and leukocyte esterase positive
LR = 28
Probability of UTI ~75 to 90 percent
Urine dipstick nitrite and leukocyte esterase negative
LR = 0.2
Probability of UTI <2 percent
Urine dipstick nitrite or leukocyte esterase positive
LR = 6
Probability of UTI ~15 to 34 percent
Urine dipstick nitrite and leukocyte esterase positive
LR = 28
Probability of UTI ~46 to 71 percent


Verbal child with urinary or abdominal symptoms
Females and uncircumcised males
Probability of UTI ~8 percent
Circumcised males
Probability of UTI <1 percent
Dysuria or frequency?
Consider urinalysis and culture only if multiple signs or symptoms of UTI
Yes
No
LR ~2.5
Probability of UTI ~18 percent
Abdominal pain, back pain, or new-onset incontinence?
Yes
No
LR ~5
Probability of UTI ~30 percent
UTI unlikely; consider other conditions in the differential diagnosis
Obtain urinalysis and urine culture
Urine dipstick nitrite and leukocyte esterase negative
LR = 0.2
Probability of UTI 4 to 8 percent
Urine dipstick nitrite or leukocyte esterase positive
LR = 6
Probability of UTI 60 to 70 percent
Urine dipstick nitrite and leukocyte esterase positive
LR = 28
Probability of UTI 86 to 92 percent

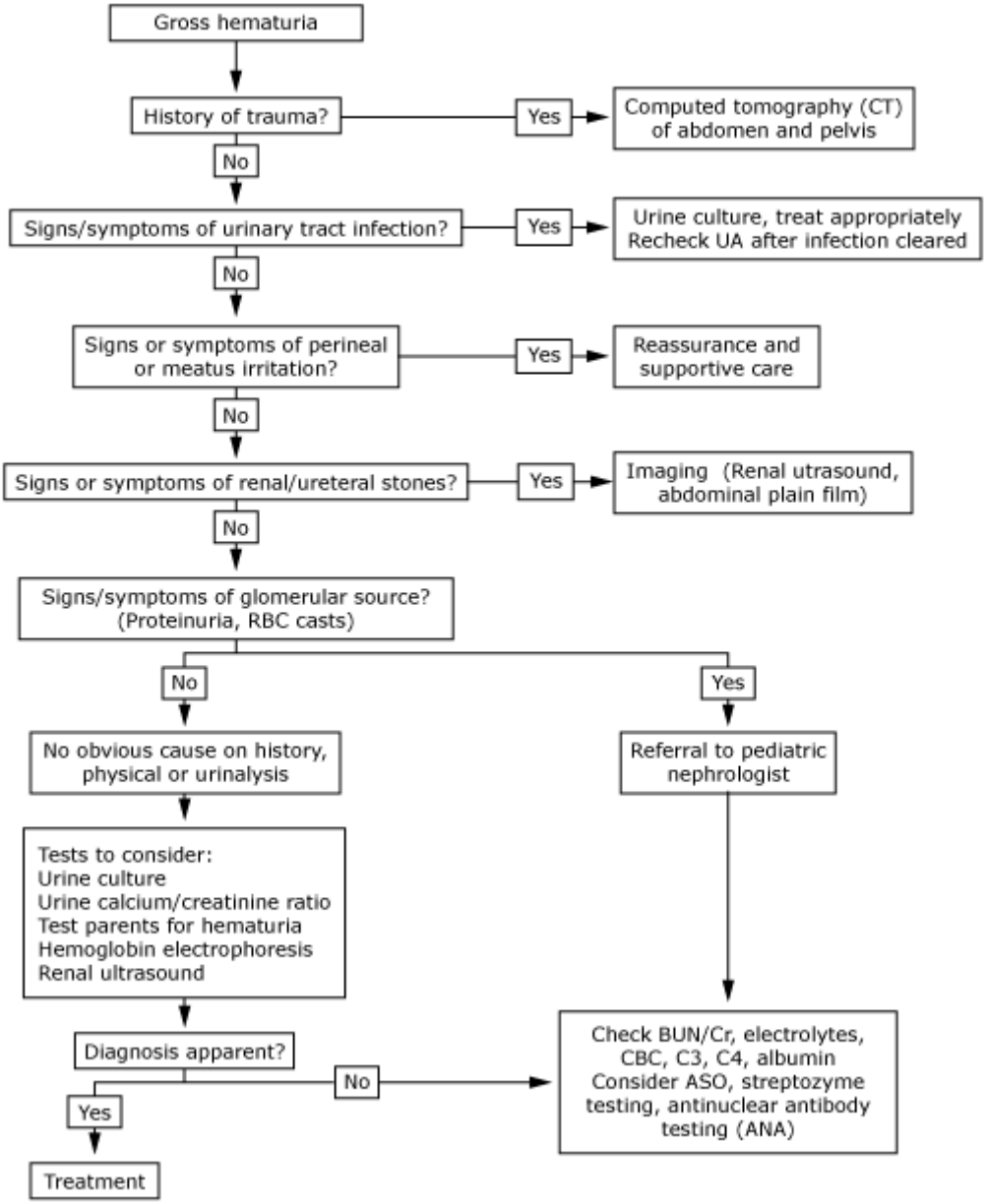

ENFANT SEPTIQUE
ENFANT NON SEPTIQUE
dès prélèvemt stérile fait
PAS DE FIEVRE
PAS D'UROPATHIE
FIEVRE et/ou
UROPATHIE CONNUE
IV
< 3 mois
> 3 mois
< 6 mois
> 6 mois
RUSU +
sur prélèvmt STERILE
en attente résultat CU
doute car RUSU +
sur prélèvmt NON stérile
doute car RUSU +
sur prélèvmt NON stérile
IV
RUSU +
sur prélèvmt STERILE
en attente résultat CU
RUSU +
sur prélèvmt STERILE
en attente résultat CU
PO
IV


NON STERILE
STERILE
SAC COLLECTEUR
- MI-JET après désinfection
- SONDAGE VESICAL
- PONCTION SUS-PUBIENNE
RUSU
RUSUCU
FIABLE SSI
- enfant > 6 mois
- temps pose max 30'

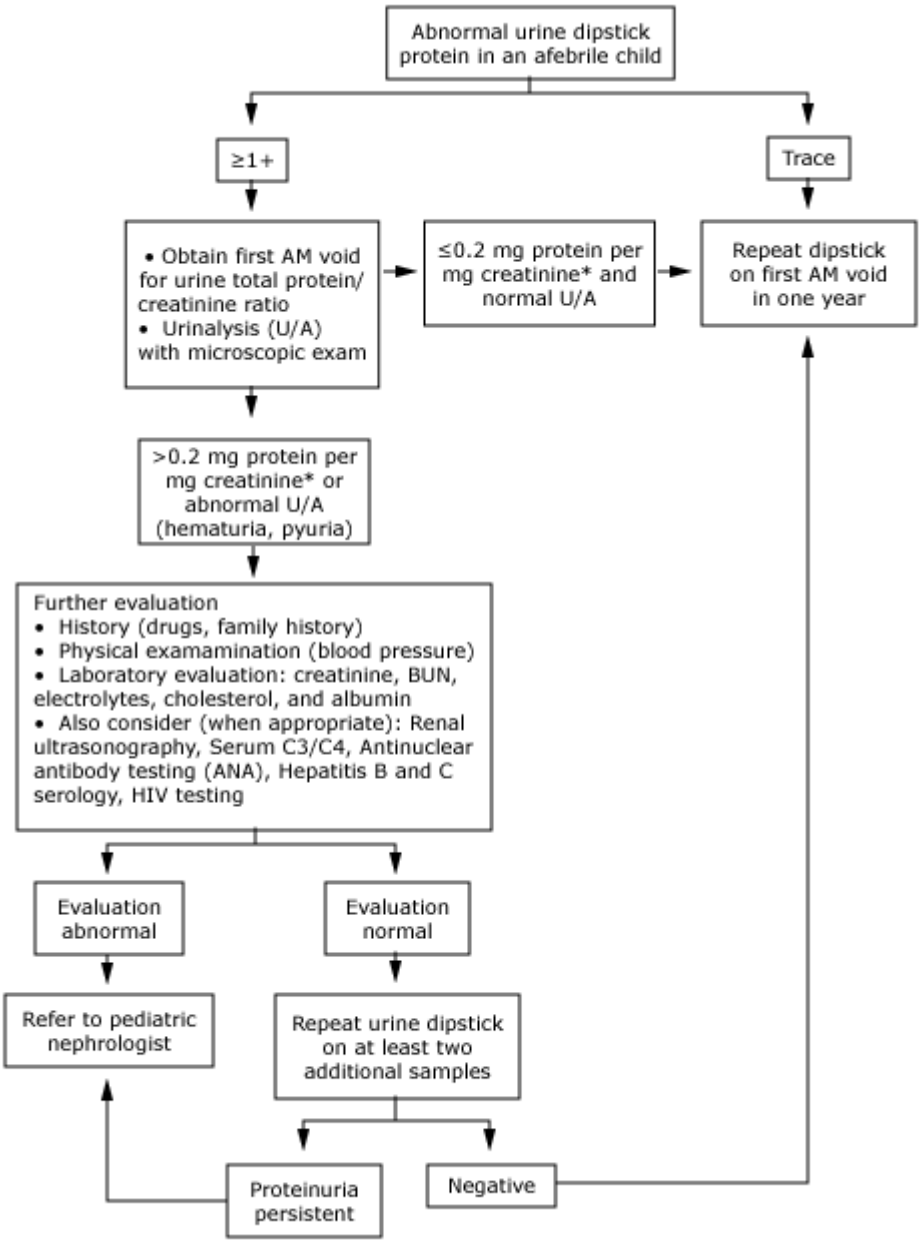

ENFANT SEPTIQUE
ENFANT NON SEPTIQUE
STERILE
PAS DE FIEVRE
PAS D'UROPATHIE
FIEVRE
et/ou
UROPATHIE CONNUE
< 3 mois
> 3 mois
< 6 mois
> 6 mois
STERILE
NON STERILE
STERILE
NON STERILE
GB ou Ni ou Sg (+)
GB et Ni et Sg (-)
STERILE
ATTENDRE
se méfier < 6 mois



Hématurie microscopique



Hématurie macroscopique



Protéinurie

Dilatation des voies urinaires

Syndrome de la jonction pyélo-urétérale

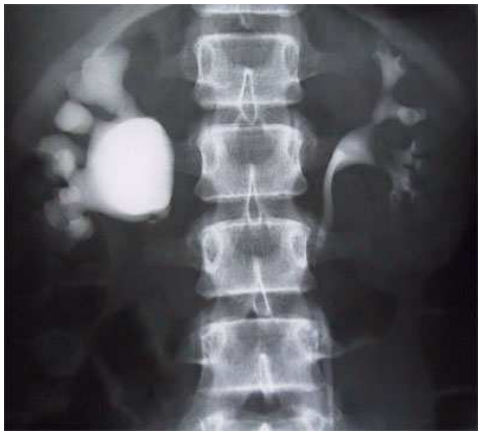


Fig. 1 : Jonction pyélo-urétérale droite (UIV)

Anomalies structurales des reins

- **Dysplasie multikystique**

.

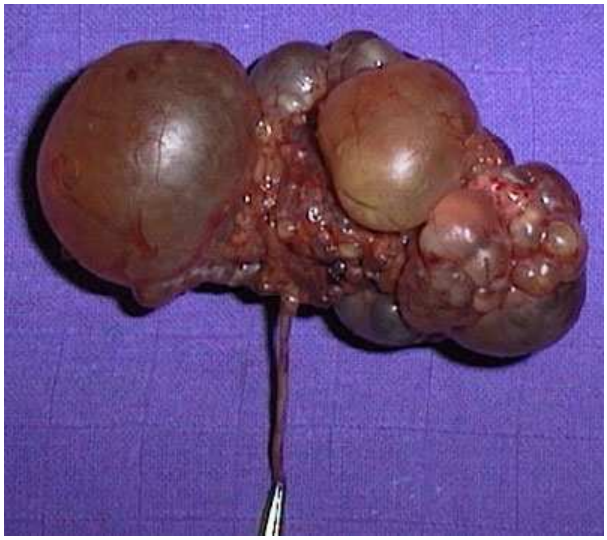
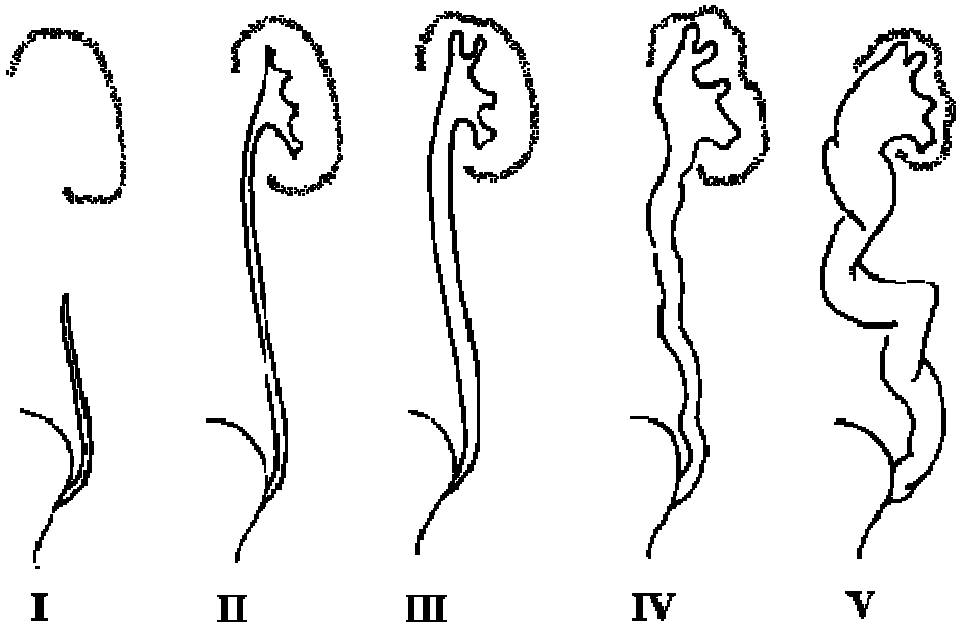


Fig. 2 : Dysplasie multikystique du rein

Examens complémentaires

La cystographie rétrograde est l'examen clé. Elle seule permet d'affirmer le reflux et de le classifier (Fig. 3). Il faut attendre au moins un mois après une infection urinaire pour la réaliser. L'échographie est obligatoire, elle permet d'apprécier le retentissement rénal. L'urographie intraveineuse n'a plus guère d'indications, sauf si on suspecte un reflux secondaire à une duplicité. La scintigraphie rénale au DMSA est l'examen de référence pour apprécier l'existence ou non de séquelles rénales .



- **Classification des reflux vésico-urétéraux**

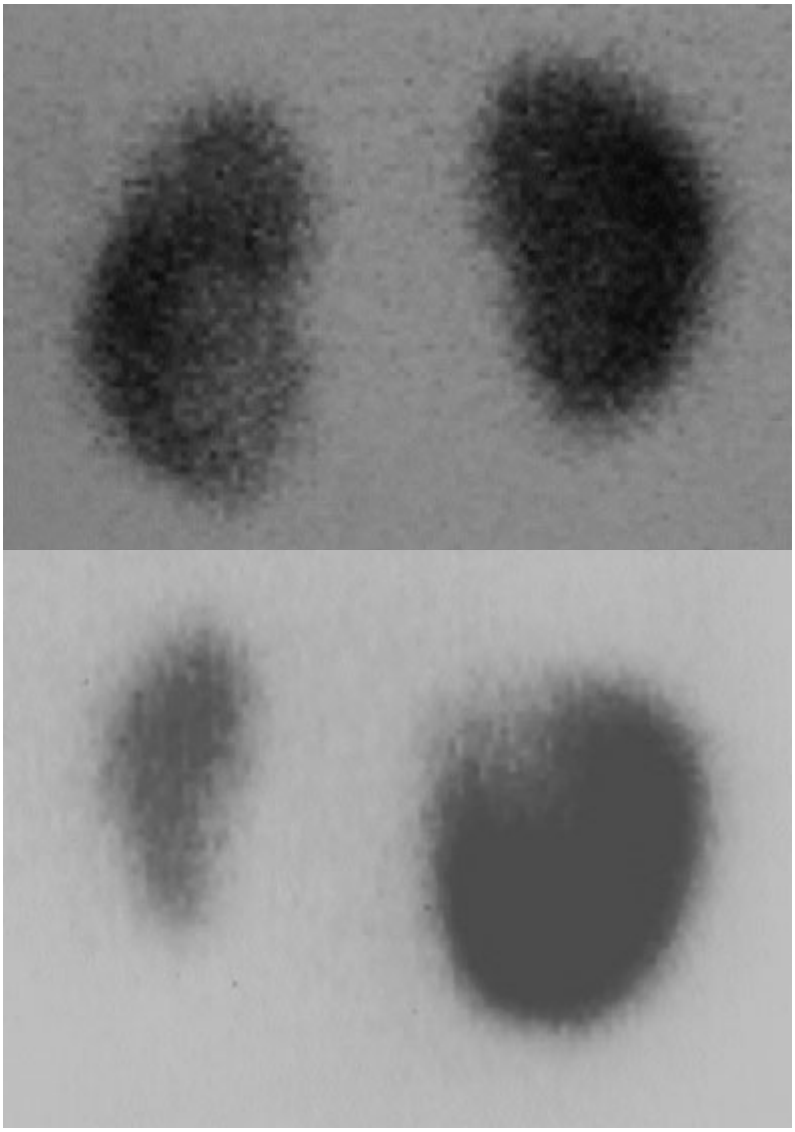


Fig. 10 : Deux exemples de scintigraphie (Cystographie sus pubienne) au DMSA.
Hypofixation du pôle inférieur droit en haut, atrophie rénale droite globale en bas.

- **Uretères doubles**

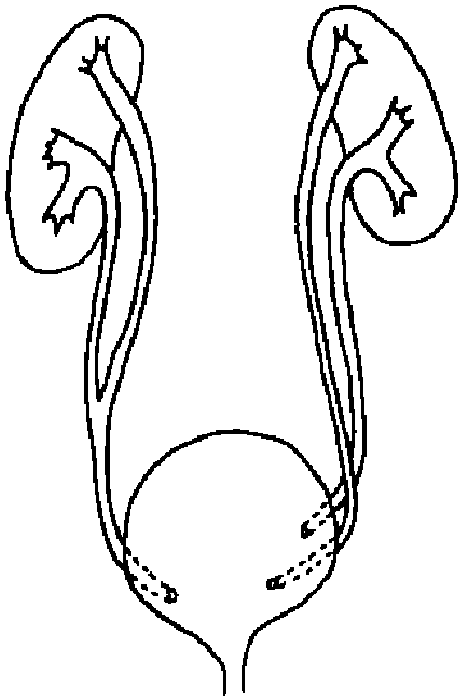


Fig. 4 : Bifidité droite, duplicité gauche

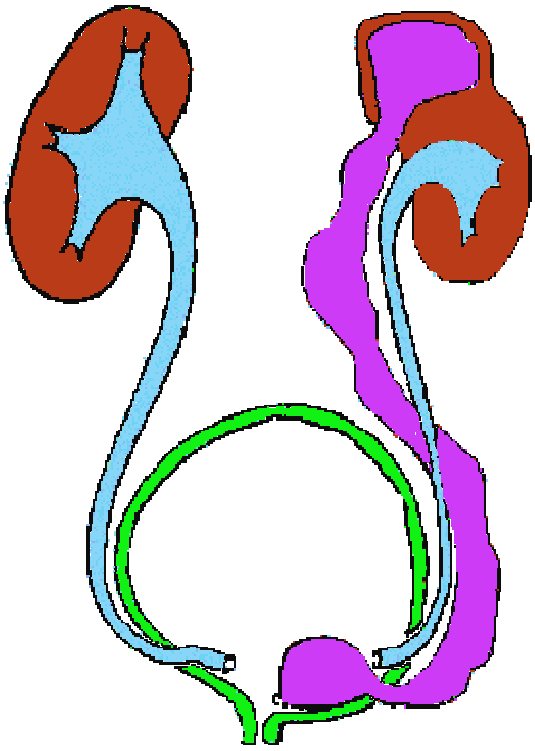


Fig. 5 : Urétérocèle sur duplicité

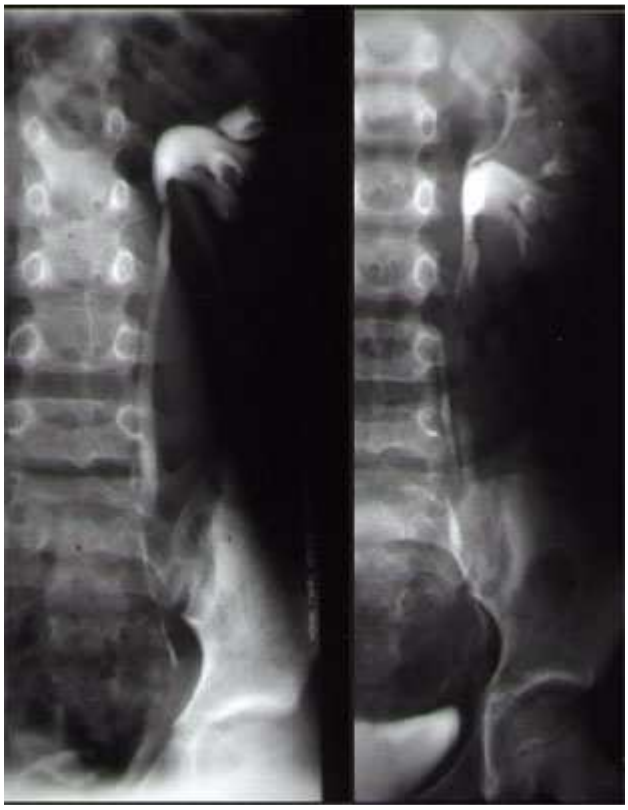


Fig. 6 : Reflux dans un pyélon inférieur de duplicité
(Cystographie rétrograde à gauche, UIV à droite)

Le bilan des duplicités repose sur l'échographie et la cystographie rétrograde. L'urographie intraveineuse est souvent utile. Les duplicités pathogènes nécessitent en règle un traitement chirurgical.

- Mégauretère primitif obstructif

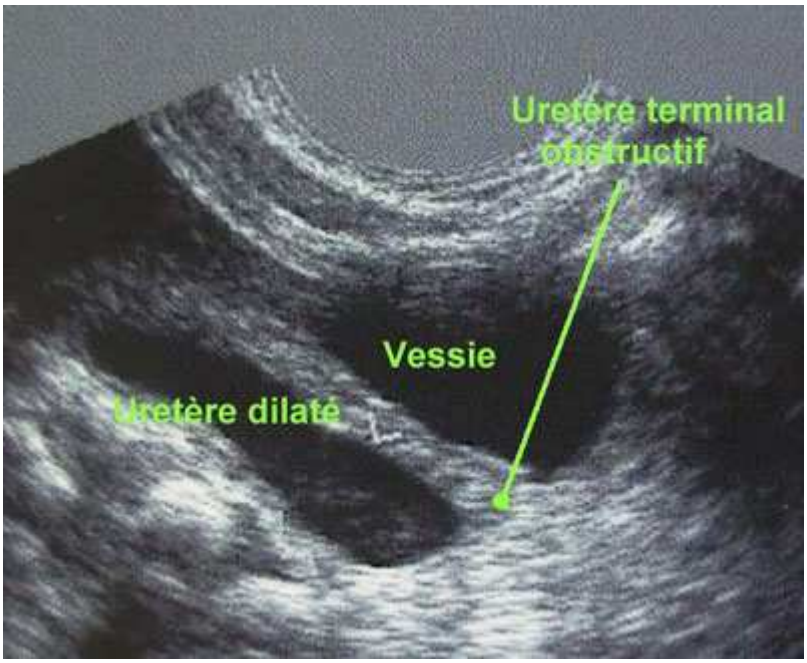


Fig. 7 : Mégauretère primitif obstructif congénital (aspect en échographie)

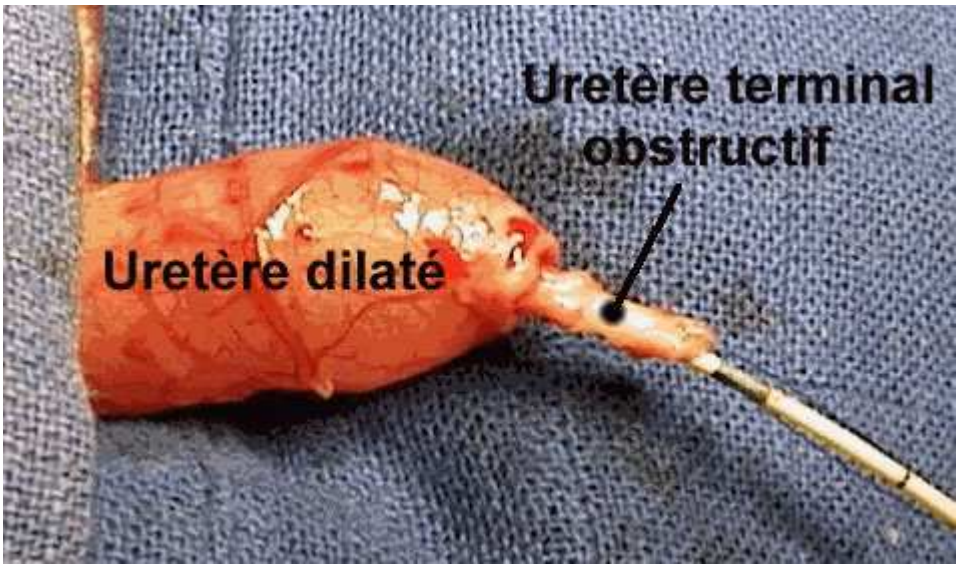


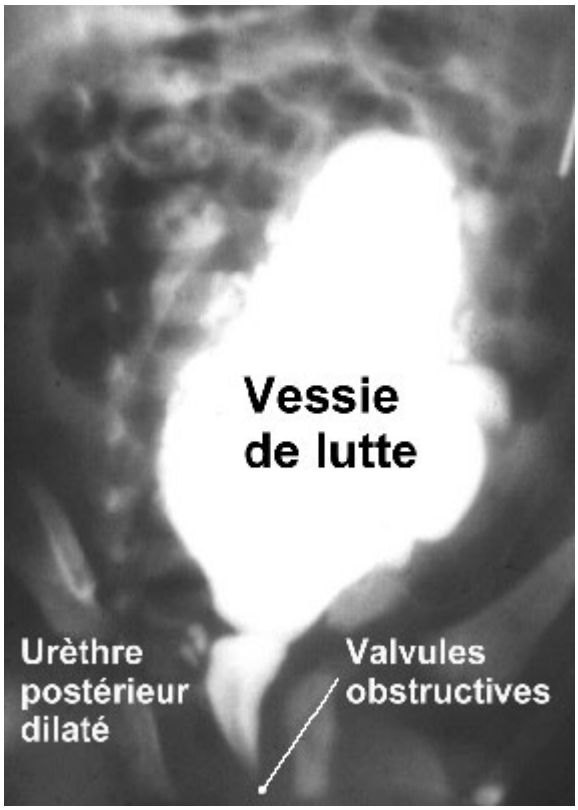
Fig. 8 : Mégauretère primitif obstructif congénital (vue per-opératoire)

- Valves de l'urèthre postérieur

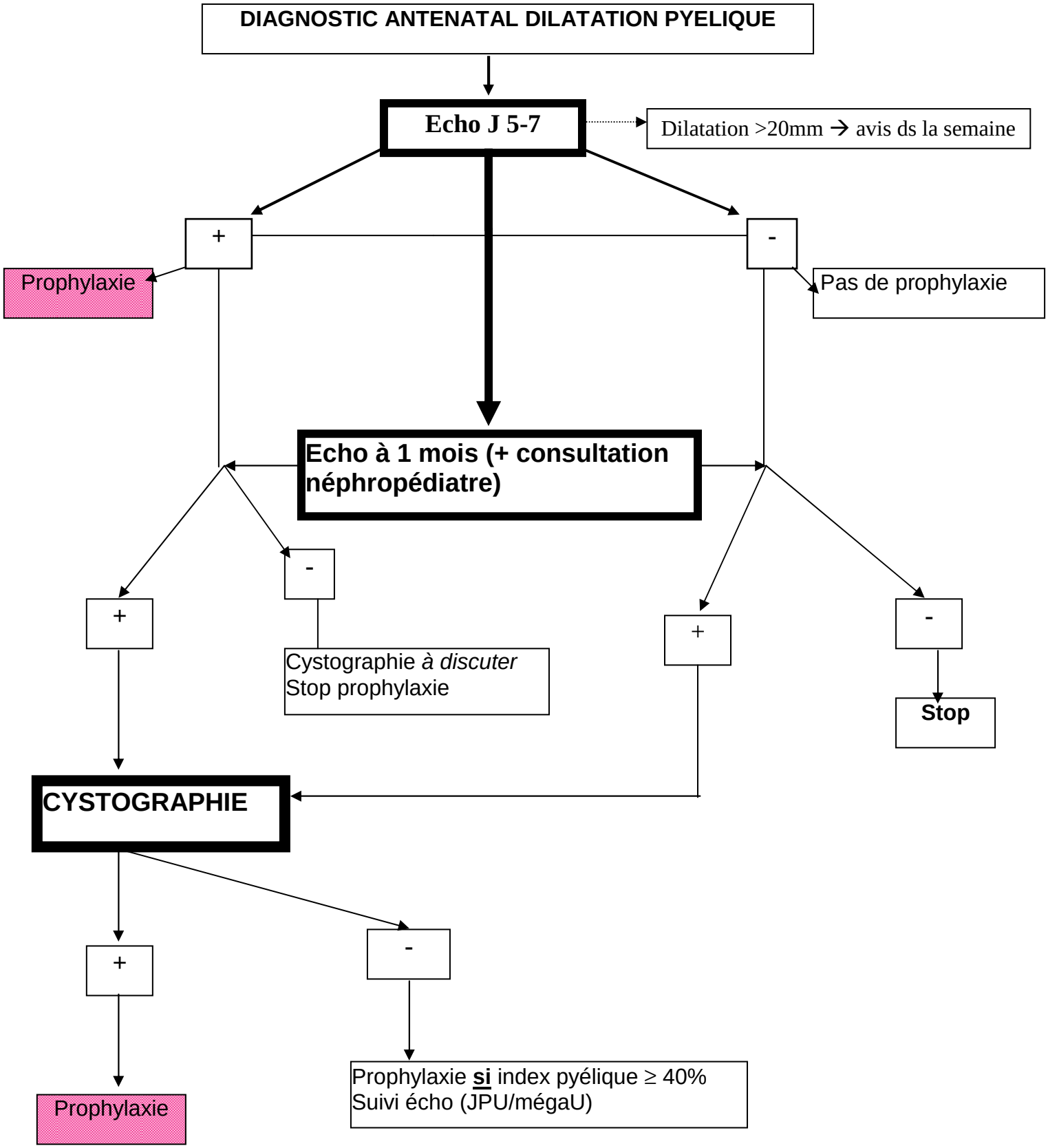
Elles réalisent un obstacle sous-vésical d'installation très précoce pendant la vie embryonnaire induisant un retentissement d'amont parfois très important. Atteignant uniquement les enfants de sexe masculin, c'est potentiellement une des uropathies les plus graves. Ces valves sont des replis muqueux en nid de pigeon au niveau du veru montanum, elles empêchent l'écoulement normal des urines dans l'urèthre postérieur. Elles s'accompagnent d'une vessie de lutte, d'une dilatation urétéro-rénale bilatérale avec souvent un reflux massif. Le parenchyme rénal peut être aminci et dysplasique, la fonction rénale peut être altérée précocement et de façon importante.

Le diagnostic est presque toujours suspecté par les échographies fœtales. A un stade précoce, des critères de mauvais pronostic (dilatation majeure, oligoamnios, hyperéchogénicité rénale, biochimie défavorable des urines fœtales prélevées par ponction sous échographie) peuvent parfois amener à proposer une interruption médicale de grossesse.

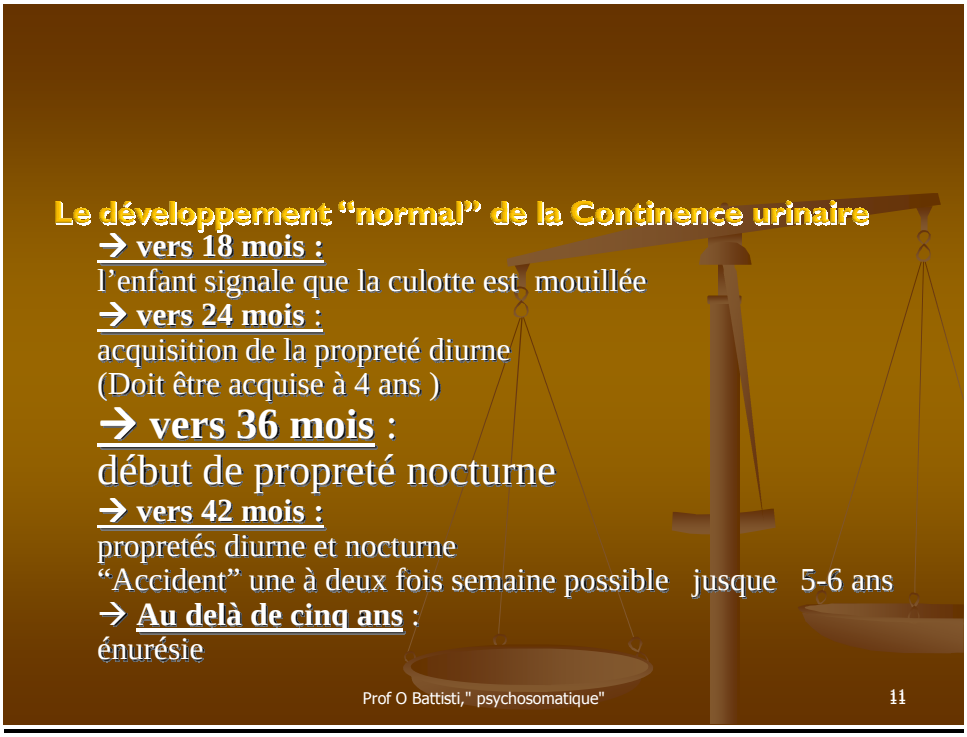
Après la naissance, on observe en général un globe vésical et l'absence de mictions normales. On peut parfois palper deux gros reins. Il s'agit d'une urgence néonatale, imposant un transfert rapide en milieu spécialisé de chirurgie pédiatrique. Un traitement antibiotique préventif est instauré. Le diagnostic des valves est confirmé par une cystographie, que l'on réalise au mieux par ponction sus pubienne directe de la vessie (Fig. 9). Le traitement consiste en une section des valves par voie endoscopique. La fonction rénale doit être surveillée étroitement pendant de longues années. Les problèmes de continence, fréquents pendant l'enfance, s'arrangent généralement après la puberté.



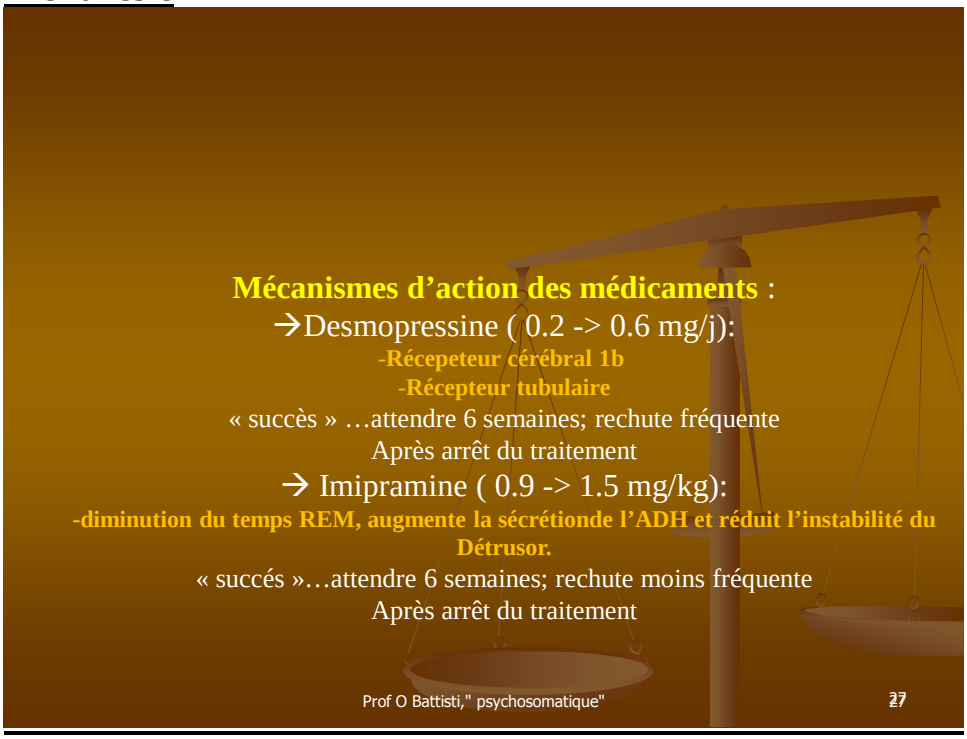
Valvules de l’urèthre postérieur



Prophylaxie : Clamoxyl 25 mg/Kg/j en 2x ou TMP 2 mg/Kg/j en 2x


Le développement "normal" de la Continence urinaire
→ vers 18 mois :
l'enfant signale que la culotte est mouillée
→ vers 24 mois :
acquisition de la propreté diurne
(Doit être acquise à 4 ans)
→ vers 36 mois :
début de propreté nocturne
→ vers 42 mois :
propretés diurne et nocturne
"Accident" une à deux fois semaine possible jusque 5-6 ans
→ Au delà de cinq ans :
énurésie
Prof O Battisti," psychosomatique"
11

L'énurésie


Mécanismes d'action des médicaments :
→Desmopressine (0.2 -> 0.6 mg/j):
-Récepeteur cérébral 1b
-Récepteur tubulaire
« succès » …attendre 6 semaines; rechute fréquente
Après arrêt du traitement
→ Imipramine (0.9 -> 1.5 mg/kg):
-diminution du temps REM, augmente la sécrétionde l'ADH et réduit l'instabilité du Détrusor.
« succés »…attendre 6 semaines; rechute moins fréquente
Après arrêt du traitement
Prof O Battisti," psychosomatique"
27

➔Difficultés respiratoires, circulatoires et troubles du rythme

Laryngites : score de WESTLEY *(Am J Dis Child 1978 ;132 :484-7)*

	0	1	2	3	4	5
Stridor	absente	à l'agitation	au calme			
Tirage	absent	léger	modéré	sévère		
Entrée d'air	normale	diminuée	très diminuée			
Cyanose	absente				à l'agitation	au calme
Conscience	normale					altéré

- Forme sévère = score supérieur ou égal à 8

Crises d'asthme : score de WOOD *(Wood DW, et al. Am J Dis Child 1972 ;123 :227- 8)*

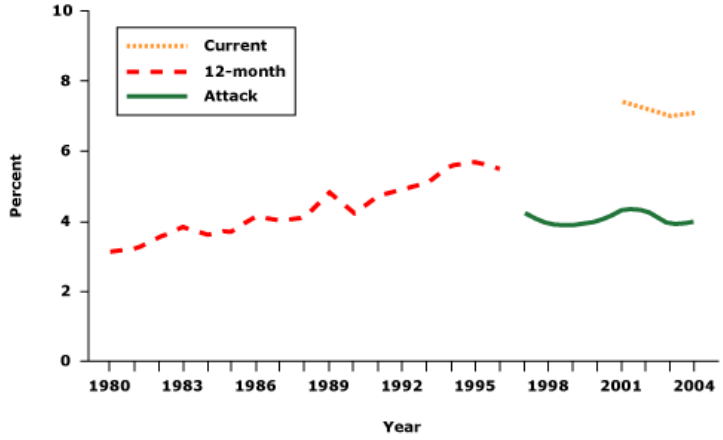
	0	1	2
Wheezing	Aucun	modéré	marqué
Tirage	Aucun	modéré	sévère
Entrée d'air	Normale	diminuée	très diminuée
Cyanose (ou PaO2<70) (*)	absente	à l'air ambiant	sous FiO2 40%
Conscience	normale	altérée ou agitation	coma

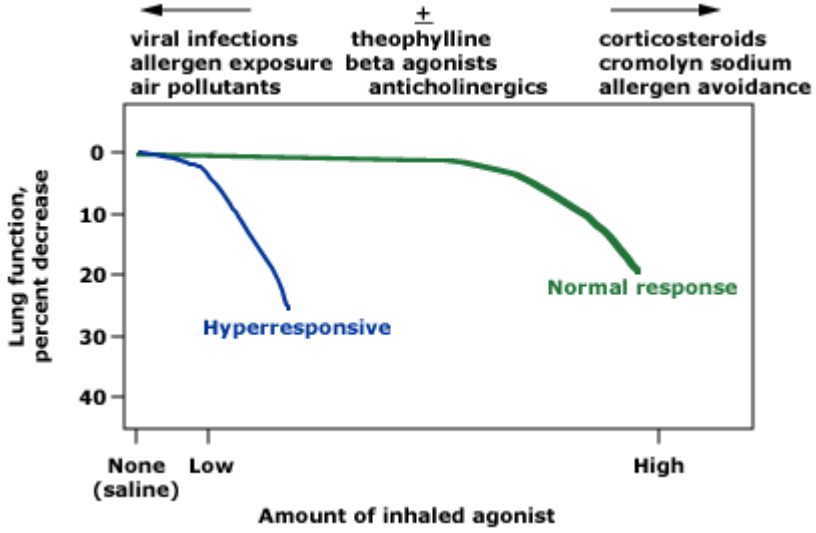
(*) La mesure de la PaO2 pourrait être remplacée par la mesure de la saturation : Sat°<90%

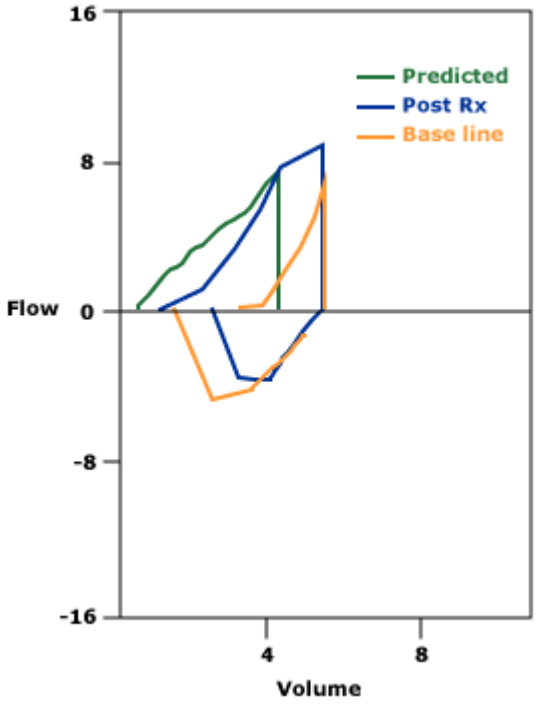
- Crise sévère = score supérieur à 4
- Insuffisance respiratoire aigue = score supérieur à 7
  et PaCO2>65 ou PaO2<100 sous Fi02 100%

Bronchiolites : score de GADOMSKi *(Pediatrics 1994 : 93 ;907-911)*

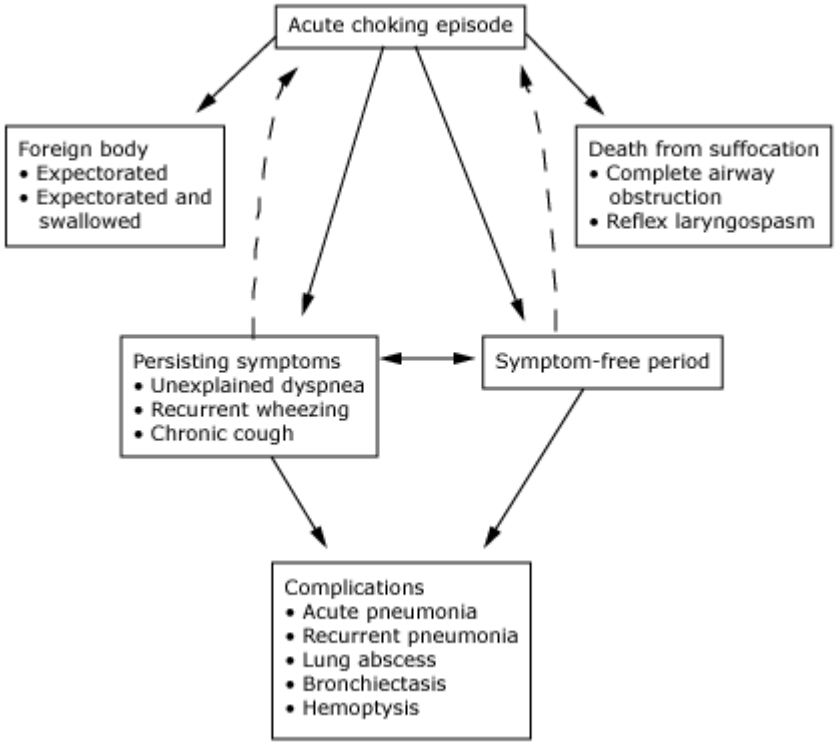
	0	1	2	3
Battement des ailes du nez	aucun		modéré ou intermittent	marqué et persistant
Grunting	aucun	intermittent	persistant	
Tirage	aucun	discret	modéré	sévère
Entrée d'air	normale			diminuée
Cyanose (ou PaO2<70) (*)	absente	à l'air ambiant	sous FiO2 40%	
Conscience	normale	altérée ou agitation	coma	


Current
12-month
Attack
Percent
10
8
6
4
2
0
1980
1983
1986
1989
1992
1995
1998
2001
2004
Year

