Cardiopathies acquises

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Plan du cours

- Maladie de Kawasaki
- Endocardite infectieuse
- Myocardite

Maladie de Kawasaki

Kawasaki – Les points clés

- Maladie décrite en 1967 – seulement 50 ans de recul !
- **Vascularite systémique** qui touche essentiellement **les artères de moyen calibre** avec un tropisme électif pour **les artères coronaires** (gravité de la maladie)
- Les complications coronaires surviennent dans **15 à 25 %** des cas chez les enfants non traités
- L'administration précoce d'immunoglobulines humaines par voie intraveineuse a transformé le pronostic **en diminuant par 5** le risque d'anévrisme coronaire
- **Risque de mortalité 0,015 %** (Japon), surtout entre le 15-45eme jour (thrombocytose concomittante avec la vascularite)

Epidémiologie

- **Première cause de cardiopathie acquise de l'enfant dans les pays développés**

- **Tous les âges pédiatriques (80 % des cas avant 5 ans)**
- Les patients de **moins de 1 an** ou **de plus de 8 ans** sont rares mais ont un risque plus élevé d'anévrisme coronaire

- **Formes de l'adulte:** première fois décrite en 1977
- symptômes majeurs décrits identiques

- **Kawasaki atteint chaque année:**
  - 265/100.00 enfants < 5 ans au Japon
  - extrapolation française: 600 nouveaux cas par an en France

Newburger JACC 2016

Caractéristiques cliniques: critères majeurs

La fièvre de plus de 5 jours et au moins 4 critères suivants

- La **conjonctivite** bulbaire non purulente

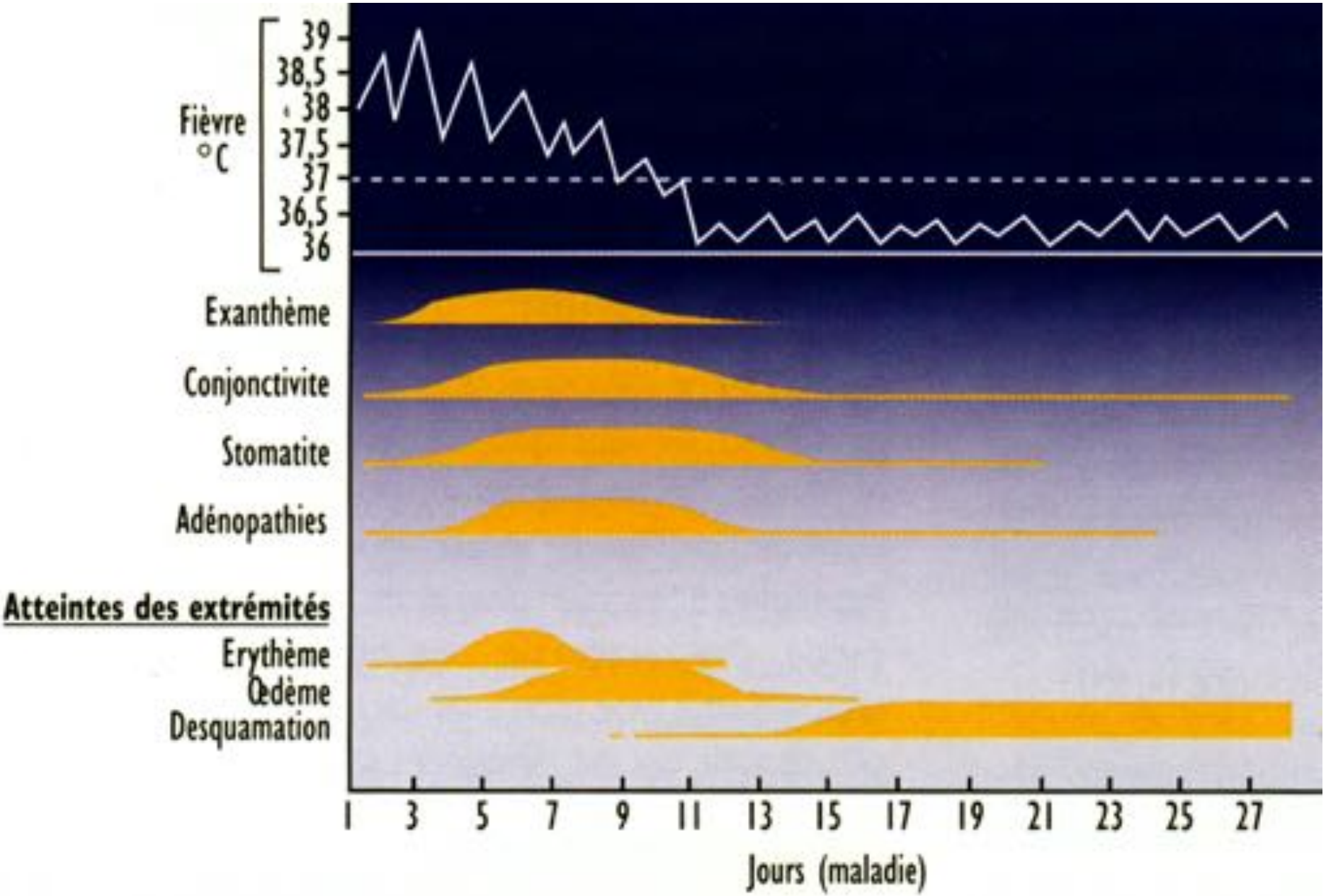
- **L'atteinte muqueuse** : la pharyngite, la chéilite, la langue framboisée, la stomatite

- **L'exanthème polymorphe du tronc**

- **L'atteinte des extrémités** : un érythème des paumes des mains et/ou des plantes des pieds, l'œdème palmo-plantaire, la desquamation palmo-plantaire secondaire

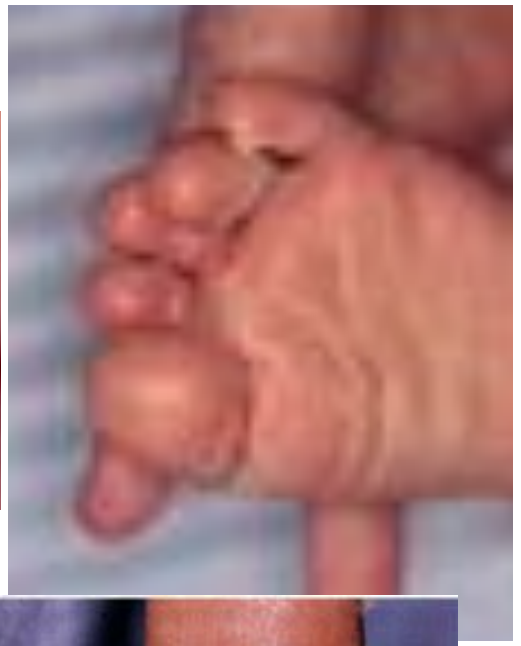
- **L'atteinte unilatérale des ganglions cervicaux**, de plus de 1.5 cm de diamètre

Critères majeurs définis par l'American Heart Association Committee On Rheumatic Fever, Endocarditis, and Kawasaki Disease


Fièvre °C
39
38,5
38
37,5
37
36,5
36
Exanthème
Conjonctivite
Stomatite
Adénopathies
Atteintes des extrémités
Erythème
Œdème
Desquamation
1
3
5
7
9
11
13
15
17
19
21
23
25
27
Jours (maladie)

Faciès typique d'un enfant avec une maladie de Kawasaki

Atteinte muqueuse et desquamation palmo-plantaire



Formes rares: forme psoriasiforme/ BCGite

Atteinte cardiovasculaire

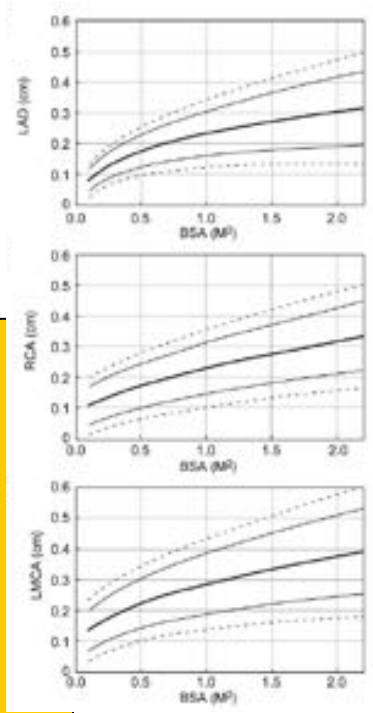
- **Pas d'atteinte coronaire dans 75% des cas !**
- **Anomalie ECG ou échocardiographique:**
  - **Dilatation des artères coronaires (20%)**
  - Anévrismes coronaires
  - Infarctus
  - Myocardite avec possible insuffisance ventriculaire gauche sévère
  - Péricardite, épanchement péricardique
  - Fuites valvulaires par inflammation des valves cardiaques et particulièrement la valve mitrale (1 %)
  - Troubles conductifs et troubles du rythme par inflammation du tissu de conduction

Les anévrismes coronaires

- **Entre le 10ème et le 25ème jour d'évolution**
- 5 % lorsque le traitement est fait précocement

  = < 10 jours du début des symptômes
- Souvent multiples et siègent habituellement dans la partie proximale des artères coronaires

- **Pronostic cardiaque dépend essentiellement de leur taille.**
- Nouveauté 2016: plus la taille en mm mais le Z score !