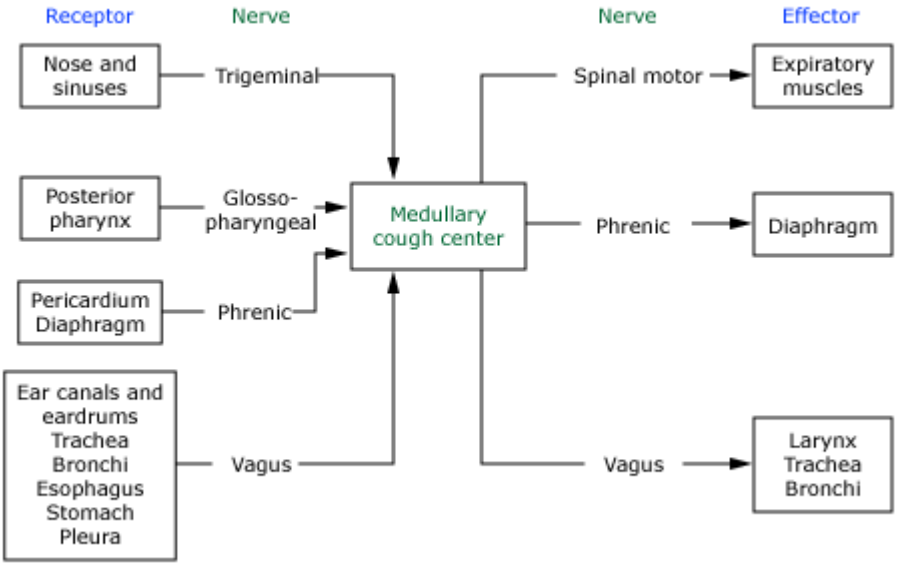

viral infections
allergen exposure
air pollutants
±
theophylline
beta agonists
anticholinergics
corticosteroids
cromolyn sodium
allergen avoidance
Lung function, percent decrease
0
10
20
30
40
Hyperresponsive
Normal response
None (saline)
Low
High
Amount of inhaled agonist


Predicted
Post Rx
Base line
16
8
Flow
0
-8
-16
4
8
Volume

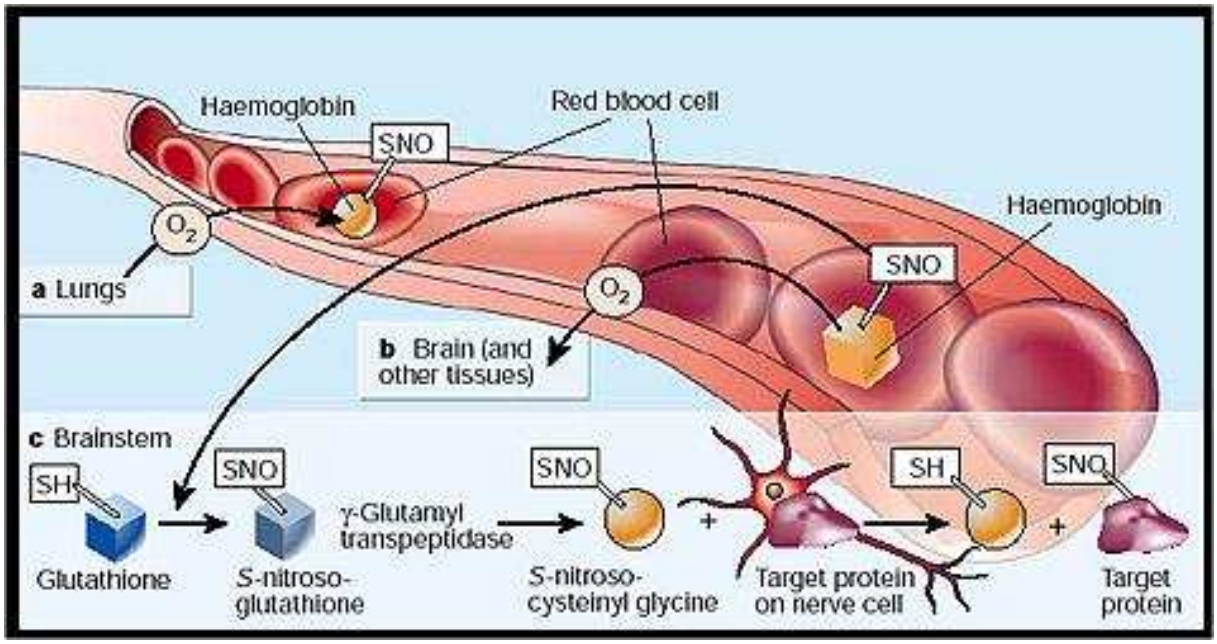
Corps étranger des voies respiratoires

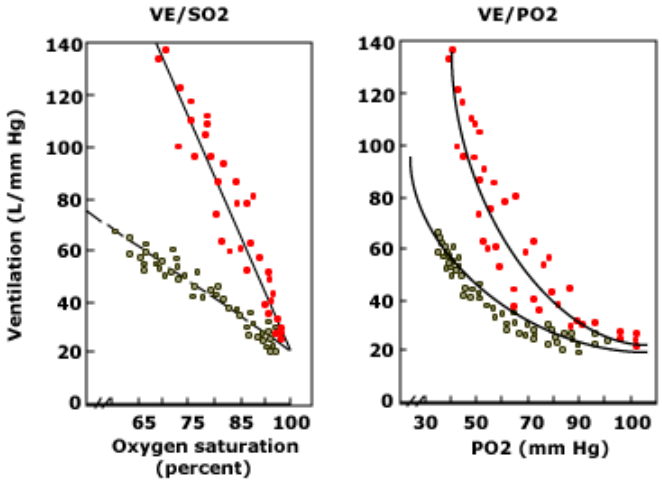


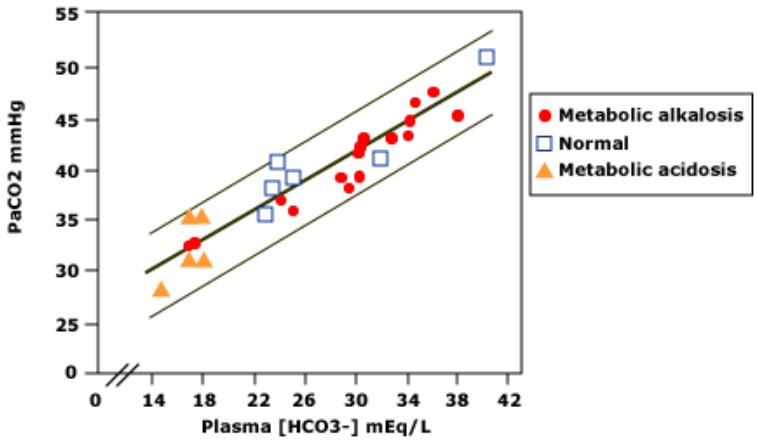
La toux

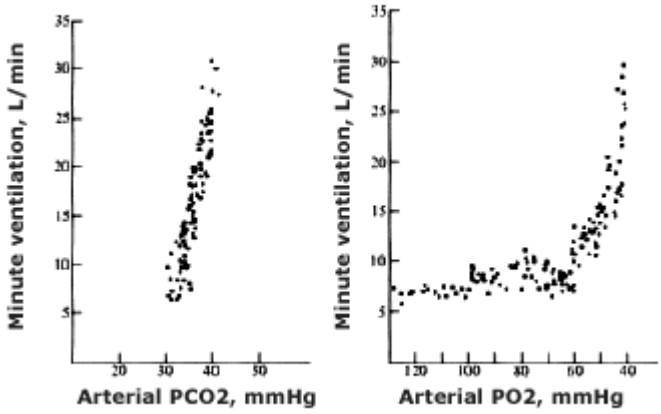


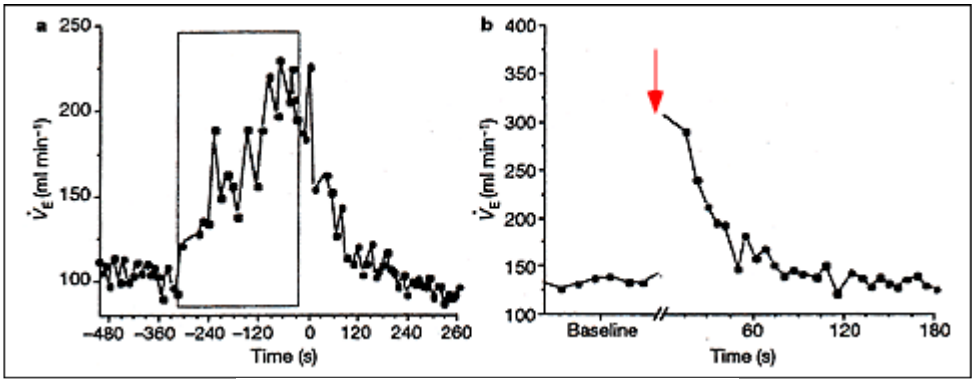
La ventilation : mécanismes de contrôle

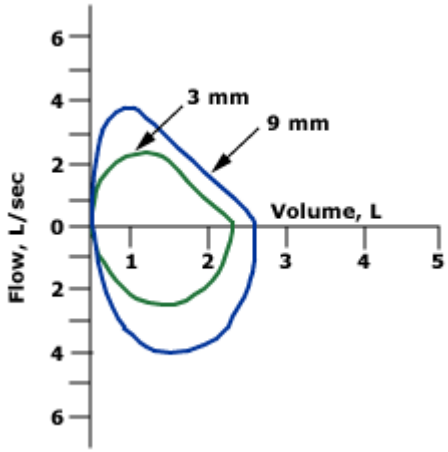


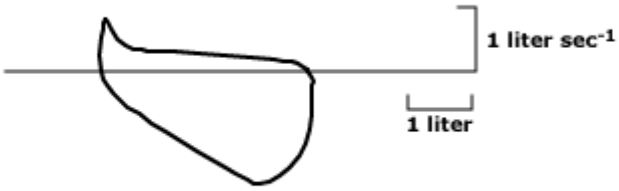


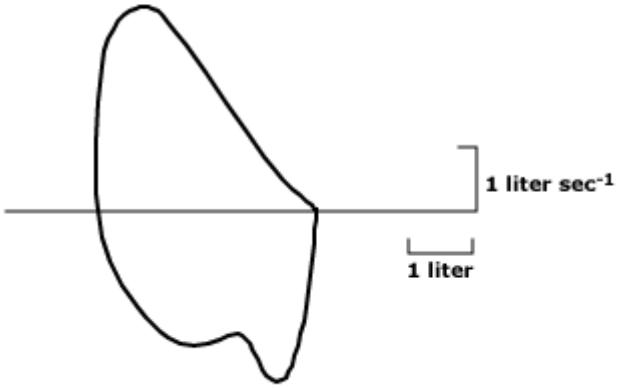

PaCO2 mmHg
Plasma [HCO3-] mEq/L
Metabolic alkalosis
Normal
Metabolic acidosis

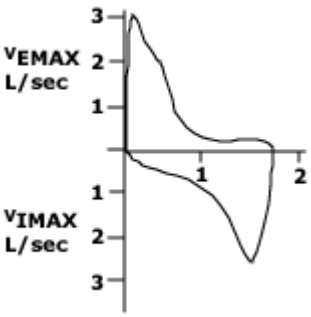

Minute ventilation, L/min
Arterial PCO2, mmHg
Minute ventilation, L/min
Arterial PO2, mmHg

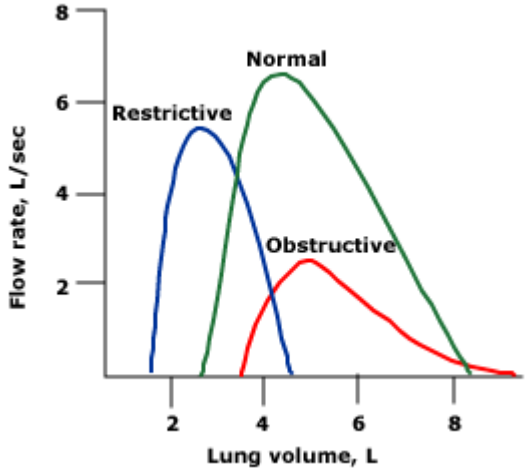

a
b
Time (s)
Baseline
Time (s)

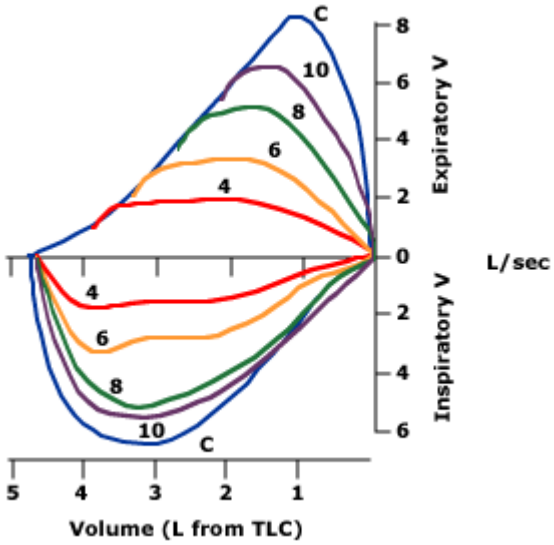

3 mm
9 mm
Volume, L
Flow, L/sec

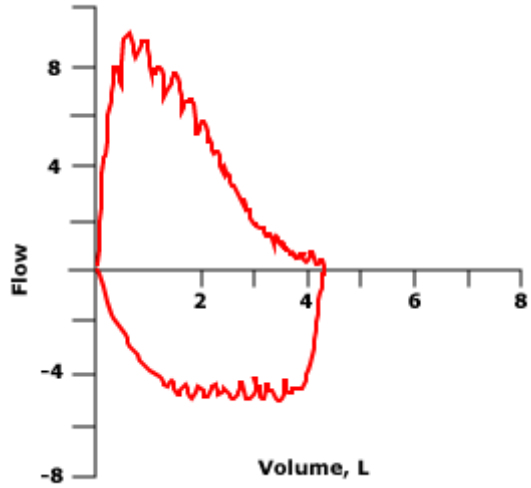

1 liter sec-1
1 liter

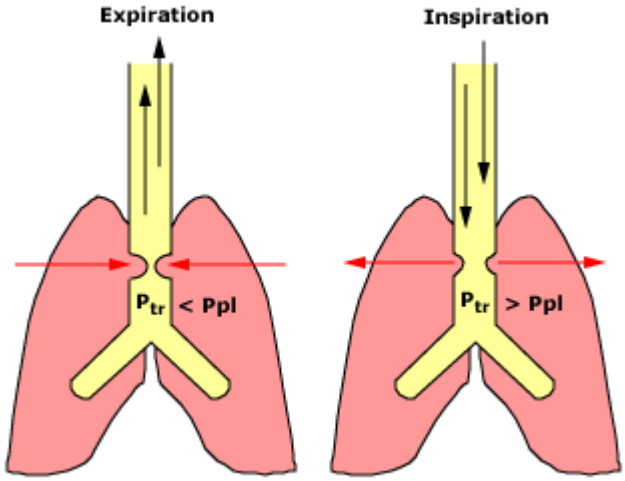

1 liter sec-1
1 liter

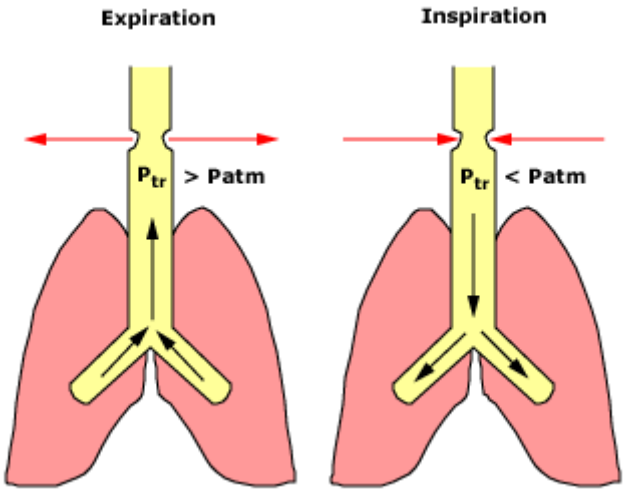

VEMAX L/sec
VIMAX L/sec
1
2
3
2
1
1
2
3


Normal
Restrictive
Obstructive
Flow rate, L/sec
Lung volume, L
8
6
4
2
2
4
6
8

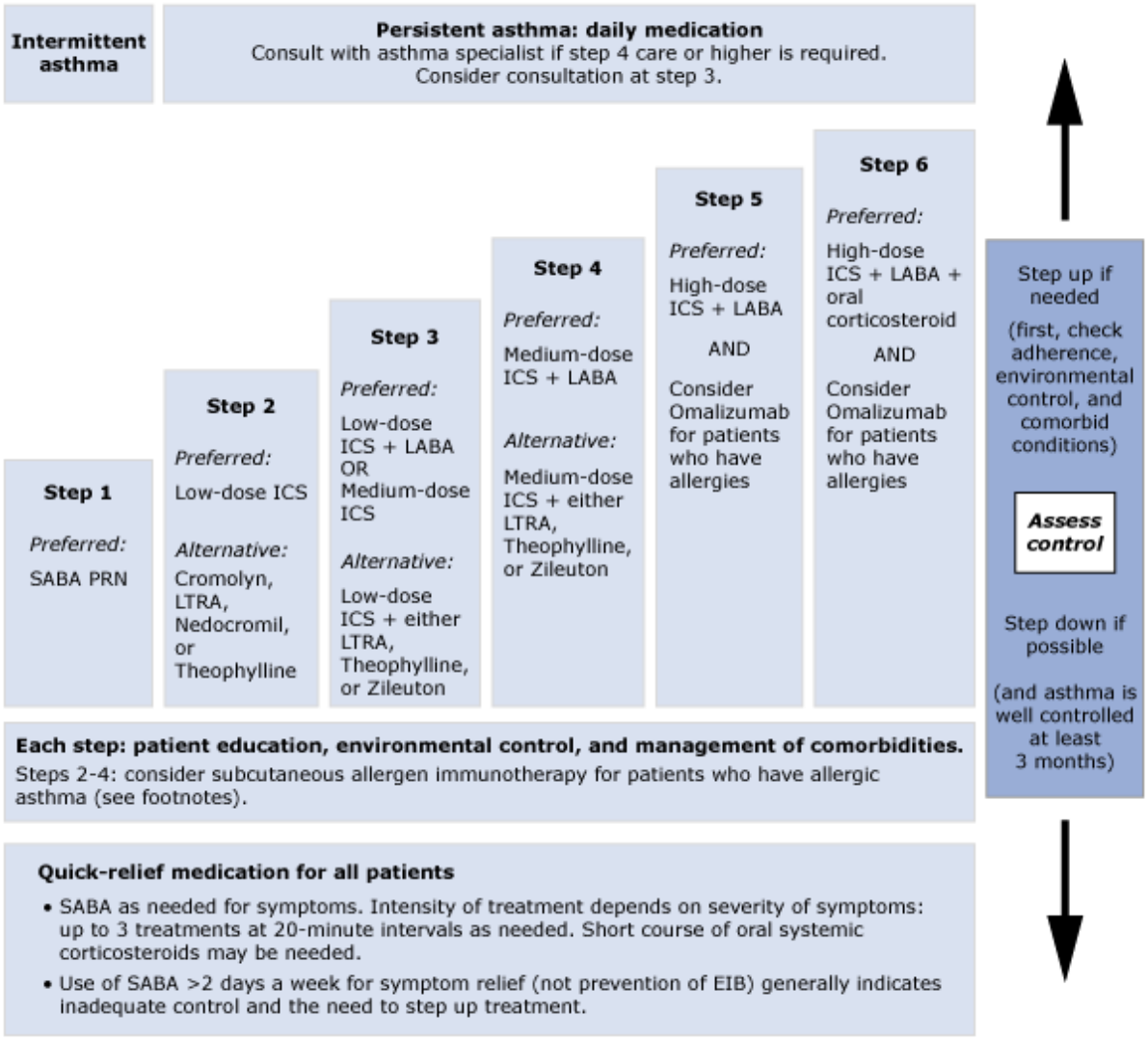

C
10
8
6
4
4
6
8
10
C
Expiratory V
Inspiratory V
L/sec
Volume (L from TLC)
5
4
3
2
1


8
4
-4
-8
2
4
6
8
Flow
Volume, L


Expiration
Inspiration
Ptr < Ppl
Ptr > Ppl


Expiration
Inspiration
Ptr > Patm
Ptr < Patm

L'asthme bronchique



This survey was designed to help you describe your asthma and how your asthma affects how you feel and what you are able to do. To complete it, please mark an X in the box that best describes your answer.

1. In the **past 4 weeks**, how much of the time did your asthma keep you from getting as much done at work or at home?

All of the time	Most of the time	Some of the time	A little of the time	None of the time
☐ 1	☐ 2	☐ 3	☐ 4	☐ 5

2. During the **past 4 weeks**, how often have you had shortness of breath?

More than once a day	Once a day	3 to 6 times a week	Once or twice a week	Not at all
☐ 1	☐ 2	☐ 3	☐ 4	☐ 5

3. In the **past 4 weeks**, how often did your asthma symptoms (wheezing, coughing, shortness of breath, chest tightness or pain) wake you up at night or earlier than usual in the morning?

4 or more nights a week	2 to 3 nights a week	Once a week	Once or twice	Not at all
☐ 1	☐ 2	☐ 3	☐ 4	☐ 5

4. In the **past 4 weeks**, how often have you used your rescue inhaler or nebulizer medication (such as Albuterol, Ventolin®, Proventil®, Maxair®, or Primatene Mist®)?

3 or more times per day	1 or 2 times per day	2 or 3 times per week	Once a week or less	Not at all
☐ 1	☐ 2	☐ 3	☐ 4	☐ 5

5. How would you rate your asthma control during the **past 4 weeks**?

Not controlled at all	Poorly controlled	Somewhat controlled	Well controlled	Completey controlled
☐ 1	☐ 2	☐ 3	☐ 4	☐ 5

Using an inhaler seems simple, but most patients do not use it the right way. When you use your inhaler the wrong way, less medicine gets to your lungs.

For the next few days, read these steps aloud as you do them or ask someone to read them to you. Ask your doctor or nurse to check how well you are using your inhaler.

Use your inhaler in one of the three ways pictured below. A or B are best, but C can be used if you have trouble with A and B. Your doctor may give you other types of inhalers.

Steps for using your inhaler

Getting ready

1. Take off the cap and shake the inhaler.
2. Breathe out all the way.
3. Hold your inhaler the way your doctor said (A, B, or C below).

Breathe in slowly

4. As you start breathing in slowly through your mouth, press down on the inhaler one time. (If you use a holding chamber, first press down on the inhaler. Within 5 seconds, begin to breathe in slowly.)
5. Keep breathing in slowly, as deeply as you can.

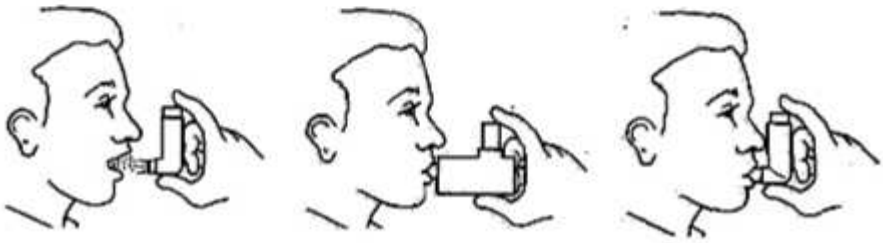
Hold your breath

6. Hold your breath as you count to 10 slowly, if you can.
7. For inhaled quick-relief medicine ($beta_2$-agonists), wait about 15-30 seconds between puffs. There is no need to wait between puffs for other medicines.

A. Hold inhaler 1 to 2 inches in front of your mouth (about the width of two fingers).

B. Use a spacer/holding chamber. These come in many shapes and can be useful to any patient.

C. Put the inhaler in your mouth. Do not use for steroids.



Clean your inhaler as needed, and know when to replace your inhaler. For instructions, read the package insert or talk to your doctor, other health care provider, or pharmacist.

Fréquence respiratoire

Age	Normale	Tachypnée
nouveau-né	30-50	> 60
nourrisson	20-40	> 50
jeune enfant	20-30	> 50
> 5 ans	15-20	> 30

PRISE EN CHARGE D'UNE CRISE D'ASTHME AUX URGENCES

1. Prise en charge initiale (MIN 0)

EVALUATION :

- ATCD, R/ de fond, Compliance
- Examen clinique, Sat° O2
- Asthme aigu grave AAG ?
- Score de Wood .
- (Gazo artér. si Sat° O2 < 92% sous FiO2 > 40%)

TRAITEMENT :

- O2 pour Sat° > 94%
- Ventolin NEB /20min x 3 ou AD /5min x3
- Atrovent NEB ou AD une seule fois
- Corticoïdes PO (ou) IV si crise sévère et pas de réponse rapide au ventolin

2. Réévaluation et adaptation du traitement (MIN 20-60)

Bonne réponse au R/ et Wood < 4	Réponse partielle au R/ et/ou Wood > 4	Pas de réponse au R/ et/ou Wood > 7 et/ou AAG
- Ventolin NEB ou AD / 60min - Atrovent NEB ou AD / 4h - Corticoïdes PO ?	- O2 pour Sat° > 94% - Ventolin NEB ou AD / 30-60min - Atrovent NEB ou AD / 4h - Corticoïdes IV ou PO	- O2 pour Sat° > 94% - Ventolin NEB / 20min - Atrovent NEB / 4h - Corticoïdes IV - Adrénaline NEB et/ou SC ? - Ventolin IVC ? - Sulfate de Mg IV ?

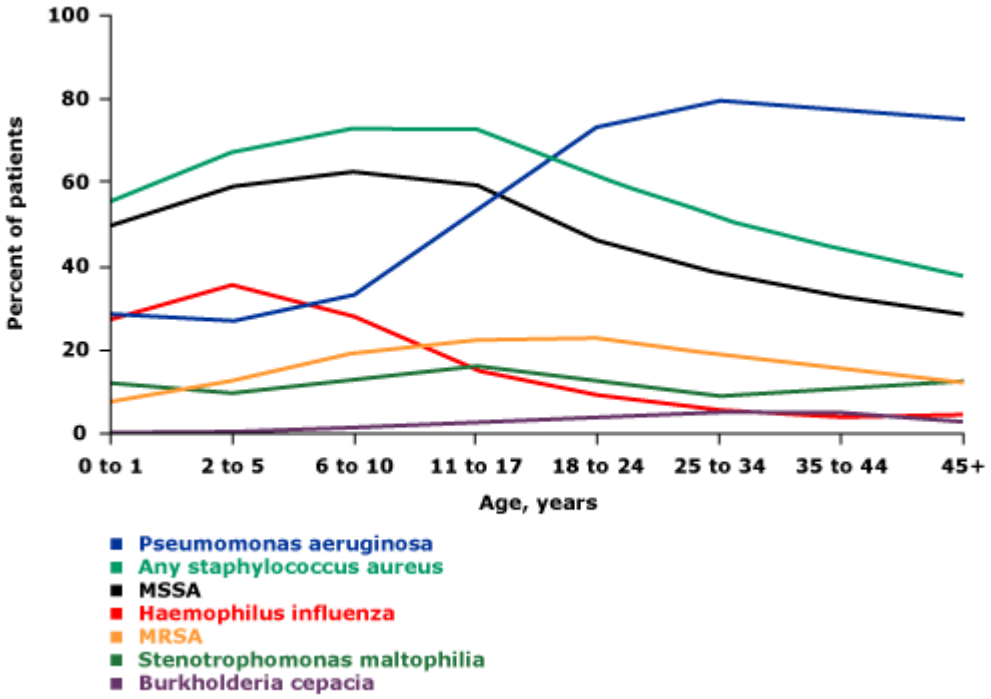
(voir tableau 3 pour posologies)

3. Hospitalisation ?

Bonne réponse au R/ et Wood < 4	Réponse partielle au R/ et/ou Wood > 4 (*)	Pas ou peu de réponse au R/ et/ou Wood > 7 et/ou Sat° < 94% sous O_2-lunettes
(1)	(2)	(2)
Retour à domicile	Pédiatrie	Soins Intensifs

(1) : dépend aussi de la capacité de l'enfant et des parents à gérer le R/ à la maison
(2) : dépend aussi du taux d'occupation des salles et de la présence ou non des parents

La mucoviscidose : germes pathogènes



Deux scores utilisés dans la mucoviscidose

Shwachman-Kulczycki Score

Reference Shwachman H & Kulczycki LL. Long-term study of 105 patients with cystic fibrosis. Am J Dis Child 1958;96:6-15.

This is a general score of clinical severity, which is assessed at annual review. Score each of the 4 parameters out of 25, and add up the 4 scores to give the total score (out of 100). Excellent-86-100; Good-71-85; Mild- 56-70;Moderate -41-55; Severe - <40

POINTS	GENERAL ACTIVITY	PHYSICAL EXAMINATION	NUTRITION	X-RAY FINDINGS
25	Full normal activity. Plays ball, goes to school regularly.	No cough, clear lungs, normal HR & RR, good posture.	Weight and height above 25th centile, Normal stool, good muscle mass and tone.	Normal, clear lung fields.
20	Lacks endurance, tires at end of day, good school attendance.	Rare cough, normal HR, minimal hyperinflation, clear lungs, no clubbing.	Wt and Ht 15-20th centile, stool slightly abnormal, fair muscle tone and mass.	Minimal accentuation of bronchovascular markings, early hyperinflation.
15	May rest voluntarily, tires easily after exertion, fair school attendance, tires after exertion.	Occasional cough/wheeze, increased RR, mild hyperinflation, early clubbing.	Wt and Ht above 3rd centile, stools often abnormal, large and poorly formed, minimal abdominal distension, reduced muscle mass and poor tone.	Mild hyperinflation, patchy atelectasis, increased bronchovascular markings.
10	Home teacher, dyspnoeic after short walk, rests frequently.	Frequent cough, often productive, clubbing, chest retraction, moderate hyperinflation, wheezes and crackles, moderate clubbing.	Wt and Ht below 3rd centile, bulky Offensive stool, mild to moderate Abdominal distension, flabby muscles and Reduced mass.	Moderate hyperinflation, widespread atelectasis and areas of infection. minimal bronchiectasis.
5	Orthopnoeic, stays in chair or bed.	Tachypnoea, tachycardia, severe coughing spells, extensive crackles, cyanosis, signs of heart failure, severe clubbing.	Marked malnutrition With protuberant Abdomen, rectal Prolapse, large foul Frequent fatty stools.	Severe hyperinflation, lobar atelectasis and bronchiectasis, nodules / cysts. pneumothorax, cardiac enlargement.

<u>Score de Brasfield</u>: sévérité maximale si = 25
Ref: Am J Radiol, 1980, 134, 1195-1198
→ Piégeage linéaire :

distension pulmonaire généralisée avec bombement sternal, dépression diaphragmatique et/ou cyphose dorsale:
0 si absent, 1 à 5 selon l'intensité;

→ opacités linéaires :

denses, en rail, +- armorisées, ou à extrémité semi-circulaire :
0 si absent, 1 à 5 selon l'intensité ;

→ opacités nodulaires ou kystiques :

multiples, finement cerclées de 0.5 cm ou plus avec centre clair ou opaque (quand elles sont confluentes, elles ne sont pas codées « à lésions étendues ») :
0 si absent, 1 à 5 selon intensité ;

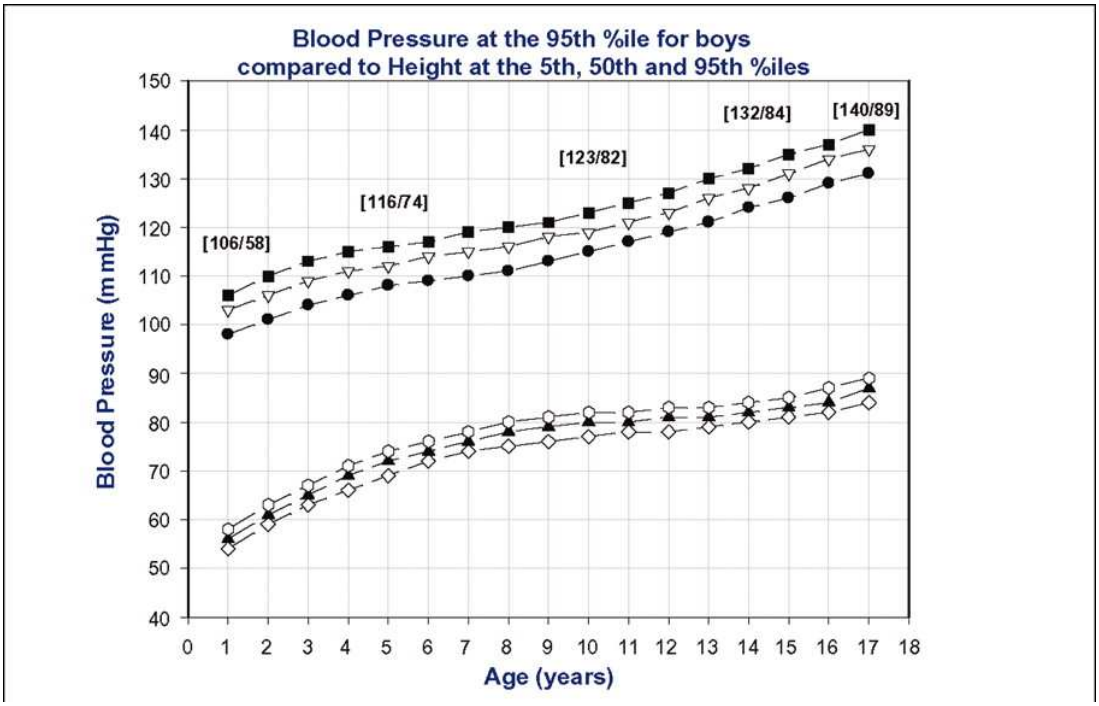
→ lésions étendues :

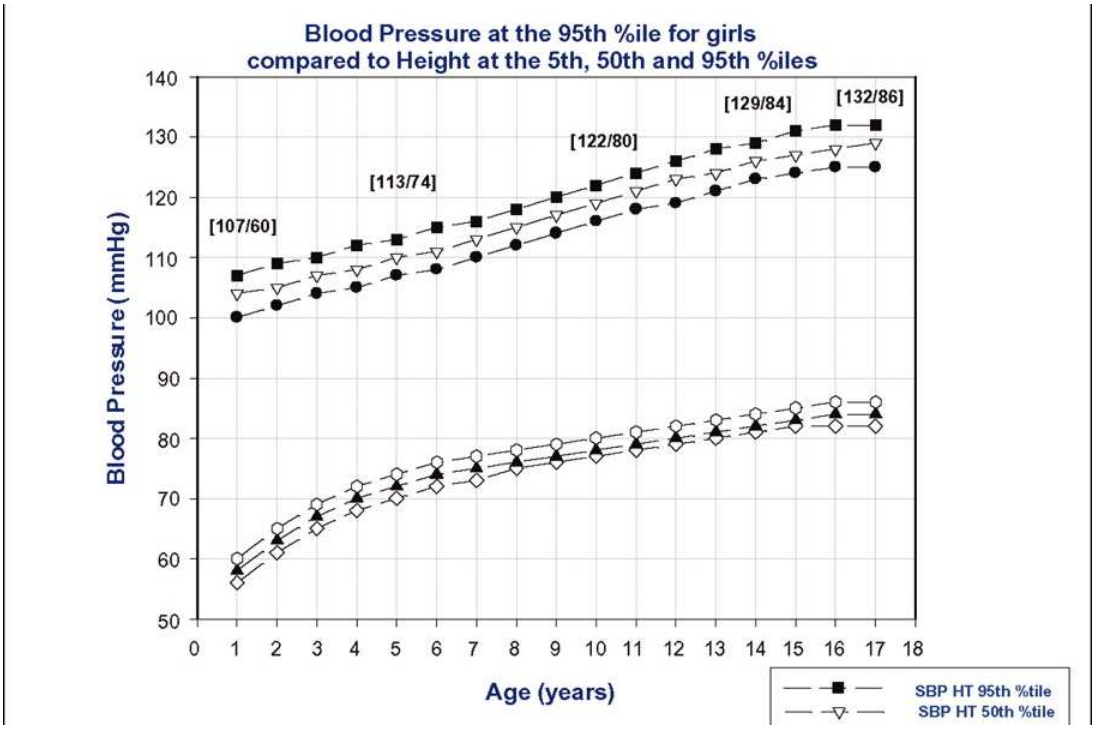
Atélectasies lobaires ou segmentaires non rétractile, syndrome de condensation lobaire ou segmentaire (pneumopathie aiguë non incluse) :
0 si absent ; 1 à 5 selon l'intensité ; 5 si complication telle une cardiomégalie, un pneumothorax

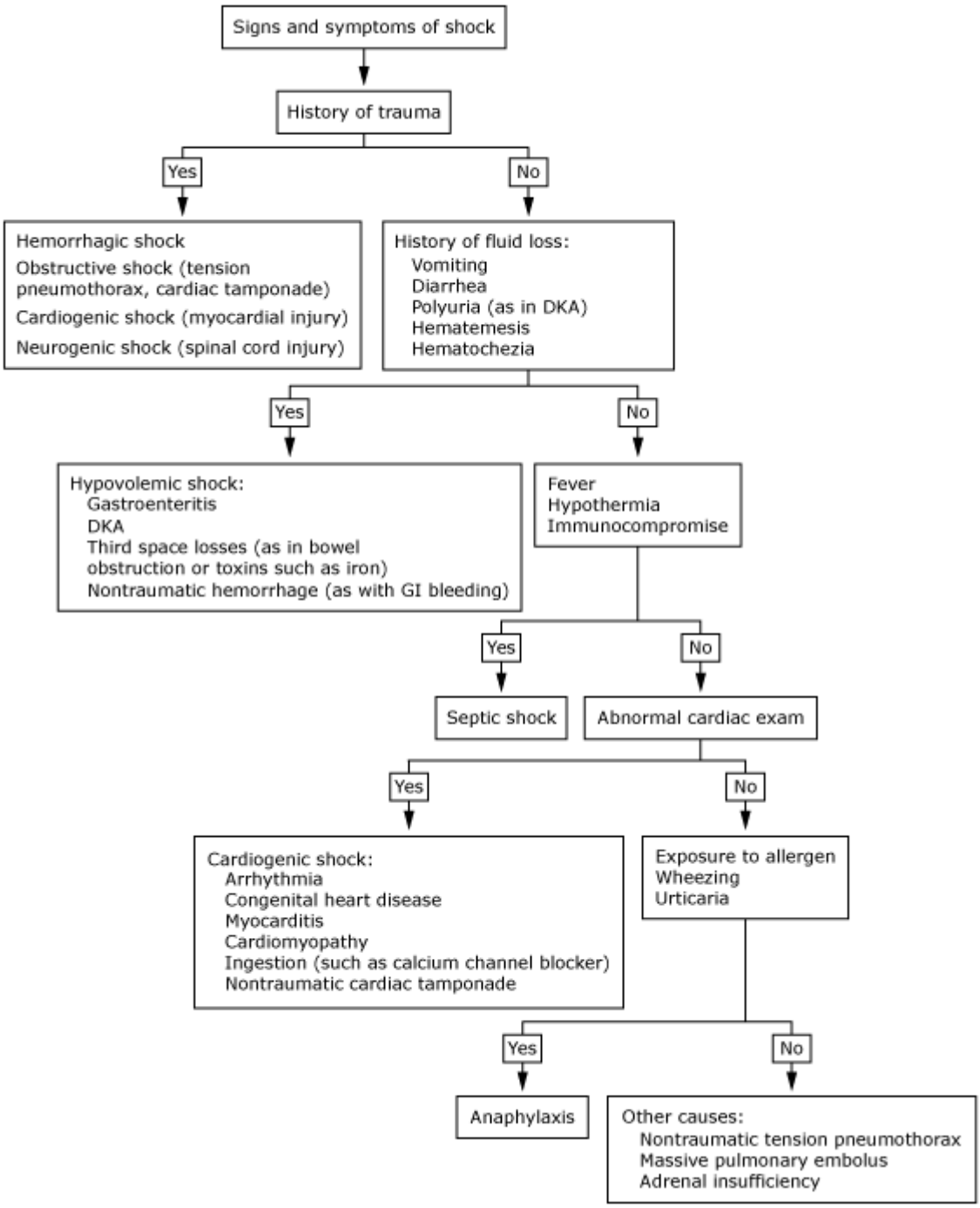
Valeurs normales : Fréquence Cardiaque , Tension Artérielle

Age	Battements/min
< 1 an	110-160
2-5 ans	95-140
5-12 ans	80-120
> 12 ans	60-100

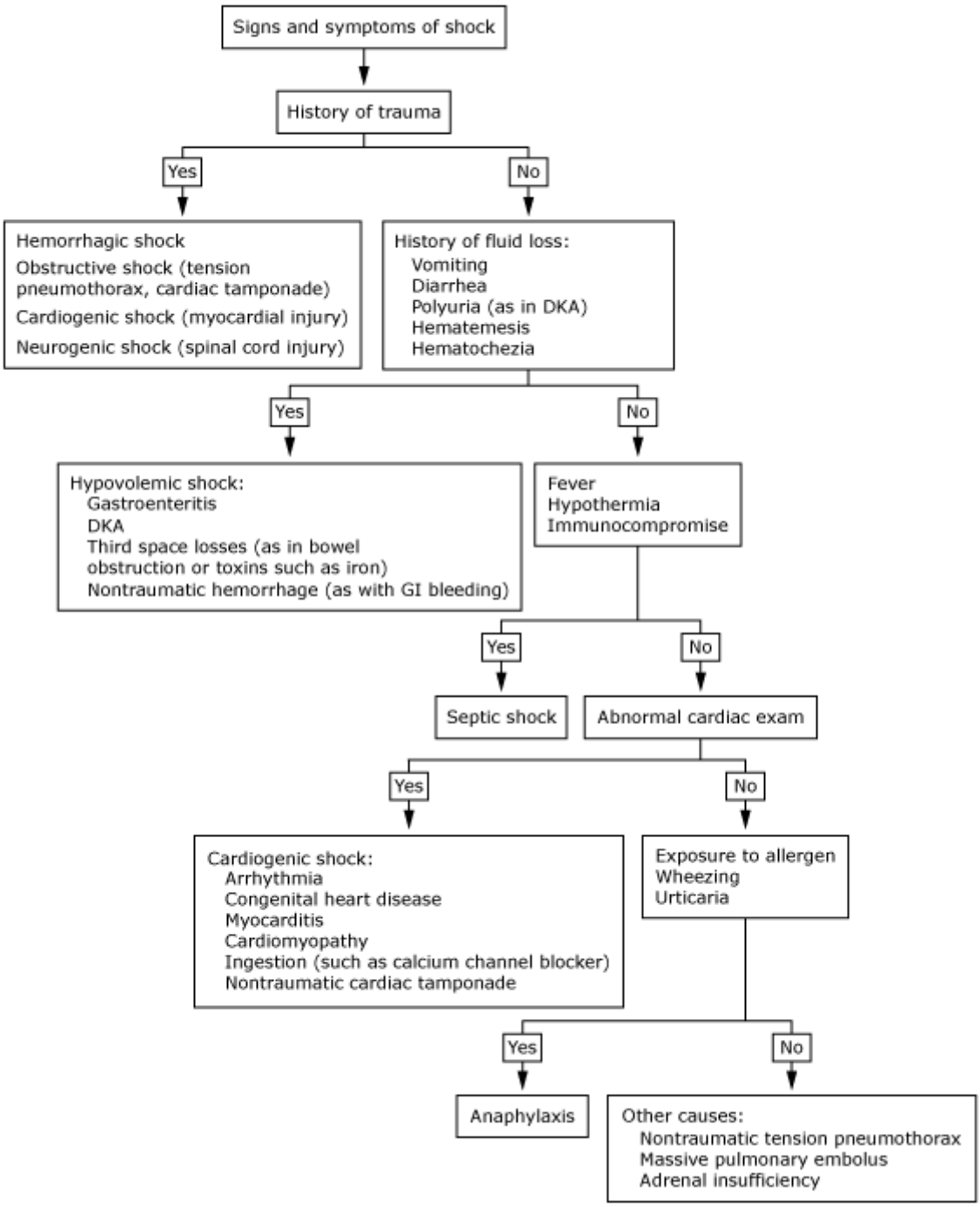
Âge	**systolique normale**	**Systolique minimale**
0-1 mois	**> 60**	**> 50**
1 – 12 mois	**> 80**	**> 70**
1 – 10 ans	**90 + [2x années]**	**70 + [2x années]**
> 10 ans	**110-130**	**> 90**

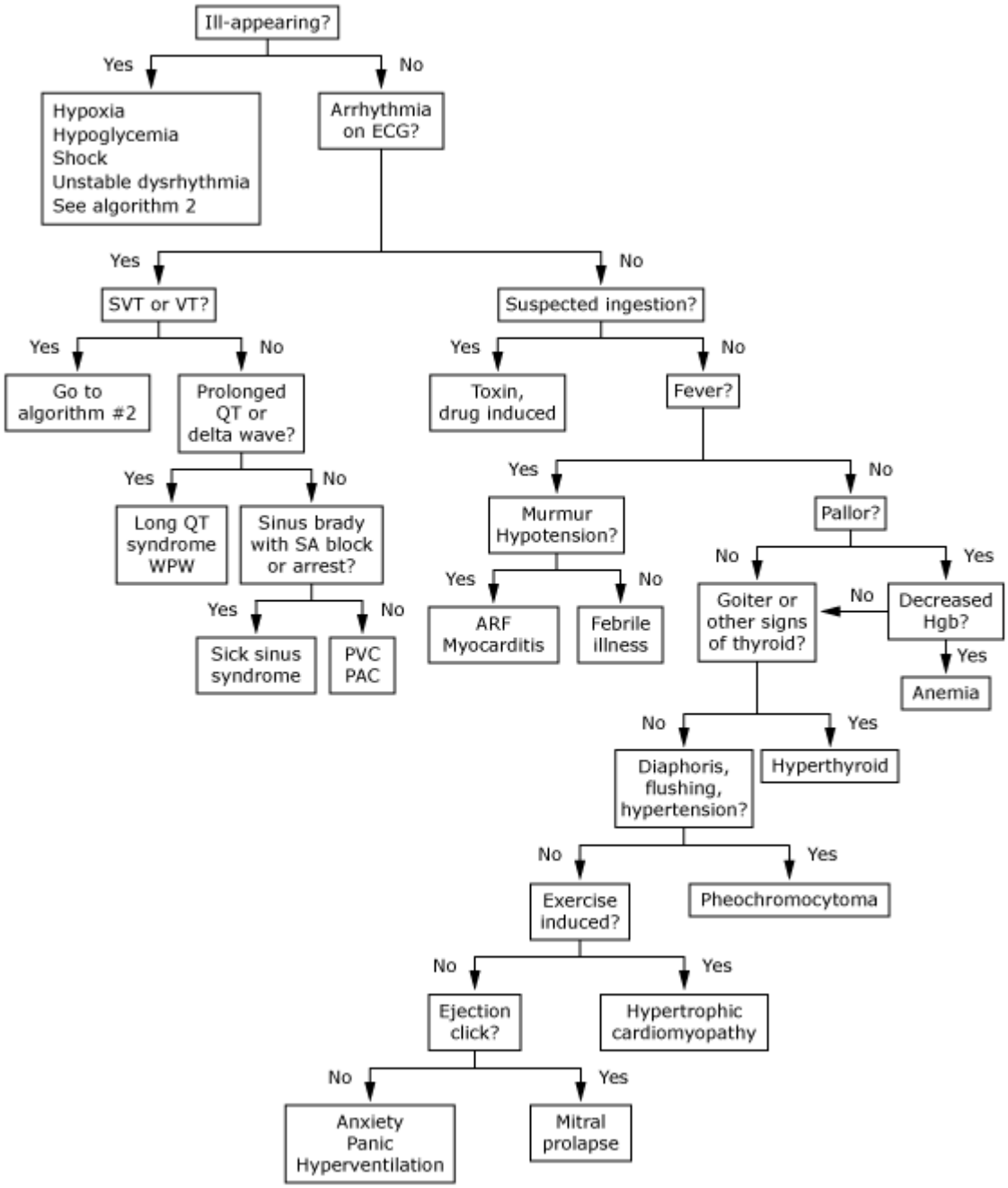

Blood Pressure at the 95th %ile for boys
compared to Height at the 5th, 50th and 95th %iles
Blood Pressure (mmHg)
Age (years)
[106/58]
[116/74]
[123/82]
[132/84]
[140/89]


Blood Pressure at the 95th %ile for girls
compared to Height at the 5th, 50th and 95th %iles
Blood Pressure (mmHg)
Age (years)
[107/60]
[113/74]
[122/80]
[129/84]
[132/86]
SBP HT 95th %tile
SBP HT 50th %tile

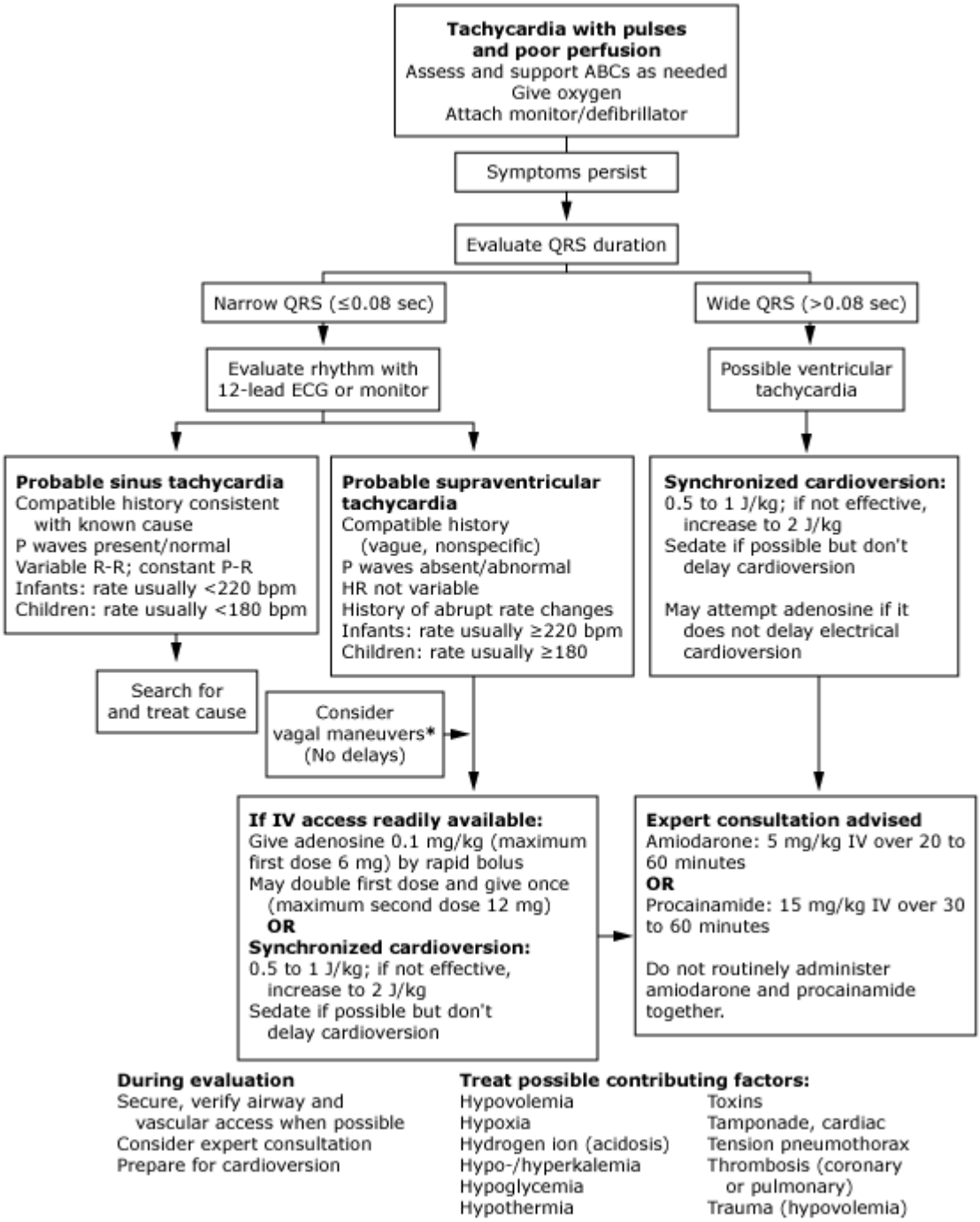


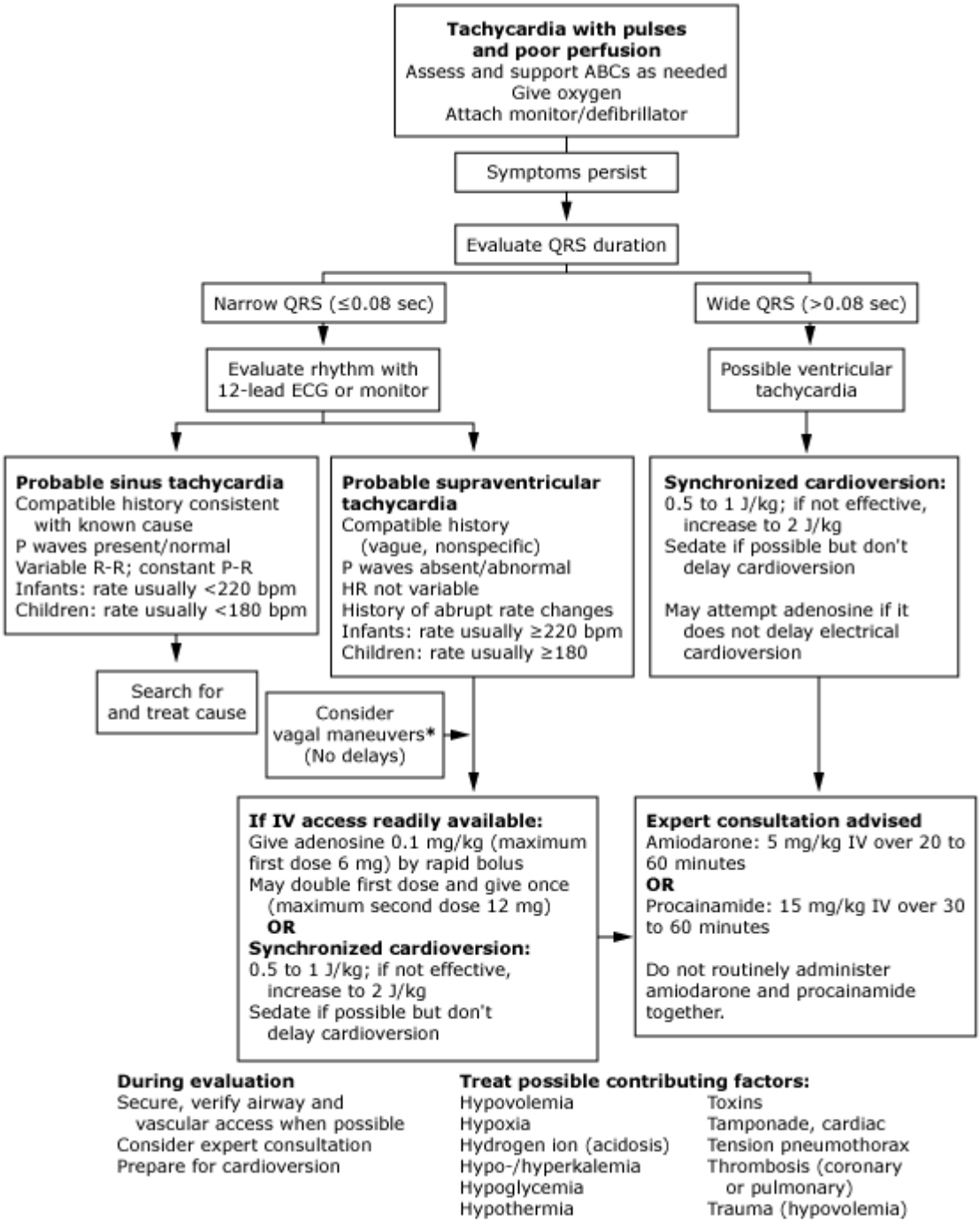
Etat de choc circulatoire

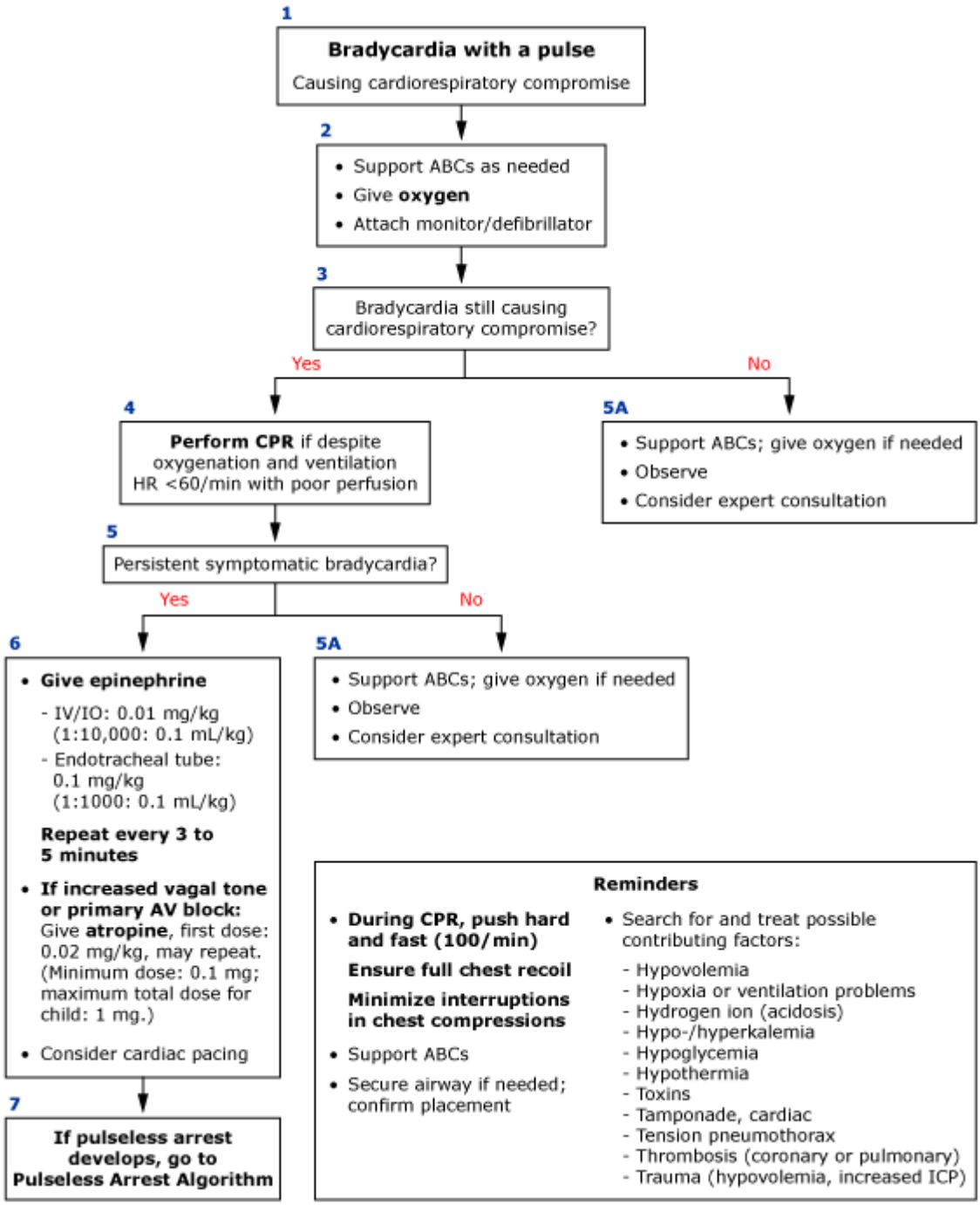

Signs and symptoms of shock
History of trauma
Yes
No
Hemorrhagic shock
Obstructive shock (tension pneumothorax, cardiac tamponade)
Cardiogenic shock (myocardial injury)
Neurogenic shock (spinal cord injury)
History of fluid loss:
Vomiting
Diarrhea
Polyuria (as in DKA)
Hematemesis
Hematochezia
Yes
No
Hypovolemic shock:
Gastroenteritis
DKA
Third space losses (as in bowel obstruction or toxins such as iron)
Nontraumatic hemorrhage (as with GI bleeding)
Fever
Hypothermia
Immunocompromise
Yes
No
Septic shock
Abnormal cardiac exam
Yes
No
Cardiogenic shock:
Arrhythmia
Congenital heart disease
Myocarditis
Cardiomyopathy
Ingestion (such as calcium channel blocker)
Nontraumatic cardiac tamponade
Exposure to allergen
Wheezing
Urticaria
Yes
No
Anaphylaxis
Other causes:
Nontraumatic tension pneumothorax
Massive pulmonary embolus
Adrenal insufficiency


Ill-appearing?
Yes
No
Hypoxia
Hypoglycemia
Shock
Unstable dysrhythmia
See algorithm 2
Arrhythmia on ECG?
Yes
No
SVT or VT?
Suspected ingestion?
Yes
No
Go to algorithm #2
Prolonged QT or delta wave?
Toxin, drug induced
Fever?
Yes
No
Long QT syndrome WPW
Sinus brady with SA block or arrest?
Murmur Hypotension?
Pallor?
Yes
No
Sick sinus syndrome
PVC PAC
ARF Myocarditis
Febrile illness
Goiter or other signs of thyroid?
Decreased Hgb?
Anemia
Hyperthyroid
Diaphoris, flushing, hypertension?
Pheochromocytoma
Exercise induced?
Ejection click?
Hypertrophic cardiomyopathy
Anxiety Panic Hyperventilation
Mitral prolapse

➔ tachycardie

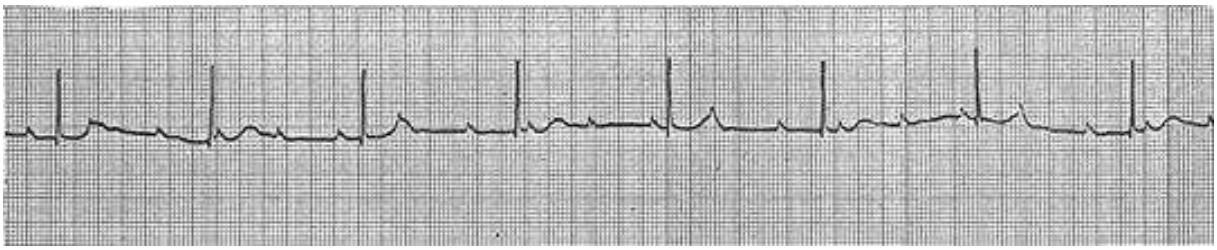


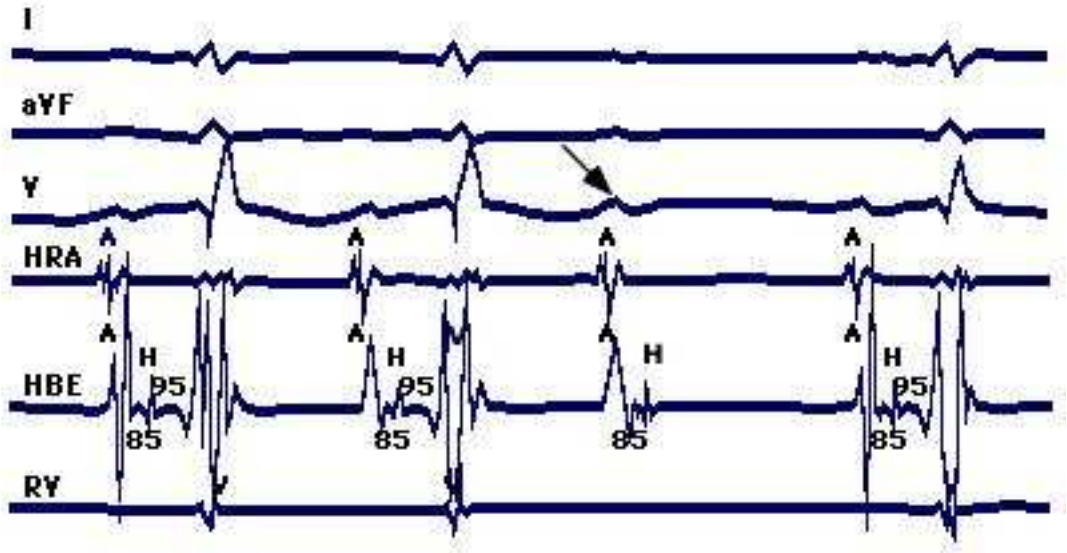

Tachycardia with pulses and poor perfusion
Assess and support ABCs as needed
Give oxygen
Attach monitor/defibrillator
Symptoms persist
Evaluate QRS duration
Narrow QRS (≤0.08 sec)
Wide QRS (>0.08 sec)
Evaluate rhythm with 12-lead ECG or monitor
Possible ventricular tachycardia
Probable sinus tachycardia
Compatible history consistent with known cause
P waves present/normal
Variable R-R; constant P-R
Infants: rate usually <220 bpm
Children: rate usually <180 bpm
Probable supraventricular tachycardia
Compatible history (vague, nonspecific)
P waves absent/abnormal
HR not variable
History of abrupt rate changes
Infants: rate usually ≥220 bpm
Children: rate usually ≥180
Synchronized cardioversion:
0.5 to 1 J/kg; if not effective, increase to 2 J/kg
Sedate if possible but don't delay cardioversion
May attempt adenosine if it does not delay electrical cardioversion
Search for and treat cause
Consider vagal maneuvers* (No delays)
If IV access readily available:
Give adenosine 0.1 mg/kg (maximum first dose 6 mg) by rapid bolus
May double first dose and give once (maximum second dose 12 mg)
OR
Synchronized cardioversion:
0.5 to 1 J/kg; if not effective, increase to 2 J/kg
Sedate if possible but don't delay cardioversion
Expert consultation advised
Amiodarone: 5 mg/kg IV over 20 to 60 minutes
OR
Procainamide: 15 mg/kg IV over 30 to 60 minutes
Do not routinely administer amiodarone and procainamide together.
During evaluation
Secure, verify airway and vascular access when possible
Consider expert consultation
Prepare for cardioversion
Treat possible contributing factors:
Hypovolemia
Hypoxia
Hydrogen ion (acidosis)
Hypo-/hyperkalemia
Hypoglycemia
Hypothermia
Toxins
Tamponade, cardiac
Tension pneumothorax
Thrombosis (coronary or pulmonary)
Trauma (hypovolemia)

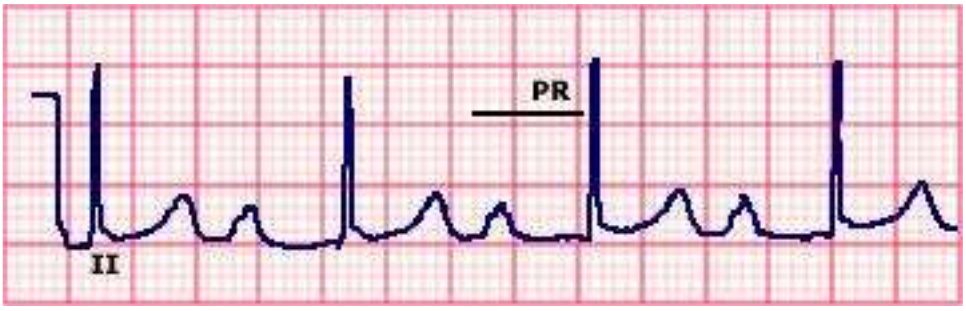


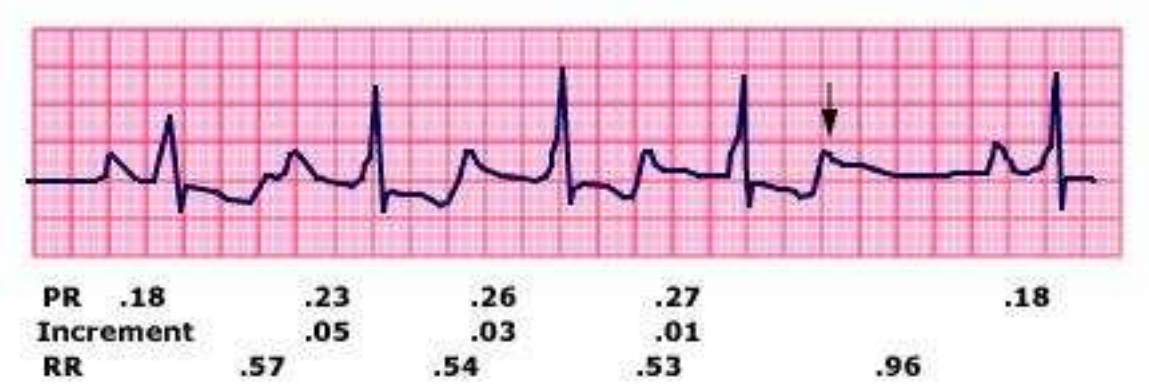
➔ **bradycardie**

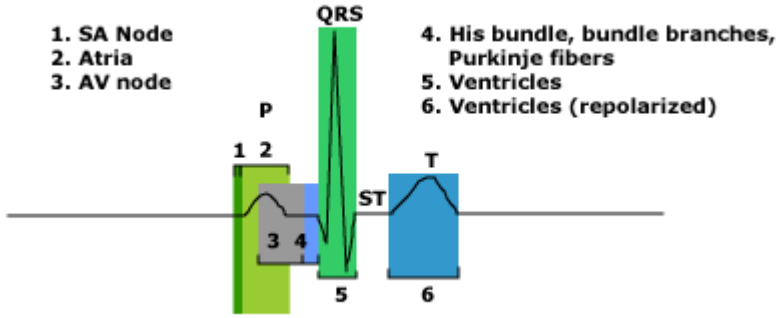
➔ Blocs AV



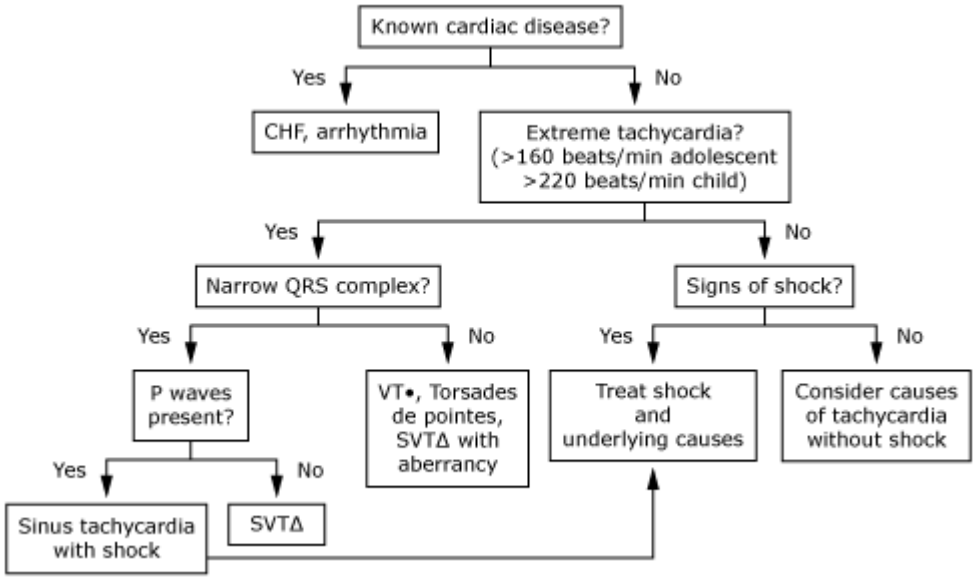


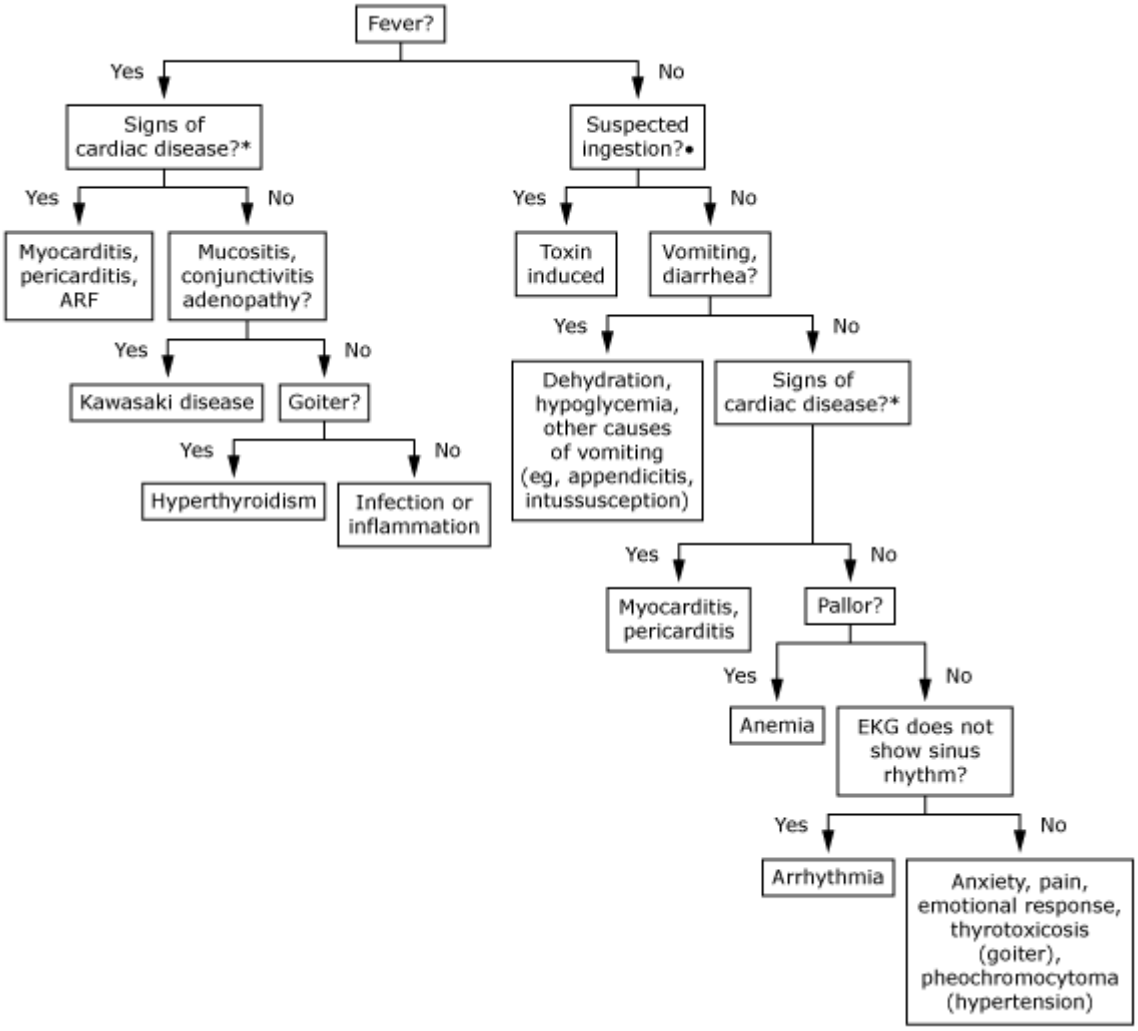




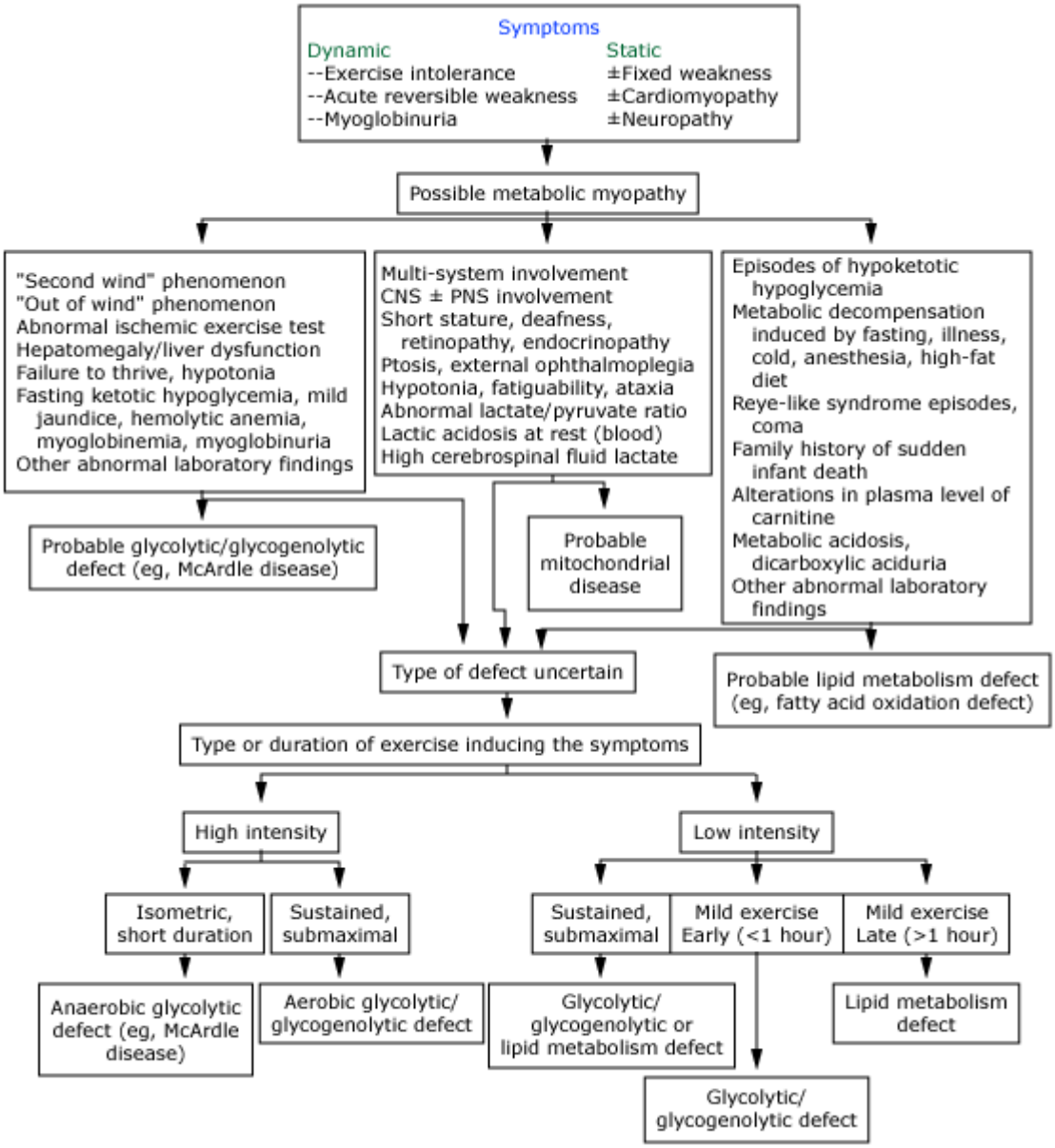


➔ **Troubles du rythme**




Fever?
Yes
No
Signs of cardiac disease?*
Suspected ingestion?•
Yes
No
Yes
No
Myocarditis, pericarditis, ARF
Mucositis, conjunctivitis adenopathy?
Toxin induced
Vomiting, diarrhea?
Yes
No
Yes
No
Kawasaki disease
Goiter?
Dehydration, hypoglycemia, other causes of vomiting (eg, appendicitis, intussusception)
Signs of cardiac disease?*
Yes
No
Yes
No
Hyperthyroidism
Infection or inflammation
Myocarditis, pericarditis
Pallor?
Yes
No
Anemia
EKG does not show sinus rhythm?
Yes
No
Arrhythmia
Anxiety, pain, emotional response, thyrotoxicosis (goiter), pheochromocytoma (hypertension)

➔ **Tolérance à l'effort**



Choc cardiogénique

Etiologie : cardiopathie congénitale

1ère semaine de vie:

Hypoplasie coeur gauche, transposition des grands vaisseaux, retour veineux pulmonaire anormal total obstructif, insuffisance tricuspide sévère (souffrance foetale), fistule artério-veineuse large.

>2e semaine de vie:

Sténose aortique/pulmonaire sévère, coarctation de l'aorte ou interruption de l'arc aortique, cardiomyopathie (maladie métabolique).
DD Sepsis du nouveau-né

>2 mois:

Gros shunt G-D: Communication inter ventriculaire large, canal artériel large (Néonat), canal atrio-ventriculaire complet, anomalie de l'implantation des coronaires.
Choc septique.
Défaillance multisystémique.
ARDS, Hypertension Artérielle Pulmonaire.
Crise hypertensive.
Troubles du rythme (Tachycardie ou Bloc Auriculo-Ventriculaire).
Myocardite virale.
Anémie sévère (hydrops foetal, drépano).

Clinique

Décompensation aiguë compensée: polypnée, oligurie, pâleur, extrémités froides, sueurs froides, pouls filant et diminués, tachycardie, tension artérielle limite inf de la normale.
Décompensation aiguë décompensée: troubles de la conscience, pouls imprenable, marbrures, tension artérielle diminuée, hépatomégalie, stase jugulaire.

Traitement

Repos voire sédation si nécessaire (attention BZD hypotension artérielle).
O^2, ventilation artificielle si nécessaire.
Traiter troubles associés : troubles ioniques et acidose (bic si BE <-6), infection, anémie, HTAP (100 % FiO2 + NO).

Catécholamines

Dobutamine

5-20 µg /kg /min.
Inotrope +, vasodilatation périphérique.

Adrénaline
0.01-0.5-2 µg /kg /min.
Inotrope +, à petite dose via récepteur beta vasodil périph, à + haute dose via récepteur alpha.
Vasoconstriction.

Noradrénaline
0.01-0.1-1 µg /kg /min.
inotrope + (mais moins que Ad) et vasoconstriction périph (+que Ad).

Dopamine
0.5-3 µg /kg /min : vasodil rénale.
3-6 µg /kg /min inotrope +.
>10 µg /kg /min vasoconstriction périphérique (max 20 µg /kg /min).

Inhibiteur des Phosphodiestérases III

Milrinone (Corotrope)

50 µg/kg en bolus puis 0.5-0.75 µg /kg /min
Inotrope +, Vasodilatation périphérique et pulmonaire

Sildenafil (Viagra) per os !!
0.25-0.5 mg /kg /dose /6h

Diurétiques
Furosémide
Bolus : 0.5-2 mg /kg /dose IV 2 min ou per os /6-12h
IVC 0.1-1 mg /kg /h

Prostaglandines si suspicion d'une cardiopathie ductodépendante
PGE1 (Alprostadil)
Dilution: 0,12cc x pds en kg + G5 % ad 10cc => 1cc /h = 0,1µg /kg /min
ou 0,3cc x pds +G5% ad 24cc => 1cc/h = 0,1µg /kg /min.
Débuter à 0.1µg /kg /min puis diminuer à 0.01µg /kg/min.

Traitement des troubles du rythme si nécessaire.

Abord en phase aiguë de la Tachycardie supra ventriculaire chez l'enfant

Définition : tachycardie régulière à QRS fins (200 à 280/min et plus).
- Attention :
Si tachycardie à QRS larges = TV jusqu'à preuve du contraire, et en cas de décompensation hémodynamique, administrer un choc électrique externe.

- Traitement de la crise :
Réaliser un ECG en continu (rythme avec 3 dérivations au choix) pendant les manoeuvres pour ramener l'enfant en rythme sinusal dans l'espoir d'enregistrer la cardioversion (important pour le diagnostic précis).

< 2 ans

Si Décompensation :
1. Icebag
2. Adénosine
3. Cardioversion par 0,5-1 joule/kg et (2)
4. Digoxine puis réessai (1) et (2)

Cardioversion : sous Diprivan ou Hypnomidate.

si Bon état hémodynamique
1. Icebag
2. Adénosine
3. Digoxine puis réessai (1)

<u>> 2 ans</u>

Si Décompensation :	si Bon état hémodynamique
1. Réaction vagale (Valsalva,...)	1. Réaction vagale
2. Adénosine	2. Adénosine
3. Cardioversion	3. β-bloquant
4. Digoxine	
5. β-bloquant	

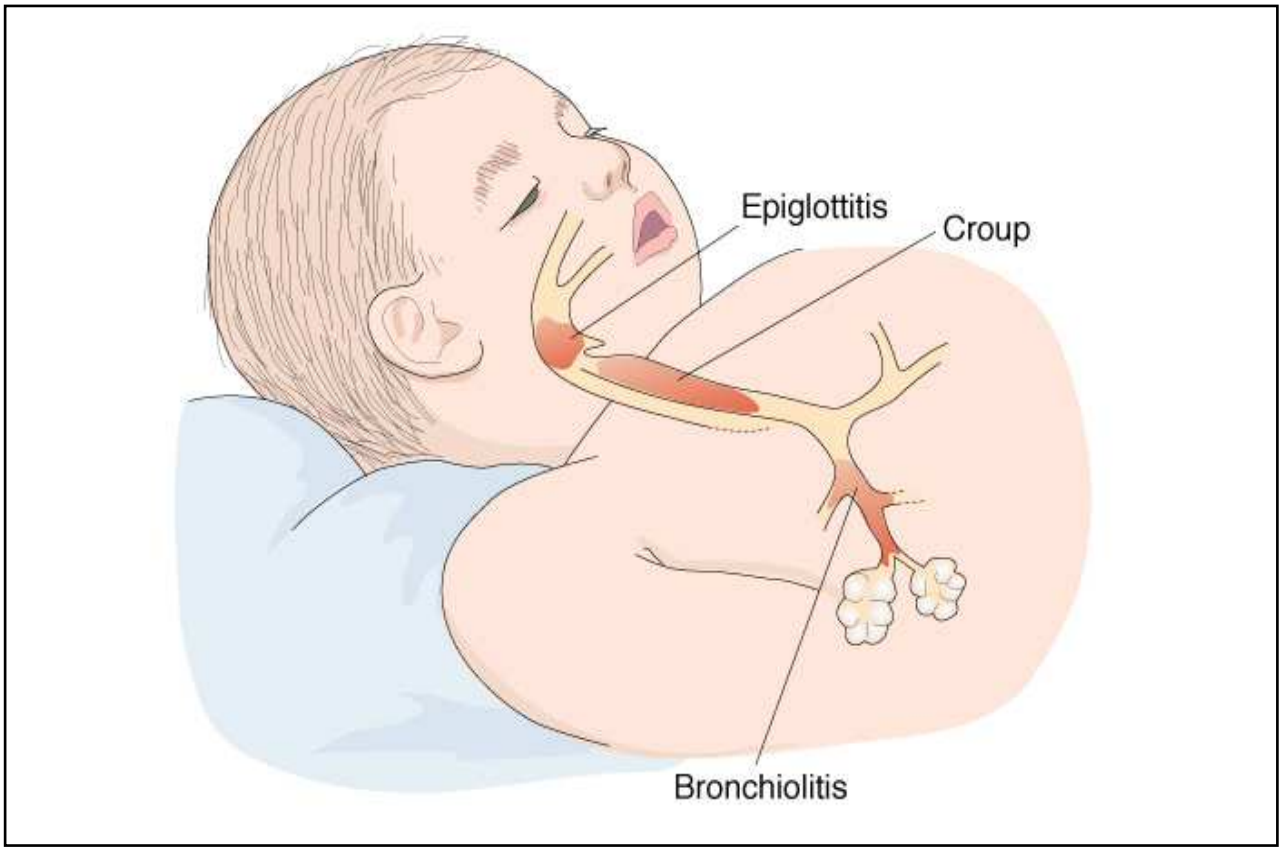
Autres possibilités
- Amiodarone

Remarques :
- En cas de Wolff-Parkinson-White (visible sur l'ECG en rythme sinusal uniquement), éviter la Digoxine.

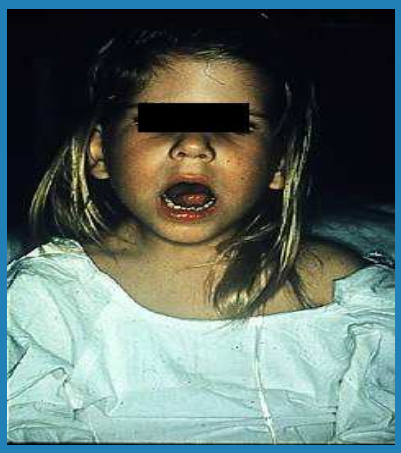
- Jamais de Vérapamil chez l'enfant < 2 ans (risque de mort subite).

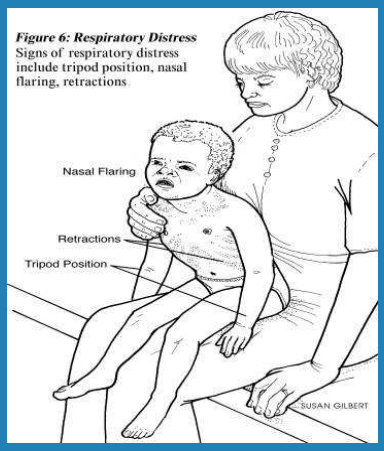
Les détresses respiratoires

Localisation d’une difficulté respiratoire selon le trajet anatomique



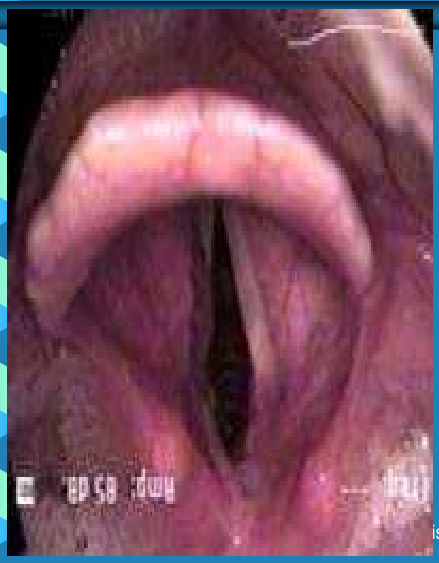
Epiglottite

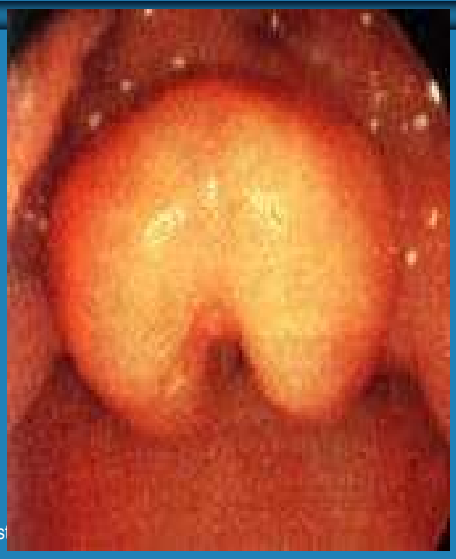




Prof O Battisti, détresses respiratoires

Epiglottite





→ Intubation par une main experte, avec prémédication et aérosol ½ LP ½ adrénaline et antibiothérapie IV (Claforan).

-

Bronchioliolite ou laryngite sous-glottique

Bronchiolites : score de GADOMSKi

	0	1	2	3
Battement des ailes du nez	aucun		modéré ou intermittent	marqué et persistant
Grunting	aucun	intermittent	persistant	
Tirage	aucun	discret	modéré	sévère
Entrée d'air	normale			diminuée
Cyanose (ou PaO2<70) (*)	absente	à l'air ambiant	sous FiO2 40%	
Conscience	normale	altérée ou agitation	coma	

Forme sévère = score supérieur ou égal à 8
score de **Westley,** dans les laryngites

	0	1	2	3	4	5
Stridor	absente	à l'agitation	au calme			
Tirage	absent	léger	modéré	sévère		
Entrée d'air	normale	diminuée	très diminuée			
Cyanose	absente				à l'agitation	au calme
Conscience	normale					altéré

Crises d'asthme : score de WOOD

crise sévère si score > 4

	0	1	2
Wheezing	**aucun**	**modéré**	**marqué**
Tirage	**aucun**	**modéré**	**sévère**
Entrée d'air	**normale**	**diminuée**	**très diminuée**
Cyanose (ou PaO2<70) (*)	**absente**	**à l'air ambiant**	**sous FiO2 40%**
Conscience	**normale**	**altérée ou agitation**	**coma**

PRISE EN CHARGE INITIALE D'UN CHOC SEPTIQUE

Prise en charge initiale (MINUTE 0)

CHOC ? **Tachycardie**, conscience, compensation respiratoire, pouls centraux et périph., temps de recoloration >3sec.	**ABC** (**O_2 100%** d'office) intubation ? **VOIE D'ACCES** 2 périphériques si très rapide Centrale si hypoTA et/ou échec périphérique

DECOMPENSE ? OUI si hypotension Intra osseuse si échec ou si absence de pouls
(voir normes hypotension chez l'enfant)

Biologie : HEMOC (y compris si via intra osseuse), hémato, CRP, glycémie, protéines, enzymes hépatiques, coagulation et PDF, pH, fonction rénale, ions, calcium, phosphore, lactate, cortisol (tube sec 2ml).

Soutien volémique agressif (MINUTE 5)

Antibiothérapie !
(Après hémoculture ! ! !)

20cc/kg IV rapide
en moins de 20 minutes

LP ou Voluven 6% (SSPP si < 1an)
Cfr annexe

⇩

Appel garde REA
(Tachycardie, TA, conscience…)

Réévaluer

⇩

Répéter si nécessaire
20cc/kg IV rapide
(Durant la première heure).