<u>Selon les guideslines américaines:</u>

Dilatation coronaire	**Z score > 2 < 2,5**
Petit anévrysme coronaire	**Z score > 2,5 <5**
Anévrysme coronaire moyen	**Z score 5-10**
Anévrysme géant	**Z score > 10 ou > 8 mm**

Newburger JACC 2016

Formes atypiques

Tableau clinique dominé par un symptôme inhabituel:

« convulsions, œdème pulmonaire, diarrhée sanglante, ascite, obstruction des voies aériennes supérieures, épiglottite, adénopathies cervicales compressives ou hémolyse et défaillance multi-viscérale, syndrome néphrotique, hyponatrémie…. »

Formes de l'adulte

- Troubles digestifs, atteinte hépatique, signes articulaires et encéphalites sont plus fréquent

Les formes incomplètes

Patients ayant eu une fièvre depuis **au moins 5 jours** et
au **moins deux critères cliniques de Kawasaki**, sans cause évidente,
et des critères biologiques en faveur d'une inflammation systémique

- **Différent de la « forme atypique »**
- Manque **un** ou **plusieurs des cinq critères** diagnostiques majeurs
- **Plus fréquentes chez les enfants les plus jeunes, à risque d'anomalies coronaires**

Diagramme décisionnel proposé par l'American Academy of Paediatrics pour aider à la prescription d'IgG dans les formes incomplètes

Newburger JW et al., Circulation. 2004

Critères cliniques et biologiques supplémentaires

Cardiovasculaires : dilatation des artères coronaires, anévrismes coronaires, infarctus, myocardite avec possible insuffisance cardiaque congestive, péricardite, épanchement péricardique, fuites valvulaires, troubles conductifs et troubles du rythme, anévrismes des vaisseaux du cou, des artères rénales, spléniques, hépatiques, pancréatiques, génitales, gangrènes distales et pseudo-Raynaud

Digestifs : diarrhées, vomissements, douleurs abdominales, hydrocholécyste, dysfonction hépatique

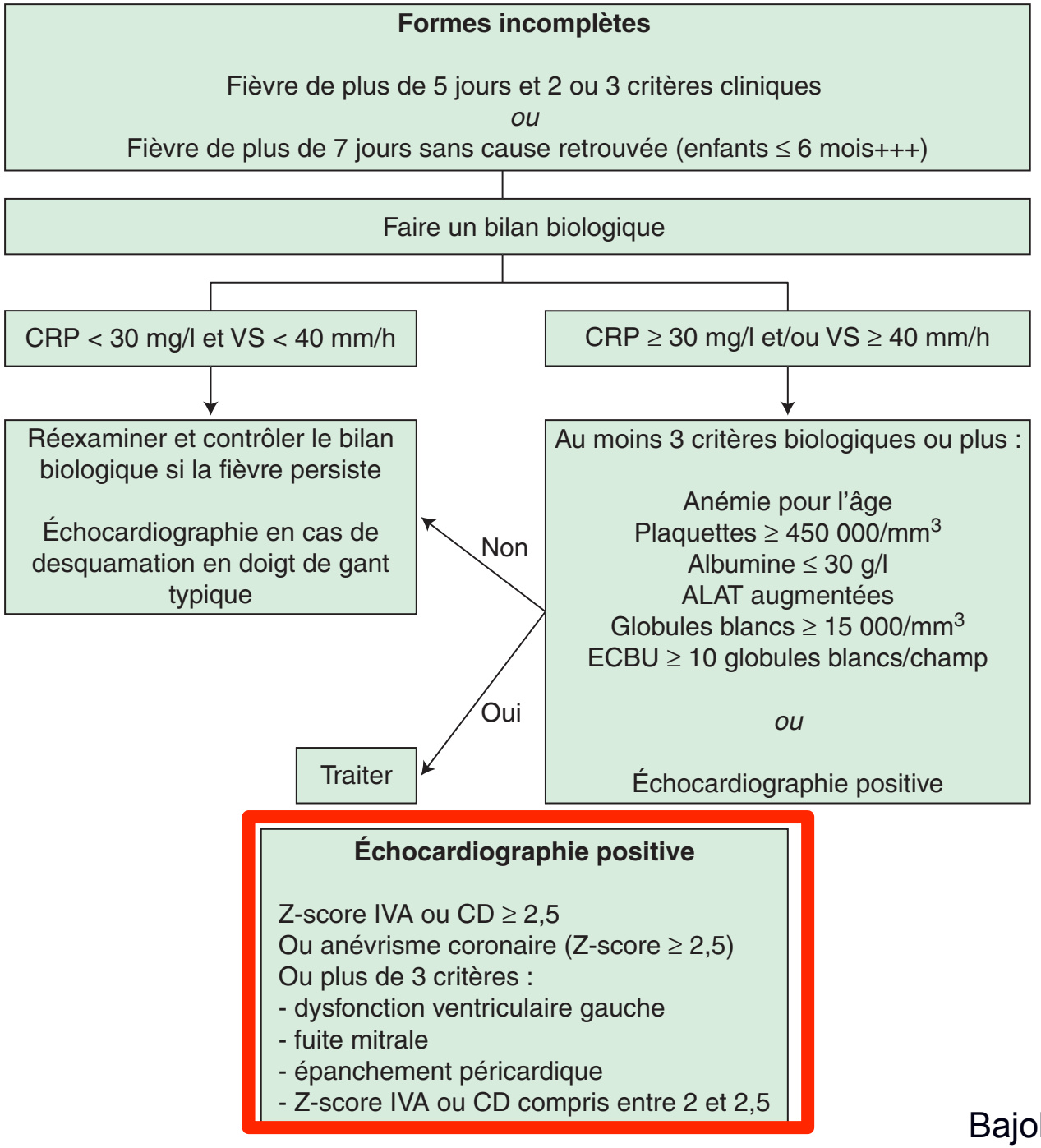
Respiratoires : toux et rhinorrhée

Neuro-méningés : troubles de la conscience avec irritabilité, apathie, état grognon, hypoacousie

Articulaires : arthrite, arthralgies
Autres : uvéite, érythème au niveau de la cicatrice de BCG, desquamation de l'aine

Albumine < ou égal 3g/dl
Anémie pour l'âge
Plaquettes ≥ à 450 000/ mm^3 à J7
Globules blancs ≥ à 15 000 / mm^3
ECBU ≥ à 10 globules blancs/ champ

D'après Newburger JW et al., Circulation. 2004



Bajolle et al. 2018 EMC

Evolution naturelle

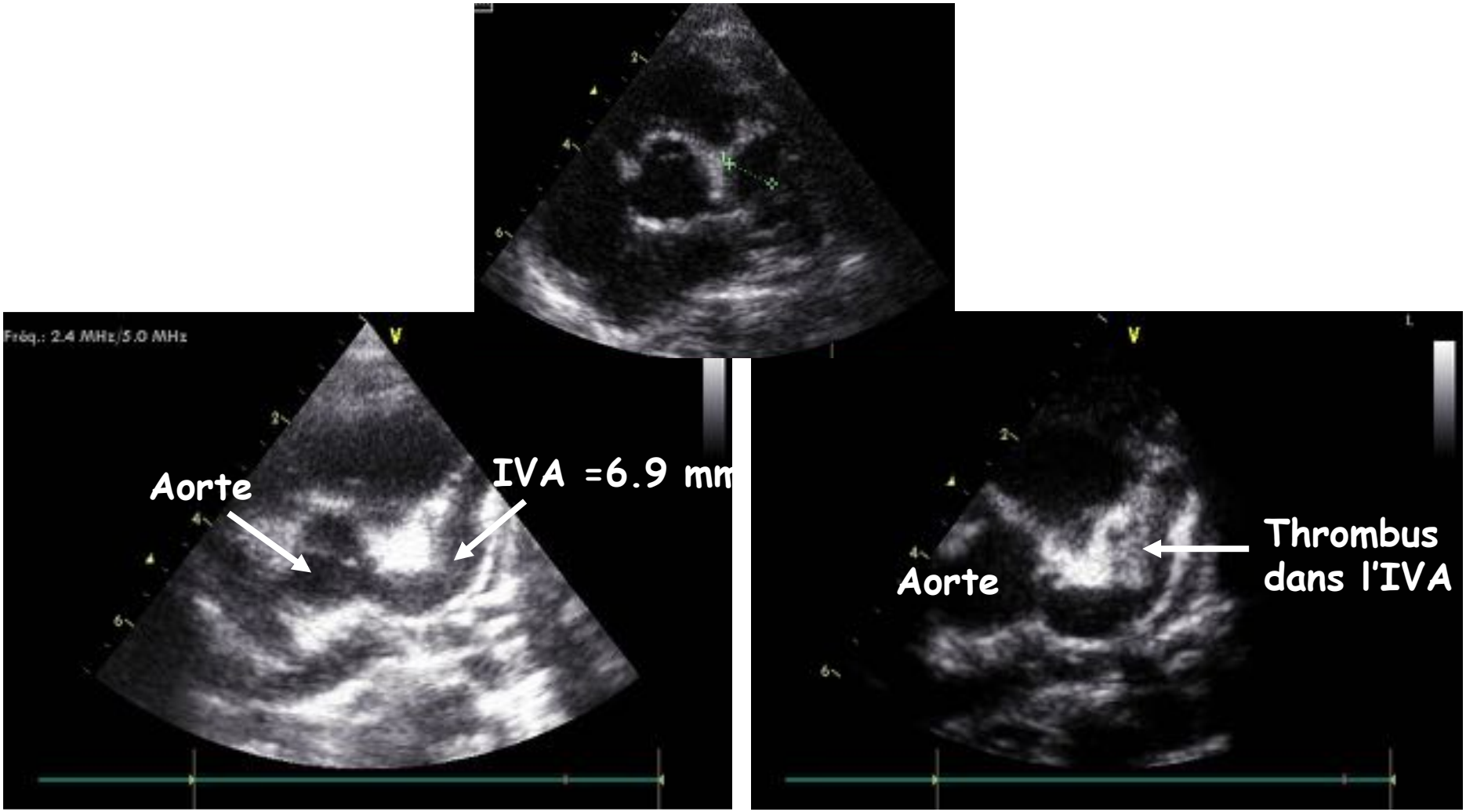
1°) La phase aiguë (J0-J10) : **atteinte cardiaque rare**