NB : Soutien volémique agressif parfois nécessaire (> 40-60 cc /kg)

⇩

Après 2x 20 cc /kg (MINUTE 10-20)

⇩

ABC
franche

Choc persistant – **TA corrigée**

Choc sévère - **hypoTA**

Conscience ? Intubation?

Voie centrale d'office

Monitoring invasif PA ?!

⇩

Dopamine (Dynatra®) IVC

10-20 µg /kg /min
Continuer remplissages !

⇩

Choc réfractaire à la dopamine ou choc sévère d'emblée

ABC	Adrénaline IVC : 0.1 – 1µg/kg/min

Intubation d'office — Noradrénaline (Lévophed®) IVC : 0.1 – 1µg/kg/min

PA invasive d'office — *Choix à adapter selon choc hyperdynamique* (Noradré) *ou choc « froid »* (Adré)

⇩

Continuer remplissages !

Choc réfractaire aux catécholamines

ABC — Envisager Hydrocortisone* — **Voir annexe si purpura fulminans*

Annexes

Définitions **selon l'âge** :

AGE	**PA systolique minimale** (mmHg)	**Tachycardie** (bpm)
0-1 mois	50	>180
1 mois – 1 an	70	
1 ans – 10 ans	70 + (2 x âge en années)	>160
> 10 ans	90	

Solutés de **remplissage** :

- Pas de supériorité démontrée des solutions colloïdes (SSPP, Voluven, Albumine 5 %) par rapport au LP en terme de morbidité et de mortalité, mais nécessité de donner des volumes plus importants en LP ?

- Voluven (6 %) premier choix parmi les colloïdes après un an.

- Dans la pratique :

LP si plus rapidement disponible	
< 1an	*SSPP*
> 1an	*Voluven*

Administration de plasma frais congelé à envisager si diathèse hémorragique ou selon bilan de coagulation. Globules rouges concentrés à envisager si choc compensé et Hb <7g /dl ou choc décompensé et Hb <10g /dl.

Antibiothérapie empirique :

< 3 mois	Ampicilline = PENTREXYL® (≠ CLAMOXYL®) +Cefotaxime = CLAFORAN® (Pas d'Amukin systématique)
> 3 mois	Ceftriaxone = ROCEPHINE ®

Préparation des **drogues hémodynamiques** :
DOPAMINE (et DOBUTAMINE)
50mg /50 ml → Poids /3 en ml = 5µg /kg /min *(ex: 6 kg → 2ml /kg=5 µg /kg /min)*

ADRENALINE et NORADRENALINE
1mg/50ml → Poids/3 en ml = 0.1µg /kg /min

HydrocortisoneU : (=Solucortef®) (≠Solumedrol)
Purpura Fulminans : 2mg/kg/8h d'office pendant 3 jours (> 3 jours si cortisol bas à l'admission).
Adulte : 100 mg /8h
Purpura fulminant avec choc résistant à la dopamine : 50mg /kg (durée selon cortisolémie initiale).
Choc réfractaire aux catécholamines (adré/noradré) : 50mg /kg (durée selon cortisolémie initiale).

Place de la **ponction lombaire** :
La présence d'un choc (comme la présence de pétéchies évolutives et/ou d'une diathèse hémorragique) est une contre-indication à la réalisation d'une ponction lombaire. Celle-ci sera discutée en dehors de la phase de stabilisation.

Si **purpura fulminant** :
Frottis nasal appuyé ; AG soluble sang, urine, LCR ; PCR sur LCR.
Déclaration maladie transmissible
Indication de protéine C ?

Si **choc septique sévère** :
Discuter indication d'une hémodiafiltration continue veino-veineuse (équilibration volémique et effet sur les toxines et médiateurs inflammatoires).

Traitement empirique de la méningite

< 3 mois Néonatologie 1er jour de vie	Ampi (Listeria)= PENTREXYL® + Aminoside = AMUKIN®	OU	Cefuroxime = ZINACEF(si suspicion TD) ® Ampi = PENTREXYL® Aminoside = AMUKIN®

NB : Amukin ± 4 jours max.

< 3 mois Néonatologie Qq jours de vie	Ampi = PENTREXYL® + Cefotaxime = CLAFORAN® **(Pas d'Amukin systématique)**
> 3 mois	**Ceftriaxone = ROCEPHINE ® + (dexaméthasone si pas vacciné pour HiB ou a reçu 1 seule dose de HiB)**

TRAITEMENTS PARTICULIERS

■ Méningite tardive à streptoco β groupe B [± 1 mois, sérotype 3 (80 %)]

- Empirique : AMPI =PENTREXYL + CEFTRIAXONE
- Si identification : CEFTRIAXONE (car Ampi. modifie trop la flore).
- Pas de gammaglobuline I.V.

■ **Si choc septique** : cortisolémie à l'admission *(voir protocole purpura fulminans)* + 2 mg /kg /8 h d'hydrocortisone (= SOLUCORTEF®) pendant 3 jours (≠ prednisolone).

DUREE

NEONATO	**ENFANT**
(strepto B, bacille Gram-) **14 à 21 j** en fonction - de la vitesse de négativation de la CRP - de l'évolution clinique	Méningo 7 j si pas de Pneumo 10 j *HiB* *10 j* *complication*

Remarques : Infection invasive HiB * 1-2 cas / partie francophone Belgique /an
* Immunisation si ≥ 2 injection si < 1 an
* Des cas sporadiques > 4-5 ans et adultes

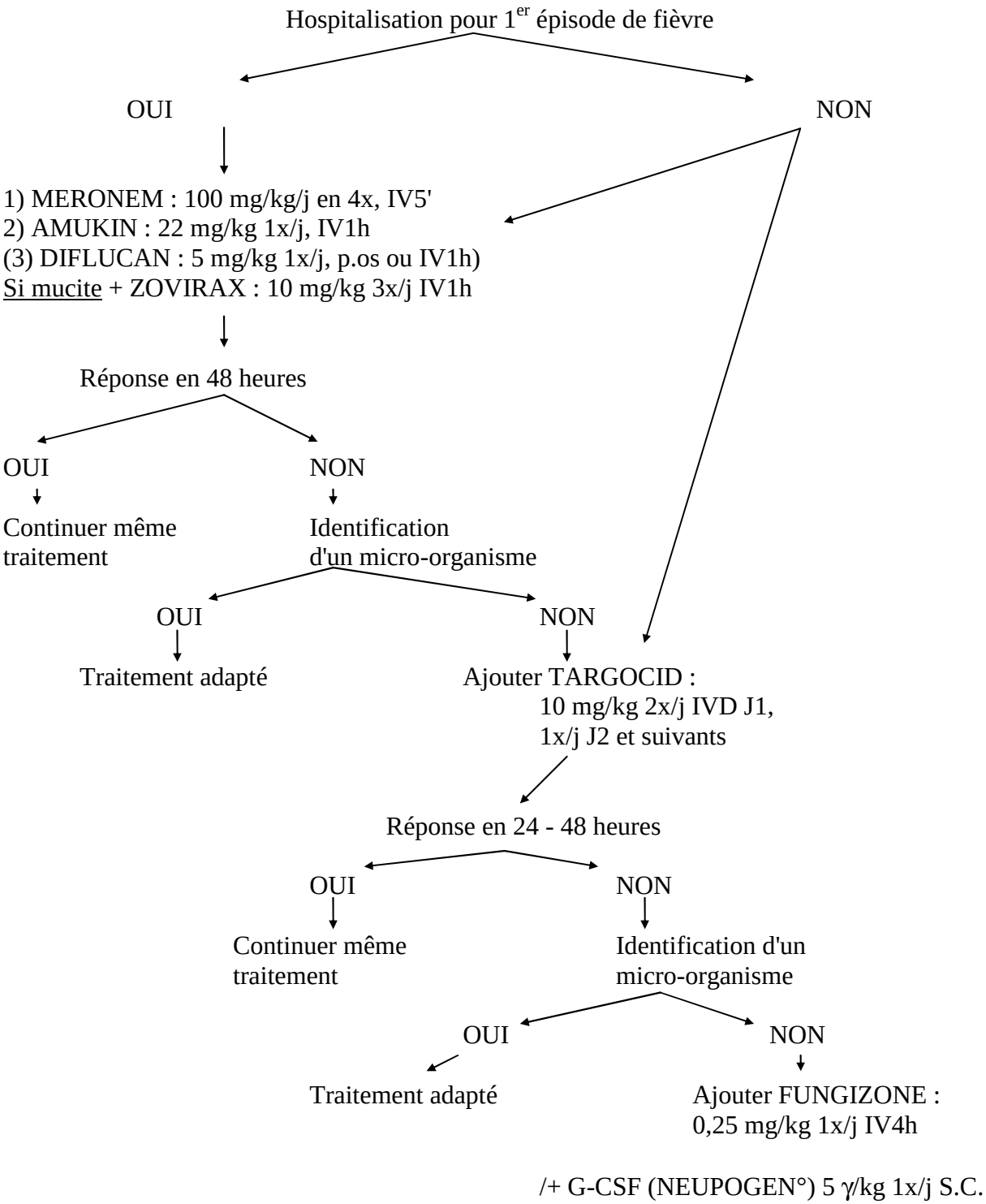
=> dose vaccin HiB à tout âge chez l'enfant si pas vacciné

Doses :

Ampicilline	200 mg /kg /jour en 6x /jour !
Ceftriaxone	100 mg /kg /jour en 1x /jour !
Cefuroxine	200 mg /kg /jour en 6x /jour !
Amukin	20 mg /kg /jour en 1x /jour IV 30 min.
Cefotaxime	200 mg /kg /jour en 6x /jour

Le patient neutropénique fébrile

NEUTROPENIE < (500 -) 1000 / mm^3
Fièvre > 38°



/+ G-CSF (NEUPOGEN°) 5 γ/kg 1x/j S.C.

PRISE EN CHARGE INITIALE D'UN JEUNE NOURRISSON FEBRILE

DEFINITIONS :

Jeune nourrisson : Agé de moins de trois mois.
Fébrile : Température mesurée en rectale, et supérieure ou égale à 38°C.

PRINCIPES :

La fièvre dans cette tranche d'âge ne doit jamais être considérée comme un symptôme banal ; le risque d'infection bactérienne invasive est plus important que chez l'enfant plus âgé.
Les difficultés diagnostiques à cet âge tiennent au caractère non spécifique et souvent paucisymptomatique à leur début, d'infections potentiellement sévères. Les signes sont d'autant moins spécifiques que l'enfant est plus jeune.
Les 2/3 à 3/4 de ces enfants ont une infection virale. Dans 20 à 25 % des cas, les infections sont d'origine bactérienne. Dans 5 à 10 %, ces nourrissons fébriles ont une bactériémie avec ses risques de complications. L'infection bactérienne la plus fréquente dans cette tranche d'âge est la pyélonéphrite aiguë.

Haut risque d'infection potentiellement sévère (un seul signe suffit):
Troubles du comportement
Troubles de la vigilance et/ou du tonus
Anomalies du cri
Anomalies de la réactivité (envers l'entourage familier)
Irritabilité et/ou inconsolabilité
Difficultés d'alimentation
Anomalies de l'hémodynamique
Anomalies de la coloration
Signes de détresse respiratoire
Signes de déshydratation
Signes en faveur d'une infection des parties molles ou du squelette
Purpura

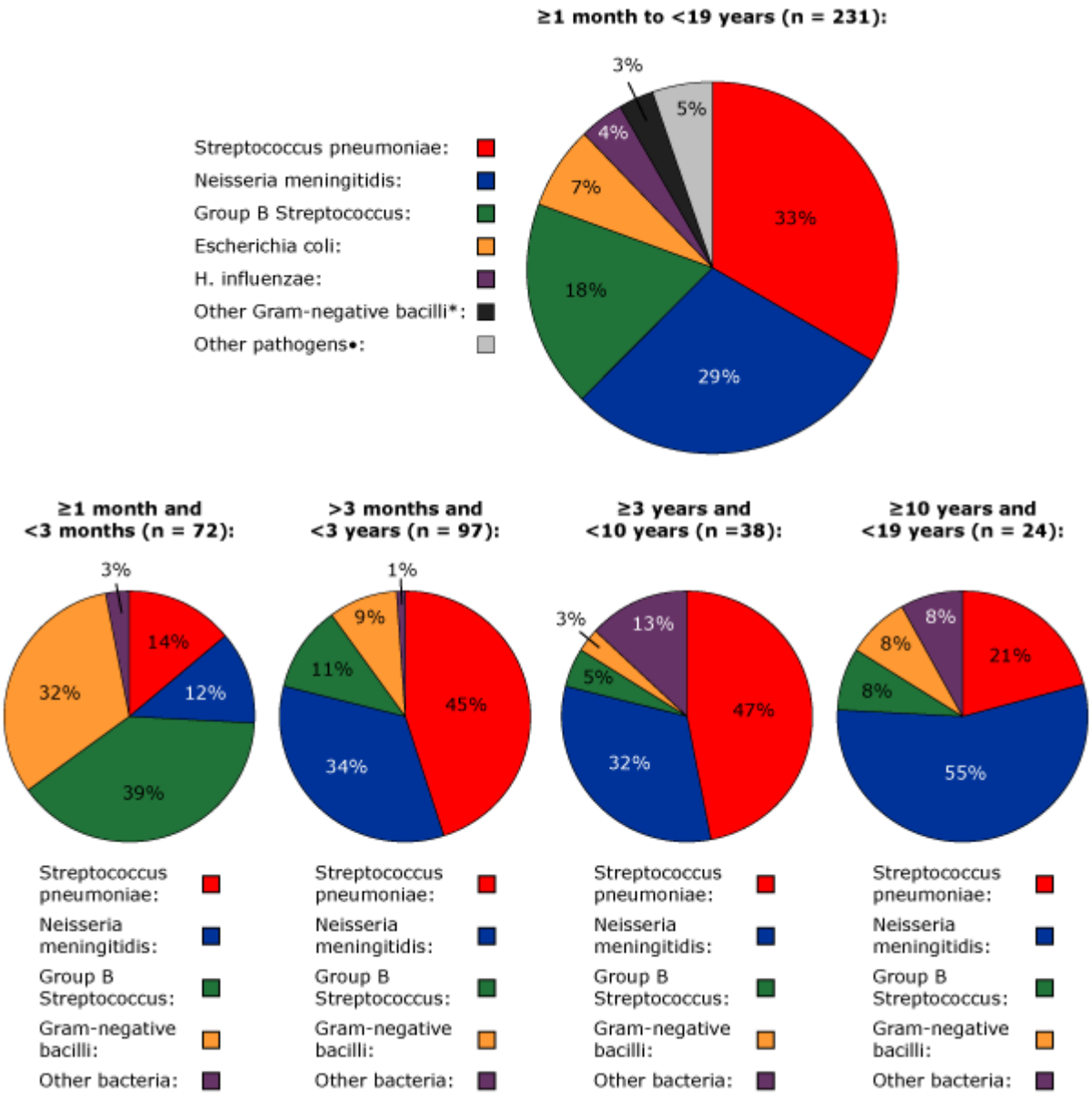
Chez les nourrissons ne présentant aucun de ces signes, l'évaluation clinique ne permet pas à elle seule dans cette tranche d'âge d'exclure une infection bactérienne sévère, et des examens complémentaires sont indispensables

Les nourrissons à bas risque d'infection bactérienne sont définis comme ceux qui ne présentent aucun signe clinique définissant le haut risque et aucun signe biologique en faveur d'une infection bactérienne : globules blancs compris entre 5 000 et 15 000 par mm^3, pas de syndrome inflammatoire : CRP < 10 mg /l, examen direct des urines (RUSU) fiable normal (ce qui n'exclut pas une infection urinaire !).

Il est par ailleurs possible de garder quelques heures en observation à l'HP (avec mesure répétée de la température rectale) un nourrisson avec une fièvre non quantifiée par les parents ou objectivée par les parents mais non retrouvée aux urgences, et dont l'état général et l'examen physique sont sans particularité. On envisagera un retour à domicile après avis du senior si l'enfant est resté apyrétique et stable et que sa biologie n'est pas perturbée.

AGE :	MISE AU POINT :	ATTITUDE :
< 1 mois	Biologie + hémoculture Rx thorax si symptômes respiratoires Ponction sus-pubienne / sondage vésical PL d'office +/- PCR Herpès sur LCR [$]	Hospitalisation d'office Antibiothérapie d'office : CLAFORAN PENTREXYL AMUKIN +/- ZOVIRAX
1 mois à 3 mois	Biologie + hémoculture Rx thorax si symptômes respiratoires Ponction sus-pubienne / sondage vésical PL d'office sauf avis senior	Hospitalisation et antibiothérapie selon résultats et avis senior : CLAFORAN PENTREXYL Et AMUKIN si RUSU +

[$] La PCR Herpès sur le LCR est à réaliser chez tout nourrisson de moins de un mois, qui présente une anamnèse ou des signes cliniques cutanés, oculaires ou buccaux d'infection herpétique, ou qui présente des convulsions ou un état septique. Un traitement empirique par Zovirax sera dans ce cas instauré jusqu'à obtention des résultats de la PCR


≥1 month to <19 years (n = 231):
Streptococcus pneumoniae:
Neisseria meningitidis:
Group B Streptococcus:
Escherichia coli:
H. influenzae:
Other Gram-negative bacilli*:
Other pathogens•:
33%
29%
18%
7%
4%
3%
5%
≥1 month and <3 months (n = 72):
14%
12%
39%
32%
3%
>3 months and <3 years (n = 97):
45%
34%
11%
9%
1%
≥3 years and <10 years (n =38):
47%
32%
5%
13%
3%
≥10 years and <19 years (n = 24):
21%
55%
8%
8%
8%
Streptococcus pneumoniae:
Neisseria meningitidis:
Group B Streptococcus:
Gram-negative bacilli:
Other bacteria:

CONVULSION => ETAT DE MAL CONVULSIF

0-5 minutes

ABC → **BENZOS** → **« DON'T »**

TEMESTA (*)
IR : 0.1mg/kg
IV lente 1min : 0.05-0.1mg/kg
Max 4mg/dose

ou

VALIUM (*)
IR : 0.5mg /kg

Thiamine ? Pyridoxine ?

IV lente 1 min: 0.1-0.3 mg /kg
Max 10 mg /dose

D extro
O 2
N arcan ?
T : Trouble ionique ?
Trauma ?
Température ?

(*) : 2ème dose après 5 minutes si nécessaire

Biologie : Sang complet, CRP, pH, glycémie, iono (avec Ca et Mg), bilan hépatique et rénal, HbCO ? toxico ?, dosage médicaments (traitement de fond)?

5-20 minutes

ABC → **< 2 A** — **> 2 A** → **Etiologie ? ***

MIN 10

LUMINAL (IV)

20 mg /kg IV 20 min
max. 1 mg /kg /min

max. 300 mg /dose

Dilué ds LP0.9 % ou G5 %

DEPAKINE (IV)

<u>Pas de Dépakine en traitement de fond :</u>
IV (5 min) 20 mg /kg
(10-15 mg /kg chez adulte)

<u>Dépakine en traitement de fond :</u>
IVC 0.5 mg /kg /h (max. 2.5 g /24h)
Dilué dans LP0.9 % ou G5% (100 mg/1ml)

*Ex complémentaires :	Traitement étiologique :
CT cérébral ?	*Glucose ?*
PL ?	*Antipyrétiques ?*
Bilan métabolique ?	*Antibiotiques ?*
EEG ?	*Zovirax ?*
Bactériologie, virologie ?	*Thiamine ?*
Toxicologie ?	*Pyridoxine ?*

20-40 minutes

ABC → **< 2 A** — **> 2 A** → **Etiologie ***

MIN 30

Si persistance convulsion :

PHENOBARBITAL (IV)
+ 10 mg /kg IV 10min

MIN 40

DIPHANTOINE (IV)

20 mg /kg IV 20-30min
max. 0.5-1 mg/kg/min
max. 1000 mg/dose

Dilué dans LP 0.9 % uniquement

DEPAKINE (IV)

Pas de Dépakine en traitement de fond :
25 min après fin du bolus :
IVC : 1mg/kg/h
(max 1gr /24h chez l'enfant et
2.5 gr /24h chez l'adulte)
Dépakine en traitement de fond :
IVC : 0.5 => 1mg /kg /h si convulse encore

PHENOBARBITAL (IV)

20 mg /kg IV 20 min
max. 1 mg /kg /min
max. 300 mg /dose

Alternative = DIPHANTOINE

La déshydratation

= déficits
→ Eau
→ NaCl et KCl
→ Énergies

Le raisonnement dépendra de :
- l'importance de ces déficits, corrélé à l'examen clinique, au pH et à l'ionogramme,
- de la vitesse d'installation,
- de la fragilité préalable (métabolique, endocrinienne, rénale, intestinale, hépatique, cardiaque),
- de la corpulence (anorexique ou obésité) du sujet.

Perfuser si :

- perte de > 5 % en 1 jour ;
- échec du traitement entéral ;
- si fragilité antérieure (maladie métabolique ou endocrinienne, intestin court, iléostomie, infirmité motrice, insuffisance rénale ou cardiaque ou hépatique, si réserves faibles : mucoviscidose,..) et perte > 5 % ;
- si perte de > 10 % ;
- si Natrémie > 155 ou < 125 mEq / L ;
- CALCUL DU TROU ANIONIQUE DANS LES DIARRHEES :
  Trou anionique=290-(Na+K)
  <50 mOsm /l et Na>90 mEq /l =diarrhée sécrétoire.
  >100 mOsm /l et Na <60 mEq /l =diarrhée osmotique (Na et K fecaux en mEq/l).

PRINCIPES DE BASE DE LA REHYDRATATION I.V.

0 – 1 h — **15-20 ml /kg de Physio Na Cl 0,9 % ou Hartmann non glucosé**

Correction
Vol. circulant
« Déchoquage initial » (Si choc ++ : HAES-STERIL)

Exclure ! ! : - méningite
- SHU
- Diabète

1 – 25 h analyse

Maintenance

Règle des 100-50-20

+

Perte de poids

à corriger en: 24 h ou 48 h

+

Pertes excessives selon

dig. Haut => LP
dig. Bas =>Hartmann

Na < 130 mEq /l	**Na 130 – 150 mEq /l**	**Na > 150 mEq /l**
• 2/3 LP ou Hartmann 1/3 Glucosé 10% (ou NaCl 0,9% G5%) • Réhydratation en 24 h (1/4 en4h, puis 3/4 en20h) • Si convulsions (Na < 120) : NaCl 3% 4 ml/kg = 2 mEq/kg en 15-20 min	• 1/2 LP ou Hartmann 1/2 Glucosé 10% • Réhydratation en 24 h (1/4 en 4 h, puis 3/4 en 20 20h)	• 2/5 LP ou Hartmann 3/5 glucosé 10% • Réhydratation en 48 h (1/2 perte de poids / 24 h) • ↓ Na+ < 0,5 mEq Na/L/h sinon risque de convulsion • Dialyse si > 180 ? ! • Garder une [Na] perf d'autant + grde que le [Na] sang est élevé Na 35 à 75 mEq /L

Suivi de l'évolution

Iono, urée, PS, pH / 4 h **Si urée reste ↑ => ↑ apports liquidiens**

Si Na^+ reste ↑ => ↓ apports en Na^+

Si Na^+ ↓↓↓ => restriction hydrique et ↑ apports Na^+
+ K : ssi diurèse

NOTES

1. REGLES DES 100 – 50 – 20 kg/j ou des 4 – 2 – 2 (cc /kg /h)

< 10 kg :	100 ml /kg /jour (4)
10-20 kg :	1000 ml + 50 ml /kg /jour (2) pour les 10 – 20 kg
> 20 kg :	1500 ml + 20 ml /kg /jour (1) pour les > 20 kg

2. HYPER Na et convulsions pendant la correction

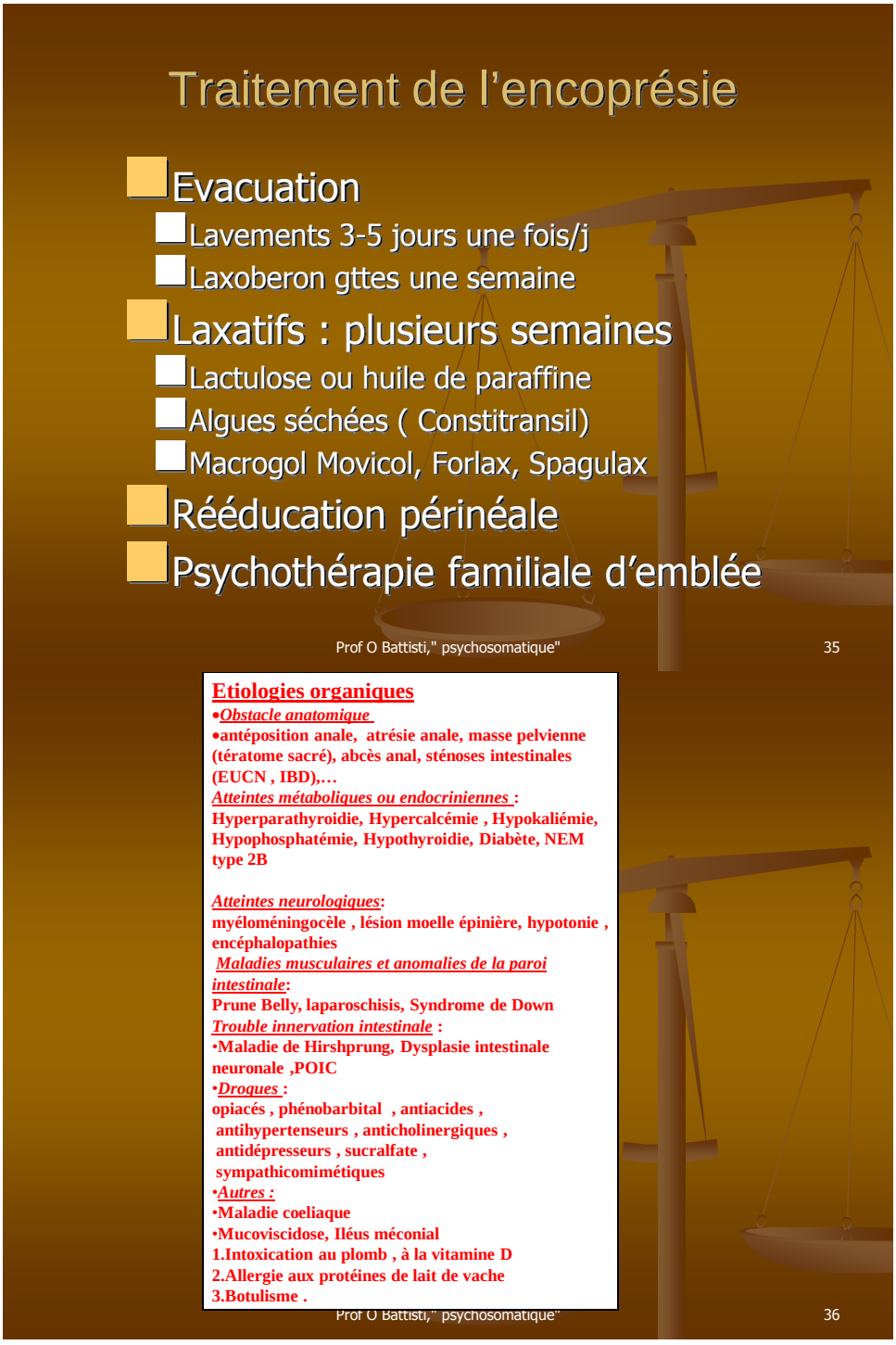
3. K^+

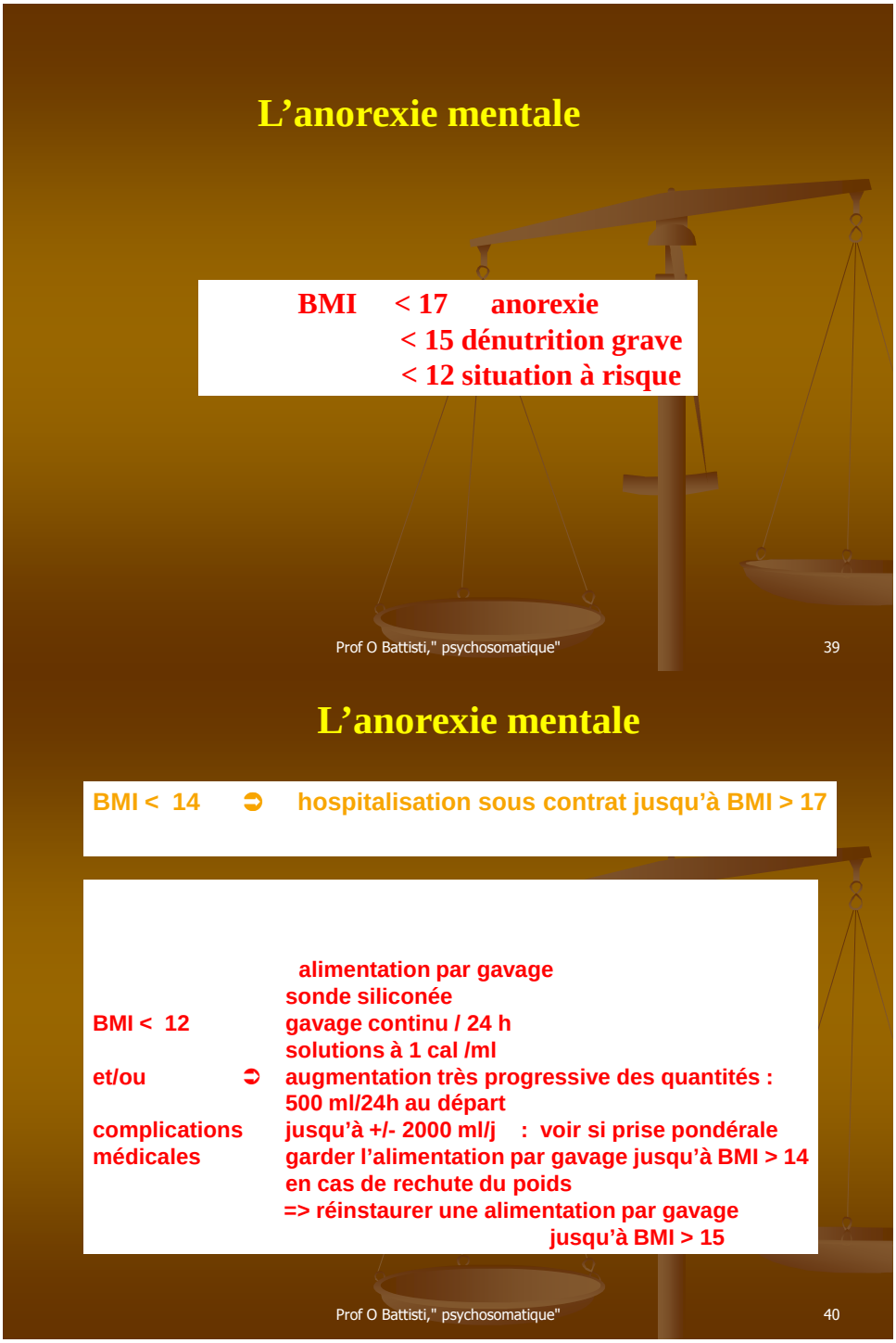
si nl :	+ 10 – 20 mEq /L de soluté ssi diurèse +
si ↑ :	attendre normalité et reprise diurèse
si ↓ :	+ 30-40 mEq /L de soluté

Ca^{++} si hypoCa (+ fréquent si hyperNa^+) => gluconate Ca 10 %

20 ml /L de soluté

! ! Incompatible avec bicarbonate
Compatible avec lactate


Traitement de l'encoprésie
Evacuation
Lavements 3-5 jours une fois/j
Laxoberon gttes une semaine
Laxatifs : plusieurs semaines
Lactulose ou huile de paraffine
Algues séchées (Constitransil)
Macrogol Movicol, Forlax, Spagulax
Rééducation périnéale
Psychothérapie familiale d'emblée
Prof O Battisti," psychosomatique"
35
Etiologies organiques
•Obstacle anatomique
•antéposition anale, atrésie anale, masse pelvienne (tératome sacré), abcès anal, sténoses intestinales (EUCN , IBD),…
Atteintes métaboliques ou endocriniennes :
Hyperparathyroidie, Hypercalcémie , Hypokaliémie, Hypophosphatémie, Hypothyroidie, Diabète, NEM type 2B
Atteintes neurologiques:
myéloméningocèle , lésion moelle épinière, hypotonie , encéphalopathies
Maladies musculaires et anomalies de la paroi intestinale:
Prune Belly, laparoschisis, Syndrome de Down
Trouble innervation intestinale :
•Maladie de Hirshprung, Dysplasie intestinale neuronale ,POIC
•Droques :
opiacés , phénobarbital , antiacides , antihypertenseurs , anticholinergiques , antidépresseurs , sucralfate , sympathicomimétiques
•Autres :
•Maladie coeliaque
•Mucoviscidose, Iléus méconial
1.Intoxication au plomb , à la vitamine D
2.Allergie aux protéines de lait de vache
3.Botulisme .
Prof O Battisti," psychosomatique"
36

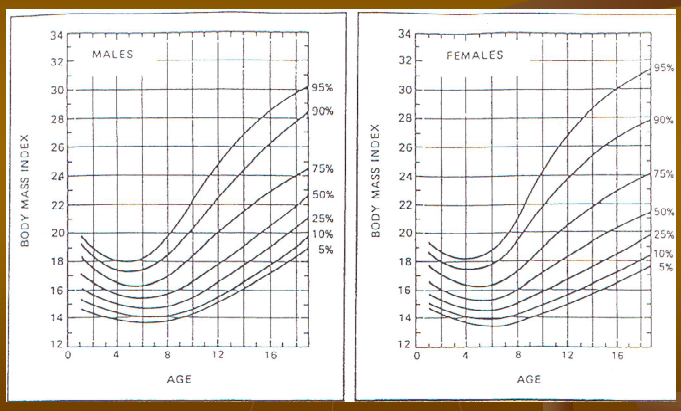

L'anorexie mentale
BMI < 17 anorexie
< 15 dénutrition grave
< 12 situation à risque
Prof O Battisti," psychosomatique" 39
L'anorexie mentale
BMI < 14 ➲ hospitalisation sous contrat jusqu'à BMI > 17
alimentation par gavage
sonde siliconée
BMI < 12 gavage continu / 24 h
solutions à 1 cal /ml
et/ou ➲ augmentation très progressive des quantités :
500 ml/24h au départ
complications jusqu'à +/- 2000 ml/j : voir si prise pondérale
médicales garder l'alimentation par gavage jusqu'à BMI > 14
en cas de rechute du poids
=> réinstaurer une alimentation par gavage
jusqu'à BMI > 15
Prof O Battisti," psychosomatique" 40

L'anorexie mentale

Surveillance :
Ions Na, K, Ca, P
ECG
Echocardiographie
(! ! ! décompensation cardiaque, congestion)
Supplémentation
en phosphates lors de la rénutrition
de 780 à 1560 mg de phosphore élément PO par jour

Prof O Battisti," psychosomatique" 41

L'anorexie « mentale »



Prof O Battisti," psychosomatique" 37

Atrésie de l' œsophage classification anatomique

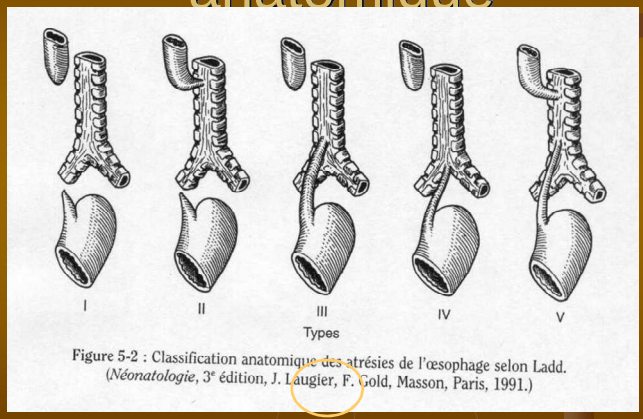
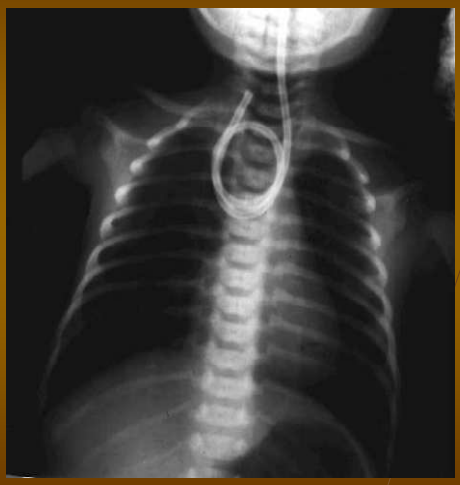
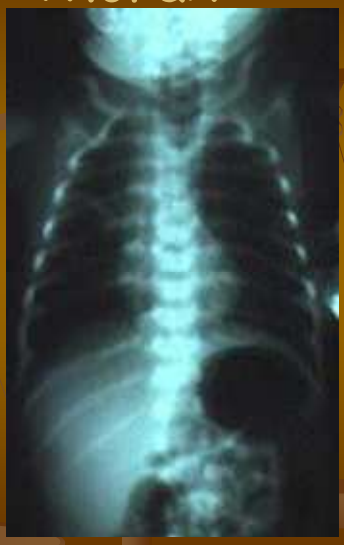


Figure 5-2 : Classification anatomique des atrésies de l'œsophage selon Ladd.
(*Néonatologie*, 3e édition, J. Laugier, F. Gold, Masson, Paris, 1991.)

Prof O Battisti, semio-techniques gastro 23

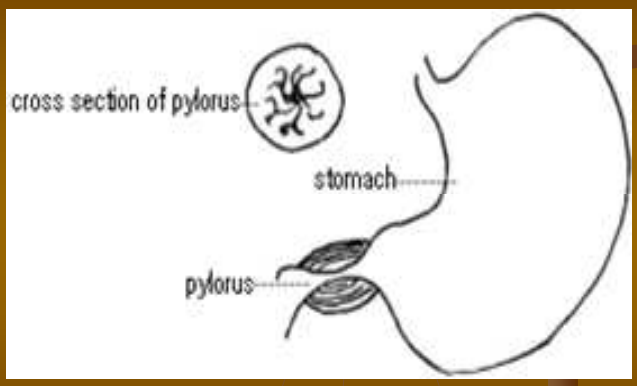
Radiographie de Thorax





Prof O Battisti, semio-techniques gastro 24

Sténose du pylore



Hypertrophie concentrique
de la musculeuse du pylore

Prof O Battisti, semio-techniques gastro 40

Vomissements

Perte H+, Cl-, K+
Déshydratation EC

Alcalose métabolique hypochlorémique

Excrétion NaHCO3

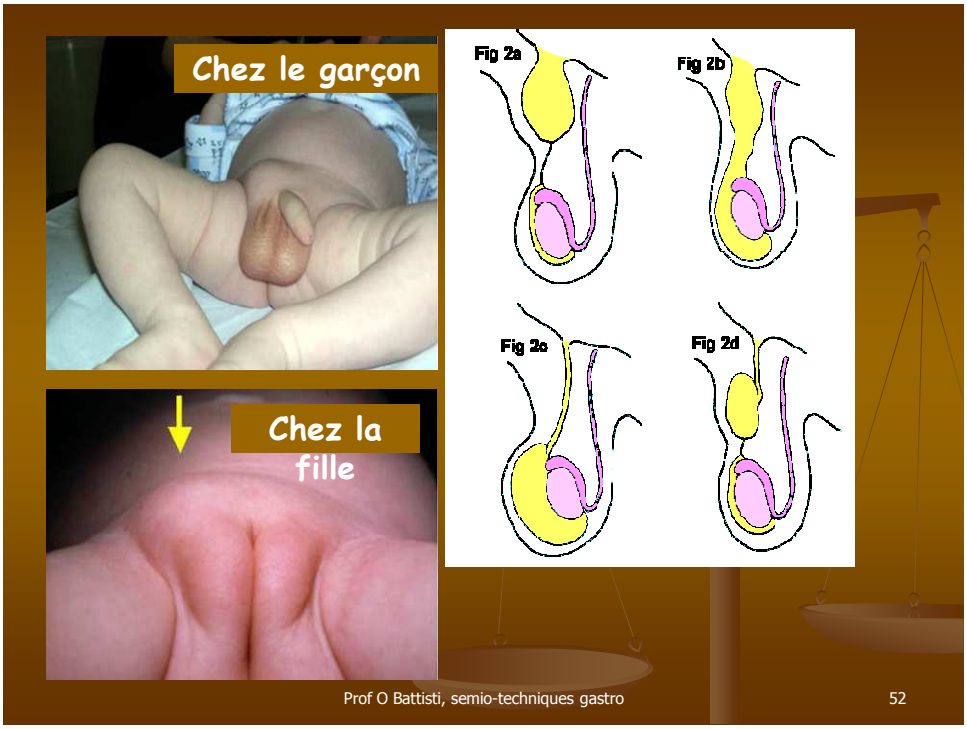
Urine alcaline

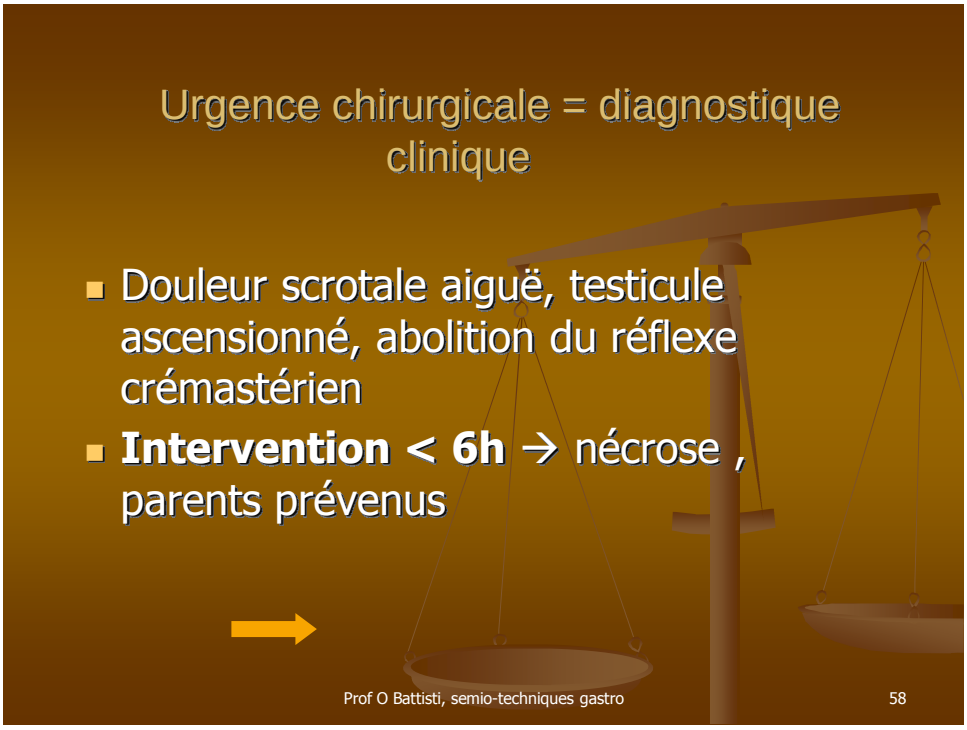
Persistance

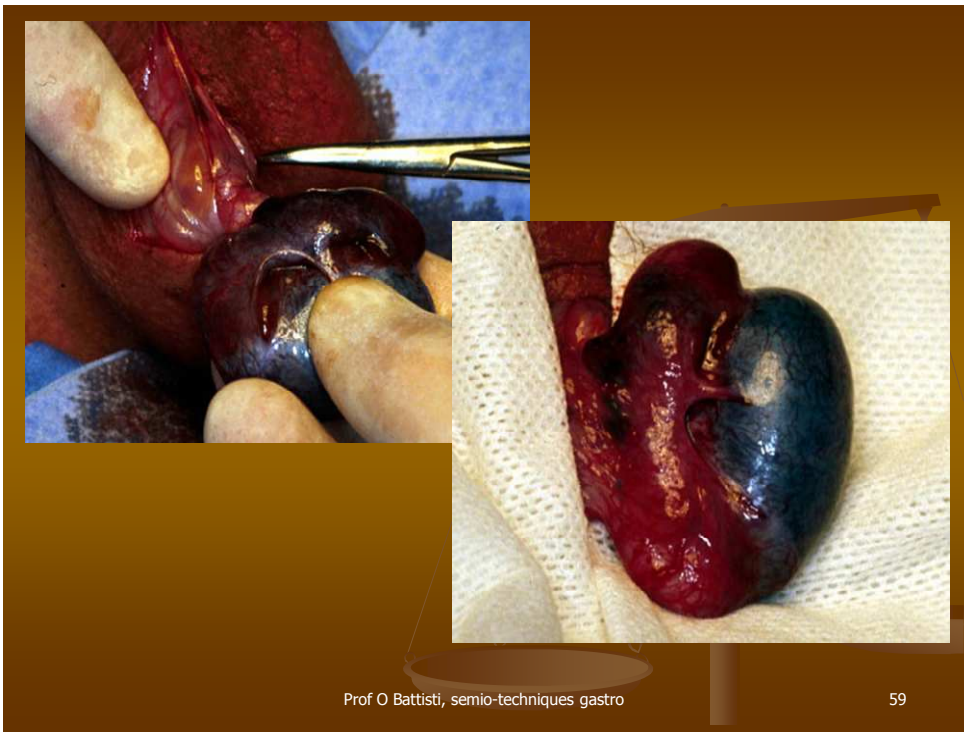
Réabsorption NaCl, NaCOH3
excrétion K+

Acidurie paradoxale Chlore U < 20 mEq/l

Prof O Battisti, semio-techniques gastro 44


Chez le garçon
Chez la fille
Fig 2a
Fig 2b
Fig 2c
Fig 2d
Prof O Battisti, semio-techniques gastro
52