2°) La phase subaiguë (J10-J20) : **diagnostic de complication coronaire**

3°) La phase de convalescence (J20-J70) : **constatation d'anévrysmes et de sténoses cicatricielles** en cas de complication coronaire à la deuxième phase

Exemples d’atteinte coronaires

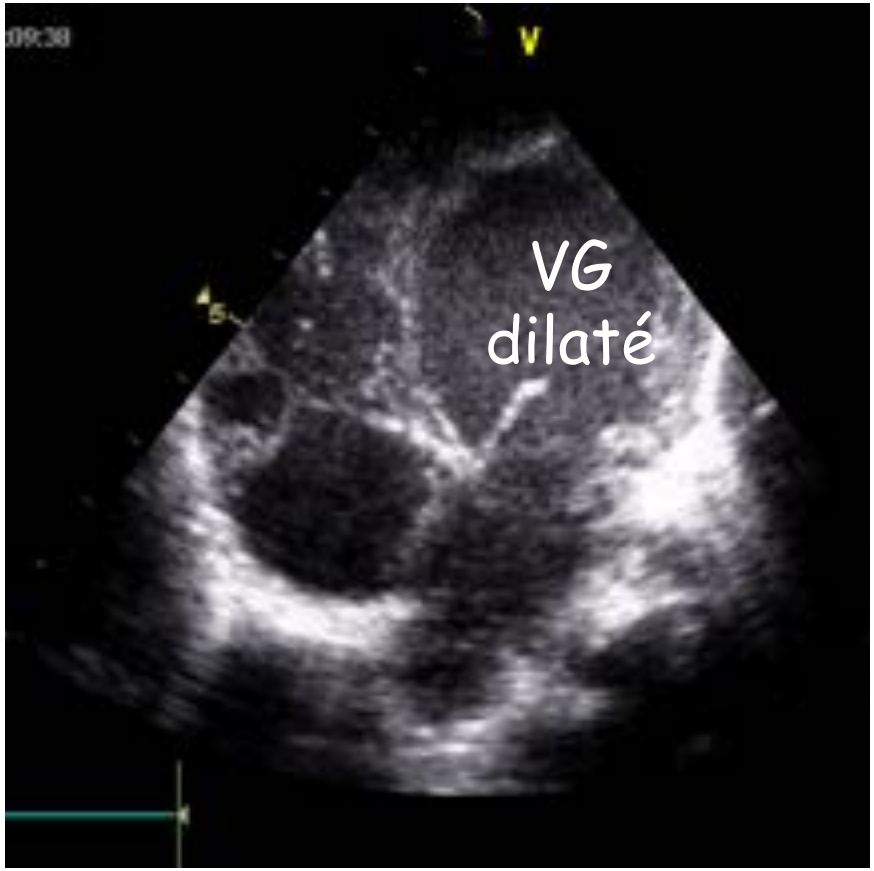
Dilatation anévrysmale des artères coronaires

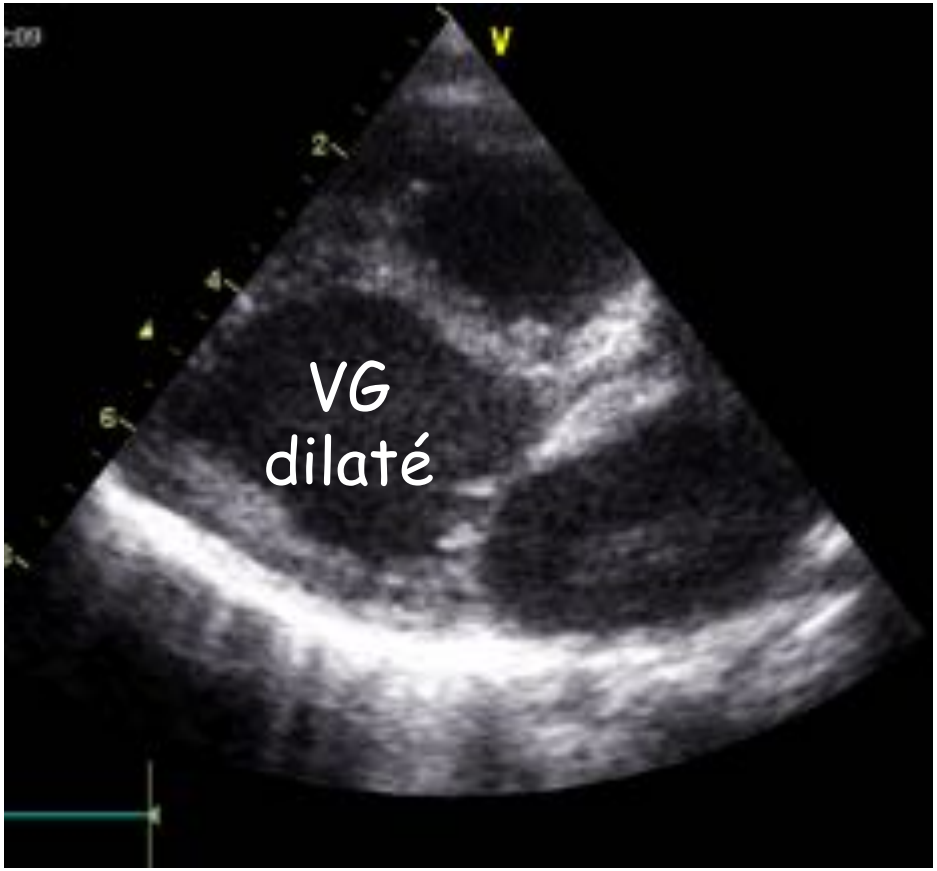


(A) IVA=interventriculaire antérieure
(B) thrombus dans l'IVA

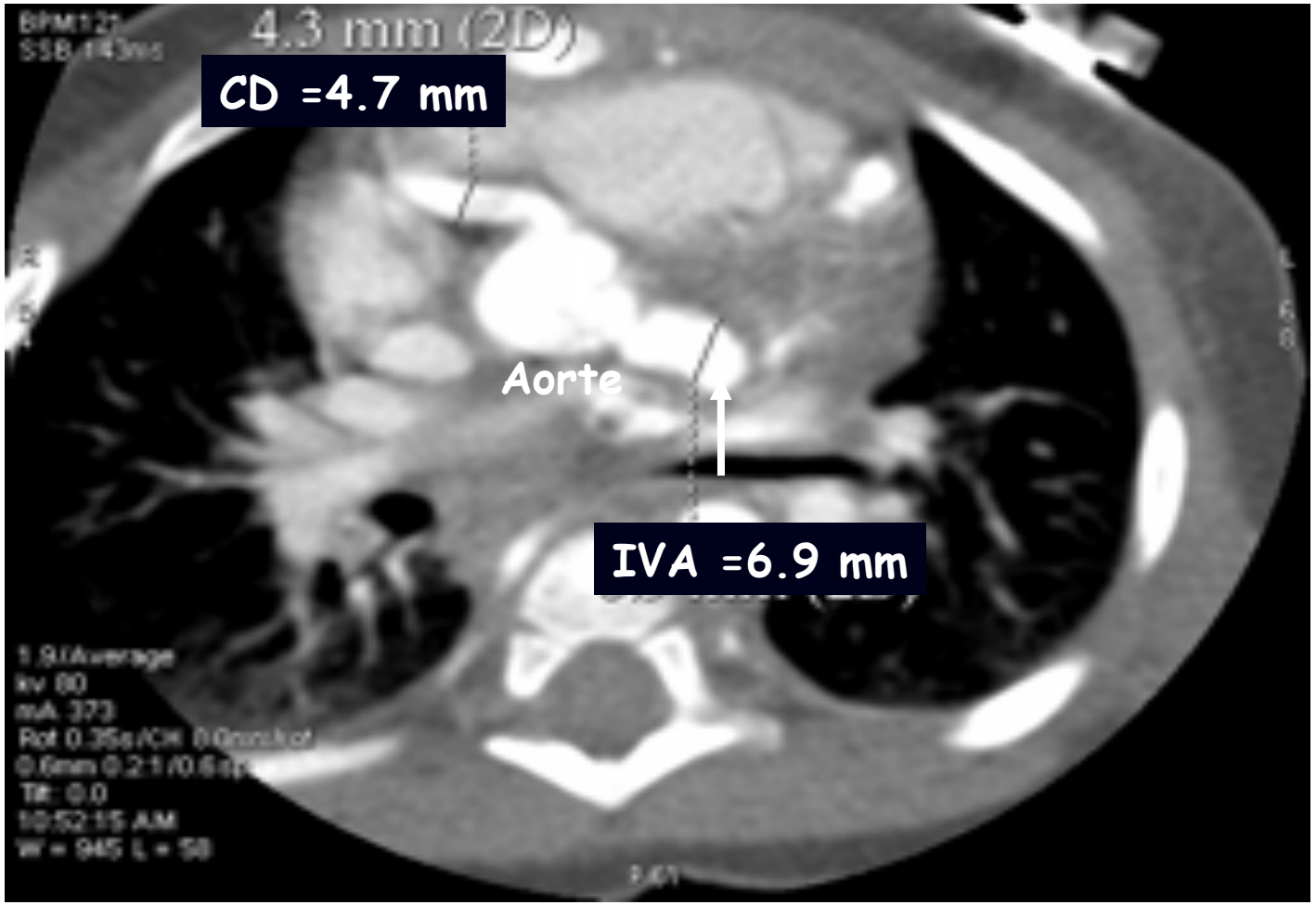
Dysfonction ventriculaire gauche sévère en échographie