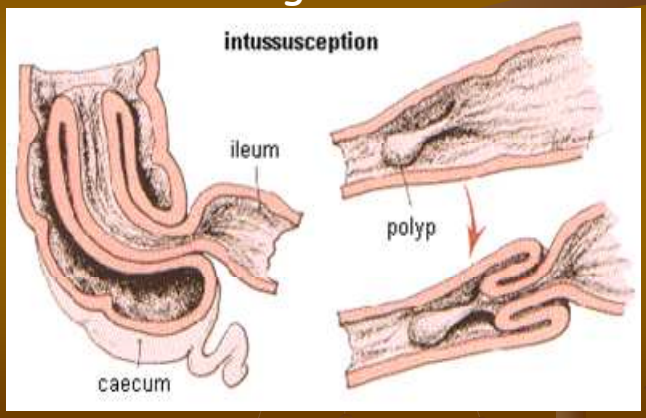

Urgence chirurgicale = diagnostique clinique
Douleur scrotale aiguë, testicule ascensionné, abolition du réflexe crémastérien
Intervention < 6h → nécrose , parents prévenus
Prof O Battisti, semio-techniques gastro
58


Prof O Battisti, semio-techniques gastro
59


Ischémie digestives et gonadiques
Prof O Battisti, semio-techniques gastro
61

Invagination intestinale

Passage d'un segment intestinal d'aval dans le segment d'amont



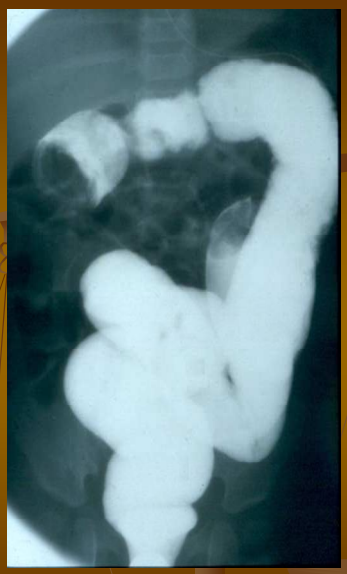
Invagination intestinale aiguë

- Formes
  - Idiopathiques: hyper péristaltisme, GG mésentériques, hyperplasie des plaques de Peyer
  - Secondaires: Meckel, tumeur, purpura rhumatoide
- Types d 'invagination :
  - Iléo-iléale (la plus simple)
  - Iléo-cæcale (la plus fréquente)
  - Iléo-colique (transvalvulaire)

Examens Radiologiques

- Cliché abdomen sans préparation
  - Boudin d 'invagination : opacité allongée (50 à 60 %)
  - Contenu aérique et fécal colique pauvre
  - Disparition du granité cæcal, niveaux HA
- **Échographie abdominale**
  - Examen diagnostic de référence
  - boudin = coupe transversale: aspect de "cible", coupe longitudinale: aspect en " pince de crabes"
- **Lavement opaque** (baryte, iode, air)

  = diagnostic et traitement



Vomissements du nouveau-né

CAUSES CHIRURGICALES	CAUSES MEDICALES DIGESTIVES	CAUSES EXTRADIGESTIVES
▫ Occlusion néonatale ▫ Atrésie duodénale ▫ Atrésie du grêle ▫ Iléus méconial ▫ Atrésie colique ▫ Hirschsprung	▫ R.G.O ▫ I P L V ▫ Infection ▫ Erreur de régime	▫ Infections - méningite - infection urinaire - etc... ▫ Neurologiques : - hémorragie méningée - H S D ▫ Endocriniennes - hyperplasie congénitale des surrénales ▫ Métaboliques - Aminoacidopathies - H Calcémie - Intoxication médicamenteuse ▫ Rénales

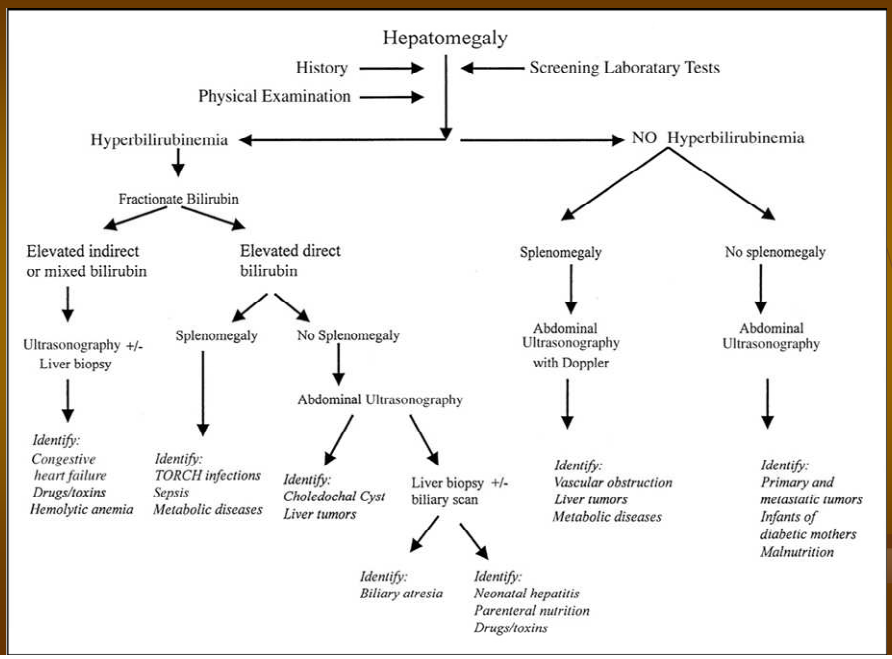
Prof O Battisti, semio-techniques gastro 93

Vomissements du nourrisson et de l'enfant

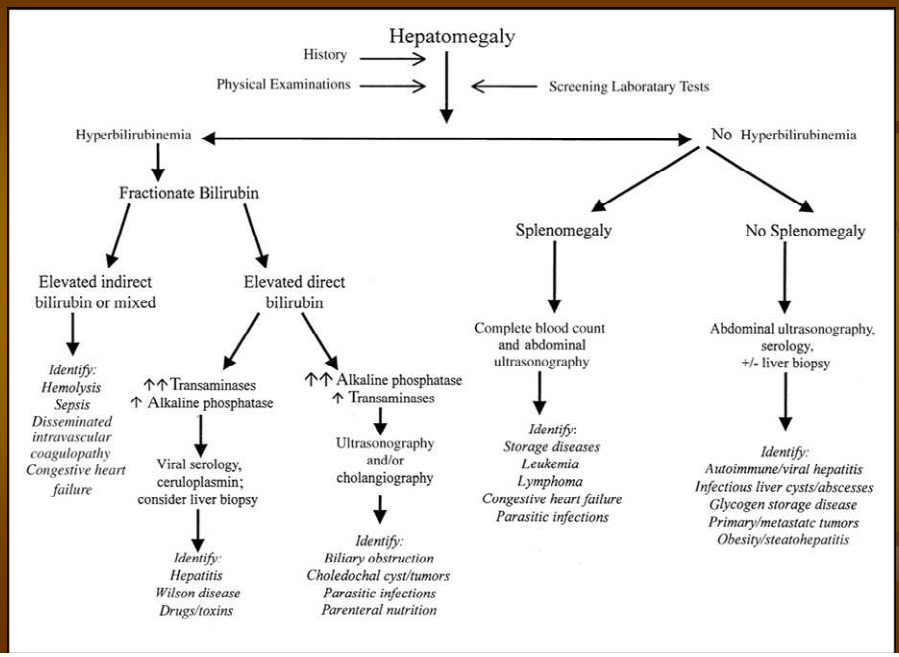
URGENCES CHIRURGICALES	CAUSES DIGESTIVES	CAUSES EXTRADIGESTIVES	CAUSES FONCTIONNELLES
▫ I I A ▫ Appendicite ▫ Péritonite ▫ Volvulus ▫ Hernie étranglée	▫ Sténose du pylore ▫ R.G.O ▫ I P L V ▫ Infection (GEA)	▫ Infections ▫ Neulogogiques - H.S.D - tumeur ▫ Endocriniennes - hyperplasie congénitale ▫ Métaboliques - hypercalcémie - intoxication ▫ Rénales	▫ Erreur de régime ▫ Vomissements acétoniques ▫ Vomissements psychogènes

Prof O Battisti, semio-techniques gastro 94

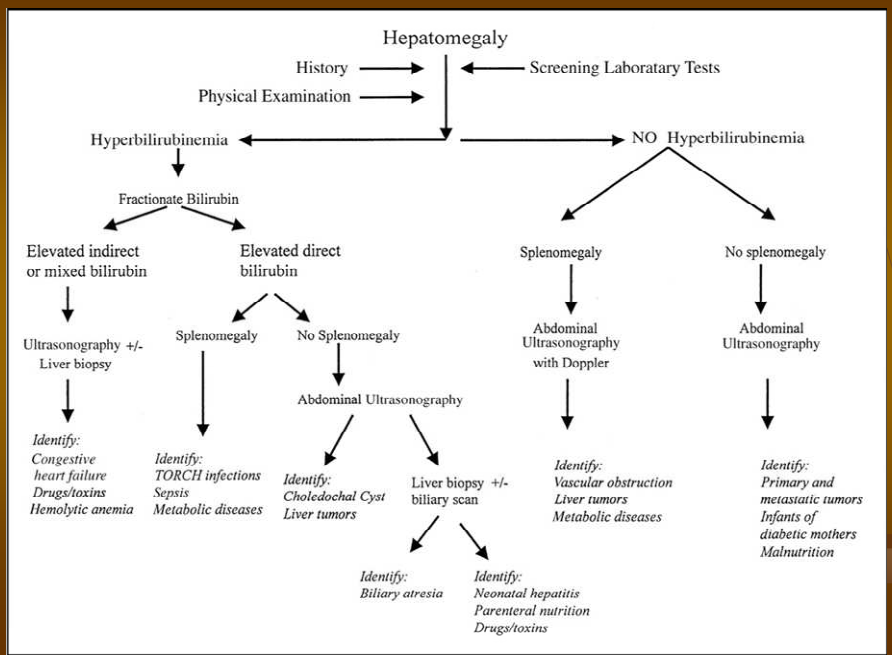
L'enfant de moins d'un an



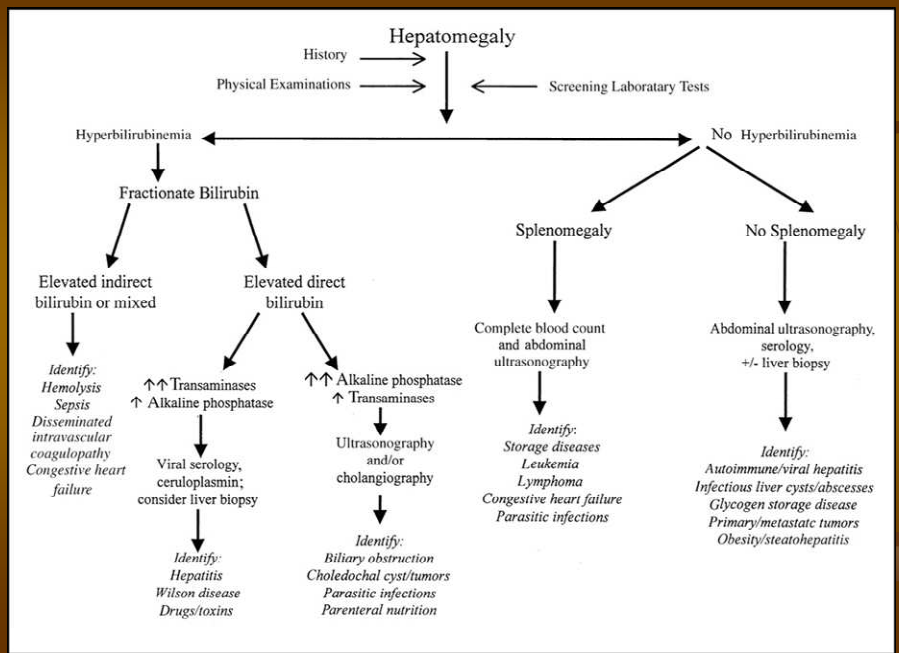
L'enfant de plus d'un an



L'enfant de moins d'un an



L'enfant de plus d'un an



Dysrégulations immunitaires et maladies inflammatoires du tube digestif

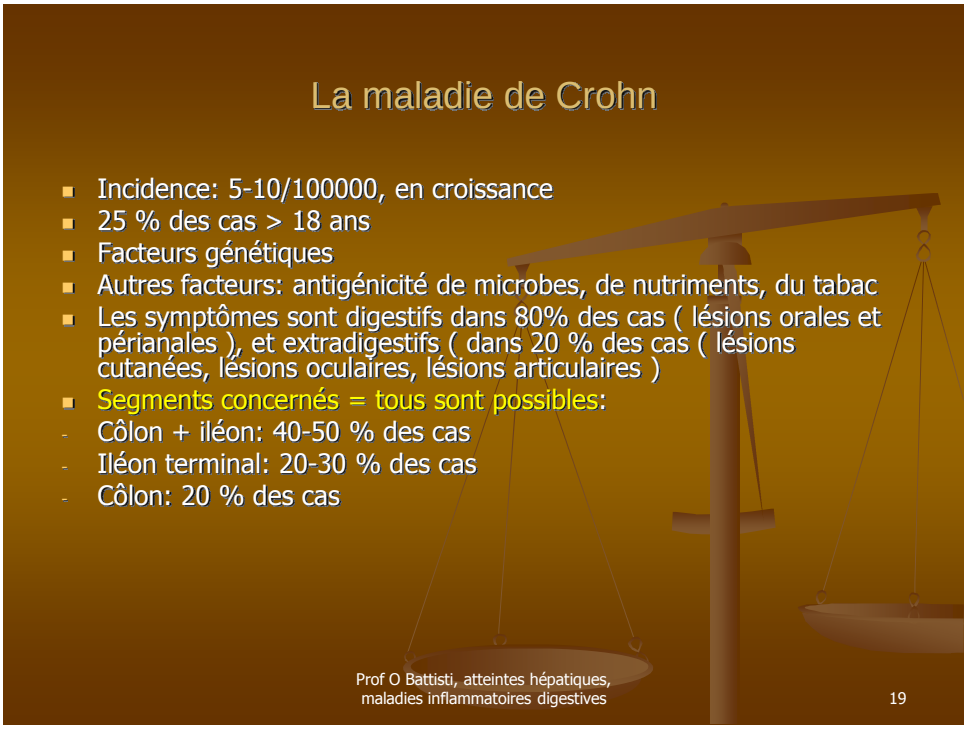
- Concept de GALT ou "gut associated lymphoïd tissue"
- Dans CU ou UC: Anticorps P-ANCA (antinuclear cytoplasmic antibody)
- Dans Crohn ou CD: Anticorps ASCA (antisaccharomyces cerevisae antibody)
- Mucus anormal
- Sécrétions anormales de produits inflammatoires
- Présence de récepteurs épithéliaux anormaux
- Exagération du "homing" et de la diapédèse

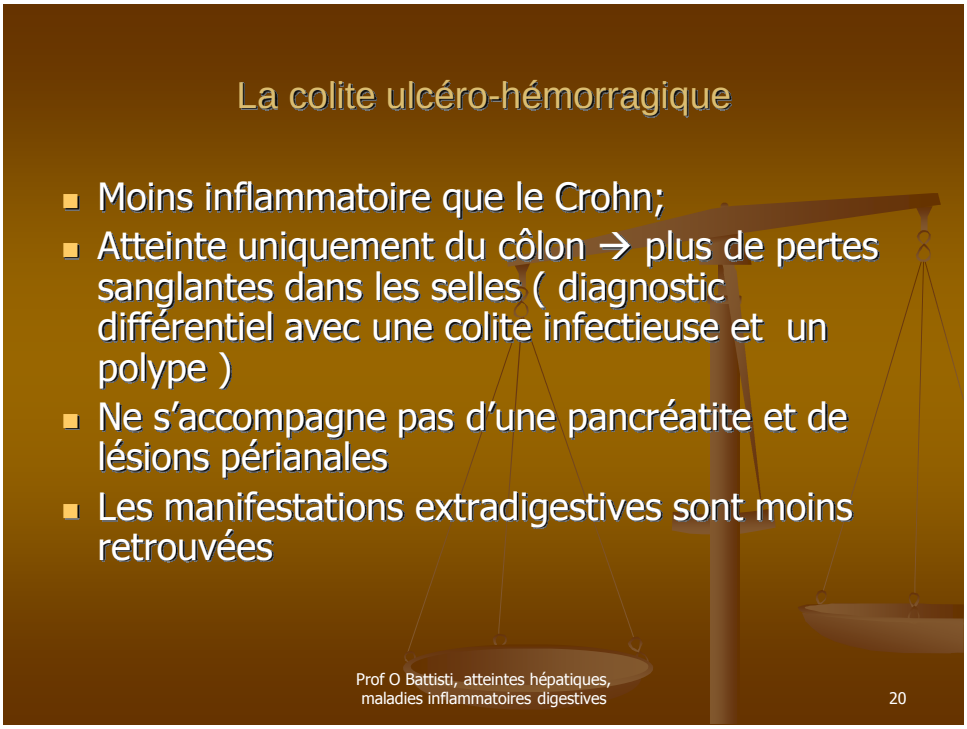
Prof O Battisti, atteintes hépatiques, maladies inflammatoires digestives 17

Symptomatologie et signes

- Douleurs abdominales
- Perte de poids, diarrhée, retard de croissance,
- pertes de sang dans les selles
- lésions de la bouche, lésions articulaires, lésions oculaires, lésions cutanées, arthrites
- Pâleur, troubles de l'humeur
- Recherche de répercussions biologiques, ostéoarticulaires, hématologiques, vitaminiques et hépatiques.

Prof O Battisti, atteintes hépatiques, maladies inflammatoires digestives 18

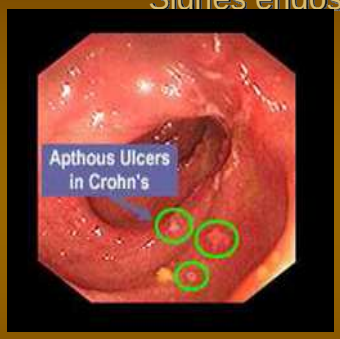

La maladie de Crohn
Incidence: 5-10/100000, en croissance
25 % des cas > 18 ans
Facteurs génétiques
Autres facteurs: antigénicité de microbes, de nutriments, du tabac
Les symptômes sont digestifs dans 80% des cas (lésions orales et périanales), et extradigestifs (dans 20 % des cas (lésions cutanées, lésions oculaires, lésions articulaires)
Segments concernés = tous sont possibles:
- Côlon + iléon: 40-50 % des cas
- Iléon terminal: 20-30 % des cas
- Côlon: 20 % des cas
Prof O Battisti, atteintes hépatiques, maladies inflammatoires digestives
19

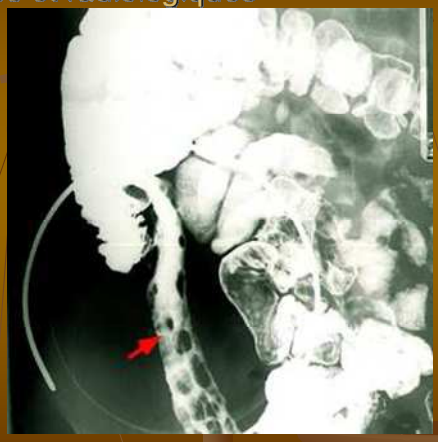

La colite ulcéro-hémorragique
Moins inflammatoire que le Crohn;
Atteinte uniquement du côlon → plus de pertes sanglantes dans les selles (diagnostic différentiel avec une colite infectieuse et un polype)
Ne s'accompagne pas d'une pancréatite et de lésions périanales
Les manifestations extradigestives sont moins retrouvées
Prof O Battisti, atteintes hépatiques, maladies inflammatoires digestives
20

Monitoring

- clinique (index d'activité)
- biologique
- et par imagerie

Signes endoscopiques et radiologiques





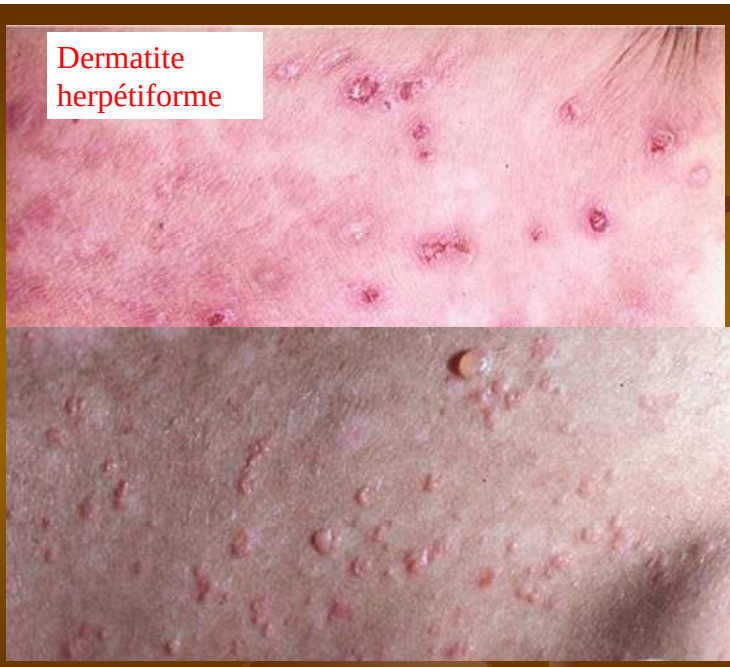
Traitements médicamenteux
il s'agit d'une maladie inflammatoire !

- Corticoïdes: prednisone et budesonide
- Aminosalicylates: sulfazaline, mesalamine
- Immunosuppresseurs: azathioprine, 6-mercaptopurine
- Anti TNF: infliximab, natalizumab, adalimumab; Tacrolimus in loco
- Cures d'antibiotiques: ciproxine, metronidazole

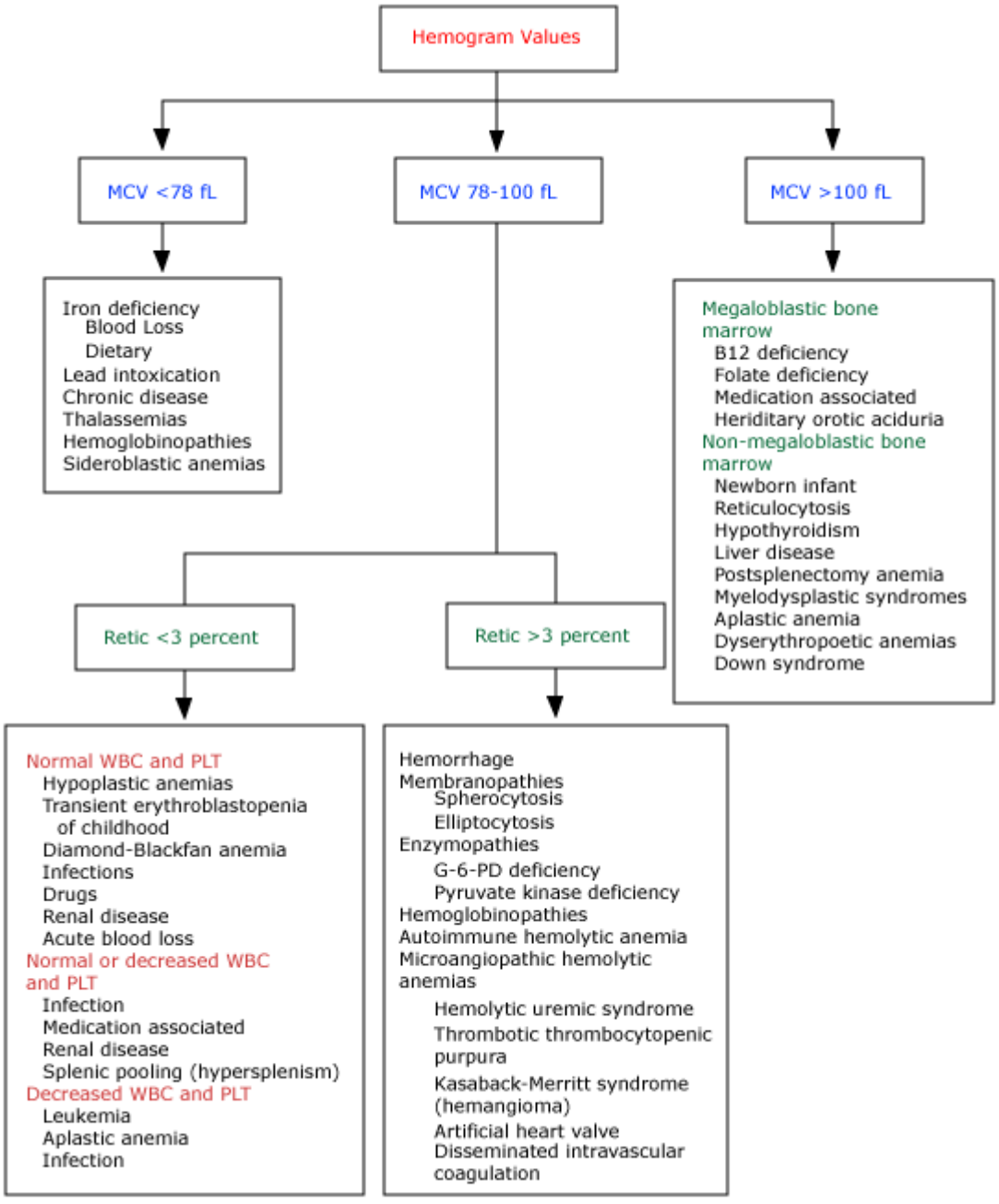
La maladie caeliaque

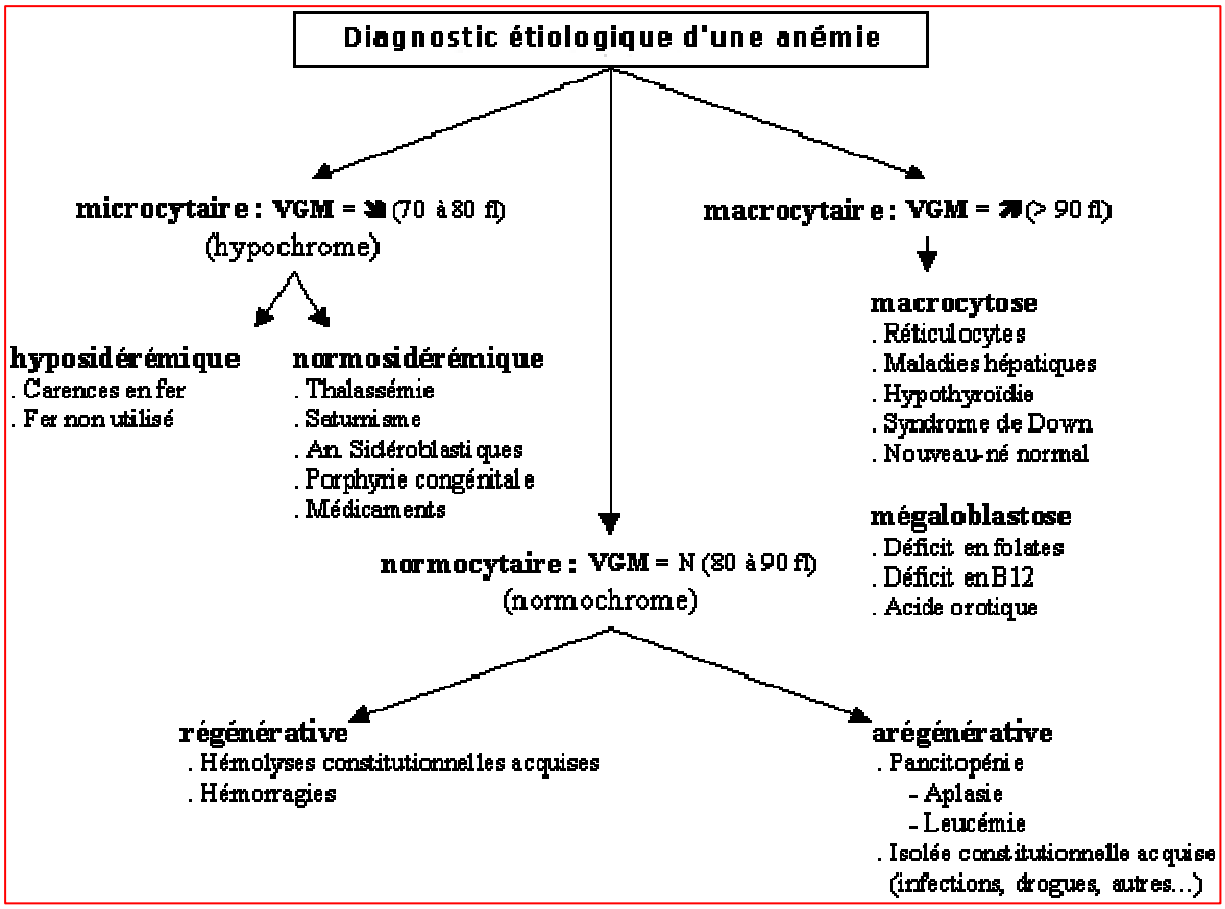
- Fréquente: 1/80 à 1/500
- Peut se déclarer à tout âge
- Forte composante génétique soit sur le système HLA (DR et DQ), mais aussi par gène (CTLA-)
- Réaction immuno-allergique à la gliadine
- Les symptomes sont digestifs et extradigestifs (notamment fonte du tissu adipeux, dermatite herpétiforme)

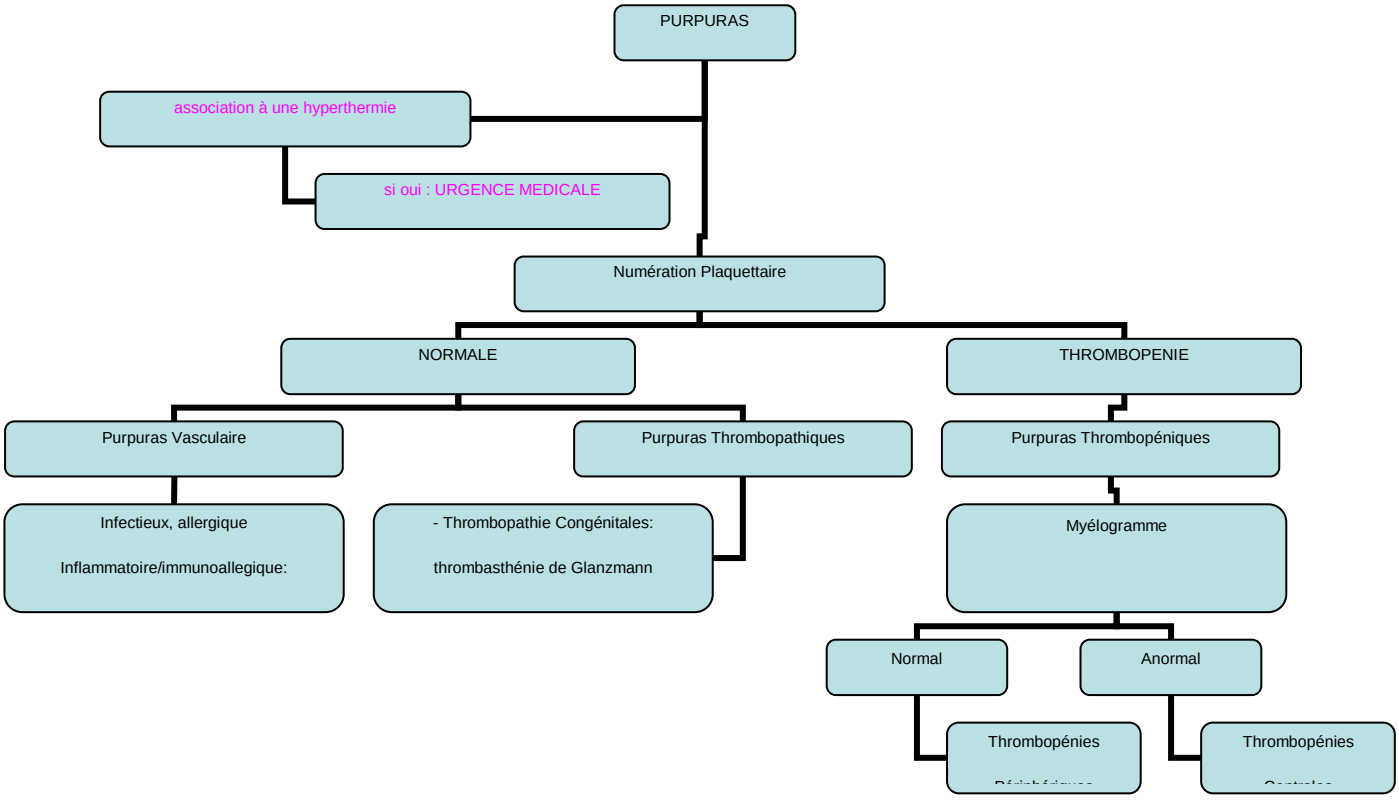
Dermatite herpétiforme

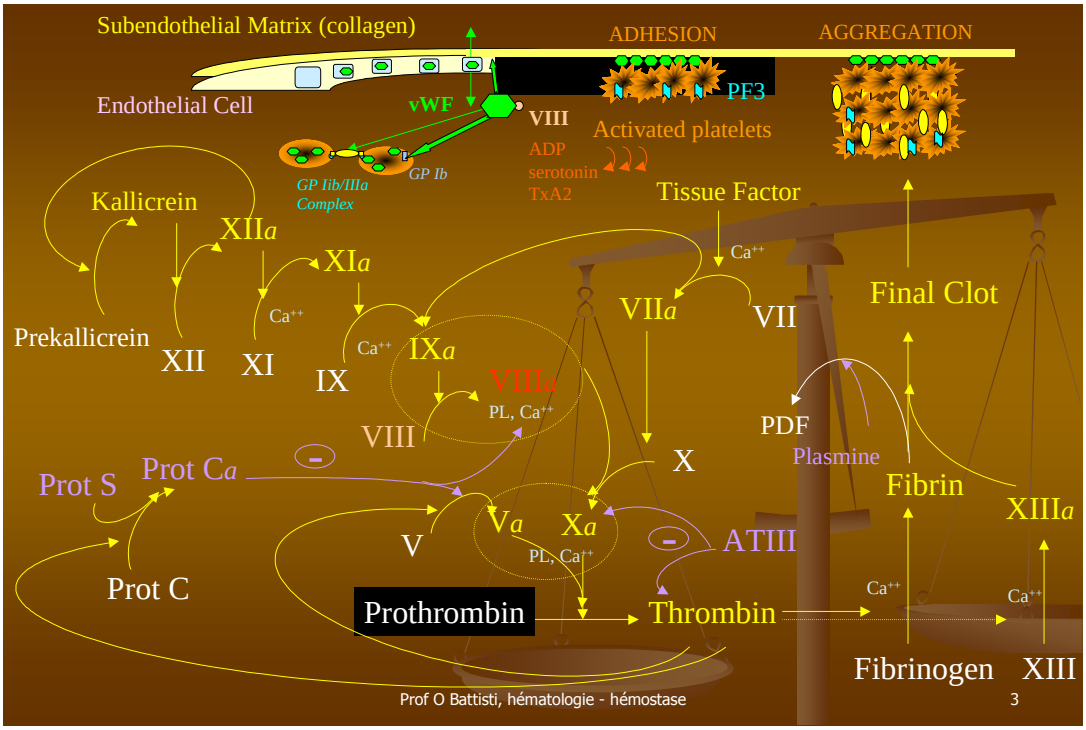


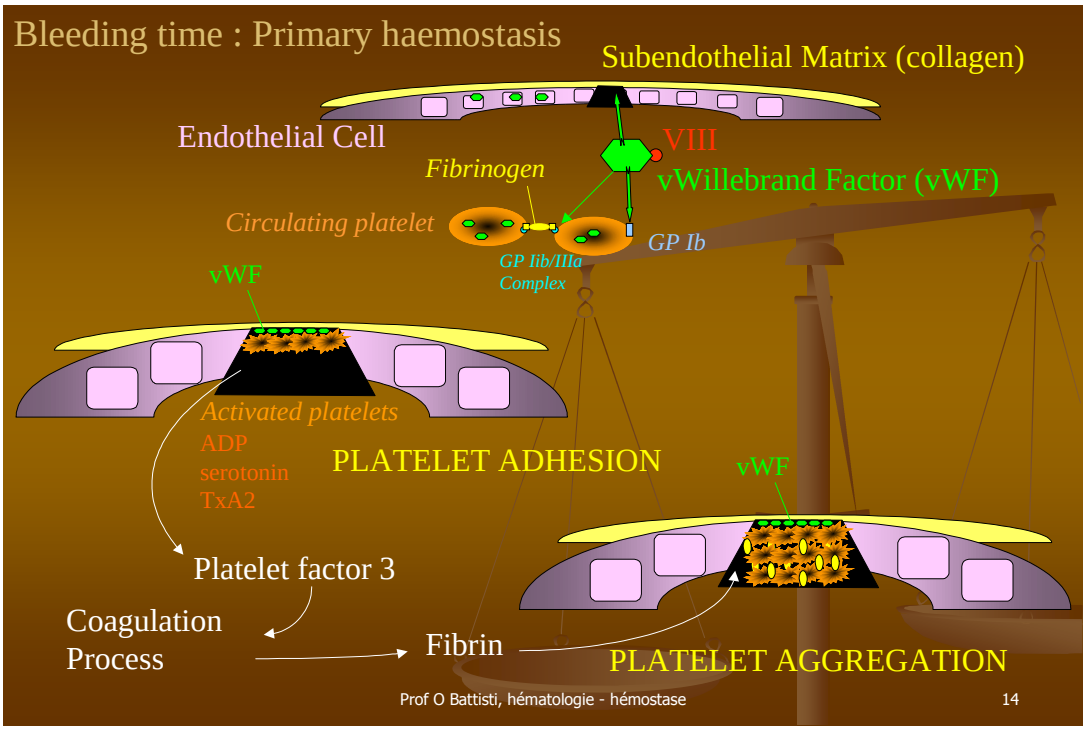
Hématologie

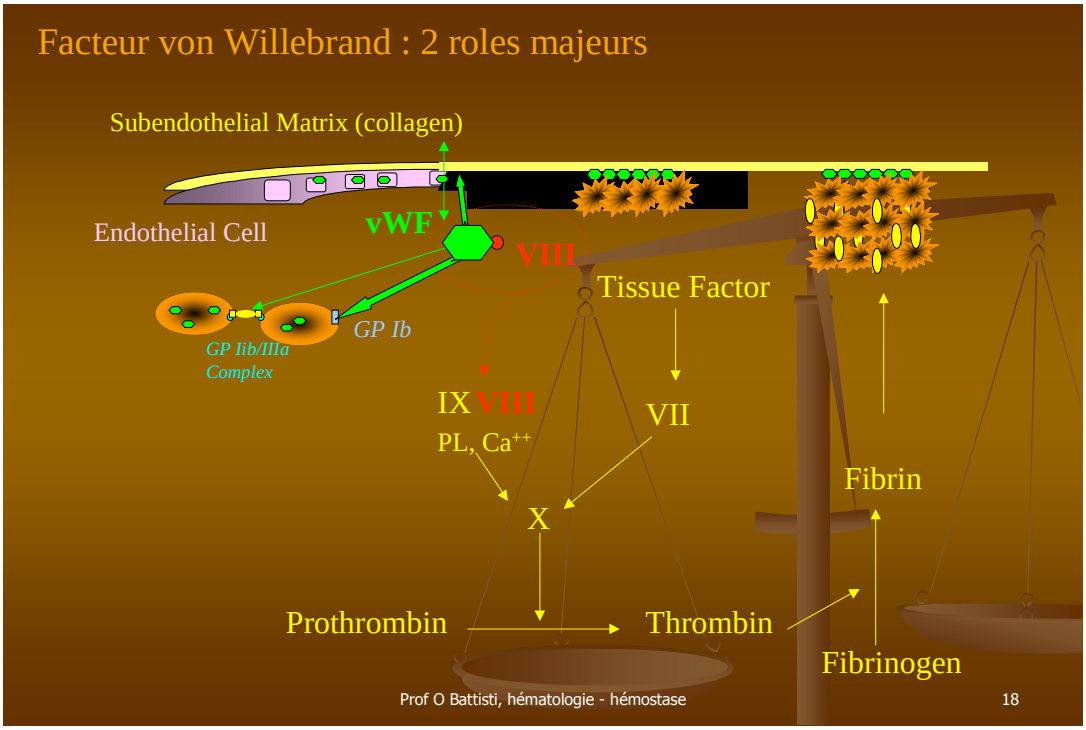


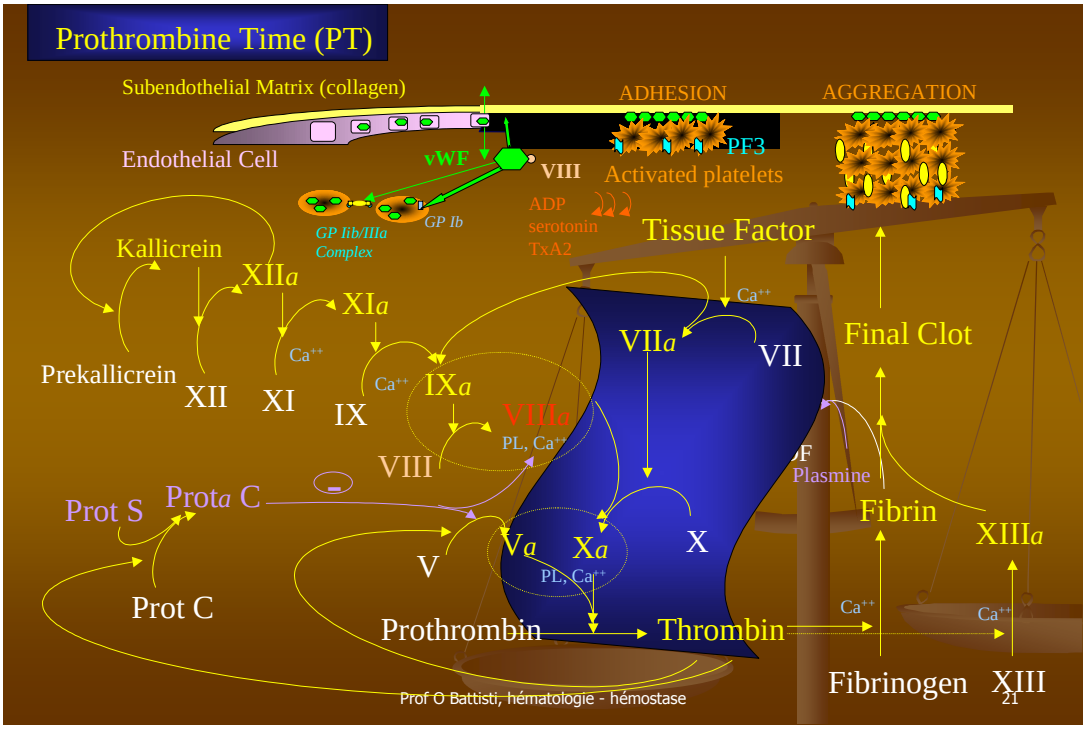

Diagnostic étiologique d'une anémie
microcytaire : VGM = ↘ (70 à 80 fl)
(hypochrome)
macrocytaire : VGM = ↗ (> 90 fl)
hyposidérémique
. Carences en fer
. Fer non utilisé
normosidérémique
. Thalassémie
. Saturnisme
. An. Sidéroblastiques
. Porphyrie congénitale
. Médicaments
macrocytose
. Réticulocytes
. Maladies hépatiques
. Hypothyroïdie
. Syndrome de Down
. Nouveau-né normal
mégaloblastose
. Déficit en folates
. Déficit en B12
. Acide orotique
normocytaire : VGM = N (80 à 90 fl)
(normochrome)
régénérative
. Hémolyses constitutionnelles acquises
. Hémorragies
arégénérative
. Pancitopénie
- Aplasie
- Leucémie
. Isolée constitutionnelle acquise
(infections, drogues, autres...)