Etat de choc dans 7% des cas de maladie de Kawasaki

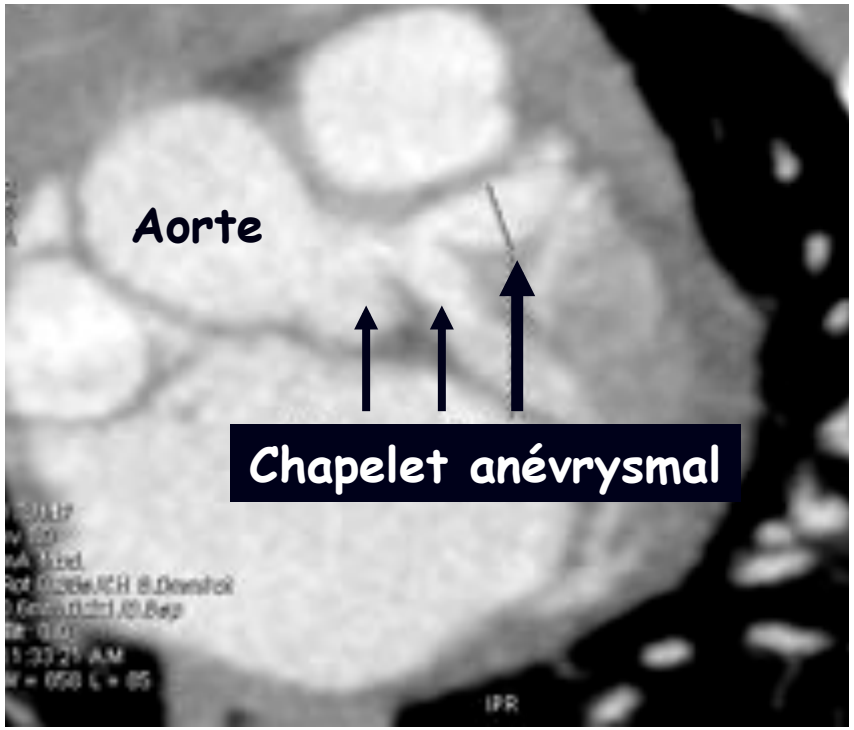


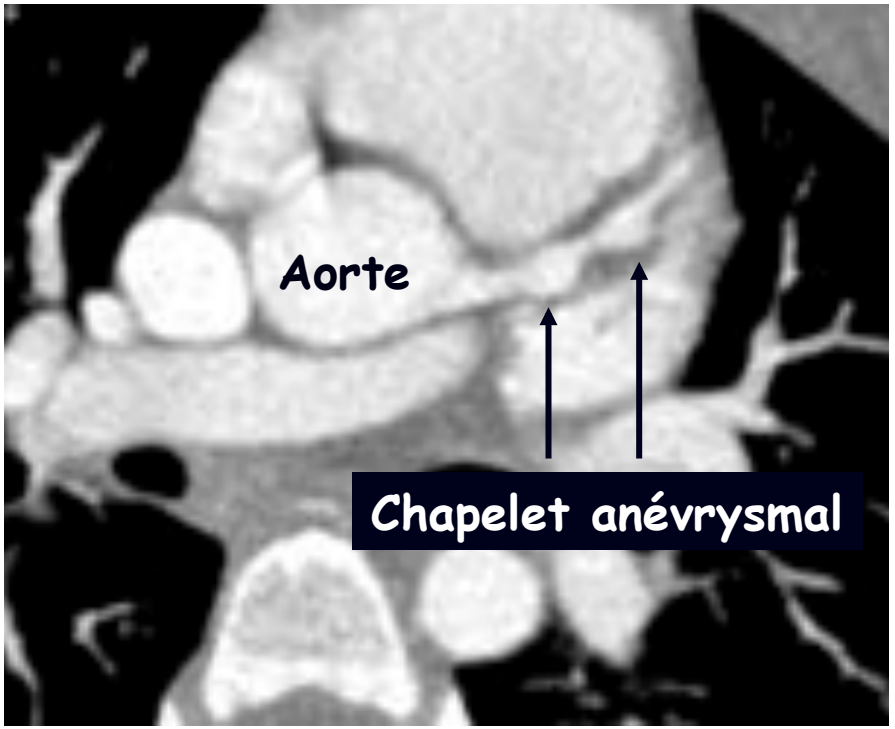


Dilatation anévrysmale des artères coronaires au scanner

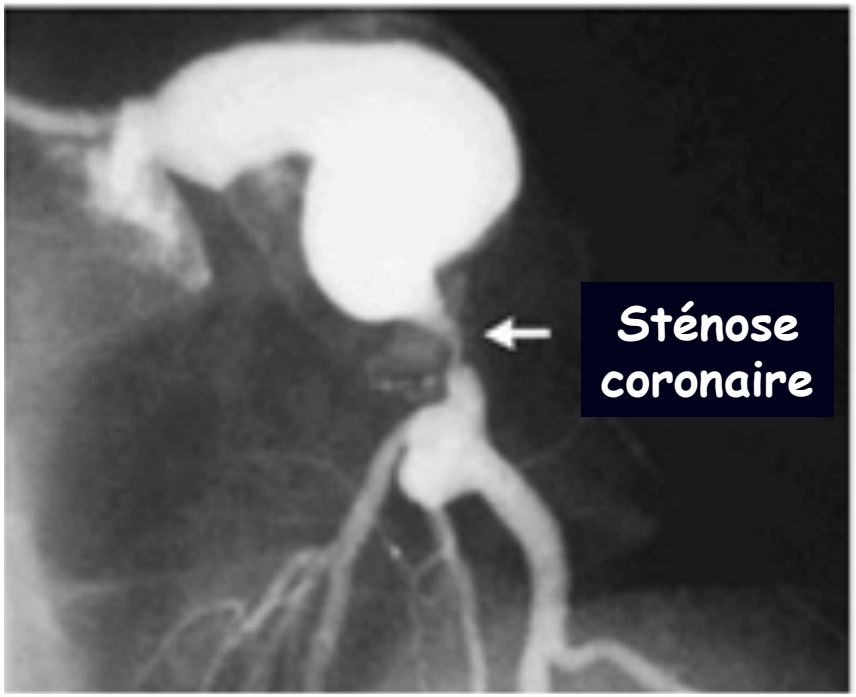


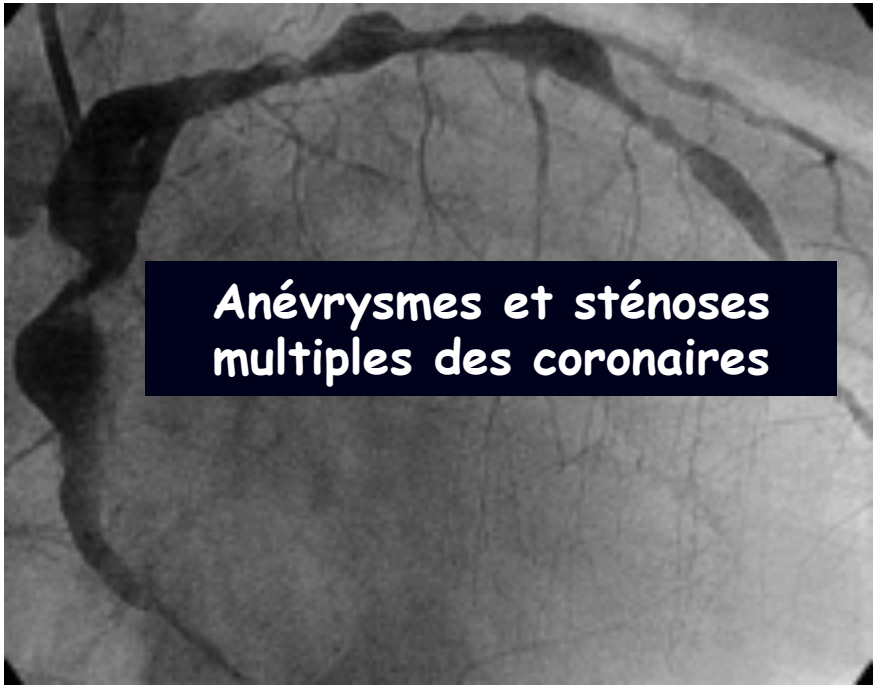
Dilatation anévrysmale de l'IVA en chapelet au scanner



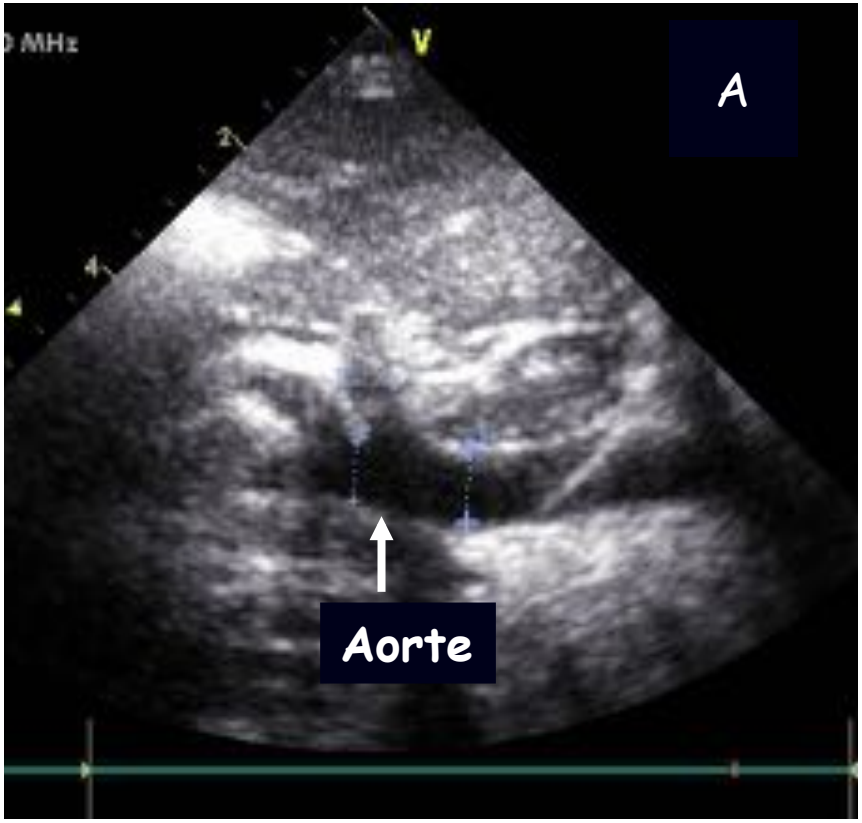


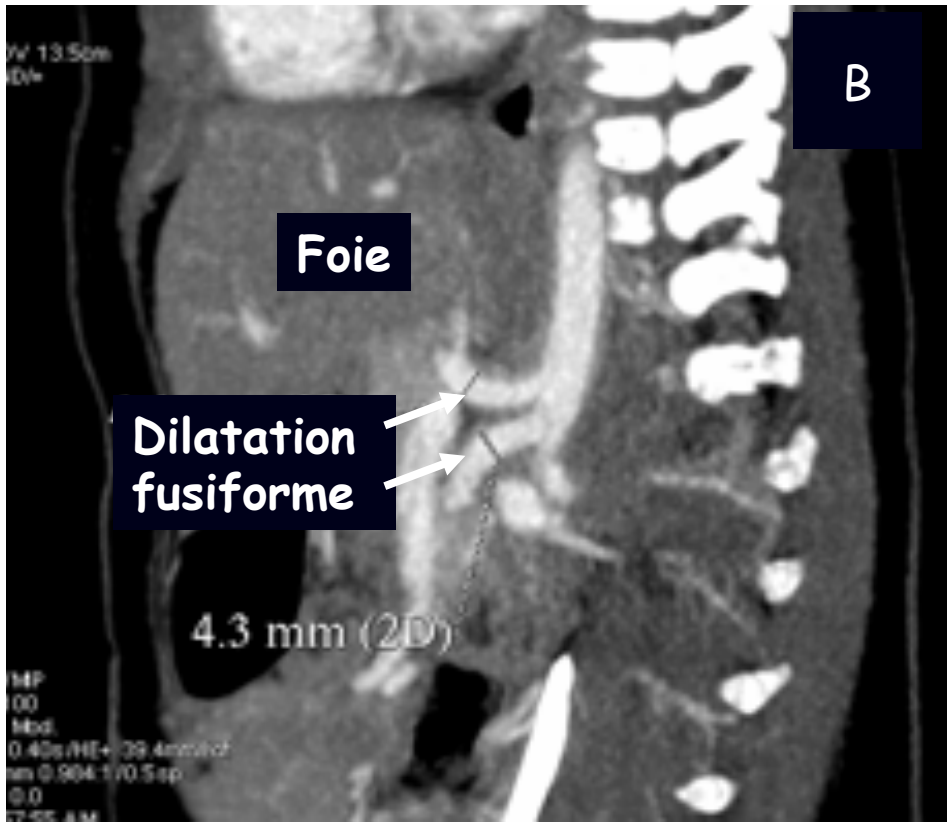
Anévrysme coronaire avec sténose coronaire au cathétérisme cardiaque





Atteinte diffuse des axes vasculaires





(A) épaississement pariétal hyperéchogène de l'aorte, de l'artère mésentérique supérieure et du tronc coeliaque en échographie

(B) dilatation fusiforme de l'artère mésentérique supérieure, du tronc caeliaque au scanner

Le traitement de 1ère intention

Immunoglobulines intraveineuses:

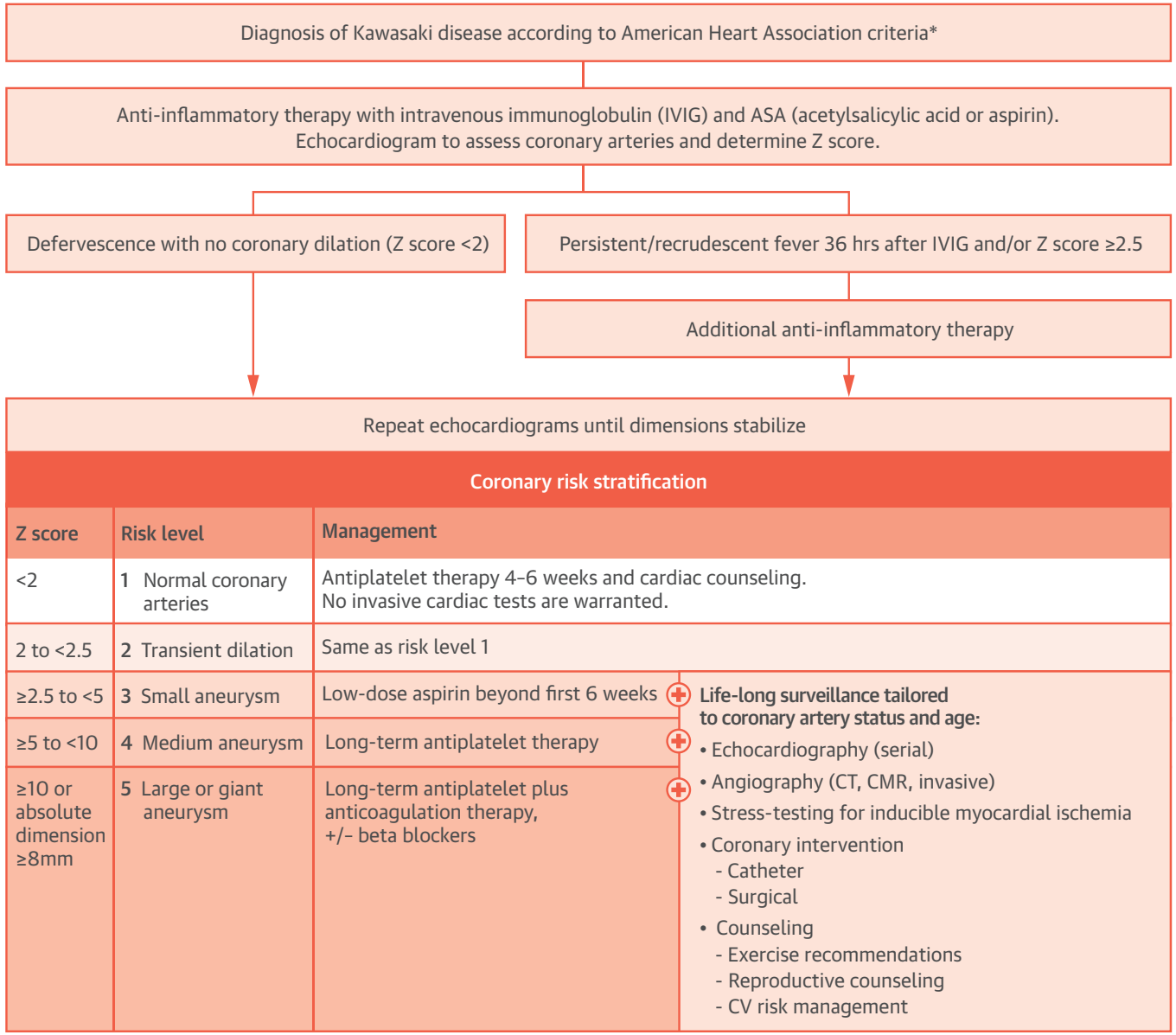
- 2g/kg en 8 à 12 heures, à posologie progressive

En association à de **l'acide acétylsalicylique (AAS) :**

- à fortes doses pour ses effets anti-inflammatoires et anti-thrombotiques **(30-50 mg/kg/j en 4 fois)** à la phase aiguë
- puis à **dose anti-aggrégante plaquettaire** rapidement 48-72 h après l'apyrexie (3-5 mg/kg/jour)

De Graeff et al 2019, Newburger 2016

CENTRAL ILLUSTRATION Management of Kawasaki Disease



Newburger, J.W. et al. J Am Coll Cardiol. 2016;67(14):1738–49.