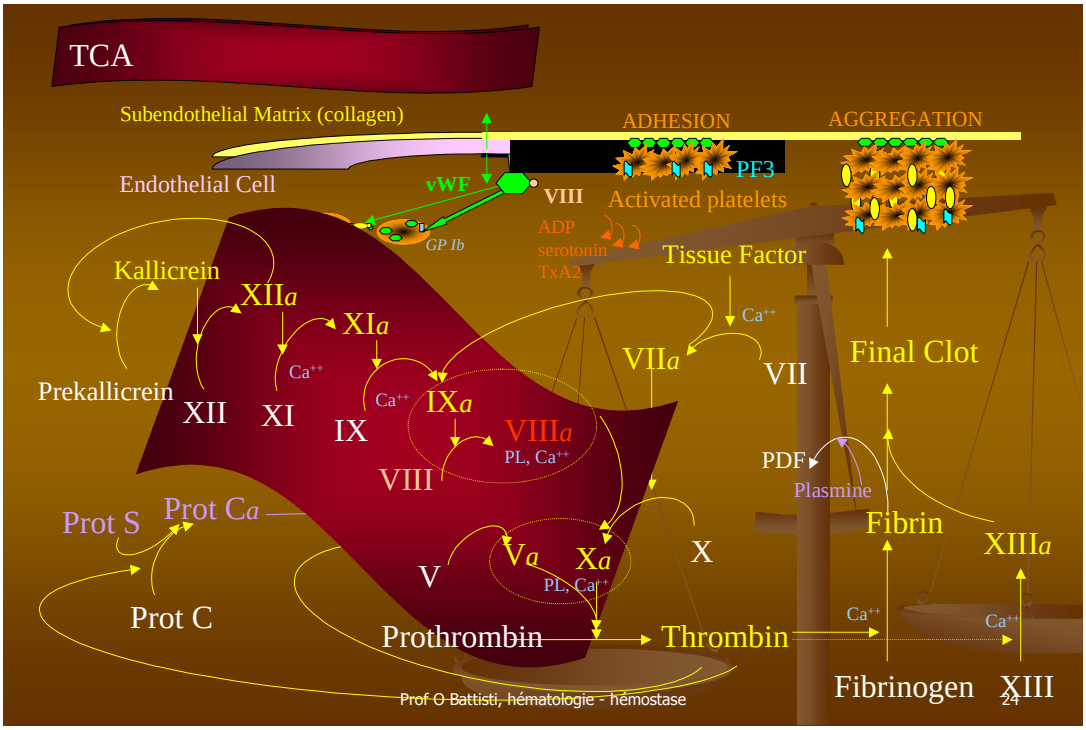

PURPURAS
association à une hyperthermie
si oui : URGENCE MEDICALE
Numération Plaquettaire
NORMALE
THROMBOPENIE
Purpuras Vasculaire
Purpuras Thrombopathiques
Purpuras Thrombopéniques
Infectieux, allergique
Inflammatoire/immunoallegique:
- Thrombopathie Congénitales:
thrombasthénie de Glanzmann
Myélogramme
Normal
Anormal
Thrombopénies
Thrombopénies

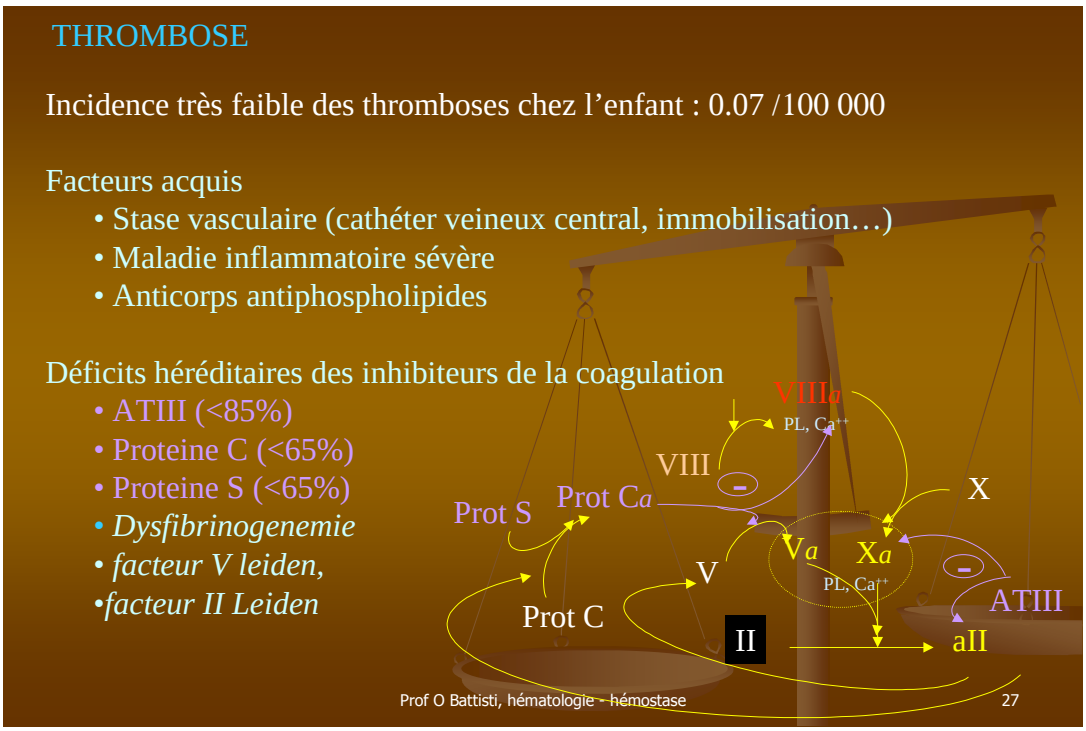

Subendothelial Matrix (collagen)
ADHESION
AGGREGATION
Endothelial Cell
vWF
VIII
PF3
Activated platelets
ADP
serotonin
TxA2
GP Ib
GP IIb/IIIa Complex
Tissue Factor
Kallicrein
XIIa
XIa
Ca++
Prekallicrein
XII
XI
IX
IXa
VIIIa
PL, Ca++
VIII
VIIa
VII
Final Clot
PDF
Plasmine
Prot S
Prot Ca
X
Fibrin
XIIIa
V
Va
Xa
ATIII
Prot C
Prothrombin
Thrombin
Fibrinogen
XIII
Prof O Battisti, hématologie - hémostase
3

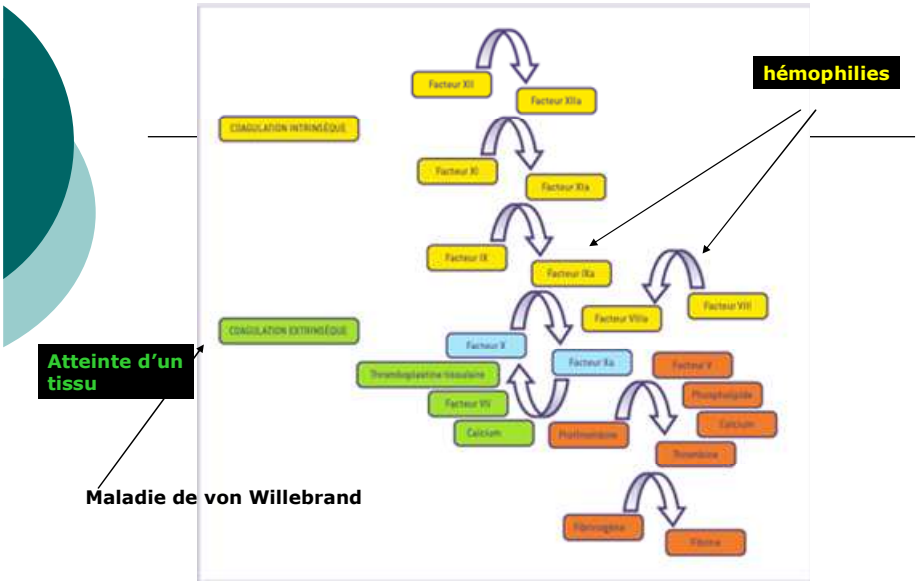

Bleeding time : Primary haemostasis
Subendothelial Matrix (collagen)
Endothelial Cell
VIII
vWillebrand Factor (vWF)
Fibrinogen
Circulating platelet
GP Ib
GP IIb/IIIa Complex
vWF
Activated platelets
ADP
serotonin
TxA2
PLATELET ADHESION
vWF
Platelet factor 3
Coagulation Process
Fibrin
PLATELET AGGREGATION
Prof O Battisti, hématologie - hémostase
14


Facteur von Willebrand : 2 roles majeurs
Subendothelial Matrix (collagen)
Endothelial Cell
vWF
VIII
Tissue Factor
GP Ib
GP IIb/IIIa Complex
IX VIII
PL, Ca++
VII
Fibrin
X
Prothrombin
Thrombin
Fibrinogen
Prof O Battisti, hématologie - hémostase
18


Prothrombine Time (PT)
Subendothelial Matrix (collagen)
ADHESION
AGGREGATION
Endothelial Cell
vWF
VIII
PF3
Activated platelets
ADP
serotonin
TxA2
GP Ib
GP IIb/IIIa Complex
Tissue Factor
Kallicrein
XIIa
XIa
Ca++
VIIa
VII
Final Clot
Prekallicrein
XII
XI
IX
IXa
VIIIa
PL, Ca++
VIII
Plasmine
Fibrin
XIIIa
Prot S
Prota C
Prot C
V
Va
Xa
X
PL, Ca++
Prothrombin
Thrombin
Fibrinogen
XIII
Prof O Battisti, hématologie - hémostase
21


TCA
Subendothelial Matrix (collagen)
ADHESION
AGGREGATION
Endothelial Cell
vWF
VIII
PF3
Activated platelets
GP Ib
ADP
serotonin
TxA2
Tissue Factor
Kallicrein
XIIa
XIa
Ca++
Prekallicrein
XII
XI
IX
IXa
VIIIa
PL, Ca++
VIII
VIIa
VII
Final Clot
PDF
Plasmine
Prot S
Prot Ca
Prot C
V
Va
Xa
X
Fibrin
XIIIa
Prothrombin
Thrombin
Fibrinogen
XIII
Prof O Battisti, hématologie - hémostase
24


THROMBOSE
Incidence très faible des thromboses chez l'enfant : 0.07 /100 000
Facteurs acquis
• Stase vasculaire (cathéter veineux central, immobilisation…)
• Maladie inflammatoire sévère
• Anticorps antiphospholipides
Déficits héréditaires des inhibiteurs de la coagulation
• ATIII (<85%)
• Proteine C (<65%)
• Proteine S (<65%)
• Dysfibrinogenemie
• facteur V leiden,
•facteur II Leiden
VIIIa
PL, Ca++
VIII
Prot S
Prot Ca
X
Va
Xa
V
ATIII
Prot C
II
aII
Prof O Battisti, hématologie - hémostase
27



Prof O Battisti, hématologie non oncologique 25

Facteur IX → BENEFIX
Facteur VIII → RECOMBINATE

Hémorragies		
Degré d'hémorragie	Activité	Fréquence des perfusions
Saignements modérés dans les articulations, les muscles ou sous la peau	20 – 40 UI / kg	/ 12 à 24h pdt 1 à 3 jours
Hémarthroses Hémorragies musculaires Hématomes plus importants	30 – 60 UI / kg	/ 12 à 24h pdt 3 jours
Saignements menaçant la vie	60 - 100 UI / kg	/ 8 à 24h → disparition de l'hémo.
Interventions chirurgicales		
Mineures (y compris les extractions dentaires)	60 – 80 UI / kg	1 seule injection pré -op.
Majeure	80 – 100 UI / kg	Pré – post op. / 8 à 24h en fonction évolution

HEMOPHILIES en abrége

Hémophilie A : déficit fact. VIII XR
Hémophilie B : déficit fact. IX XR

Sévère si < 1 %. Modéré : 2-5 %. Frustre : 6-40 %
Risque de saignement spontané si < 2 %
Si saignement : faire remonter à > 30 %

½ vie	Effet	Tt préventif
Fact. VIII :	8 - 12 h.	0,5 U/kg ⇒ 1 % fact. 20 U/kg/2-3 j
Fact. IX :	12 - 24 h.	20 U/kg/3-4 j

von WILLEBRAND

AD
1-2 % de la population

PFA – TCK – Fact. VIII coagulant – Fact. Von Willebrand Ag – Cofacteur Ristocétine
vW > si infection, post-op, stress, grossesse, ...
vW < si groupe sanguin O

Type I :	AD – 70 % Quantitatif
Type II :	AD ou AR Qualitatif II A – B – C - D –E – F – G – H – N
Type III :	AR vW = 0 ⇒ fact. VIII <<

MINIRIN® = DDAVP = ADH = Vasopressine

I.nasal 100 µg/ml ou 10 µg/spray
IV/IM/SC 4 µg/ml

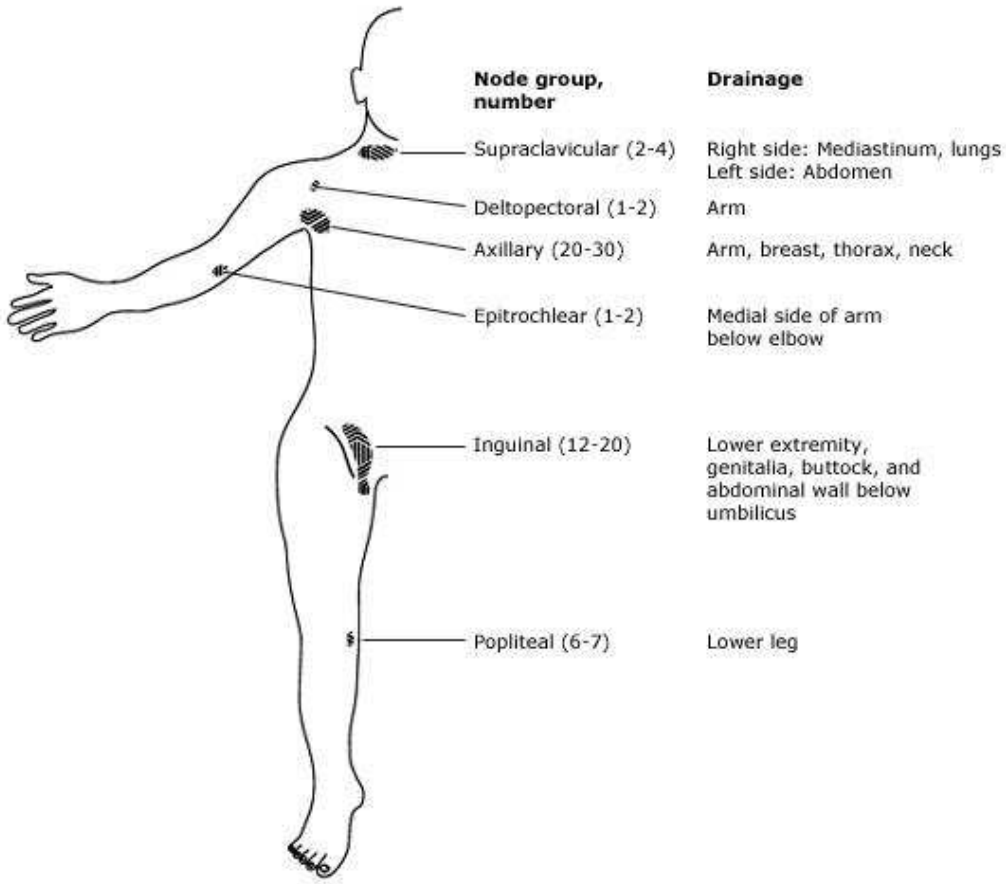
Prof O Battisti, hématologie non oncologique 23

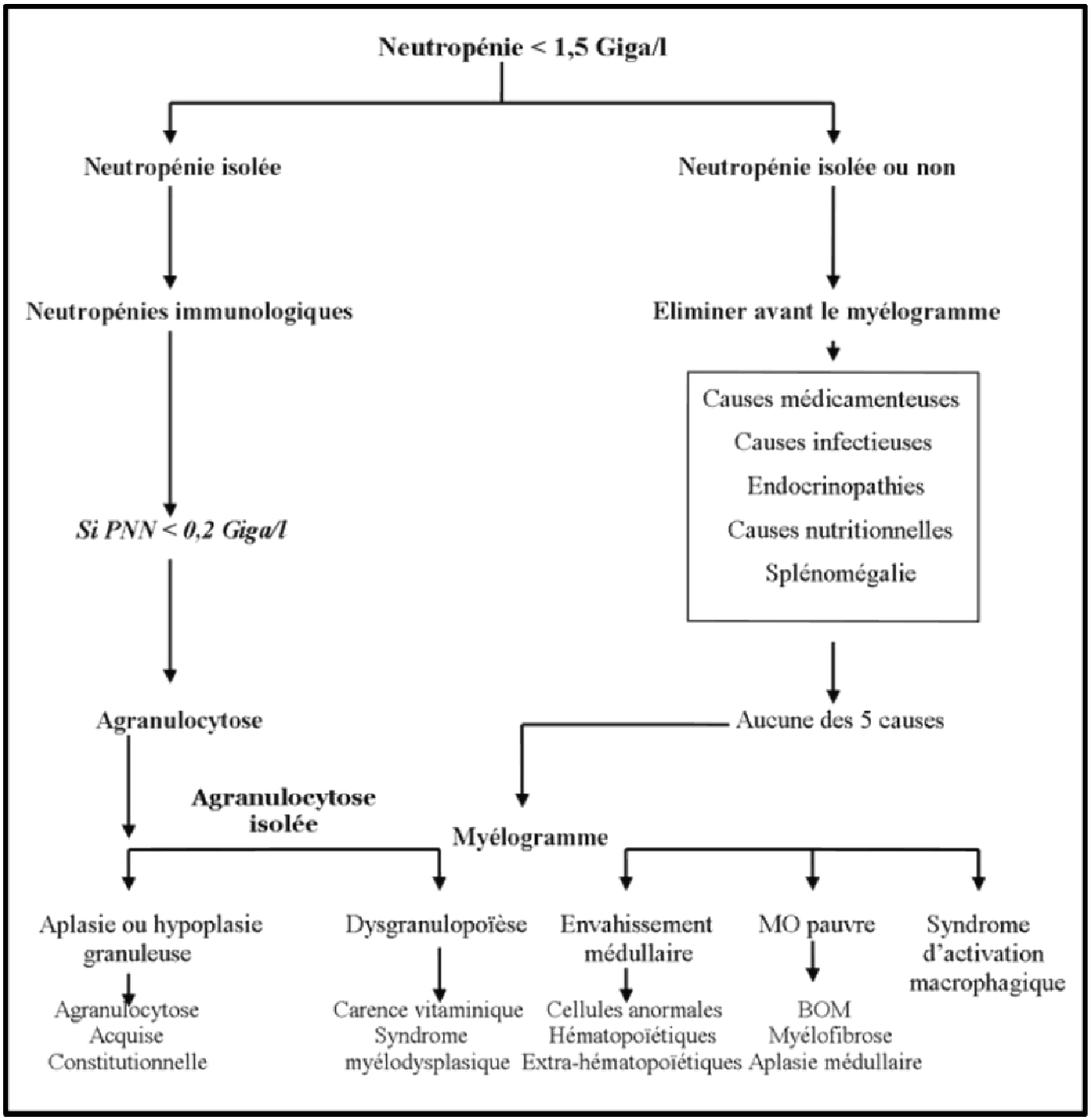
Facteur von Willebrand SD® (DCF) :

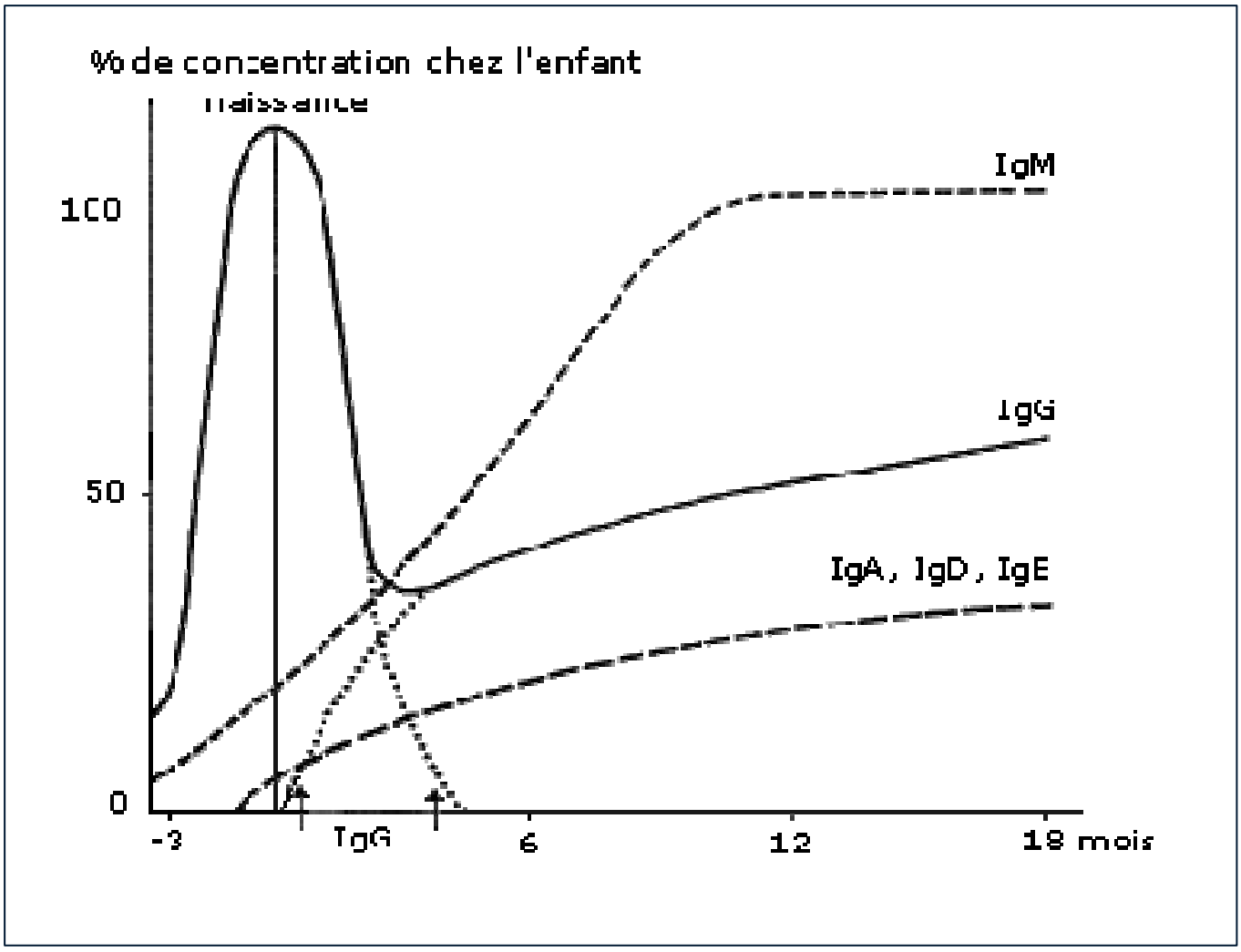
Lyophilisé + 40 ml AD
40 UI vW et 20 UI fact. VIII / 1 ml ⇒
800 UI vW et 400 UI fact. VIII / flacon

20 – 50 U/kg/8-12 h selon la gravité du d

Prof O Battisti, hématologie non oncologique 24


Node group, number
Drainage
Supraclavicular (2-4)
Right side: Mediastinum, lungs
Left side: Abdomen
Deltopectoral (1-2)
Arm
Axillary (20-30)
Arm, breast, thorax, neck
Epitrochlear (1-2)
Medial side of arm below elbow
Inguinal (12-20)
Lower extremity, genitalia, buttock, and abdominal wall below umbilicus
Popliteal (6-7)
Lower leg


Neutropénie < 1,5 Giga/l
Neutropénie isolée
Neutropénie isolée ou non
Neutropénies immunologiques
Eliminer avant le myélogramme
Causes médicamenteuses
Causes infectieuses
Endocrinopathies
Causes nutritionnelles
Splénomégalie
Si PNN < 0,2 Giga/l
Agranulocytose
Aucune des 5 causes
Agranulocytose isolée
Myélogramme
Aplasie ou hypoplasie granuleuse
Dysgranulopoïèse
Envahissement médullaire
MO pauvre
Syndrome d'activation macrophagique
Agranulocytose Acquise Constitutionnelle
Carence vitaminique Syndrome myélodysplasique
Cellules anormales Hématopoïétiques Extra-hématopoïétiques
BOM Myélofibrose Aplasie médullaire


% de concentration chez l'enfant
Naissance
100
50
0
IgM
IgG
IgA, IgD, IgE
-3
IgG
6
12
18 mois

Approach to chest pain in children

Authors
Robert L Geggel, MD
Erin E Endom, MD

Section Editors
David R Fulton, MD
Gregory Redding, MD
Jan E Drutz, MD
Gary R Fleisher, MD

Deputy Editor
James F Wiley, II, MD, MPH

Last literature review version 17.2: May 2009

INTRODUCTION — Chest pain is a common presenting complaint in children. Although the etiology is benign in most cases, this symptom may lead to school absences and restriction of activities and causes considerable anxiety in patients and their families. A thorough history and physical examination usually can determine the cause and identify patients who require acute intervention and those who can be managed with reassurance and continued follow-up. Laboratory testing is necessary only in a small number of patients [1] . In the absence of associated symptoms of illness, positive findings on physical examination related to the cardiac or respiratory systems, or symptoms during exertion, a serious organic cause is unlikely.

The emergent evaluation of respiratory distress in children and adolescents, with or without chest pain, is discussed separately. (See "Emergent evaluation of acute respiratory distress in children").

EPIDEMIOLOGY — Chest pain is a common symptom in children and adolescents [2,3] . In an urban black adolescent population, for example, this symptom was the seventh most common reason for seeing a physician [4] . Children younger than 12 years old also are affected, accounting for approximately one-half of patients with this symptom who presented to an emergency department [1] . There is no gender predilection. An equal number of males and females present with chest pain.

PRESENTATION — Chest pain can be acute or chronic. Estimates of the relative proportions depend upon the setting in which patients are evaluated. In a report of 407 children with chest pain seen in an emergency department, symptoms were acute (less than 48 hours duration) in 43 percent and chronic (more than six months duration) in 7 percent [2] . In contrast, in a study of 100 adolescents with chest pain seen in a general pediatric clinic, pain was present longer than six months in 36 percent [5] . Some patients with idiopathic chest pain report symptoms for longer than one year [1,5-8] .

Chest pain has important functional consequences because it may result in restriction of activities and school absences. In the review of 100 adolescents noted above, restriction of activities was reported in 69 percent and 41 percent had absences from school because of pain [5] .

Chest pain causes considerable anxiety in patients and their families. Because of its association with fatal heart disease in adults, this symptom often is viewed as a harbinger of serious cardiac disease (show table 1) [5,6,9] . This interpretation is more common after the occurrence of sudden death involving an athlete in the community or at the professional level.

Adolescents with chest pain often are worried about their symptoms. In the series of adolescent patients seen in a general pediatric clinic, nearly all of whom had no serious illness, 44 percent thought they were having a heart attack or were worried about heart disease (12 percent) or cancer (12 percent) [5] .

CAUSES — Causes of chest pain in children vary among reports and depend in part upon whether patients were seen in an emergency department with acute symptoms or in a pediatric or cardiology setting with a more chronic complaint (show table 2) [1,2,5,6,8-14] . In both settings, patients typically have no serious underlying organic medical condition [6-8,15] .

In contrast to perceptions of the patient and family, cardiac disease is uncommon (1 to 6 percent). A diagnosis cannot be established in a substantial proportion of cases (21 to 45 percent); these are considered idiopathic. Similar to most patients with chest pain, those with idiopathic pain typically have no serious underlying medical condition.

Musculoskeletal conditions — Conditions affecting the musculoskeletal system are identified as the cause of chest pain in 15 to 31 percent of cases [2,5] . These comprise the largest category of known etiologies.

Musculoskeletal pain can be traumatic or nontraumatic, although the latter is more common. Trauma may result in a rib fracture, bruise, or, rarely, hemopericardium. Nontraumatic conditions include costochondritis, myalgia, and slipping rib syndrome, and are known collectively as the chest wall syndrome [16] .

Costochondritis — Costochondritis is a frequent cause of chest pain. In one report, tenderness of the costal cartilages was the only abnormal finding in 79 of 100 adolescents who complained of chest and upper abdominal pain [3] . Costochondritis typically was

unilateral, occurring more frequently on the left side. The left fourth sternocostal cartilage was affected most often. One type of injury producing strain of chest wall muscles and ligaments is lifting a heavy school bag and carrying it over one shoulder.

Slipping rib syndrome — Slipping rib syndrome involves the eighth, ninth, and tenth ribs, which are not attached by costal cartilage to the sternum but are attached to each other by fibrous tissue [17-20] . If these fibrous connections are weakened or ruptured by trauma, the ribs can slip and impinge on the intercostal nerve, producing pain.

Precordial catch — An uncommon and benign etiology of musculoskeletal chest pain in children is precordial catch, also known as Texidor's twinge [10,13,14,21,22] . This condition consists of brief episodes (seconds to a few minutes) of sharp pain that can be localized with the fingertip to one interspace at the left sternal border or cardiac apex. The pain has a sudden onset, typically at rest or during mild activity, and increases with inspiration. The cause is unknown, although it has been associated with poor posture.

Psychogenic causes — Chest pain may have a psychogenic etiology in as many as 30 percent of cases [5,9,11] . This cause is more common in children older than 12 years of age [1,2] . The pain may reflect anxiety or a conversion disorder triggered by stressful events [11,23,24] . In one series, approximately one-third of children with chest pain seen in a general pediatric clinic had a history of stressful events, including recent death, illness or accident in the family, family separations, or school changes [5] . Most children with psychogenic chest pain have other recurrent somatic complaints, including headache or abdominal or extremity pain [25,26] . Approximately one-third have significant sleep disturbances [25] .

Hyperventilation can result in chest pain that frequently is accompanied by lightheadedness or paresthesias. The mechanism is uncertain. Possibilities include spasm of the diaphragm resulting from rapid, repetitive use, gastric distension arising from aerophagia, or coronary artery vasoconstriction caused by hypocapneic alkalosis (the latter tested in adults with ischemic coronary disease) [27] .

Breast causes — The breast can be a source of chest pain in adolescent patients, although it typically accounts for less than 5 percent of complaints. Pain may be a presenting symptom in males with gynecomastia. These patients also may have anxiety about the size of their breasts. Painful conditions of the breast in females include mastitis, fibrocystic disease, thelarche, or tenderness associated with pregnancy. Worries about cancer are often present in these patients [5] .

Respiratory disorders — Respiratory disorders, including pneumonia, bronchitis, and reactive airway disease, are common causes of acute chest pain. Exercise-induced bronchoconstriction appears to be a frequent cause of chest discomfort even in patients without audible wheezing. This was illustrated by a study of pulmonary function testing before and after exercise in 88 otherwise healthy children and adolescents with chest pain [28] . Exercise decreased forced expiratory volume in one second or peak expiratory flow rate in 72.7 percent of the children. Subjective or objective improvement occurred in 97 and 70 percent, respectively, of 36 children given inhaled albuterol. In this condition, air hunger typically precedes the chest pain.

Less common causes are pleuritis, pleural effusion, pneumothorax, and pneumomediastinum. Air leak (pneumothorax or pneumomediastinum) often is a complication of trauma or an underlying disorder, such as reactive airway disease [29-31] , cystic fibrosis [32] , or Marfan syndrome [33] , but can be idiopathic [14] . Other respiratory causes of chronic pain in cystic fibrosis are pleuritis and rib fracture [34] . (See "Cystic fibrosis: Clinical manifestations and diagnosis").

Gastrointestinal disorders — Gastrointestinal disorders that cause chest pain can affect the esophagus (the most common site), stomach, bowel, biliary tract, and pancreas. In one series, among 21 children with a gastrointestinal cause of chest pain, 16 had esophagitis (typically caused by gastroesophageal reflux), four had gastritis, and one had diffuse esophageal spasm [35] . (See "Clinical manifestations and diagnosis of gastroesophageal reflux disease in children and adolescents").

Esophageal endoscopy and manometry may detect abnormalities in children with chest pain, even without other gastrointestinal symptoms. This was demonstrated in a study of 83 children with chest pain who underwent these procedures [36] . Esophageal histology and motility were normal in 47 patients (57 percent). Among the others, 15 (18 percent) had esophagitis with normal motility, 13 (16 percent) had normal histology and dysmotility, and 8 (9 percent) had both esophagitis and dysmotility. The most common motility disorders were diffuse esophageal spasm and achalasia, which occurred in seven and four patients, respectively.

In addition to those mentioned above, esophageal disorders causing chest pain include strictures, foreign body, and caustic ingestions. Conditions affecting the stomach and bowel include ulcer and irritable bowel. Cholecystitis may cause symptoms that suggest angina in adults, but is uncommon in children. Biliary (eg, gallstones) and pancreatic disorders are rare causes [35,37] .

Pulmonary vascular disease — Causes of chest pain related to the pulmonary vascular system include pulmonary embolism and pulmonary hypertension. Children with sickle cell disease may develop acute chest syndrome. (See "Pulmonary complications of sickle cell disease", section on Acute chest syndrome).

Pulmonary embolism — Pulmonary embolism is unusual in children but can present with chest pain. In a retrospective study in which 18 affected patients were identified over a 15-year period, the incidence of pulmonary embolism was estimated as 78 per 100,000 hospitalized adolescents [38] . In this series, major risk factors were oral contraceptive use and pregnancy termination (75 percent of females) and trauma (67 percent of males). Other predisposing causes include immobility, ventriculoatrial shunts for hydrocephalus,

central venous catheters, solid tumors, heart disease, infection, dehydration, hypercoagulable states, low cardiac output, or obesity [39,40] .

Pulmonary hypertension — Pulmonary hypertension can be secondary to lung disease, congenital heart disease, or other systemic disorders, or have no identified cause (idiopathic pulmonary arterial hypertension) (show table 3). It may cause chest pain and other symptoms including fatigue, lethargy, and dyspnea or syncope with exertion. The mechanism of chest pain is uncertain.

Acute chest syndrome — Acute chest syndrome is a serious and potentially fatal cause of chest pain in patients with sickle cell disease. It occurs in almost one-half of patients with the disorder. In addition to chest pain, acute chest syndrome is characterized by the presence of a new pulmonary infiltrate involving at least one complete lung segment (not atelectasis), temperature >38.5ºC, and tachypnea, wheezing, or cough [40,41] . (See "Pulmonary complications of sickle cell disease").

Cardiac conditions — Cardiac conditions are a rare but potentially serious cause of chest pain in children. Cardiac disease is more likely if chest pain occurs during exertion and is recurrent. Most conditions will be associated with an abnormal cardiac examination or coexisting symptoms. In patients with known heart disease, chest pain may indicate progression of the underlying condition. Conditions that may present with chest pain include:

- Severe left ventricular outflow tract obstruction caused by aortic stenosis (subvalvar, valvar, or supravalvar), obstructive cardiomyopathy, or coarctation of the aorta [42,43] .

- Aortic root dissection associated with Marfan syndrome, Turner syndrome, type IV Ehlers-Danlos syndrome, chronic systemic hypertension, homocystinuria, rare familial aortopathies [44] , or cystic medial necrosis [40] . (See appropriate topics).

- Pericarditis, which often is idiopathic, or may be caused by an infectious agent or associated with an underlying condition such as collagen vascular disorder, uremia, neoplasm, or trauma. Pericarditis can occur as part of the postpericardiotomy syndrome after cardiac surgery. (See "Pericardial and postpericardial injury syndromes").

- Myocarditis, in which chest pain typically occurs with concomitant pericarditis (see "Clinical manifestations and diagnosis of myocarditis in children")

- Coronary artery abnormalities, including congenital disorders or acquired conditions (eg, coronary artery aneurysm or stenosis caused by Kawasaki disease) (show table 4) (show figure 1). Anomalous origin of the left coronary artery from the main pulmonary artery usually presents in infancy, but can become symptomatic later in childhood. In that disorder, left ventricular ischemia usually results in cardiomyopathy and mitral regurgitation.

- Ruptured sinus of Valsalva aneurysm, a rare condition caused by congenital absence of media in the aortic wall behind the sinus of Valsalva. The aneurysm typically ruptures into the right ventricle or right atrium but can affect the other chambers or pulmonary artery [45] .

- Tachyarrhythmias (eg, supraventricular tachycardia with or without underlying Wolff-Parkinson-White syndrome, ventricular tachycardia) or palpitations (caused by ventricular premature beats).

- Coronary vasospasm (variant angina) and myocardial infarction are rare causes of chest pain in children and adolescents. Coronary vasospasm is associated with transient ischemic changes on ECG, normal coronary arteriography, reversible septal akinesia by echocardiography, and cardiac enzyme elevation [46-48] . (See "Variant angina").

In the United States, the incidence of acute myocardial infarction in adolescents is estimated as 6.6 events per 1 million patient-years resulting in 157 events per year [49] . Myocardial infarctions occur more frequently in males and are associated with substance abuse and smoking. (See "Toxic exposure" below).

Mitral valve prolapse — Whether mitral valve prolapse is associated with chest pain is controversial. In one study of 119 children, 18 percent had atypical chest pain [50] . However, in the Framingham Heart Study, the incidence of chest pain was not increased in 83 adults with mitral valve prolapse, compared to 3403 without the abnormality [51] . Similarly, in a report of 813 children aged 9 to 14 years, 31 of whom had mitral valve prolapse, the incidence of chest pain was similar in affected and unaffected patients [52] .

Toxic exposure — Exposure to vasoconstrictive agents, such as cocaine, can cause chest pain that is likely of ischemic origin [53,54] . Systemic hypertension, myocardial infarction, ventricular arrhythmias, myocarditis, and sudden death may occur [55] . Chest pain also has been associated with the use of marijuana [56] , methamphetamines, and sympathomimetic decongestants [10] .

Cigarette smoking has been associated with chest pain in adults. In one study, chest pain reported by questionnaire in a sample of 70,208 adults was greater in smokers than nonsmokers (average excess 1.6-fold in men, 1.3-fold in women) [57] .

Neurologic disorders — Chest pain rarely is caused by a neurologic disorder. Herpetic neuralgia in a dermatomal distribution on the chest can cause pain, which may be manifest before lesions appear. (See "Postherpetic neuralgia"). Spinal cord compression, which may be caused by tumor or vertebral collapse, or epidural abscess may cause radicular pain.

Idiopathic — A substantial proportion (21 to 45 percent) of cases of chest pain have no obvious cause after a thorough evaluation and are considered idiopathic [2,5-8] . Although repetitive episodes of pain may occur, symptoms typically resolve over time. In one report of 31 children with idiopathic chest pain followed for an average of 4.1 years, persistent symptoms were reported by 45 percent [7] . However, pain resolved in 81 percent of patients followed for more than three years.

HISTORY — The history should include a description of the pain and any associated symptoms. The patient and/or parents should be asked about precipitating events, such as trauma, muscle strain, and foreign body ingestion, or stressful factors, such as a recent death in the family.

Underlying medical conditions that may be associated with chest pain should be noted. These include asthma, cardiac disease, Kawasaki disease, and sickle cell disease.

Family history should be obtained for genetic conditions with features that may lead to chest pain. Disorders including Marfan syndrome, Turner syndrome, and type IV Ehlers-Danlos syndrome predispose to aortic root dissection. Hypertrophic cardiomyopathy is a rare genetic disorder with familial and sporadic forms that may present with exertional chest pain. (See "Clinical manifestations of hypertrophic cardiomyopathy").

History should be sought about possible substance abuse, including cocaine ingestion and tobacco use. Use of other vasoactive drugs should be documented.

Description of chest pain — Chest pain should be characterized by a thorough description. This includes the time course and duration, quality, location, radiation, severity, precipitating factors, and associated symptoms.

Temporal elements — The duration and time course of the onset of chest pain may be a useful distinguishing feature. Chronic pain is unlikely to have a serious underlying cause and often has a musculoskeletal or psychogenic origin or is idiopathic.

Acute pain is more likely to be caused by a medical condition. As examples, pain caused by pulmonary conditions (eg, reactive airway disease or pneumothorax) or vascular events (eg, aortic dissection or acute pulmonary embolism) typically has an abrupt onset. Ischemic myocardial pain, which is unusual in children, may have a gradual onset with increasing intensity over time.

Quality — A description of the quality of the pain may be helpful. The pain associated with costochondritis typically is described as midsternal in location and sharp in quality with minimal radiation. It occurs mainly at rest and lasts for seconds to a few minutes. The pain may increase in intensity with deep inspiration because of stretching of the costochondral junctions or muscle fibers.

Patients with slipping rib syndrome may describe "something slipping or giving away," "a popping sensation," or "hearing a clicking sound" [17] . In that condition, pain occurs with bending over or deep breathing [21] .

Ischemic cardiac pain may be described as squeezing, tightness, pressure, constriction, burning, or fullness in the chest. Pain caused by aortic root dissection is extremely severe. It is described as tearing in quality, and typically radiates to the back.

Chest caused by to pericarditis is sharp in quality, usually retrosternal in location, often with radiation to the left shoulder. It is more severe in the supine position or with deep inspiration (because of pericardial distention). Pain may increase with swallowing if the pericardium near the esophagus is involved.

Location — Pain that localizes to a small area on the chest more likely is of chest wall or pleural origin rather than visceral. Ischemic pain is a diffuse discomfort that may be difficult to localize.

Radiation usually is associated with causes of pain that are uncommon in children. As an example, the pain of myocardial ischemia may radiate to the neck, throat, lower jaw, teeth, upper extremity, or shoulder. Other unusual causes include acute cholecystitis, which can present with right shoulder pain (although concomitant right upper quadrant or epigastric pain is more typical), aortic dissection, which may be associated with chest pain that radiates between the scapulae, or pericarditis that can radiate to the left shoulder.

Precipitating factors — The patient should be asked about factors that induce or make the pain worse:

- Body position or movement, as well as deep breathing, may exacerbate chest pain of musculoskeletal origin.

- Pain made worse by swallowing likely is of esophageal origin. Discomfort that occurs with eating also may suggest upper gastrointestinal disease.

- Chest discomfort provoked by exertion often has a cardiac or respiratory cause.

- Pleuritic chest pain is worsened by breathing and may be exacerbated when lying down.

- Pain associated with coronary artery anomalies typically occurs with exertion due to limitations in myocardial oxygen delivery.

Associated symptoms — Associated symptoms may help determine etiology.

- Fever, especially when associated with tachypnea or cough, may suggest a respiratory infection. Fever also can be present in patients with pericarditis, myocarditis, or Kawasaki disease (see appropriate topic reviews)

- Dyspnea may indicate pulmonary disorders, including pathology of the airways, lung parenchyma, or pulmonary vasculature, or be a sign of cardiac disease. In addition to chest pain, for example, patients with pulmonary embolism can develop dyspnea, hypoxemia, apprehension, cough, and diaphoresis [41] . Patients with myocarditis may have dyspnea and fatigue. (See "Clinical manifestations and diagnosis of myocarditis in children").

- Vomiting or regurgitation, painful swallowing, or heartburn associated with feeding suggest gastrointestinal disease, such as gastroesophageal reflux and esophagitis.

- Recurrent somatic complaints, including headache or abdominal or extremity pain, occur in most children with psychogenic chest pain [25,26] . Approximately one-third have significant sleep disturbances [25] .

- Lightheadedness or paresthesias frequently accompany chest pain resulting from hyperventilation.

- Syncope or palpitations suggest an underlying cardiac disorder. (See "Emergent evaluation of syncope in children and adolescents").

EXAMINATION — A thorough physical examination should be performed to detect findings associated with organic disease. Physical signs may indicate genetic conditions that include cardiac abnormalities. Fever suggests an infectious etiology.

The remainder of the examination should focus on the chest. Patients with acute onset of chest pain and who are in distress require immediate management. This should be initiated while the evaluation proceeds.

Chest wall — Musculoskeletal disorders, including costochondritis, are a major cause of chest pain in children and result in chest wall tenderness. Bruises associated with trauma sometimes are seen.

Examination should include palpation of the costochondral junctions, the insertion site of the pectoralis major muscle group (by grasping the head of the muscle between the examiner's fingers and thumb), the inframammary area, and other regions of the chest where pain is reported. Involvement of the costochondral junctions often is asymmetric, with the left side more frequently affected [3] . The diagnosis is confirmed when palpation reproduces the discomfort.

Diagnosis of slipping rib syndrome can be confirmed by the "hooking" maneuver in which the examiner's fingers are placed around the lower costal margin. Lifting anteriorly will elicit a click and reproduce pain [20] .

Tenderness on palpation of breast tissue often can be elicited in patients who complain of breast pain. Tender subareolar masses may be palpable.

Respiratory signs — Signs of a respiratory etiology of chest pain may include tachypnea and respiratory distress. Diminished breath sounds over affected areas of the lung or rales may indicate pneumonia, whereas wheezing may be audible in patients with asthma. Pneumomediastinum may produce subcutaneous emphysema that is detected as crepitus on palpation of the supraclavicular or neck region [30] .

Hyperventilation, associated with chest pain of psychogenic origin, may be present at the time of the examination. If not, it may be possible to reproduce the chest pain by asking the patient to hyperventilate. In general, hyperventilation must continue for at least 20 minutes to cause symptoms [5] .

The emergent evaluation of children and adolescents with respiratory distress is discussed separately. (See "Emergent evaluation of acute respiratory distress in children").

Cardiac signs — A cardiac cause of chest pain may be suggested by auscultation of abnormal heart sounds or a cardiac murmur or abnormal pulse or blood pressure.

Left ventricular outflow obstruction — Signs of left ventricular outflow obstruction include a systolic ejection murmur at the right upper sternal border and occasionally along the left sternal border. Coarctation of the aorta is associated with elevated blood pressure in the arms and a lower blood pressure in the legs. If the coarctation is long-standing (present for more than five to seven years), collateral vessels may form that connect the upper and lower portions of the aorta; these vessels create a continuous murmur over the lateral aspect of the ribs. (See "Clinical manifestations and diagnosis of coarctation of the aorta").

Pericarditis — In patients with pericarditis, pain increases when manual pressure is applied to the sternal region. The pain typically improves with sitting up and leaning forward. Signs of pericarditis depend upon the size of the pericardial effusion. Patients with a small effusion typically have an audible pericardial friction rub, caused by rubbing together of the inflamed parietal and visceral pericardial surfaces. The rub often is continuous in systole and diastole. It is easier to hear with the diaphragm of the stethoscope when the patient is sitting and leaning forward.

A rub will not be heard if the effusion is large because the two pericardial surfaces of the pericardium are not in contact with each other. A large effusion may result in cardiac tamponade, manifested by a narrow pulse pressure, elevated pulsus paradoxus (>10 mmHg), elevated jugular venous pressure, distant heart sounds, hepatomegaly, ascites, and peripheral edema.

Signs of myocarditis include tachycardia, muffled heart sounds, a gallop rhythm, and a murmur of mitral regurgitation, usually accompanied by fever. Patients often complain of dyspnea and fatigue.
(See "Clinical manifestations and diagnosis of myocarditis in children").

Pulmonary hypertension — Patients with pulmonary hypertension typically have a right ventricular heave and a single loud S2. They may have a murmur consistent with tricuspid or pulmonary regurgitation.

Mitral valve prolapse — Signs of mitral valve prolapse are a constant, mid-systolic apical click and occasionally an apical systolic murmur of mitral regurgitation. Both auscultatory findings are more prominent when the patient is in the standing rather than supine position.

Arrhythmia — Tachycardia may suggest supraventricular tachycardia. Cocaine ingestion may present with tachycardia, hypertension, and anxiety.

DIAGNOSTIC STUDIES — Most patients with chest pain have a normal physical examination or findings consistent with a musculoskeletal etiology. Further investigations are not needed in those cases.

Diagnostic studies may help establish a diagnosis in patients with abnormal physical findings or with associated symptoms that suggest organic disease. Although cardiac causes of chest pain are uncommon in children, patients with dyspnea, palpitations, anginal pain, pain with exertion that cannot be attributed to respiratory disease, or syncope should be referred to a pediatric cardiologist for further evaluation (show figure 2 and show figure 3).

Chest radiograph — A chest radiograph should be obtained in children in whom a cardiac or pulmonary disorder is suspected. Signs of cardiac enlargement may be apparent in conditions causing left ventricular outflow obstruction, heart failure, myocarditis, pericarditis, or pericardial effusion.

Patients with pulmonary hypertension may have prominent main and central pulmonary arteries. The peripheral lung fields may be dark if pulmonary vascular resistance is chronically elevated.

In patients with suspected pulmonary disorders, a chest radiograph may show infiltrates caused by pneumonia or areas of atelectasis and air trapping caused by aspiration of a foreign body. Hyperinflation typically is seen in patients with asthma. In addition, pneumothorax, pneumomediastinum, or pleural effusions can be detected.

Electrocardiogram — An ECG should be obtained if cardiac disease is suspected. In patients with a rapid pulse rate or palpitations, an ECG can identify the type of arrhythmia. If the arrhythmia is intermittent, a Holter monitor or event monitor recording may be needed.

Other ECG abnormalities can help with diagnosis and indicate the need for further testing.

- Patients with left ventricular outflow obstruction may have evidence of left ventricular hypertrophy or strain. Formal exercise tolerance testing may be needed to assess the development of arrhythmia or ischemia during exertion.