Recommandations américaines

TABLE 1 Principles in Acute Management of KD

1. The goal of therapy is to reduce systemic and tissue-level inflammation as rapidly as possible. For this reason, patients should be treated as soon as diagnosis can be confidently established.
2. All patients within the first 10 days of fever onset should be treated with IVIG. Patients diagnosed after 10 days should receive IVIG treatment if they are still febrile, have markedly elevated inflammatory parameters, or have coronary artery dilation.
3. Recrudescent fever at least 36 h after the end of IVIG infusion without other explanation is a marker for persistent inflammation and should prompt immediate and aggressive anti-inflammatory therapy
   a. Antibody-mediated hemolysis has become common in KD patients who have received IVIG retreatment and have type A or B blood; rescue therapies other than IVIG (e.g., infliximab, corticosteroids) should be considered.
4. Patients with coronary artery dilation (*z*-score >2.0) should be followed with a repeat echocardiogram at least twice a week until dimensions stabilize; additional anti-inflammatory therapy should be considered.
5. Patients with giant aneurysms should have frequent echocardiograms in the first 3 months of illness for thrombus surveillance, even after dimensions stabilize.
6. Infants under 6 months of age are at extremely high risk of aneurysm formation, even with timely therapy. They require echocardiograms every few days until dimensions have stabilized.
7. Patients with giant CAA (*z*-score ≥10) are at highest risk for thrombosis during the first 3 months after fever onset
   a. Systemic anticoagulation together with an antiplatelet agent should be administered until coronary dimensions improve.
   b. Low-molecular-weight heparin is easier to regulate than warfarin in infants, as well as in patients of any age, during the acute phase of illness or until hsCRP normalizes.

CAA = coronary artery aneurysm; hsCRP = high-sensitivity C-reactive protein; IVIG = intravenous immunoglobulin; KD = Kawasaki disease.

Newburger JACC 2016

Surveillance à court terme

Pour les patients sans complication coronaire:

- 1 echo entre 1-2 semaines et 1 echo entre 4-6 semaines (Classe 1)

Pour les patients avec Z score coronaire > 2,0 à la phase aigue:

- 2 echos par semaines jusqu'à l'arret de la progression

Pour les patients avec anévrismes géants:

- 2 échos par semaine tant que les lésions progressent
- 1 écho/sem pdt 45 jours puis
- 1 écho/mois pendant 3 mois (Casse IIA)

Newburger JACC 2016

Antiagrégation et anticoagulation

Patients sans atteinte coronaire:

- Aspirine pendant 6 semaines (Cl 1)

Patients avec atteinte coronaire d'aggravation rapide:

- Hospitalisation pour mise sous heparine (AntiXa 0,5-1)
- Arret si stabilisation; Z score < 10 ou 8 mm
- Aspirine 12 mois

Patients avec anévrysmes géants:

- Hospitalisation pour Heparine et relais AVK (INR 2-3)
- Aspirine à vie

Que faire en cas de persistance de la fièvre après une première cure d'IgIV?

- **Résistants:** 15 à 20 % des cas
- Associée à un risque plus élevé d'atteinte coronaire

- Deuxième dose d'IVG
- associée à un bolus de corticoides (20-30 mg/kg/jour IV methylprednisolone)

- **Discuter:** prednisolone p.o. plus prolongé 2-3 semaines
- Alternative: infliximab
- En cas d'échec: ciclosporine

Newburger JW et al., Circulation. 2004

Egami K et al., J Pediatr 2006

Comment identifier les résistants ?

Score d'Egami (2006)	Score de Kobayashi (2006)	Score de Sano (2007)
Age ≤ 6 mois (2 points)	Age ≤ 12 mois (1 point)	Bilirubine totale ≥ 0.9mg/dL (1 point)
≤ 4 jours de fièvre (1 point)	Traitement dans les 4 premiers jours de fièvre (2 points)	CRP ≥ 7mg/dL (1 point)
Plaquettes ≤ 300.10^9/L (1 point)	Plaquettes ≤ 300.10^9/L (1 point)	ASAT ≥ 200 U/L (1 point)
CRP ≥ 8mg/dL(1 point)	CRP ≥ 10mg/dL (1 point)	
ALAT > 100 U/L (1 point)	ASAT ≥ 100 U/L (1 point)	
	≥ 80% neutrophiles (2 points)	
	Na+ ≤ 133 mmol/L (2 points)	
Haut risque si ≥ 3 points	Haut risque si ≥ 5 points	Haut risque si ≥ 2 points

Scores issus de la population japonaise, spécifique mais non sensible

European consensus-based recommendations for the diagnosis and treatment of Kawasaki disease – the SHARE initiative

Rheumatology 2019;58:672–682

8. Corticosteroid treatment should be given to patients with severe KD[a]:	1A[b]
(a) Who are IVIG resistant, that is, with ongoing fever and/or persistent inflammation or clinical signs ⩾48 h after receiving IVIG as a single dose of 2 g/kg. A second dose of IVIG is at the discretion of the treating physician.	1A 3 4
(b) Kobayashi score ⩾5 (see Supplementary Table S5, available at *Rheumatology* online)	4
(c) With features of HLH	4
(d) With features of shock	4
(e) Who are under the age of 1 year	
(f) Who present with coronary and/or peripheral aneurysms	
9. If corticosteroids are indicated, the following regimens would be reasonable:	2A

Regimen 1: methylprednisolone 0.8 mg/kg BD i.v. for 5–7 days or until CRP normalizes; then convert to oral prednisone/prednisolone 2 mg/kg/day and wean off over next 2–3 weeks.
Regimen 2: methylprednisolone 10–30 mg/kg (up to maximum of 1g/day) once daily for 3 days followed by oral prednisone/prednisolone 2 mg/kg per day until day 7 or until CRP normalizes; then wean over next 2–3 weeks.

Nouveauté 2019: corticoides IV puis p.o. en phase aigue pour les patients à risque élévé d'emblée ou avec résistance

Quelle est l'histoire naturelle des complications coronaires de la MK?

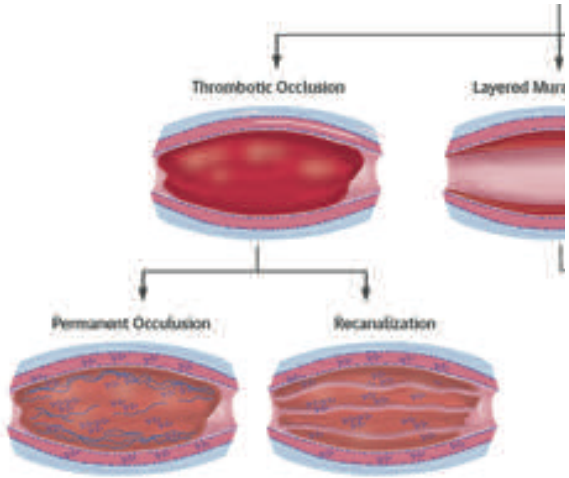
- Disparition complète dans plus de 50% des cas même en cas d'anévrysme (sauf géant) dans les 2 ans

- Occlusion coronaire; sténoses localisées ou multiples parfois très tardives…

- Gravité des lésions tardives car multiples et chirurgie difficile

Anévrysme géants (1%)

- Mortalité et morbidité +++
- Survie à 30 ans: 88-90%



- Cardiac event free à 30 ans : 30%
- 26% infarctus myocardique
- Risque accru dans les 2 ans après le diagnostic
- 50% de bypass coronaire à 30 ans

Surveillance à long terme

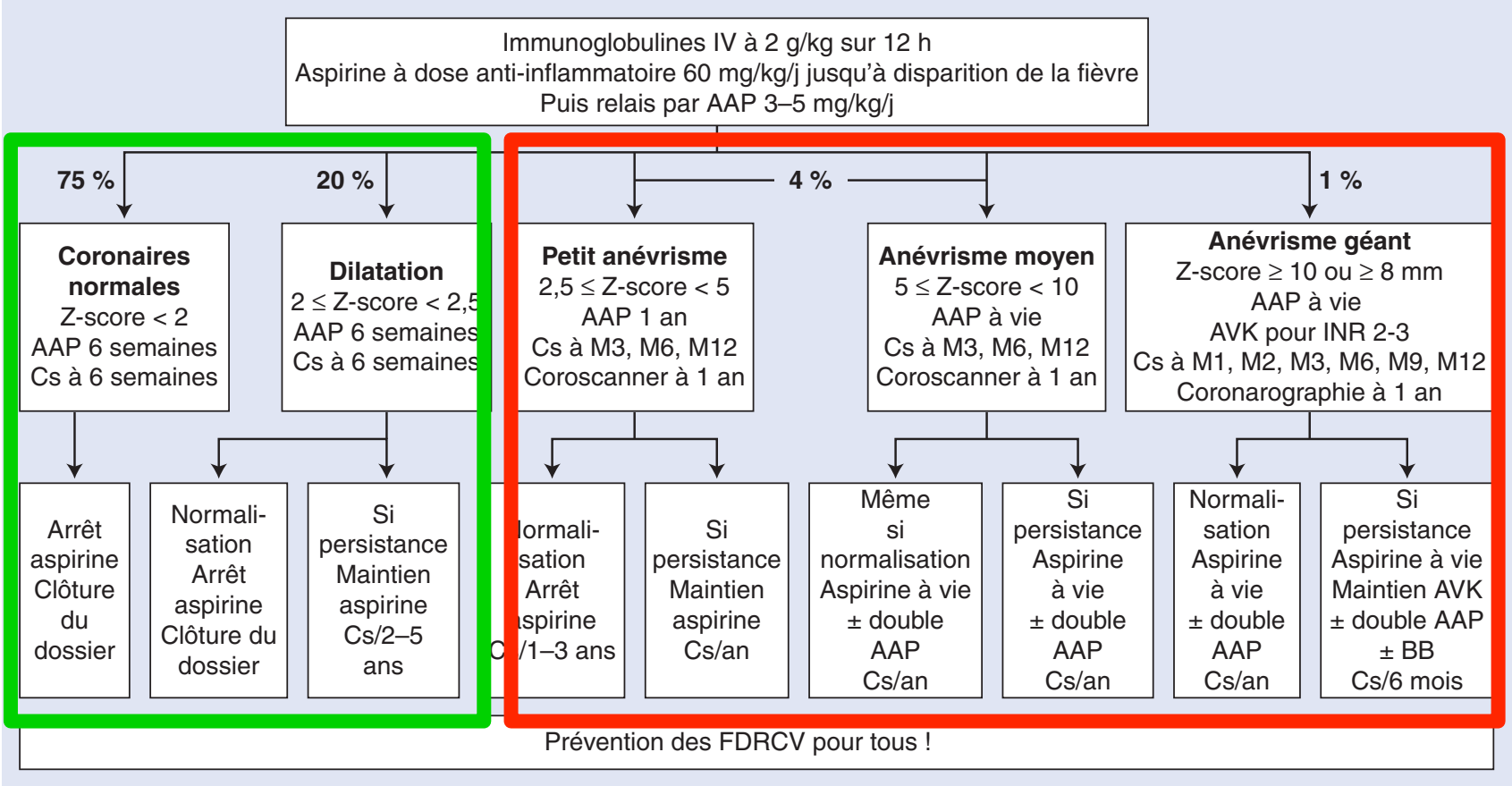


Figure 2. Arbre décisionnel. Prise en charge proposée par le centre de référence Malformations Cardiaques Congénitales Complexes (M3C) Necker. IV : p voie intraveineuse ; AAP : aspirine à dose antiagrégante plaquettaire ; Cs : consultation ; AVK : antivitamine K ; INR : *international normalized ratio* ; FDRCV facteurs de risques cardiovasculaires.

Bajolle et al. 2018 EMC

Devenir à long terme

TABLE 2 Principles in the Long-Term Management of Patients With KD

1. On the basis of available data, patients with no demonstrated coronary artery dilation by echocardiogram with excellent visualization of all arterial segments during the first weeks of illness appear to have normal cardiovascular status in early adulthood.
2. Remodeling (so-called regression) of aneurysms, especially if moderate or large, to normal internal lumen diameter is often accompanied by luminal myofibroblastic proliferation and abnormal vascular reactivity.
3. Patients with persistent CAA are at lifelong risk of progressive coronary artery stenosis or occlusion and worsening ischemia.
4. Patients with CAA documented at any stage require lifelong cardiovascular surveillance tailored to disease severity and age.
5. Testing should minimize exposure to ionizing radiation whenever possible.
6. Sedentary life-style should be avoided.
7. Women with coronary aneurysms can carry pregnancy successfully, but should have reproductive counseling.
8. Monitoring and counseling regarding traditional CV risk factors is appropriate to reduce the likelihood of later atherosclerosis.

Newburger JAC 2016

Endocardite

Endocardite infectieuse

Def: Infection/inflammation de l'endocarde = valves cardiaques

Dg: Echographie trans-thoracique voire ETO

- EI des VAV: sur le versant auriculaire

- EI des valves sigmoïdes: sur le versant ventriculaire

- Hémocultures: au moins 3!!!!!!!!!!! (au mieux 6)
- Pas d'ATB à l'aveugle
- Scanner total body (cérébral, thoracique et abdominal)
- Examen ophtalmologique, bandelette urinaire
- Recherche porte d'entrée: examen dentaire, ORL, cutané, digestif, urinaire, KTC…

Table 11 Modified Duke criteria for the diagnosis of infective endocarditis (adapted from Li et al.[94])

MAJOR CRITERIA

Blood cultures positive for IE:

- Typical microorganisms consistent with IE from two separate blood cultures:
  Viridans streptococci, *Streptococcus bovis*, HACEK group, *Staphylococcus aureus*; or
  Community-acquired enterococci, in the absence of a primary focus;

or

- Microorganisms consistent with IE from persistently positive blood cultures:
  At least two positive blood cultures of blood samples drawn > 12 h apart; or
  All of three or a majority of ≥ 4 separate cultures of blood (with first and last sample drawn at least 1 h apart)

or

- Single positive blood culture for *Coxiella burnetii* or phase I IgG antibody titer > 1 : 800

Evidence of endocardial involvement

- Echocardiography positive for IE
  Vegetation - Abscess - New partial dehiscence of prosthetic valve
- New valvular regurgitation

MINOR CRITERIA

- Predisposition: predisposing heart condition, injection drug use
- Fever: temperature > 38 °C
- Vascular phenomena: major arterial emboli, septic pulmonary infarcts, mycotic aneurysm, intracranial haemorrhages, conjunctival haemorrhages, Janeway lesions
- Immunologic phenomena: glomerulonephritis, Osler's nodes, Roth's spots, rheumatoid factor
- Microbiological evidence: positive blood culture but does not meet a major criterion or serological evidence of active infection with organism consistent with IE

Diagnosis of IE is definite in the presence of	Diagnosis of IE is possible in the presence of
2 major criteria, or *1 major and 3 minor criteria, or* *5 minor criteria*	*1 major and 1 minor criteria, or* *3 minor criteria*

Adapted from Li JS, Sexton DJ, Mick N, Nettles R, Fowler VG, Jr., Ryan T, Bashore T, Corey GR. Proposed modifications to the Duke criteria for the diagnosis of infective endocarditis. *Clin Infect Dis* 2000;**30**:633–638.

Endocardite: germes

- Streptocoques ++ 40%
- Staphylocoques 40%
- Autres : 10%
  - Escherichia Coli
  - BGN
  - HACEK
- Hémocultures négatives: 5 à 10%

Table 12 Predictors of poor outcome in patients with IE

Patient characteristics
- Older age
- Prosthetic valve IE
- Insulin-dependent diabetes mellitus
- Comorbidity (e.g frailty, previous cardiovascular, renal or pulmonary disease)

Presence of complications of IE
- Heart failure
- Renal failure
- Stroke
- Septic shock
- Periannular complications

Microorganism
- S. aureus
- Fungi
- Gram-negative bacilli

Echocardiographic findings
- Periannular complications
- Severe left-sided valve regurgitation
- Low left ventricular ejection fraction
- Pulmonary hypertension
- Large vegetations
- Severe prosthetic dysfunction
- Premature mitral valve closure and other signs of elevated diastolic pressures

Endocardite: Traitement médical

- **Principes généraux**
  - Bi-thérapie ATB
  - Bactéricide
  - Intraveineux
  - Prolongé: 4 à 6 semaines
  - Adaptée (antibiogramme)
  - Taux sériques efficaces

Endocardite: Traitement chirurgical

- **Indications**
  - Complications hémodynamiques
  - Sepsis non contrôlé
  - Embol gauche
  - Végétation > 10mm
  - Abcès
- **Types de chirurgie**
  - Eviter prothèse mécanique
  - Plastie, Ross, homogreffe

Endocardite: prévention

Table 4 Cardiac conditions at highest risk of infective endocarditis for which prophylaxis is recommended when a high risk procedure is performed

Recommendations: prophylaxis	Class[a]	Level[b]
Antibiotic prophylaxis should only be considered for patients at highest risk of IE 1. Patients with a prosthetic valve or a prosthetic material used for cardiac valve repair 2. Patients with previous IE 3. Patients with congenital heart disease a. cyanotic congenital heart disease, without surgical repair, or with residual defects, palliative shunts or conduits b. congenital heart disease with complete repair with prosthetic material whether placed by surgery or by percutaneous technique, up to 6 months after the procedure c. when a residual defect persists at the site of implantation of a prosthetic material or device by cardiac surgery or percutaneous technique	IIa	C
Antibiotic prophylaxis is no longer recommended in other forms of valvular or congenital heart disease	III	C

[a]Class of recommendation.
[b]Level of evidence.

Endocardite: prévention

Table 5 Recommendations for prophylaxis of infective endocarditis in highest risk patients according to the type of procedure at risk

Recommendations: prophylaxis	Class[a]	Level[b]
A - Dental procedures: **Antibiotic prophylaxis should only be considered for** dental procedures requiring manipulation of the gingival or periapical region of the teeth or perforation of the oral mucosa	IIa	C
Antibiotic prophylaxis is not recommended for local anaesthetic injections in non-infected tissue, removal of sutures, dental X-rays, placement or adjustment of removable prosthodontic or orthodontic appliances or braces. Prophylaxis is also not recommended following the shedding of deciduous teeth or trauma to the lips and oral mucosa	III	C
B - Respiratory tract procedures[c]: **Antibiotic prophylaxis is not recommended for respiratory tract procedures, including** bronchoscopy or laryngoscopy, transnasal or endotracheal intubation	III	C
C - Gastrointestinal or urogenital procedures[c]: **Antibiotic prophylaxis is not recommended for** gastroscopy, colonoscopy, cystoscopy or transoesophageal echocardiography	III	C
D - Skin and soft tissue[c]: **Antibiotic prophylaxis is not recommended for any procedure**	III	C

[a]Class of recommendation.
[b]Level of evidence.
[c]For management when infections are present, please refer to text.