- In pericarditis with effusion, the ECG changes during the clinical course. Generalized ST segment elevation involving limb and precordial leads is seen in the initial 10 to 14 days (show ECG 1). This is followed by T wave flattening or inversion that can

persist for an additional two weeks. Voltages will be reduced in amplitude if the effusion is large. Abnormal Q waves typically are not seen in pericarditis. ST-T wave abnormalities occur in myocarditis. (See "Clinical manifestations and diagnosis of myocarditis in children").

- In children with anomalous origin of the left coronary artery, the typical ECG pattern is of an anterolateral infarction with deep and wide Q waves and T wave inversions in leads I, aVL, V5, and V6 (show figure 4).

- Patients with pulmonary hypertension typically have signs of right ventricular hypertrophy and right axis deviation. The ECG may show right ventricular strain (abnormal T waves in the anterior leads).

- ECG findings in pulmonary embolism usually are nonspecific ST-T segment changes or sinus tachycardia. If acute right ventricular hypertension occurs, the classic pattern of S wave in lead 1, Q wave, and T wave inversion in lead III can be seen [41] .

Echocardiogram — An echocardiogram will establish a diagnosis in cardiac disorders and help assess severity. This technique can be used to

- Establish the diagnosis of pulmonary hypertension and evaluate ventricular function and associated structural abnormalities.

- Determine the severity and site of left ventricular outflow obstruction and the level of ventricular function.

- Assess the size of a pericardial effusion and signs of tamponade, which include variation in Doppler peak velocity across the valves during the cardiac cycle, atrial free wall collapse, or ventricular septal paradoxical motion into the left ventricle during inspiration.

- Identify coronary artery abnormalities, including abnormal origin or course, fistula, or aneurysm or stenosis caused by Kawasaki disease.

- Diagnose aortic root dissection (show ECG 2). Alternatively, the diagnosis can be made with magnetic resonance imaging (show radiograph 1), computed tomography, or aortography (show radiograph 2 and show radiograph 3A-3B). The most efficient imaging technique available at the institution should be used to establish the diagnosis.

- Diagnose ruptured sinus of Valsalva aneurysm.

Gastrointestinal evaluation — Children with chest pain and gastrointestinal symptoms should be referred to a pediatric gastroenterologist. Evaluation of the upper gastrointestinal tract may reveal esophagitis, gastritis, and/or motility disorders such as diffuse esophageal spasm or achalasia. These abnormalities may be detected in some children with chest pain and no gastrointestinal symptoms [36] . Thus, a gastrointestinal evaluation should be considered in children with persistent chest pain and no obvious etiology.

Other tests — Additional testing should be based upon associated signs and symptoms and the results of the initial evaluation. Further evaluation may include pulmonary function testing in patients with respiratory disorders, a ventilation-perfusion scan to confirm the diagnosis of pulmonary embolism, or cardiac catheterization to evaluate coronary arteries or arrhythmia. An exercise tolerance test can be useful in patients with chest pain induced by exercise. Patients suspected of having an infection should have a complete blood count with differential and appropriate cultures. Toxicology screening should be performed if substance abuse is suspected.

MANAGEMENT — Children with acute onset of chest pain and severe distress, abnormal vital signs, or hypoxemia require immediate intervention. Specific disorders (eg, asthma, pneumonia) should be treated appropriately. (see appropriate topics).

Most patients with chronic chest pain require minimal intervention. Musculoskeletal pain typically responds to analgesics and rest, although frequently no medication is necessary.

Referral — Patients with chronic chest pain and associated symptoms should be referred to an appropriate specialist for further evaluation. Children with gastrointestinal symptoms, or those with persistent or recurrent pain with no apparent etiology, should be referred to a gastroenterologist. Those with known cardiac disease or who have pain with exertion, syncope, dizziness, or palpitations should be referred to a cardiologist.

Reassurance — Reassurance is an important component of management. Although the etiology usually is benign, this symptom causes considerable anxiety in patients and their families because of the more serious implications of chest pain in adults. Cases with a psychogenic etiology may need additional counseling.

Follow-up — Chest pain may persist or recur, although the natural history is not well described. One study reported follow-up of 407 patients with chest pain who were seen in an emergency department [8] . Of the initial cohort, 149 (37 percent) were seen for at least six months and 51 (13 percent) for at least two years. Chest pain persisted in 43 percent of the group that was followed. A previously unrecognized organic etiology was identified in 12 patients, only one of whom had a cardiac abnormality (mitral valve prolapse); three had asthma. In another report of children with idiopathic chest pain, symptoms persisted in 19 percent followed for more than three years [7] . Follow-up should be provided until symptoms resolve.

GRAPHICS

Family's or patient's understanding of symptom of chest pain

Cause	Prevalence, percent
Cardiac	52 to 56
Muscoskeletal	13
Respiratory tract	10
Skin infection	3
Breast	3
Cancer	0 to 12
Unsure	10 to 19

Data from Pantell, RH, Goodman, BW Jr, Pediatrics 1983; 71:881and Driscoll, DJ, Glicklich, LB, Gallen, WJ, Pediatrics 1976; 57:648.

Causes of pediatric chest pain

Cause	Prevalence, percent
Idiopathic	21 to 45
Muscoskeletal	15 to 31
Hyperventilation/psychiatric	0 to 30
Breast related	1 to 5
Respiratory	2 to 11
Gastrointestinal	2 to 8
Cardiac	1 to 6
Miscellaneous	9

Data from Selbst, SM, Pediatrics 1985; 75:1068, Selbst, SM, Ruddy, RM, Clark, BJ, et al., Pediatrics 1988; 82:319, Pantell, RH, Goodman, BW Jr, Pediatrics 1983; 71:881, Fyfe, DA, Moodie, DS, Clin Pediatr (Phila) 1984; 23:321, Selbst, SM, Ruddy, R, Clark, BJ, Clin Pediatr (Phila) 1990; 29:374, Driscoll, DJ, Glicklich, LB, Gallen, WJ, Pediatrics 1976; 57:648, Kocis, KC,

Pediatr Clin North Am 1999; 46:189, Tunaoglu, FS, Olgunturk, R, Akcabay, S, Oguz, D, Pediatr Cardiol 1995; 16:69, Anzai, AK, Merkin, TE, Am Fam Physician 1996; 53:1682, Zavaras-Angelidou, KA, Weinhouse, E, Nelson, DB, Pediatr Emerg Care 1992; 8:189, and Selbst, SM, Pediatr Rev 1986; 8:56.

Etiology of pulmonary hypertension

Cardiac
Increased pulmonary blood flow associated with left-to-right shunts at the atrial, ventricular, or great vessel level
Examples: atrial septal defect, patent ductus arteriosus, transposition of the great arteries, truncus arteriosus
Left-sided obstructive lesions associated with pulmonary venous congestion.
Examples: coarctation of aorta, aortic stenosis, mitral stenosis, pulmonary vein stenosis or atresia, cor triatriatum
Cardiomyopathy
Pulmonary
Idiopathic: primary pulmonary hypertension, veno-occlusive disease.
Hypoxic vasoconstriction
Alveolar hypoventilation: sleep disorders, upper airway obstruction, neuromuscular disorder, chest wall deformity
Obstructive or restrictive lung disease: cystic fibrosis, bronchopulmonary dysplasia, interstitial fibrosis
Pneumonia
High altitude
Pulmonary hypoplasia: primary, associated with congenital diaphragmatic hernia or renal dysplasia
Peripheral pulmonary stenosis
Persistent pulmonary hypertension of the newborn
Thromboembolic
Ventriculoatrial shunt, indwelling catheters, depe vein thrombosis
Parasitic disease: filariasis, schistosomiasis
Hematologic: sickle cell disease, polycythemia
Hepatic disease
Cirrhosis, portal hypertension
Collagen vascular disease
Scleroderma, systemic lupus erythematosus, rheumatoid arthritis, mixed connective tissue disease
Granulomatous disease
Sarcoidosis

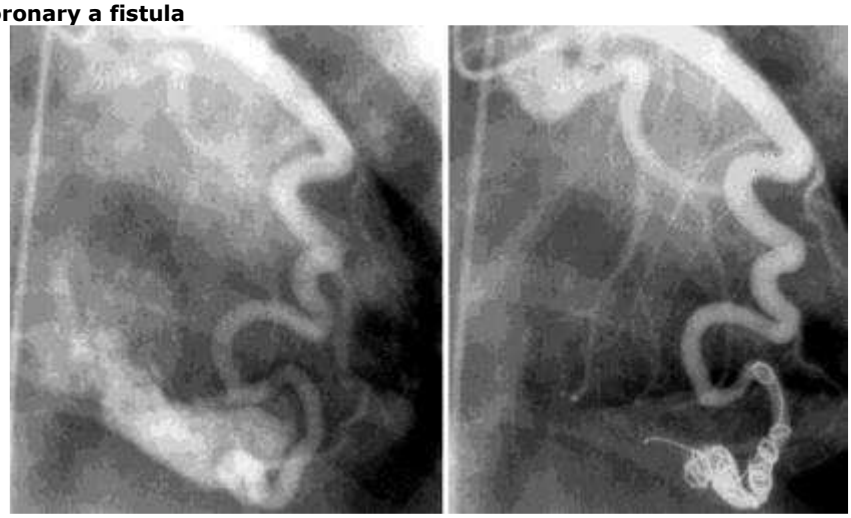
Courtesy of Robert L Geggel, MD.

Coronary artery abnormalities causing pediatric chest pain

1. Anomalous origin of the left coronary artery from the main pulmonary artery coronary artery fistula
2. Anomalous origin of the left coronary artery from the right sinus of Valsalva
3. Anomalous origin of the right coronary artery from the left sinus of Valsalva
4. Coronary ostial stenosis or atresia
5. Premature arteriosclerosis (progeria), familial hyperlipidemia
6. Kawasaki disease complicated by coronary artery aneurysm or stenosis
7. Coronary artery spasm
8. Coronary thrombosis associated with paradoxical embolus or hypercoagulable state

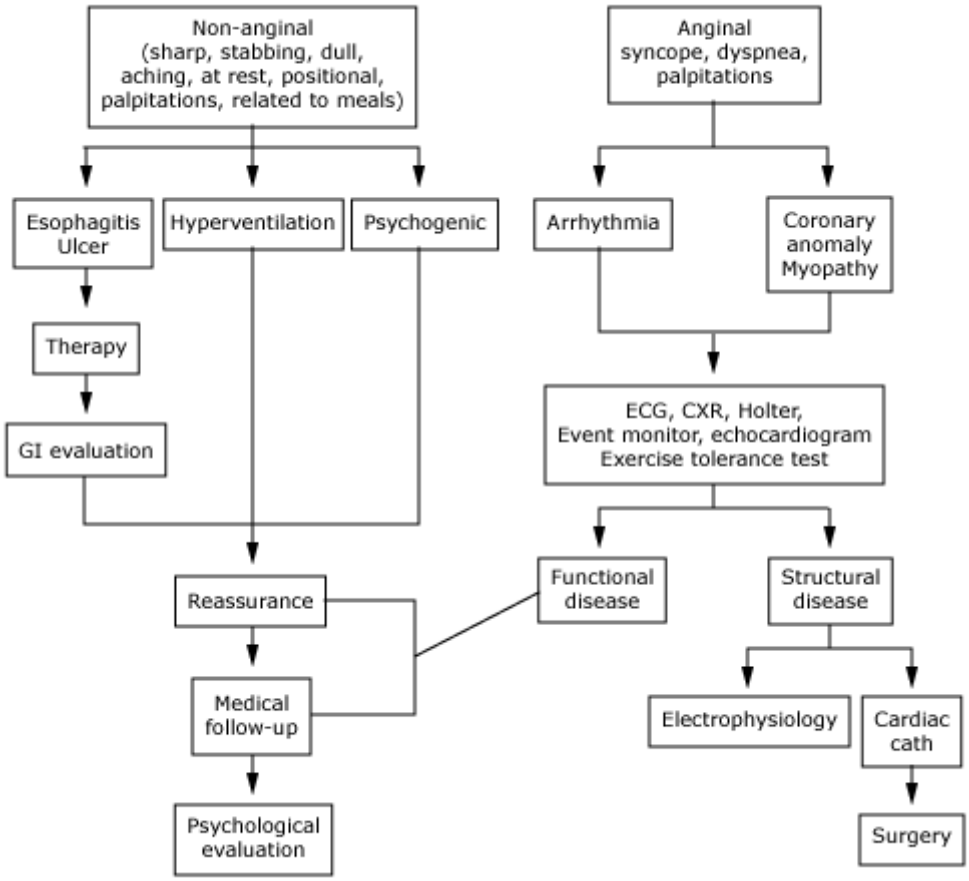
Courtesy of Robert L Geggel, MD.

Angiogram coronary a fistula



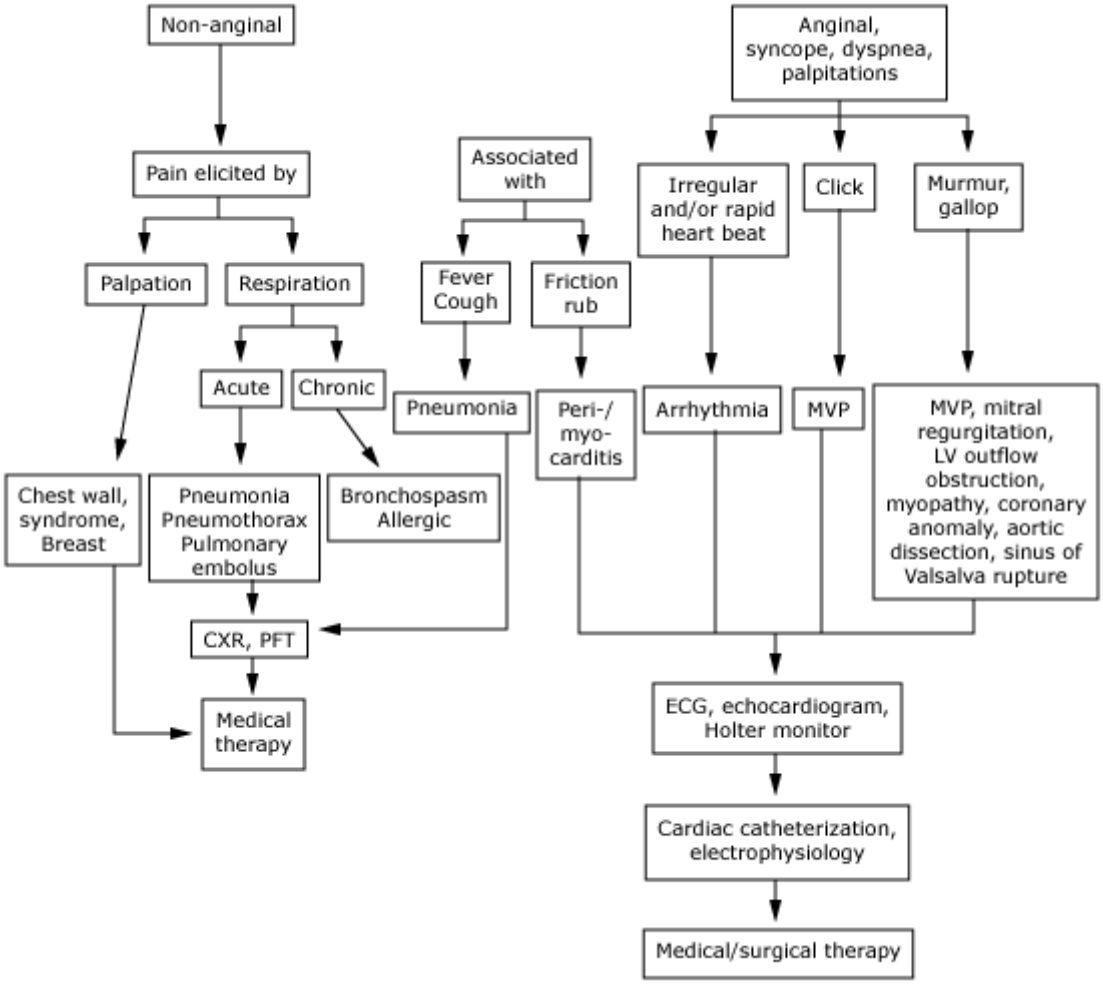
Right anterior oblique view of left coronary angiogram showing fistulous connection of left anterior descending coronary artery to the right ventricle (left panel). This connection was occluded by transcatheter placement of occluding coils (right panel). *Reproduced with permission from: Okubo, M, Nyaken, D, Benson, LN. Outcomes of transcatheter embolization in the treatment of coronary artery fistulas. Cathet Cardiovasc Intervent 2001; 52:510. Copyright © 2001 John Wiley &Sons, Inc.*

Normal physical examination



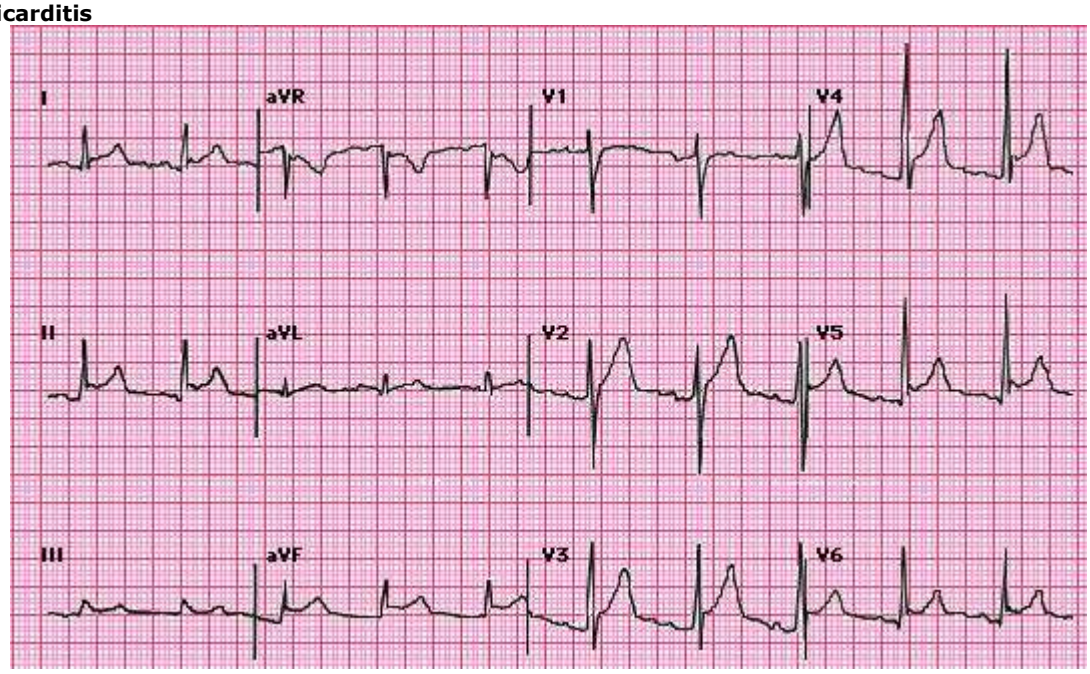
Schematic approach to evaluation of pediatric patient with chest pain in whom the physical examination is normal. For details about the various conditions, see text. *Data from: Brenner, JI, Ringel, RE, Berman, MA. Cardiologic perspectives of chest pain in childhood: a referral problem? To whom? Pediatr Clin North Am 1984; 31:1241.*

Evaluation of pediatric chest pain - abnormal physical examination



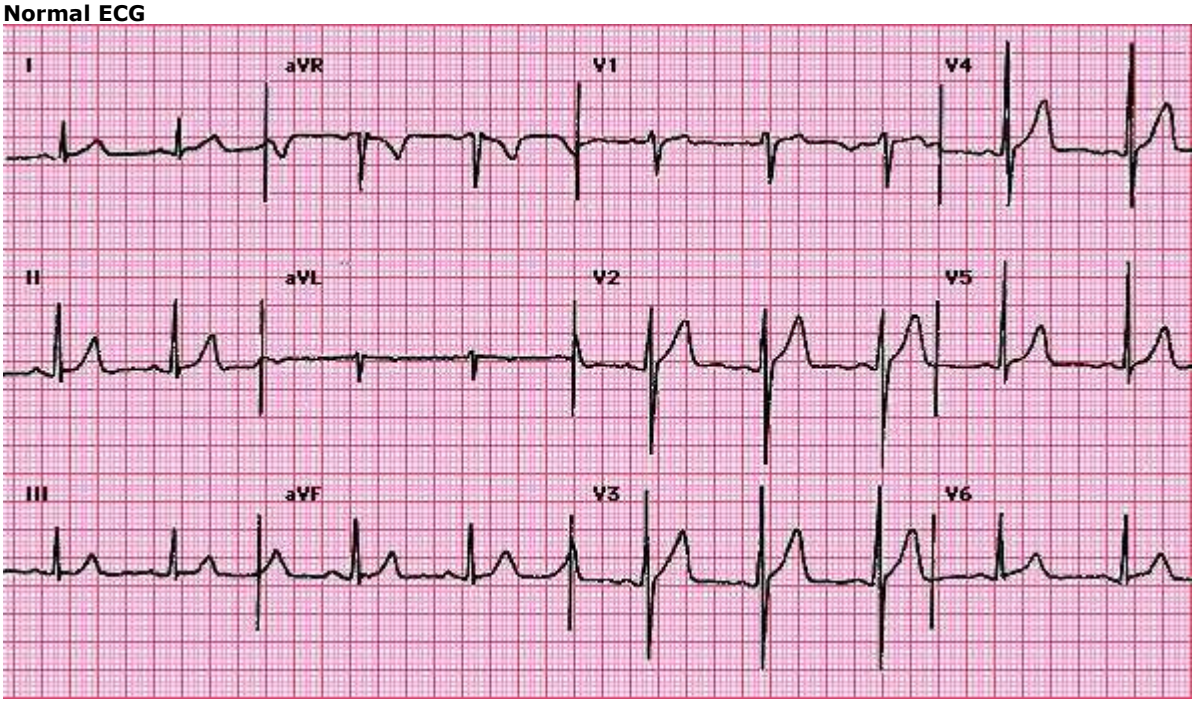
Schematic approach to evaluation of pediatric patient with chest pain in whom the physical examination is abnormal. For details about the various conditions, see the text. *Data from: Brenner, JI, Ringel, RE, Berman, MA. Cardiologic perspectives of chest pain in childhood: a referral problem? To whom? Pediatr Clin North Am 1984; 31:1241.*

Pericarditis



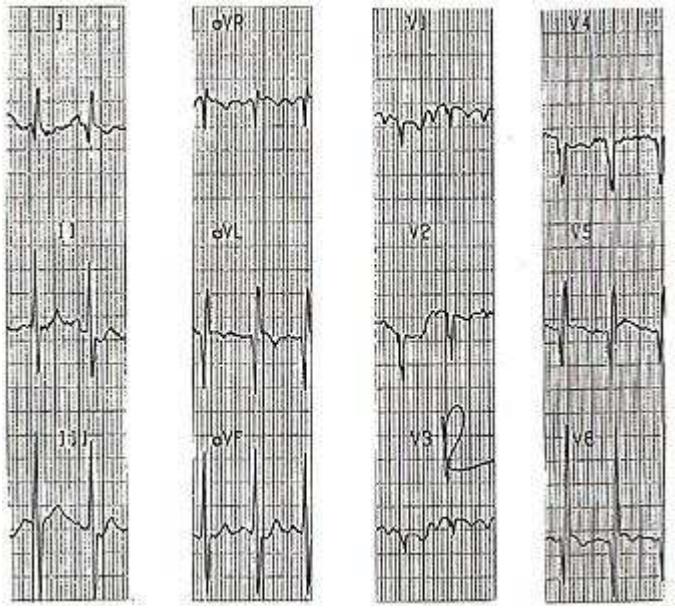
Electrocardiogram in acute pericarditis showing diffuse upsloping ST segment elevations seen best here in leads II, III, aVF, and V2 to V6. There is also subtle PR segment deviation (positive in aVR, negative in most other leads). ST segment elevation is due to a ventricular current of injury associated with epicardial inflammation; similarly, the PR segment changes are due to an atrial current of injury which, in pericarditis, typically displaces the PR segment upward in lead aVR and downward in most other leads. *Courtesy of Ary Goldberger, MD.*

Normal ECG



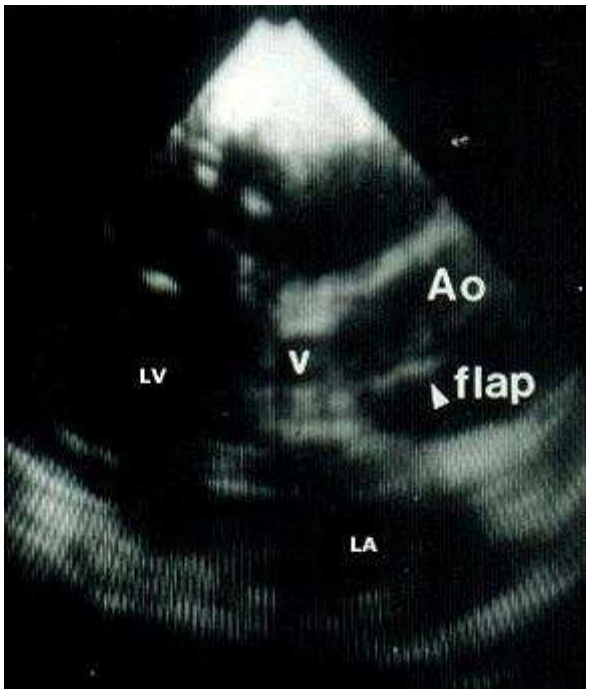
Normal electrocardiogram showing normal sinus rhythm at a rate of 75 beats/min, a PR interval of 0.14 sec, a QRS interval of 0.10 sec, and a QRS axis of approximately 75º. *Courtesy of Ary Goldberger, MD.*

An electrocardiogram obtained from a patient with anomalous left main coronary artery arising from the pulmonary artery



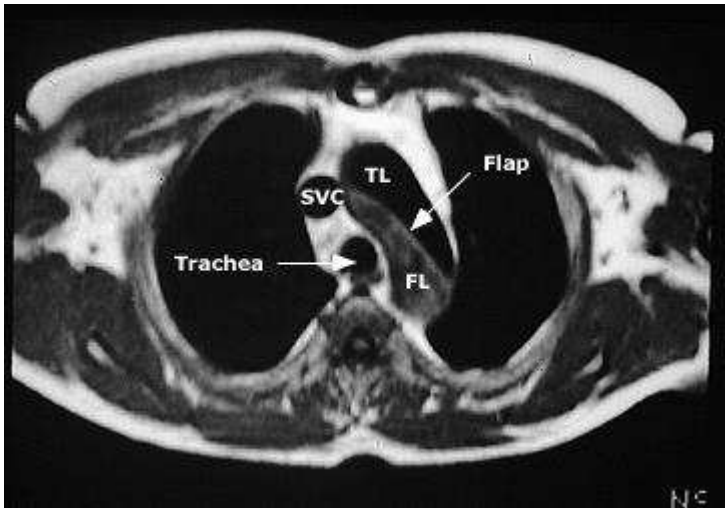
There is a pattern of anterolateral infarction with deep wide Q waves in the anterior (I, aVL) and lateral (V5, V6) leads. There are T wave inversions in these leads as well. *Courtesy of Robert L Geggel, MD.*

Proximal dissection of the aorta



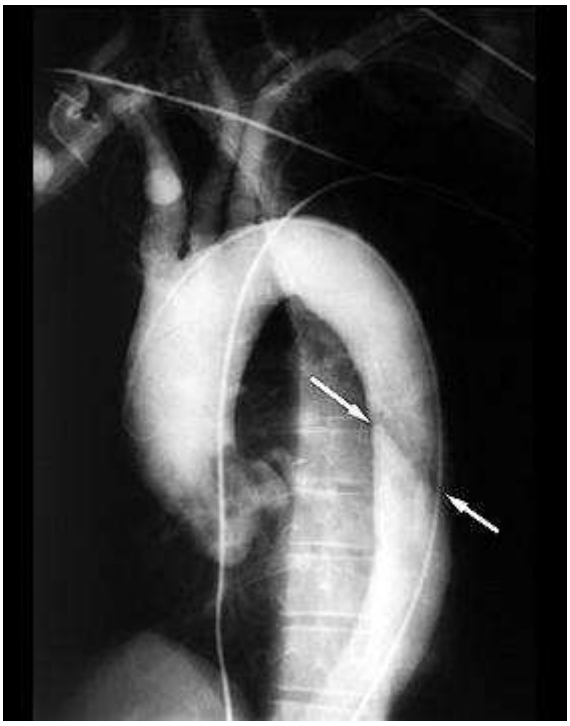
Modified long axis view shows a proximal dissection of the aortic root (Ao), with a flap extending to the aortic valve (V). This aortic pathology is seen by imaging the ascending aorta one interspace above the usual long axis precordial window. LA: left atrium; LV: left ventricle.

Aortic dissection on MRI



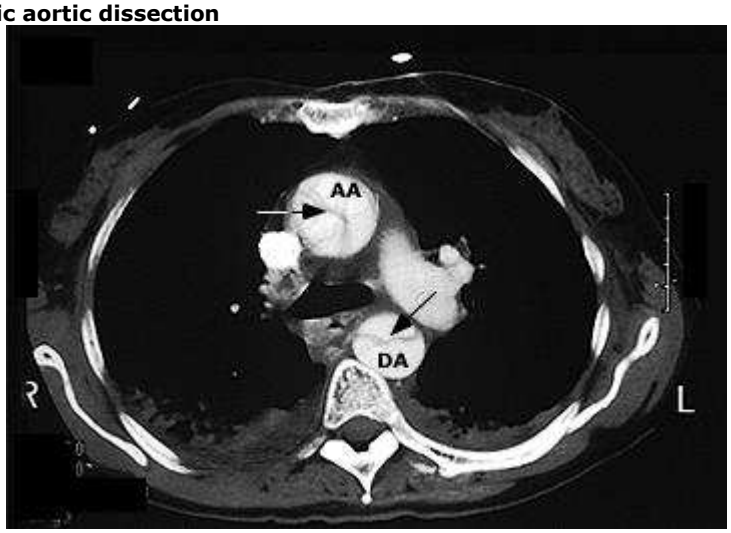
Spin-echo magnetic resonance imaging scan of an aortic dissection in the transverse plane at the level of the aortic arch. The true lumen (TL), false lumen (FL), and intimal flap can be easily identified. The trachea and superior vena cava (SVC) are also seen. *Courtesy of Warren Manning, MD.*

Descending aortic dissection



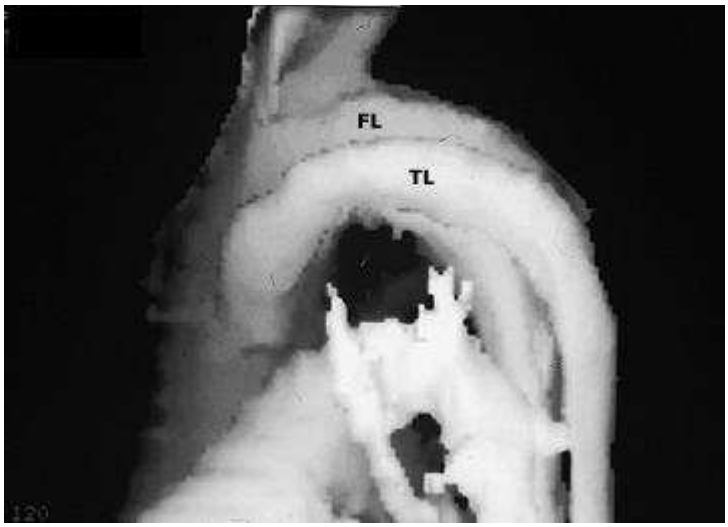
This aortogram demonstrates dissection of the descending thoracic aorta, arising immediately distal to the origin of the left subclavian artery. An oblique lucency is noted within the lumen of the aorta, which is a diagnostic feature (arrows). *Courtesy of Jonathan Kruskal, MD.*

Spiral CT of thoracic aortic dissection



Transverse plane through ascending (AA) and descending (DA) thoracic aorta showing the intimal flap (arrows) and both lumens of a type A aortic dissection. One cannot distinguish between the true and false lumens based on this view alone. *Courtesy of Vassilios Raptopoulos, MD.*

Spiral CT of thoracic aortic dissection



Sagittal plane reconstruction of transverse and descending thoracic aorta showing the true (TL) and false (FL) lumens of the same patient as the previous CT. *Courtesy of Vassilios Raptopoulos, MD.*

Approach to the child with palpitations

Authors
Robert Mazor, MD
Suzan Mazor, MD

Section Editors
Tony Woodward, MD
Jan E Drutz, MD

Deputy Editor
James F Wiley, II, MD, MPH

Last literature review version 17.2: May 2009 | **This topic last updated:** December 1, 2008 **(More)**

INTRODUCTION — Palpitations describe perception of the heartbeat that is usually concerning to the patient. In adults, palpitations occasionally herald serious underlying cardiac events [1] . However, palpitations in children typically arise from physiologic stimuli, such as fever, exercise, anxiety, or anemia, rather than life-threatening causes (eg, cardiac arrhythmia). In addition, children with serious arrhythmias may report no palpitations. This topic will review the differential diagnosis and approach to the child with palpitations.

The differential diagnosis of tachycardia in children who do not have palpitations is discussed elsewhere. (See "Approach to the child with tachycardia").

DIFFERENTIAL DIAGNOSIS — The differential diagnosis of palpitations encompasses rare etiologies that are life-threatening and common causes (show algorithm 1 and show table 1).

Life-threatening causes — Children with a serious underlying cause for their palpitations often have a history of syncope, congenital heart disease, or cardiac surgery (show table 1) [2-6] .

- Arrhythmias may be broadly classified as fast or slow. The child with tachyarrhythmia and shock should be triaged and managed per the American Heart Association (AHA) guidelines for assessment of cardiopulmonary instability espoused in the Pediatric Advanced Life Support (PALS) course [7] . (See "Approach to the child with tachycardia").

A key component of this assessment requires categorization of the QRS complex from the cardiorespiratory (CR) monitor or 12-lead ECG as narrow or wide (show algorithm 2) [2] . A 12-lead ECG before and after intervention is preferable if patient status allows.

Supraventricular tachycardia (SVT) is the most common non-sinus tachyarrhythmia of childhood and often presents with palpitations in verbal children [2,8] . Most infants with SVT are asymptomatic although feeding problems, pallor, or dyspnea, especially with feeding, may occur [8] . Ventricular tachycardia is rare in children with structurally normal hearts. (See "Management of supraventricular tachycardia in children", see "Management and evaluation of wide QRS complex tachycardia in children", and see "Sustained monomorphic ventricular tachycardia: Diagnosis and evaluation").

Symptomatic sinus bradycardia in children often arises from respiratory failure with hypoxemia. Sick sinus syndrome, Mobitz type II atrioventricular blockage, and complete atrioventricular dissociation are usually seen in children with structural heart disease. (See "Bradycardia in children").

- Myocarditis is most commonly viral in origin, with Coxsackie virus B and other enteroviruses accounting for most cases. Clinical findings of myocarditis include tachycardia out of proportion to fever, poor perfusion, and signs of heart failure. Patients with myocarditis may develop a variety of tachy- and bradyarrhythmias. Children with myocarditis may rapidly develop cardiogenic shock [9] . (See "Clinical manifestations and diagnosis of myocarditis in children").

- Hypertrophic cardiomyopathy (HCM) manifests as a hypertrophied, nondilated left ventricle and septum. Patients with HCM may present with palpitations or syncope during strenuous exercise caused by atrial fibrillation or ventricular arrhythmia. The subset of HCM patients who present with syncope are at significant risk for sudden death [10] . (See "Causes of syncope in children and adolescents" and see "Clinical manifestations of hypertrophic cardiomyopathy").

- Toxin exposures have the potential to cause life-threatening tachy- and bradyarrhythmias. The specific arrhythmia depends on the characteristics of the toxic agent (show table 2).

- Pheochromocytoma and paragangliomas are rare neoplasms in children. Tumors that arise from the adrenal medulla are termed pheochromocytomas, and those with extraadrenal origins are called paragangliomas. The classic triad of symptoms in these disorders consists of episodic headache, sweating, and tachycardia, usually accompanied by hypertension. Malignant hypertension can occur, with its associated complications (eg, increased intracranial pressure, encephalopathy). (See "Pheochromocytoma in children" and see "Hypertensive emergencies: Malignant hypertension and hypertensive encephalopathy").

Common causes — See table 1 for a differential diagnosis of palpitations (show table 1).

- Increased metabolic rate accompanies fever and anemia. The resulting sinus tachycardia and hyperdynamic cardiac activity may cause palpitations.

- Palpitations may also follow the catecholamine release associated with exercise, emotional arousal, and psychiatric distress (eg, anxiety, panic attack) [11] .

- Hyperventilation, breathing in excess of physiologic requirement to maintain oxygenation and ventilation, may occur without obvious underlying etiology in adolescents [12-15] . Patients with hyperventilation often have associated symptoms of dyspnea, chest tightness, chest pain, paresthesias, and palpitations (often described as a "racing" heart) [14] . Frequently they attribute their symptoms to a life-threatening problem and appear very anxious. In addition, children with hyperventilation may have a past medical history of asthma [13] .

While clinical findings reveal no serious cardiac or pulmonary abnormalities, patients with severe hyperventilation may display a positive Chvostek or Trousseau sign, laryngospasm, or spontaneous carpopedal spasm [12,15] . Ancillary studies (eg, venous or arterial blood gas and electrolytes) typically show respiratory alkalosis. Rarely, T wave changes, including ST segment depression, T wave flattening, and T wave inversion may appear on ECG during the hyperventilation episode [15] .

Once serious etiologies, such as status asthmaticus, metabolic acidosis pain, central nervous system disorders, and drug intoxication are excluded, treatment consists of reassurance [14,15] . Additional measures may be necessary:

- Placement of a partial rebreather oxygen mask with oxygen flow set below 5 liters per minute allows the patient to rebreathe carbon dioxide without causing hypoxia and may assist in terminating the hyperventilation.

- Benzodiazepine administration in the form of oral diazepam or lorazepam may be necessary to end the acute episode [15] .

Patients with hyperventilation need close follow-up and counseling. As many as 40 percent of patients with hyperventilation will have repeated episodes into adulthood [15] .

- Drug induced palpitations, as opposed to toxic exposures (show table 2), involve medications and substances with sympathomimetic or anticholinergic properties that increase heart rate and cardiac contractility with routine dosing [16-19] . Implicated agents include albuterol, caffeine, tobacco, cough and cold preparations, dietary supplements that contain ephedra, herbal medications, energy drinks, and isotretinoin.

- Postural orthostatic tachycardia syndrome (POTS) is defined as a form of orthostatic intolerance characterized by an excessive increase in heart rate (>30 bpm over baseline or to >120 bpm in adults) that occurs on standing without arterial hypotension. POTS is a common disorder among teenage girls that manifests as palpitations, anxiety, dizziness, and tremulousness. (See "Postural tachycardia syndrome").

- Premature atrial contractions (PACs), the most common sinus arrhythmia of childhood, result from premature depolarization of an atrial focus. Patients often describe that they feel like their heart "stops" or "flip-flops." PACs are benign [3] .

- Premature ventricular contractions (PVCs) result from premature depolarization of a ventricular focus or reentry. Once structural heart disease and ventricular dysfunction are excluded by ancillary testing, PVCs can be considered benign [3] .

Other causes — See table 1 for a differential diagnosis of palpitations (show table 1).

- Hyperthyroidism frequently presents with palpitations in children. Goiter, diaphoresis, weight loss, heat intolerance, tachycardia, and widened pulse pressure comprise common additional findings [20] . (See "Clinical manifestations and diagnosis of hyperthyroidism in children and adolescents").

- Acute rheumatic fever (ARF) is a delayed, nonsuppurative sequela of a pharyngeal infection with the group A streptococcus. Fever and pancarditis are clinical manifestations of this disease that can cause tachycardia. Other findings include arthralgia, arthritis, erythema marginatum, subcutaneous nodules, and chorea. Testing for recent streptococcal infection is essential to establish the diagnosis. (See "Clinical manifestations and diagnosis of acute rheumatic fever").

- Mitral valve prolapse is associated with a wide variety of clinical features. Palpitations are a common presenting complaint in MVP. The evaluation of patients with symptomatic palpitations is similar to that in patients without MVP. Palpitations are often associated with ventricular premature beats, but supraventricular arrhythmias can also occur. (See "Arrhythmic complications of mitral valve prolapse" and see "Mitral valve prolapse syndrome").

HISTORY — A careful history helps to identify patients at higher risk for a life-threatening cause for their palpitations.

- Palpitation description - Palpitations caused by life threatening tachyarrhythmias often start and stop abruptly (like turning a light switch on and off). Children may feel like there is a butterfly in their chest or like their heart is beating so fast that it will come out of the chest. These palpitations generally are of short duration (seconds to minutes).

Palpitations due to cardiac hyperdynamic states with sinus tachycardia are often described as a rushing or pounding in the ears, particularly when the patient is supine. Exercise, fever, anemia, pharmaceuticals, nutraceuticals, caffeine drinks, and emotional arousal may produce this sensation of increased ventricular stroke volume that often lasts minutes to hours.

Children with premature atrial or ventricular contractions may note the increased stroke volume of the compensatory heart beat as a "flip-flop" or say that their heart "stops". A similar sensation may be produced in patients with bradycardia and high grade (Type II or Type III) atrioventricular block due to escape beats.

Palpitations associated with diaphoresis, headache, flushing and hypertensions suggest pheochromocytoma.

Palpitations associated with heat intolerance and diaphoresis suggest hyperthyroidism.

- Congenital heart disease, repaired or unrepaired - Palpitations in this setting have a high likelihood of cardiac arrhythmia as an underlying cause [21] .

- Syncope - A history of palpitations with syncope, especially in association with exercise, raises concern for a primary cardiac etiology such as cardiac arrhythmia, hypertrophic cardiomyopathy, or myocardial ischemia (usually related to congenital anomaly of a coronary artery). (See "Neurocardiogenic (vasovagal) syncope").

In children with intermittent tachyarrhythmias (eg, prolonged QT syndrome or Wolff Parkinson White syndrome), palpitations with abrupt syncope may be described. Triggers for these events involve intrinsic catecholamine stimulation such as exercise, sudden exposure to cold water (diving), or sudden surprise. Unlike vasovagal syncope, patients usually fall without protecting themselves from injury. Unlike seizures, patients usually recover full mental status without a postictal phase as long as the arrhythmia is of short duration.

- Illness/fever - A history of viral illness or fever in conjunction with respiratory distress or myocardial dysfunction suggests myocarditis, especially if tachycardia is out of proportion to the degree of fever or is present after defervescence. (See "Clinical manifestations and diagnosis of myocarditis in children").

A history of a recent streptococcal infection in the febrile patient with tachycardia raises clinical suspicion for acute rheumatic fever. If signs of carditis, such as a new murmur of mitral valve regurgitation or pericardial rub are present, then the patient meets Jones criteria for the diagnosis of acute rheumatic fever. (See "Clinical manifestations and diagnosis of acute rheumatic fever").

- Family history — Family history of sudden cardiac death or deafness raises clinical suspicion for genetic disorders such as long QT syndrome and hypertrophic cardiomyopathy that may cause sudden onset of ventricular tachycardia with palpitations and syncope.

PHYSICAL EXAM — Ill-appearing patients with palpitations need rapid assessment for hypoxemia, hypoglycemia, and hemodynamic instability. Clinicians should emergently treat the patient with cardiovascular collapse per American Heart Association guidelines for cardiopulmonary resuscitation (show algorithm 1 and show algorithm 2 and show algorithm 3) [7] . (See "Bradycardia in children" and see "Approach to the child with tachycardia").

Vital signs — Tachycardia or bradycardia are important findings in children with palpitations that suggest an underlying arrhythmia:

- Tachycardia is defined as the presence of a heart rate value greater than expected for age. In addition to the values published for specific age groups [22] , clinicians may use the basic guideline of >160 BPM for infants (< 2 years), >140 BPM for children (2 to 10 years), and >100 BPM for children over 10 years of age, adolescents, and adults to define tachycardia (Show table 3).

- Conversely, bradycardia means a heart rate measurement that is less than expected for age or <90 BPM for infants, <70 BPM for children, and <60 BPM for adolescents and adults [22] .

An orthostatic increase in heart rate >30 BPM over baseline without hypotension defines postural orthostatic tachycardia.

An elevated blood pressure for age may be seen in patients with anxiety, panic attacks, hyperventilation, or pheochromocytoma.

Hypertension with a widened pulse pressure suggests hyperthyroidism.

Fever should prompt evaluation for other findings consistent with myocarditis or acute rheumatic fever.

Cardiovascular findings — Most stable patients with palpitations have a normal cardiac examination.

- Patients with fever or anemia may demonstrate short systolic flow murmurs of medium to high pitch.

- Patients with mitral valve prolapse may display a midsystolic click or multiple clicks followed by a midsystolic to late systolic murmur at the apex of the left ventricle over the mitral area show heart sound 1 show heart sound 2. These findings may be transient depending in part upon loading conditions, such as squatting that increase blood flow to the heart (show table 4). (See "Auscultation of cardiac murmurs" and see "Physiologic and pharmacologic maneuvers in the differential diagnosis of heart murmurs and sounds").

- The pancarditis of acute rheumatic fever may manifest as a pericardial rub or the murmur of mitral regurgitation. (See "Auscultation of cardiac murmurs").

- Myocarditis may be evident based on the presence of a gallop or murmur of mitral or tricuspid insufficiency in association with other signs of congestive heart failure. (See "Clinical manifestations and diagnosis of myocarditis in children").

- Patients with congenital heart disease may have a variety of cardiac findings, depending on the physiologic status of their cardiac lesion, including cyanosis, pathologic murmurs, and evidence of cardiac failure.

Noncardiovascular findings — Other important findings in patients with palpitations include:

- Conjunctival pallor that suggests anemia.

- Exophthalmos with or without goiter that indicates hyperthyroidism.

- Hyperventilation that consists of markedly rapid breathing or deep breathing, often accompanied by sighing, without an obvious etiology.

- Tachypnea with rales that implies pulmonary congestion from left sided congestive heart failure seen in patients with myocarditis or congenital heart disease.

- Hepatomegaly and jugular venous distension that suggests right sided heart failure seen in patients with myocarditis or congenital heart disease.

- Diaphoresis with flushing that is found in patients with pheochromocytoma.

- High levels of emotional distress without life-threatening findings seen in patients with anxiety, panic attack, or hyperventilation.

ANCILLARY STUDIES — In the acutely symptomatic patient with palpitations, monitoring and assessment of the cardiac rhythm is vital.

Standard bedside cardiac monitors often do not provide satisfactory rhythm tracings to allow detailed decision making. Thus, a standard 12-lead ECG and rhythm strip should be obtained upon presentation and concurrent with any attempts at cardioversion or defibrillation to guide therapy and to provide valuable data should the patient require pediatric cardiology consultation [23] .

Even in the patient who is asymptomatic on presentation, continuous monitoring during initial evaluation and a standard ECG is often helpful since an ECG is essential in the diagnosis of Wolff Parkinson White and long QT syndromes and may be helpful in the diagnosis of premature atrial and ventricular beats.

Readily available hemoglobin or hematocrit measurement can confirm clinical suspicion of anemia.

Clinical findings suggestive of hyperthyroidism can be confirmed by serum thyroid testing. Serum levels of thyroxine (T4) and triiodothyronine (T3) are elevated, and TSH secretion is suppressed in hyperthyroidism. (See "Clinical manifestations and diagnosis of hyperthyroidism in children and adolescents").

If the patient presents with an acute tachyarrhythmia, the clinician should obtain an echocardiogram to evaluate myocardial function, as well as to evaluate for underlying structural heart disease soon after normal sinus rhythm is established, preferrably within a few days. In the hemodynamically unstable patient, this study should be performed on an emergent basis as the information will impact therapeutic decision making.

Cardiac troponin levels (troponin-I and -T) may be elevated in myocarditis. However, myocardial enzymes (creatine kinase [CK], MB isoenzyme of CK [CK-MB]) levels are generally not helpful as they are rarely abnormal [9] . Serum PCR identification of a viral infection, or viral isolation from other body sites, such as the oropharynx or gastrointestinal tract, can support the diagnosis. (See "Troponins and creatine kinase as biomarkers of cardiac injury").

Pharyngeal Group A Streptococcus culture, Antistreptolysin O (ASO), anti-deoxyribonuclease B (anti-DNase B), and antihyaluronidase titers are essential for documenting antecedent streptococcal infection as part of the diagnosis of rheumatic fever [24] . (See "Clinical manifestations and diagnosis of acute rheumatic fever").

Twenty-four hour ambulatory ECG (Holter) monitors, longer term event and implantable loop recorders are helpful for sporadic events [25,26] , but prescription of these should be limited to a pediatric cardiologist. (See "Ambulatory monitoring in the assessment of cardiac arrhythmias").

ALGORITHMIC APPROACH — Ill-appearing patients require rapid assessment of airway, breathing, and circulation (show algorithm 1). The evaluation should focus on identifying and treating hypoxia, hypoglycemia, shock, and arrhythmia (show algorithm 2 and show algorithm 3). (See "Approach to the child with tachycardia").

Children with a history of palpitations, but who are not acutely symptomatic, should be placed on continuous cardiac monitoring and undergo a 12-lead ECG.

Patients with a history of syncope, congenital heart disease, or cardiac surgery are at higher risk of having a cardiac arrhythmia as the cause of their palpitations [21] . These children warrant involvement of a pediatric cardiologist early in their evaluation so that an echocardiogram looking for cardiac structural abnormality and advanced testing for cardiac rhythm disturbance occur.

Poisoned patients with palpitations are also at high risk for cardiac arrhythmia. Drugs associated with rhythm disturbance following overdose are listed in the table (show table 2).

Additional findings help distinguish among the life-threatening causes of palpitations:

- The diagnosis of Wolff-Parkinson-White syndrome is made if ventricular preexcitation (delta wave) and a short PR interval are present on 12-lead ECG [2] . (See "Electrocardiographic features of the Wolff-Parkinson-White pattern").

- A prolonged corrected QT interval (QTc) identifies the presence of long QT syndrome [22,27,28] . (See "Diagnosis of congenital long QT syndrome" and see "Acquired long QT syndrome").

- Sinus bradycardia with SA block or SA arrest identifies sick sinus syndrome. Children with pacemaker malfunction often have bradyarrhythmia. Causes include poor capture, battery failure, and lead misplacement. (See "Bradycardia in children" and see "Pacing system malfunction: Evaluation and management").

- Fever with congestive heart failure is found in children with myocarditis. (See "Clinical manifestations and diagnosis of myocarditis in children").

- The ECG may also document premature extrasystoles. If the history is convincing for periodic episodes of palpitations, but the initial workup is unrevealing, clinicians should refer the patient to a pediatric cardiologist for further evaluation and testing. Potential methods that a cardiologist will use to identify the cardiac rhythm during the palpitation episode include 24 hour ambulatory ECG (Holter) monitors, longer term event recorders, and implantable loop recorders [25,26] .

- Children with palpitations, headache, diaphoresis, and hypertension should undergo evaluation for pheochromocytoma. (See "Pheochromocytoma in children").

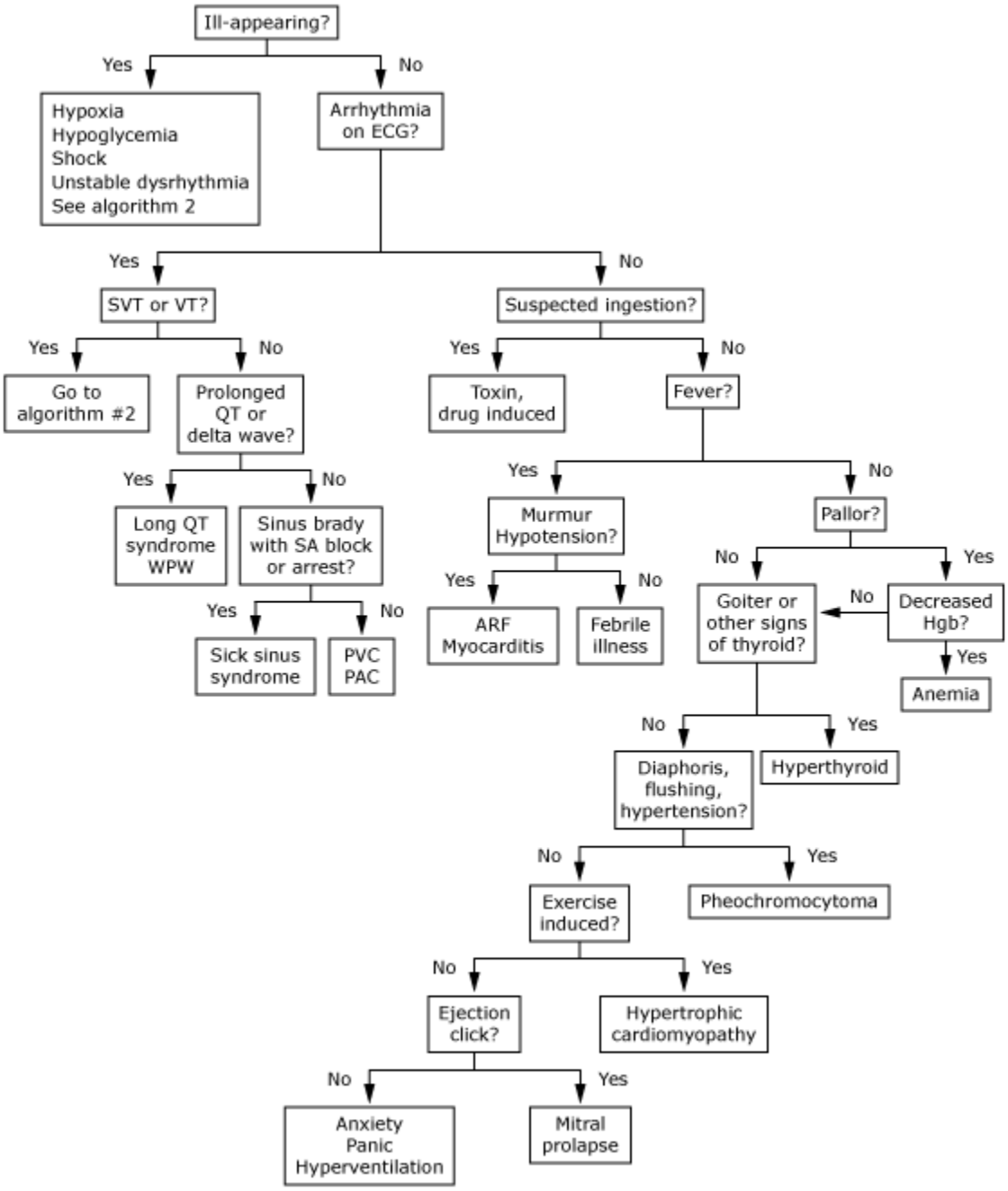
Well-appearing patients with a single episode of palpitations typically have a benign cause, such as fever, anemia, ingestion of medications and substances with sympathomimetic or anticholinergic effects, postural tachycardia syndrome, or hyperventilation (show algorithm 1).

Some patients with significant psychiatric distress due to anxiety, panic attack, or other mental illness may also present with palpitations. These patients warrant appropriate workup for potential organic causes, such as hyperthyroidism, and further mental health assessment and support.

SUMMARY AND RECOMMENDATIONS — The approach to the evaluation of palpitations is summarized in the algorithm (show algorithm 1). Assessment for ill appearance and the determination of rhythm are early decision points in the evaluation of children with palpitations. Ill-appearing patients with tachycardia or shock require immediate resuscitation (show algorithm 2 and show algorithm 3). Children with syncope or congenital heart disease are more likely to have a life-threatening dysrhythmia. The table summarizes the causes of palpitations, highlighting the most common and the most life threatening causes (show table 1). (See "Approach to the child with tachycardia").

1.

Palpitations algorithm



ECG: electrocardiogram; SVT: supraventricular tachycardia; VT: ventricular tachycardia; ARF: acute rheumatic fever; PVC: premature ventricular centraction; PAC: premature atrial centraction.

Neurocardiogenic (vasovagal) syncope

Author
Brian Olshansky, MD

Section Editor
Leonard I Ganz, MD

Deputy Editor
Robert O Blaustein, MD, PhD

Last literature review version 17.2: May 2009 | **This topic last updated:** October 13, 2008 **(More)**

INTRODUCTION — Neurally-mediated (reflex) syncope refers to a reflex response causing vasodilatation and/or bradycardia (rarely tachycardia) leading to systemic hypotension and cerebral hypoperfusion [1] . Types of neurally-mediated syncope include neurocardiogenic (vasovagal) syncope, carotid sinus syncope, situational syncope, and glossopharyngeal neuralgia (show table 1).

Neurocardiogenic syncope, also known as vasovagal syncope, is a common cause of syncope [2] . The diagnosis may be suggested by a specific history with well-known triggers, but a classic history is not required. The diagnosis can also be made by exclusion of other causes of syncope and by a characteristic response to upright tilt table testing during which the patient may pass out from bradycardia and/or hypotension.

The following discussion primarily applies to patients with recurrent syncope. Acute "vasovagal" reactions leading to syncope or presyncope are common also in a number of stressful settings, such as blood donation. They do not necessarily recur or require treatment. (See "Procedures used for blood donor screening: protection of potential blood donors and recipients", section on Vasovagal reactions).

Neurocardiogenic syncope and situational syncope will be reviewed here. General discussions of the pathogenesis, etiology, and evaluation of syncope, and issues related to carotid sinus hypersensitivity and glossopharyngeal neuralgia are discussed separately. (See "Pathogenesis and etiology of syncope" and see "Evaluation of syncope in adults" and see "Carotid sinus hypersensitivity" and see "Overview of craniofacial pain", section on Glossopharyngeal neuralgia).

There are some similarities between neurocardiogenic syncope and postural orthostatic tachycardia syndrome, which is discussed separately. (See "Postural tachycardia syndrome").

PATHOGENESIS OF NEURALLY-MEDIATED SYNCOPE — Understanding the physiology involved in neurally-mediated syncope is essential to understanding its clinical manifestations. Both neural (Bezold-Jarisch and carotid sinus reflexes) and endogenous chemical pathways are thought to be involved (show figure 1) [3] .

Alterations in autonomic activation are responsible for neurally-mediated syncope. Three types of responses are seen: a cardioinhibitory response, a vasodepressor response, and a mixed response with features of both [4] .

- The cardioinhibitory response results from increased parasympathetic tone and may be manifested by any or all of the following: sinus bradycardia, PR prolongation, and advanced atrioventricular block.

- The vasodepressor response is due to decreased sympathetic activity and can lead to hypotension. In one report, for example, the final trigger for symptomatic hypotension appeared to be the abrupt disappearance of muscle sympathetic nerve activity [5] . Reduced cardiopulmonary baroreceptor sensitivity may be a contributing factor [6] .

An individual patient may have separate syncopal events characterized by a primary vasodepressor, cardioinhibitory or mixed response. Furthermore, the response observed during tilt table testing is not necessarily the same as that recorded during clinical episodes. (See "Implantable loop recorder" below).

NEUROCARDIOGENIC SYNCOPE — Neurocardiogenic (vasovagal) syncope refers to a variety of clinical scenarios in which a neural reflex results in usually self-limited systemic hypotension characterized by bradycardia and/or peripheral vasodilation [7] . It is the most common cause of syncope (approximately 20 to 35 percent of cases), particularly in patients without apparent cardiac or neurologic disease [8-14] . However, neurally mediated syncope is the most common cause of syncope even among patients with heart disease and should be considered as a potential cause in such patients [11] .

Neurocardiogenic syncope is not a common cause of syncope in athletes. Athletes with syncope during physical activity should be evaluated for potential risk of sudden death. (See "Arrhythmia in athletes", section on Syncope).

Pathogenesis of neurocardiogenic syncope — Neurocardiogenic syncope may be caused by autonomic cardioinhibitory and/or vasodilator responses. It is likely that sensory inputs through vagal afferents, pain pathways, and central pathways (visual, temporal love, and other inputs) affect the nucleus tractus solitarius to cause sympathoinhibition and vagal efferent activation.

The most frequent mechanism for neurocardiogenic syncope is a cardioinhibitory response, as illustrated in a selected population of 111 patients with presumed neurocardiogenic syncope who received an implantable loop recorder and were followed for 3 to 15 months [15] . Syncope recurred in 34 percent; a correlation between syncope and electrocardiographic (ECG) changes was found in 84 percent. The most frequent abnormality (seen in 50 percent) was one or more prolonged asystolic pauses, primarily due to sinus arrest; bradycardia (<40 beats per min) without pauses was seen in 9 percent. The remaining patients had normal sinus rhythm or sinus tachycardia and probably had a vasodepressor response. Presyncope was usually associated with sinus rhythm, and was never associated with asystole.

Autonomic dysfunction — Autonomic dysfunction is thought to play an important role in neurocardiogenic syncope but neurocardiogenic syncope also occurs without apparent autonomic dysfunction. Bezold-Jarisch and carotid sinus reflexes may be involved [3] . Patients may have increased muscle sympathetic nerve activity at rest and a blunted response to orthostatic stress [16] .

- Bezold-Jarisch reflex — Receptors in the atria, great veins, and left ventricle exist whose activation results in the Bezold-Jarisch reflex. Activation of the mechanoreceptors in the left ventricle and stretch receptors in the great vessels with pressure or volume loading (as may occur with sympathetic stimulation) stimulate C fibers; such stimulation may result in activation of vagal afferents and then vagal efferents.

- Carotid sinus reflex — Blood pressure and heart rate are normally controlled in part by input from baroreceptors located within the carotid sinus and aortic arch. An increase in blood pressure or pressure applied to the carotid sinus enhances the baroreceptor firing rate and activates vagal efferents, thereby slowing the heart rate and reducing the blood pressure.

Central serotonergic pathways — Central serotonergic pathways appear to participate in the pathogenesis of neurocardiogenic syncope, an observation that provides the rationale for the use of selective serotonin reuptake inhibitors in this disorder (see "Other medications" below).

Drugs that enhance central serotonergic activity, such as fenfluramine and clomipramine, increase the plasma levels of prolactin and cortisol and have been used to evaluate the activity of the serotonergic system. One study administered clomipramine to 20 subjects, eight of whom had a history of neurocardiogenic syncope; those with a history of syncope had higher levels of prolactin and cortisol compared to controls without syncope [17] . Furthermore, serum concentrations of prolactin and cortisol obtained during tilt table testing (without clomipramine) were significantly elevated only in those subjects with a positive test.

Further support for the role of central serotonergic pathways comes from a study in which clomipramine therapy increased the number of patients with a history of syncope who had a positive tilt table test (80 percent versus 53 percent without therapy); the drug had no effect in controls [18] .

Adenosine — The observation that adenosine administration can provoke neurally mediated syncope raises the possibility that endogenous adenosine plays a role in the pathogenesis of neurocardiogenic syncope. (see "Adenosine administration" below).

This hypothesis was evaluated in a study of 26 patients with unexplained syncope who underwent upright tilt table testing; plasma adenosine was measured at baseline, immediately after tilting, and at 45 minutes [19] . The 11 patients with a positive tilt table test (ie, syncope was induced) had much higher plasma adenosine values at all three time periods. Plasma adenosine increased by about 50 percent above baseline during tilt-induced syncope, and the higher the value, the shorter the time to appearance of symptoms.

These observations suggest that adenosine release, perhaps mediated by mechanoreceptors in the heart, may be involved in the triggering mechanism of syncope, at least during tilt testing. Adenosine has a variety of actions including negative chronotropic and inotropic activity and vasodepression [19] .

Other — A number of other mechanisms may contribute to neurocardiogenic syncope. These include adenosine receptor activation [20] and impaired cerebral autoregulation characterized by cerebral vasoconstriction just prior to the loss of consciousness [21-23] . The latter problem is also called cerebral syncope and can occur in the absence of systemic hypotension [22,24] .

Diagnosis — The history, physical examination, and specific testing all may be important (see "Evaluation of syncope in adults").

Clinical presentation — Patients with neurocardiogenic syncope are most commonly young and otherwise healthy. Typical triggers and premonitory symptoms are strongly suggestive of neurocardiogenic syncope although these may be lacking. Women and patients younger than 40 are more likely to have typical symptoms [25] . However older patients are also frequently diagnosed with neurocardiogenic syncope [26] .

- "Classical vasovagal syncope" refers to syncope triggered by emotional or orthostatic stress such as venipuncture (experienced or witnessed), painful or noxious stimuli, fear of bodily injury, prolonged standing, heat exposure, or exertion.

- However, some patients, especially those who are older, have recurrent episodes without an identifiable cause or trigger (referred to as "non-classical" presentation).

Neurocardiogenic syncope is often associated with a prodrome and persistence of nausea, pallor, and diaphoresis, consistent with increased vagal tone. Syncope is typically of short duration and occurs in the sitting or standing position. The supine position restores adequate blood flow to the brain. However, full recovery may be delayed as the patient may feel depressed or fatigued. This course may help distinguish neurocardiogenic syncope from syncope associated with arrhythmia which is typically of abrupt onset and of short duration. Loss of consciousness may be prolonged with some other causes of syncope such as seizures and aortic stenosis but rarely with neurocardiogenic syncope. (See "Evaluation of syncope in adults").

Older patients, unlike younger patients, may have a combination of neurocardiogenic syncope, carotid sinus hypersensitivity and/or orthostatic hypotension [27,28] and are more likely to have co-existing cardiovascular disease and to be taking medications.

In patients with syncope during exertion, older patients, and other patients who may have structural heart disease, other potential causes of syncope should be excluded.

History can be very helpful in making the diagnosis [1,29] . Further evaluation with tilt table test is not indicated if diagnostic vasovagal features are identified and further testing will not alter treatment.

Upright tilt table test — The tilt table test is a commonly performed test for the evaluation of syncope in whom the diagnosis of neurocardiogenic syncope is suspected but not clear based upon initial evaluation and ruling out neurocardiogenic syncope is important (show table 2) [30,31] . However the tilt table test has limited specificity, sensitivity, and reproducibility. A detailed discussion of the tilt table test is found elsewhere (show table 3). (See "Upright tilt table testing in the evaluation of syncope").

Adenosine administration — Adenosine or the related precursor compound adenosine triphosphate (ATP) provoke a short and potent cardioinhibitory response of vagal origin, including sinus bradycardia or pauses, atrioventricular (AV) block, and vasodepression. In various studies, a positive response to adenosine or adenosine triphosphate has been attributed to various diagnoses including sick sinus syndrome, AV block and neurocardiogenic syncope. (See "Evaluation of syncope in adults" section on Adenosine and adenosine triphosphate").

Adenosine may have a complementary role to the tilt table test in the evaluation of patients with possible neurocardiogenic syncope and, as noted above, adenosine release may trigger syncopal episodes. The administration of adenosine requires less time than tilt table testing, often less than 10 minutes.

- The efficacy of adenosine (given as 6 and 12 mg boluses) was compared to head-up tilt-table testing in 85 patients presenting with syncope and in 14 normal controls [32] . A vasovagal response was defined as the development of syncope or presyncope associated with relative bradycardia and/or hypotension (decrease in systolic blood pressure ≥ 30 mmHg) occurring 15 to 60 sec after adenosine injection or during the tilt table test. The inducibility of a vasovagal response with adenosine was comparable to that with the tilt table test in patients with syncope (26 and 34 percent) and in normals (7 percent for both tests).

- A second study of 100 patients found that, despite a similar yield, the results with adenosine and routine tilt table study were discordant in 21 percent of patients [33] . However, most of the patients with a positive response to adenosine but negative tilt table study had a positive response with the use of isoproterenol, suggesting that adenosine and isoproterenol tilt testing have complementary roles.

The ATP and tilt tests may provide valuable information in different patient groups. In a study that compared the two tests in 72 patients with presumed neurocardiogenic syncope, tilt testing reproduced symptoms mostly in younger patients while the ATP test was positive primarily in older patients; the results of the two studies were not correlated with each other [34] .

The 2004 ESC syncope guidelines include recommendations for use of adenosine triphosphate (ATP) (show table 4). The use of ATP to evaluate syncope is not FDA approved and ATP infusion is not a standard test in the US at this time.

There are several reasons to be cautious about the use and interpretation of the adenosine and ATP tests. These included limited data on the specificity of positive responses and paucity of data on the impact of therapy in patients with positive responses. (See "Evaluation of syncope in adults", section on Adenosine and adenosine triphosphate tests). A limitation of ATP and tilt table tests is that both are poorly correlated with the mechanism of spontaneous syncope as detected by implantable loop recorder [35] .

Thus, further study is needed before adenosine or ATP infusion can be recommended in the routine evaluation of neurocardiogenic syncope

Implantable loop recorder — The implantable loop recorder (ILR) is a subcutaneous monitoring device for the detection of cardiac arrhythmias [36] . Such a device is typically implanted in the left pectoral region and has a battery life of 18 to 24 months. It stores events when the device is activated automatically according to programmed criteria or manually with magnet application. The ILR may be most useful in patients with infrequent symptoms and suspected arrhythmia in whom noninvasive testing is negative or inconclusive. In patients with frequent symptoms, shorter term noninvasive electrocardiographic monitoring (eg, an external loop recorder) may suffice. The use of ILR and other electrocardiographic monitoring in the diagnosis of syncope generally is discussed separately. (See "Evaluation of syncope in adults", section on Implantable loop recorder).

ILRs may more accurately establish a causative relationship between bradyarrhythmias and syncope than provocative tests (eg, upright tilt table test or ATP infusion) [35,37] . This was illustrated in a series of 392 patients without significant electrocardiographic or cardiac abnormalities who underwent ILR implantation after three or more syncopal episodes in two years [35] . Patients with orthostatic hypotension and carotid sinus syncope were excluded.

- 343 patients underwent tilt-table testing, which was positive in 164 (48 percent) and 180 underwent ATP infusion, which was positive in 53 (29 percent).
- Syncope was documented by ILR in 106 patients (26 percent) after a median of 3 months.
- Patients with positive and negative tilt tests had similar baseline characteristics, syncopal recurrence rate, and mechanism of spontaneous syncope.
- Patients with positive and negative ATP test responses had similar syncopal recurrence rates and mechanisms of syncope.

Thus, among patients with recurrent neurocardiogenic syncope, the rhythm documented by ILR during spontaneous recurrent syncope did not correlate with the results of provocative tests. These findings suggest that provocative tests (tilt table and ATP tests) have little value in guiding specific therapy.

The safety and efficacy of early ILR use to guide therapy were evaluated in an observation study of 392 patients with recurrent suspected neurocardiogenic syncope [38] . Among 106 patients with a recurrent syncopal episode recorded by ILR, 53 had therapy guided by this information (pacemakers in more than 80 percent), while the remaining patients had no specific therapy primarily because an arrhythmic cause was not identified. Recurrence rates were low among those with ILR-guided therapy (10 versus 41 percent among those who did not have an indication for and therefore did not receive ILR-guided therapy).

The risks associated with a prolonged ILR were relatively low. Seven patients (2 percent) suffered major trauma from syncope recurrence during ILR monitoring, and four patients (1 percent) developed ILR pocket infections. These risks should be balanced against the risks of early provocative testing; these include both false positive results, which may lead to unnecessary therapies, as well as false negative results which may delay diagnosis. Limited data suggests that use of an ILR is cost-effective compared to conventional approaches. (See "Evaluation of syncope in adults", section on Cost for evaluation of syncope).

Treatment — Therapy is primarily aimed at patients with recurrent syncope. Acute vasovagal reactions are common in a number of stressful settings, such as blood donation. Patients who develop syncope or other symptoms associated with the neurocardiogenic reflex should assume the supine position with legs raised at the onset of such symptoms. (See "Procedures used for blood donor screening: protection of potential blood donors and recipients", section on Vasovagal reactions).

Many of the therapies proven effective for recurrent neurocardiogenic syncope are not intuitive, and some intuitively appealing therapies have not proven effective. Therapy is particularly important in patients who have syncope in high-risk settings (eg, commercial vehicle driver, pilot) who wish to continue these activities. Patients with recurrent episodes may require restriction of activities until therapy is shown to be effective.

General measures — General treatment measures for neurocardiogenic syncope include reassurance and education regarding the nature, risks, and prognosis of the condition (show table 5) [30] . The patient should be advised to assume the supine position with legs raised at the onset of symptoms. The patient should be advised to avoid trigger events when feasible and medications that may induce hypotension should be modified or discontinued.

A study of self-reported symptom burden in 418 patients diagnosed with neurocardiogenic syncope indicated that thirty five percent were symptom-free at median 5 year follow-up regardless of presenting symptom or treatment received [39] .

Other general therapy may include support stockings (in some cases, Jobst stockings), volume expansion by liberalizing salt intake and occasionally administration of the mineralocorticoid fludrocortisone (similar to the regimen used in the treatment of orthostatic hypotension). (See "Treatment of orthostatic and postprandial hypotension").

Beta blockers — Although beta blockers have been the most commonly used drug therapy for neurocardiogenic syncope, available evidence does not support their efficacy. They may even be harmful in younger patients.

They have been postulated to act upon the ill-defined afferent limb of the reflex arc involved in the Bezold-Jarisch reflex, and to potentially also inhibit the discharge frequency of the C fibers originating from the cardiac mechanoreceptors and chemoreceptors (show figure 1).

Initial observational data suggested a lower rate of recurrent syncope with beta blocker therapy [40-42] . However, at least four randomized trials have failed to show a benefit due in part to a large placebo effect [43-46] . Benefit of any therapy is difficult to demonstrate when the placebo effect is so high.

The best data come from the POST trial, which enrolled 208 patients with recurrent syncope and an abnormal tilt table test [45] . The patients were randomly assigned to treatment with placebo or metoprolol, titrated to 200 mg daily or the highest tolerated dose (average dose 122 mg daily). At one year, the following findings were noted:

- Recurrent syncope occurred in 36 percent of both groups.
- Withdrawal rates were 22 percent in both groups.
- Prespecified analyses according to age (categorized as <42 versus ≥ 42 years) and tilt table test results did not identify any subgroups that benefited with metoprolol.

Other medications — A variety of medications other than beta blockers have been used in the management of patients with neurocardiogenic syncope. This includes: serotonin reuptake inhibitors, anticholinergics (disopyramide, scopolamine), muscarinic blockers, theophylline, midodrine, fludrocortisone, clonidine, ACE inhibitors, desmopressin, erythropoietin, indomethicin and others supported by limited data with few controlled trials.

- Midodrine (5 mg three times daily), an alpha-1-adrenergic agonist, had a beneficial effect in a small randomized trial and a number of observational studies [47-50] . The benefits have ranged from prevention of syncopal episodes in 95 percent of previously untreated patients [48] to efficacy for up to 22 months in as many as 78 percent of patients who failed to respond to a beta blocker or other conventional therapy [49,50] . However, the efficacy of midodrine is uncertain, and another alpha agonist, etilefrine, was ineffective in a placebo-controlled study of 126 patients [51] .
- There has been interest in the use of selective serotonin uptake inhibitors, such as sertraline, fluoxetine, or paroxetine [46,52,53] . Their effect is at the level of the central nervous system, but it is unclear if it is at the afferent or efferent limb of the reflex arc. The potential clinical efficacy was illustrated in a randomized trial of paroxetine versus placebo in 68 consecutive patients with recurrent syncope and a positive upright tilt test in whom beta blockers, vagolytic agents, and mineralocorticoids were ineffective or poorly tolerated [52] . Paroxetine significantly increased the likelihood of a negative tilt test at one month (62 versus 38 percent) and reduced the rate of spontaneous syncope during follow-up (18 versus 53 percent).
- Disopyramide may be useful due to its negative inotropic (inhibition of myocardial mechanoreceptors) and anticholinergic properties [54] . However, despite apparent benefit in observational studies [54,55] , a small controlled trial showed that the rate of recurrent syncope at 29 months was similar with disopyramide and placebo (27 versus 30 percent) [56] .
- A preliminary report of seven patients refractory to all other medical therapies suggested benefit from methylphenidate [57] . This agent shares some properties with the amphetamines: it is a peripheral vasoconstrictor and stimulates the central nervous system.
- Theophylline [58] and angiotensin converting enzyme inhibitors [59] have been tried in small studies with some apparent success. Theophylline may be most effective in patients with a mixed response and associated fatigue during the episodes although there are no data to support this clinical observation.

Cardiac pacemakers — Although there is usually a significant bradycardic response in neurocardiogenic syncope, there has been uncertainty about the role of pacemakers because of the severe vasodepressor reactions often found in this disorder. This is true even for those patients who have asystole during a tilt table test [60] . (See "Indications for permanent cardiac pacing").

The potential benefit of pacing in this setting was suggested by three unblinded trials, two that compared pacing to placebo, the North American Vasovagal Pacemaker Study (VPS) and the Vasovagal Syncope International Study (VASIS), and one that compared pacing to a beta blocker, the Syncope Diagnosis and Treatment trial [61-63] . In contrast, two double-blinded trials (VPS II and SYNPACE) showed little or no improvement with pacing [64,65] . A meta-analysis of nine randomized trials, including those discussed here, showed no overall benefit from pacemaker implantation and suggested that in the unblinded trials, an "expectation effect" led to an overestimation of the benefits of pacing [66] .

- The VPS randomly assigned 54 patients to pacemaker insertion or no pacemaker; the patients had at least six episodes of syncope, a tilt table test that induced syncope or presyncope, and a relative bradycardia [61] . Dual chamber pacing was used. The pacemaker had both bradycardia support and rate-drop responsiveness (rate hysteresis); it was programmed to detect a small, rapid drop in heart rate through a prespecified range and then pace at relative high rate to provide chronotropic support during a time of presumed vasodilatation. The trial was terminated prematurely because of a significant reduction in the incidence of recurrent syncope with pacing (22 versus 70 percent for no pacemaker) and a significant increase in the time to first syncope (112 versus 54 days) (show figure 2).

- The Syncope Diagnosis and Treatment trial included 93 patients over the age of 35 who had three or more syncopal episodes in the preceding two years, and positive response to tilt table testing with syncope occurring in association with relative bradycardia; the patients were randomly assigned to DDD pacing with rate hysteresis or atenolol [63] . An interim analysis revealed that DDD pacing was more effective for preventing recurrent syncope (4 versus 26 percent at 135 days) (show figure 3).

- The VPS II trial included 100 patients with over five total episodes of syncope or over two episodes in two years, a positive tilt test, and age over 19 years [64] . The patients were randomly assigned to DDD pacing (with a rate-drop response algorithm) or no pacing; therapy was blinded since all patients received a pacemaker. Based upon an intention-to-treat analysis, there was a nonsignificant trend toward less frequent syncope in the paced patients (31 versus 40 percent at six months, risk reduction 30 percent). Complications included lead dislodgement or repositioning in seven patients, and one patient each with pericardial tamponade, vein thrombosis, and infection involving the pacemaker generator. These findings do not indicate a major benefit of pacing in this population.

- In the SYNPACE trial, 29 patients with recurrent tilt-induced vasovagal syncope and at least one syncopal relapse after head-up tilt testing underwent implantation of a DDD pacemaker with rate-drop response and were randomly assigned to have their pacemaker programmed ON or OFF [65] . The trial was terminated early after the results of the VPS II trial became available. At a median of two years of follow-up, there was no reduction in the frequency of syncope among patients in the ON compared to the OFF group (50 versus 38 percent).

Given the above evidence and clinical experience, pacing therapy is NOT considered first-line therapy for most patients with neurocardiogenic syncope [1,7] . In considering pacemaker therapy, it is important to document whether the predominant cause of symptoms is cardioinhibitory or vasodepressor, if possible. Pacing is more likely to be beneficial if the episodes are cardioinhibitory and unlikely to be beneficial if the primary problem is vasodepressor. However, it may be difficult to make this distinction, since the tilt table response does not always correlate with the response during clinical episodes. In evaluating the efficacy of pacing therapy, it must be confirmed that symptomatic benefit is clearly attributed to pacing, that the benefit is maintained long-term, and that there is a consensus regarding the definition of treatment success or failure [67] .

The following recommendations are included in the 2008 American College of Cardiology (ACC)/American Heart Association (AHA)/Heart Rhythm Society (HRS) device guidelines [7] :

- Permanent pacing may be considered for significantly symptomatic neurocardiogenic syncope associated with bradycardia documented spontaneously or at the time of tilt-table testing.

- Permanent pacing is NOT indicated for situational vasovagal syncope in which avoidance behavior is effective and preferred.

The 2004 ESC guidelines limit the recommendation for cardiac pacing for neurocardiogenic syncope to patients with cardioinhibitory vasovagal syncope with a frequency >5 attacks per year or there is severe physical injury or accident and age >40 [1] .

Although the 2008 ACC/AHA/HRS and 2004 ESC guidelines refer to use of tilt-table testing to guide therapy of neurocardiogenic therapy, implantable loop recorder capture of spontaneous episodes appears to be a more accurate means of identifying whether a cardioinhibitory mechanism is responsible. Therefore, an implantable loop recorder may help guide therapy (See "Implantable loop recorder" above).

Dual-chamber permanent pacing, especially with rate-drop response, should eliminate most, if not all, symptoms in patients with a pure cardioinhibitory response. When there is a sudden drop in heart rate within a defined range and duration, rate-drop response pacemakers pace at a faster rate (eg, 90 to 100 beats per min) for a prespecified period.

In the patient with a mixed response (significant cardioinhibitory and vasodepressor components), dual-chamber permanent pacing may blunt the vasodepressor-related symptoms.

Orthostatic training program — Some patients with neurocardiogenic syncope respond poorly to general measures. Orthostatic or tilt training may be an effective approach in this group [68,69] .

The efficacy of orthostatic training started in hospital and continued at home was suggested by a controlled but non-randomized study. Forty-seven patients with recurrent syncope and a positive upright tilt table test who were refractory to traditional therapies were assigned to a tilt-training program or to continued medical therapy, depending upon their consent [68] . The training program included five daily in-hospital upright tilt table studies increasing in duration from 10 minutes to 50 minutes. The training program was continued at home with the patient instructed to stand against a wall for up to 40 minutes twice a day, under supervision of a family member.

At a mean follow-up of 18 months, nearly all patients in the training group became tilt-negative (96 versus 26 percent of the control group). None of the trained patients had spontaneous recurrent syncope compared to 57 percent of controls during 15 to 23 months of follow-up.

However, four randomized controlled studies found that home orthostatic training in patients with syncope and positive tilt table tests did not reduce tilt-positive responses or spontaneous syncopal events [70-73] . In one of these studies, there was a decrease in recurrent syncope with training in the subset of patients with vasodepressor type syncope [73] .

Physical counterpressure — Counter-pressure maneuvers such as tensing the arms with clenched fists, leg pumping, and leg-crossing may abort a syncopal episode or at least delay it long enough that patients can assume the supine position [74] . Physical counterpressure maneuvers are intended to reduce lower extremity venous pooling and therefore improve cardiac output and prevent vasovagal syncope. Such maneuvers include:

- Leg crossing with simultaneous tensing of leg, abdominal, and buttock muscles.
- Handgrip, which consists of maximum grip on a rubber ball or similar object.
- Arm tensing, which involves gripping one hand with the other while simultaneously abducting both arms.

The potential efficacy of these maneuvers was evaluated in a randomized trial of 223 patients with recurrent vasovagal syncope and recognizable prodromal symptoms [75] . Patients were randomly assigned to lifestyle modification (eg, avoidance of triggers, increasing fluid and salt intake, lying down at the onset of prodromal symptoms), or lifestyle modification plus physical counterpressure maneuvers. Patients assigned to counterpressure were instructed on the three maneuvers described above, with biofeedback to monitor blood pressure response during instruction. They were then advised to use the maneuver of their choice at the onset of symptoms and maintain it as long as tolerated, or until symptoms resolved, and to move on to a second or third maneuver if necessary. Over a mean follow-up of 14 months, patients assigned to counterpressure maneuvers were significantly less likely to have recurrent syncope compared to those assigned to lifestyle modification alone (32 versus 51 percent).

Moderate exercise training — Limited uncontrolled data suggest that moderate exercise training may increase orthostatic tolerance in patients with syncope.

Driving restrictions for patients with neurocardiogenic syncope — Although neurocardiogenic syncope generally has a benign prognosis, a frequent concern is the potential for injury, particularly during certain activities such as driving. One study evaluated the risk of driving-related injuries in 155 patients with a history of syncope in whom hypotension and syncope or presyncope was provoked during a tilt table study; 34 percent had no warning before syncope [76] . After the diagnosis of neurocardiogenic syncope was established, six patients stopped driving voluntarily. All patients were treated pharmacologically. During a median follow-up of 22 months, 3.2 percent of patients had recurrent syncope but no patient experienced syncope or injury while driving.

Further insight was provided in a second study of 245 patients undergoing a tilt table study [77] . Nine percent had experienced at least one episode of syncope while driving. After treatment, syncope or presyncope recurred in six (67 percent) of these patients, and there was one syncope-related driving incident.

In cases where patients are counseled to abstain from driving because of concerns about recurrent syncope, adherence appears to be very low [78] .

Recommendations for driving in patients with neurally mediated syncope are included in the 1996 statement from the American Heart Association (AHA) and North American Society of Pacing and Electrophysiology (NASPE, now Heart Rhythm Society, HRS) on personal and public safety issues related to arrhythmias and were unchanged in the 2007 addendum [79,80] .

Driving recommendations in both the 1996 AHA/NASPE statement and the 2004 ESC syncope update are stratified according to whether syncope is categorized as "mild" or "severe":

- Mild neurally mediated syncope is characterized by infrequent mild symptoms (usually without syncope), occurring with warning and usually only with standing, with clear precipitating causes, and infrequent occurrence.

- Severe neurally mediated syncope is characterized by severe symptoms (usually syncope), occurring without warning and in any position, with no clear precipitating causes, and/or frequent occurrence.

In the ESC guidelines, syncope during "high risk" activity (eg, driving, machine operator, flying, competitive athletics) is also considered severe.

The 1996 AHA/NASPE statement recommends no driving restrictions for private drivers with mild syncope. For commercial drivers with mild syncope driving is permitted after arrhythmia control is documented for 1 month.

For private or commercial drivers with untreated severe syncope, driving is prohibited. For private drivers treated for severe syncope, driving is permitted after arrhythmia control is documented for 3 months with adequate pacemaker follow-up as indicated. For commercial drivers treated for severe syncope, driving is permitted after arrhythmia control is documented for 6 months with adequate pacemaker follow-up as indicated.

Driving recommendations in the 2004 ESC syncope guidelines are displayed in the table (show table 6).

Legal requirements for physicians to report patients with conditions that could impair safe motor vehicle operation vary by state.

SITUATIONAL SYNCOPE — Situational syncope refers to syncope associated with specific scenarios.

Post-micturition syncope is a form of situational neurocardiogenic syncope that accounts for up to 5 percent of cases of syncope [9,81,82] . The cause for this type of syncope is probably related to an abrupt change in position combined with a strong vagal stimulus (show figure 3). Initial studies suggested a clear male predominance in this condition; however, one report found a higher incidence of syncope after evening micturition in women [83] .

Several forms of situational syncope are associated with gastrointestinal stimuli (swallowing, defecation, visceral pain).

Pathogenesis of situational syncope — Some of these situations (eg, post-micturition, coughing) appear to trigger a neural reflex; others (eg, straining, squatting) may cause syncope via mechanisms unrelated to neural reflexes. However, treatment strategies for these conditions are similar.

Diagnosis — Identification of situational syncope is generally by clinical history. Patients with situational syncope often have positive responses to carotid sinus massage and/or tilt table testing [84] , but the utility of such responses in guiding therapy has not been determined.

Treatment — Treatment of most forms of situational syncope is based upon avoiding or ameliorating the triggering activity [1] . When the activity cannot be avoided, general treatment measures include maintenance of intravascular volume, protected posture (eg, sitting rather than standing) and slow postural changes. Specific measures may be helpful for certain conditions such as use of stool softeners in those with defecation syncope, avoidance of excessive fluid intake (especially alcohol) prior to bedtime in post-micturition syncope, and avoidance of large gulps of cold drinks or boluses of food in swallow syncope.

Driving restrictions for patients with situational syncope — Recommendations for driving in patients with neurocardiogenic syncope but not situational syncope are included in the 1996 AHA/NASPE statement or in the 2007 addendum [79,80] .

Driving recommendations in the 2004 ESC syncope guideline update (as well as the 1996 AHA/NASPE statement) are stratified according to whether syncope is categorized as "mild" or "severe". (See "Driving restrictions for patients with neurocardiogenic syncope" above) (show table 6).

- If situational syncope is single/mild, no restriction is recommended for private drivers and no restriction is recommended for vocational drivers "unless syncope occurred during high risk activity." (However, the syncope would be classified as severe in that case). .

- If situational syncope is severe, driving restriction is recommended for private drivers until appropriate therapy is established and for commercial drivers until effective treatment has been established.

SUMMARY AND RECOMMENDATIONS — Diagnosis of neurocardiogenic syncope and situational syncope is suggested by history in patients who have classic triggers and premonitory symptoms. In other patients, the cause of syncope may be more difficult to identify.

- Neurocardiogenic (vasovagal) syncope is caused by cardioinhibitory (bradycardia or asystole) and/or vasodepressor mechanisms.

- Clinical features typical of neurocardiogenic syncope may be diagnostic. In such cases, further testing is indicated only when it will impact therapy.

- Provocative testing (such as upright tilt table testing, adenosine administration or adenosine triphosphate infusion) may help identify individuals with neurocardiogenic syncope but these tests have limited sensitivity and specificity. In addition, hemodynamic responses to provocative testing may not correlate with spontaneous syncopal episodes. (See "Diagnosis" above See "Diagnosis" above and see "Upright tilt table testing in the evaluation of syncope").

- Cardiac rhythms during spontaneous syncopal episodes can be captured by implantable loop recorder so that the presence or absence of a cardioinhibitory mechanism during spontaneous syncope can be accurately identified. However, acquisition of diagnostic recordings may take months if episodes are infrequent. (See "Implantable loop recorder" above).

- Therapy for neurocardiogenic and situational syncope is generally reserved for patients who have had recurrent episodes of syncope (show table 5).

- The initial approach is focused upon lifestyle modification and avoidance of medication or other triggers (see "General measures" above).

- Physical counterpressure techniques are suggested. (**Grade 2B**) (see "Treatment" above see "Treatment" above).

- Patients with recurrent syncope with a neurocardiogenic (vasovagal) bradycardic or asystolic syncopal episode documented by electrocardiographic monitoring (eg, implantable loop recorder) are considered to have neurocardiogenic syncope. We suggest permanent cardiac pacing in such patients, particularly if the frequency of attacks is >5 per year or there is severe physical injury or accident and age >40 (**Grade 2C**). In such patients dual-chamber permanent pacing with rate-drop response is recommended (see "Cardiac pacemakers" above).

- Among patients who continue to have recurrent syncope, the efficacy of further therapy is limited. (See "Treatment" above See "Treatment" above).

- In patients diagnosed with recurrent neurocardiogenic syncope, we do NOT recommend beta blocker treatment (**Grade 1A**) (see "Beta blockers" above).

- Recommendations for driving restrictions generally depend on the severity of the syncopal episodes. (See "Driving restrictions for patients with situational syncope" above).

Causes of syncope

Neurally-mediated (reflex)
Vasovagal syncope (common faint)
- Classical
- Non-classical
Carotid sinus syncope
Situational syncope
- Acute hemorrhage
- Cough, sneeze
- Gastrointestinal stimulation (swallow, defecation, visceral pain)
- Micturition (post-micturition)
- Post-exercise
- Post-prandial
- Others (eg, brass instrument playing, weightlifting)
Glossopharyngeal neuralgia
Orthostatic hypotension
Autonomic failure
- Primary autonomic failure syndromes (eg, pure autonomic failure, multiple system atrophy, Parkinson's disease with autonomic failure)
- Secondary autonomic failure syndromes (eg, diabetic neuropathy, amyloid neuropathy)
- Post-exercise
- Post-prandial
Drug (and alcohol)-induced orthostatic syncope
Volume depletion
- Hemorrhage, diarrhea, Addison's disease
Cardiac arrhythmias as primary cause
Sinus node dysfunction (including bradycardia/tachycardia syndrome)
Atrioventricular conduction system disease
Paroxysmal supraventricular and ventricular tachycardias
Inherited syndromes (eg, long QT syndrome, Brugada syndrome)
Implanted device (pacemaker, ICD) malfunction

Drug-induced proarrhythmias
Structural cardiac or cardiopulmonary disease
Cardiac valvular disease
Acute myocardial infarction/ischemia
Obstructive cardiomyopathy
Atrial myxoma
Acute aortic dissection
Pericardial disease/tamponade
Pulmonary embolus/pulmonary hypertension
Cerebrovascular
Vascular steal syndromes

Reproduced with permission from: Brignole, M, Alboni, P, Benditt, DG, et al. Guidelines on management (diagnosis and treatment) of syncope-update 2004. Europace 2004; 6:467. Copyright ©2004 Oxford University Press.