Endocardite: prévention

Table 6 Recommended prophylaxis for dental procedures at risk

		Single dose 30–60 minutes before procedure	
Situation	**Antibiotic**	**Adults**	**Children**
No allergy to penicillin or ampicillin	Amoxicillin or ampicillin*	2 g p.o. or i.v.	50 mg/kg p.o. or i.v.
Allergy to penicillin or ampicillin	Clindamycin	600 mg p.o. or i.v.	20 mg/kg p.o. or i.v.

Cephalosporins should not be used in patients with anaphylaxis, angio-oedema, or urticaria after intake of penicillin and ampicillin.
*Alternatively cephalexin 2 g i.v. or 50 mg/kg i.v. for children, cefazolin or ceftriaxone 1 g i.v. for adults or 50 mg/kg i.v. for children.

POPULATION CONGENITALE - ENFANTS

Infective Endocarditis in Children With Congenital Heart Disease

Cumulative Incidence and Predictors

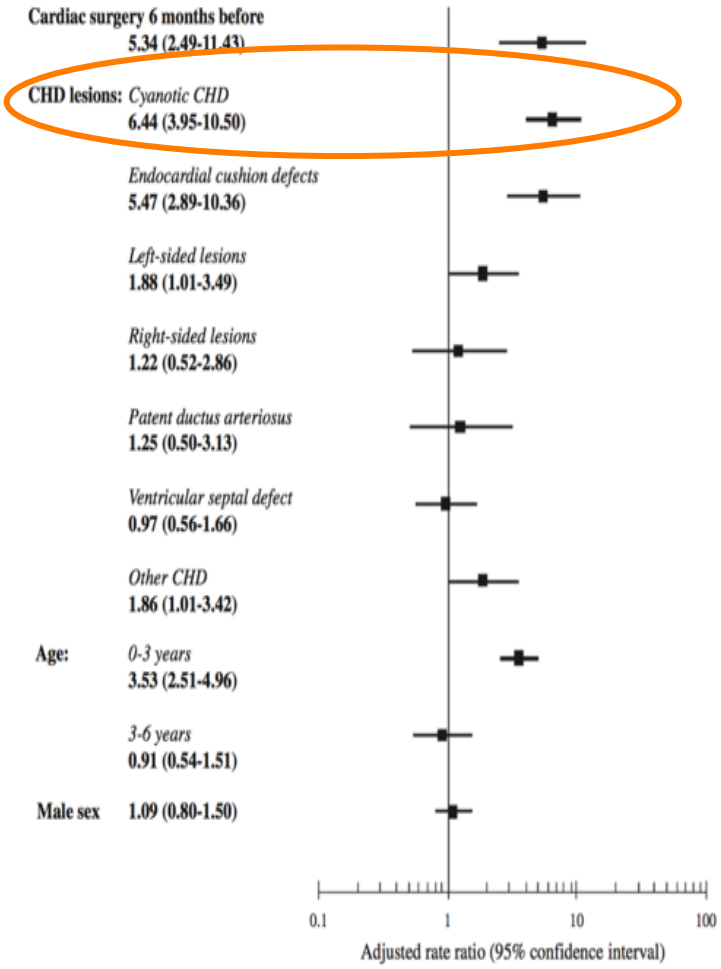
34 279 enfants avec CC suivis de 0 à 18 ans

Incidence annualisée = 4.1 / 10 000 pt-année

Table 2. Lesion Group–Specific Cumulative Incidence and Incidence Rate of IE in Children With CHD

CHD Lesions	Cumulative Incidence (95% CI) per 1000 Children			Incidence Rate (95% CI) per 10 000 Person-Years
	0–6 y	0–12 y	0–18 y	
Cyanotic CHD	16.8 (11.9–23.8)	23.3 (17.0–31.8)	31.0 (22.5–42.7)	20.7 (15.4–27.7)
Endocardial cushion defects	5.5 (2.3–13.1)	8.7 (4.1–18.6)	11.1 (5.4–22.9)	7.7 (3.9–15.4)
Left-sided lesions	2.7 (1.3–5.7)	4.8 (2.6–8.7)	7.9 (4.4–14.0)	4.4 (2.6–7.4)
Right-sided lesions	2.3 (1.0–5.5)	2.3 (1.0–5.5)	4.2 (1.5–11.5)	2.9 (1.3–6.5)
Patent ductus arteriosus	3.2 (1.4–7.1)	3.2 (1.4–7.1)	3.2 (1.4–7.1)	3.5 (1.6–7.7)
Ventricular septal defect	2.0 (1.2–3.2)	2.4 (1.5–3.8)	3.2 (1.9–5.3)	2.4 (1.5–3.7)
Atrial septal defect	1.9 (1.3–2.9)	2.2 (1.5–3.4)	3.0 (1.9–4.8)	2.3 (1.6–3.4)
Other CHD	2.9 (1.4–5.8)	3.7 (1.8–7.3)	5.5 (2.9–10.6)	3.7 (2.0–6.7)
Overall	3.2 (2.6–3.9)	4.2 (3.5–5.1)	6.1 (5.0–7.5)	4.1 (3.5–4.9)

CHD indicates congenital heart disease; CI, confidence interval; and IE, infective endocarditis.



POPULATION CONGENITALE - ADULTES

Table 4 Prediction model for developing IE, and score chart for the risk of developing IE up to 5 and 10 years

Predictor	HR(95% CI)	Points
Baseline valve-containing prosthetics	3.57(2.38–5.36)	3
Main defect[a]		
Pulmonary atresia with ventricular septal defect	4.05(1.85–8.86)	3
Double-outlet right ventricle	3.01(0.91–9.94)	2
Tetralogy of Fallot	1.81(0.99–3.33)	1
Univentricular heart	1.69(0.51–5.54)	1
Left-sided lesions	1.55(0.99–2.44)	1
Other	1	0
Multiple defects	1.68(1.15–2.46)	1
History of IE	2.21(1.22–4.01)	2
Male	1.89(1.28–2.81)	1

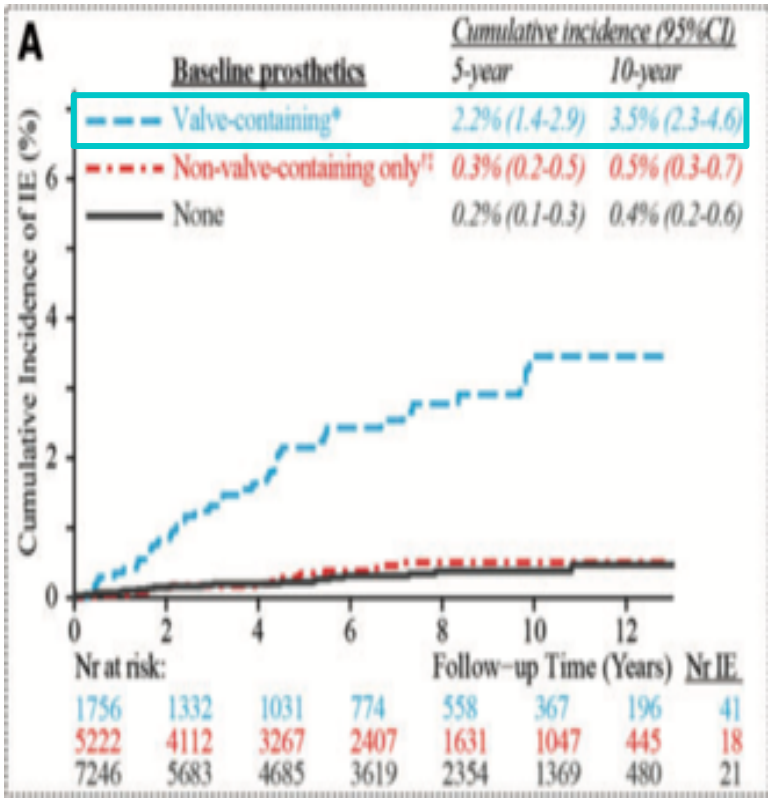
Score (sum points)

	Score									
	0	1	2	3	4	5	6	7	8	>8
Predicted 5 year risk (%)	<1	<1	1	1	1	2	3	4	7	9
Predicted 10 year risk (%)	<1	1	1	1	3	3	5	7	12	15

Registre CONCOR (14 224 patients>18 ans)

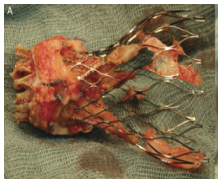
Incidence EI : **1.33/1000 pt-years**

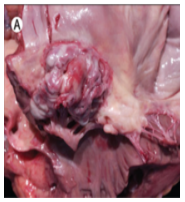
Prothèse valvulaire: **HR=3.57(2.58–5.36)**

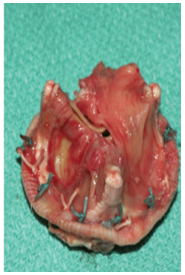


Kuijpers et al. Eur Heart Jour 2017

INCIDENCES COMPARATIVES





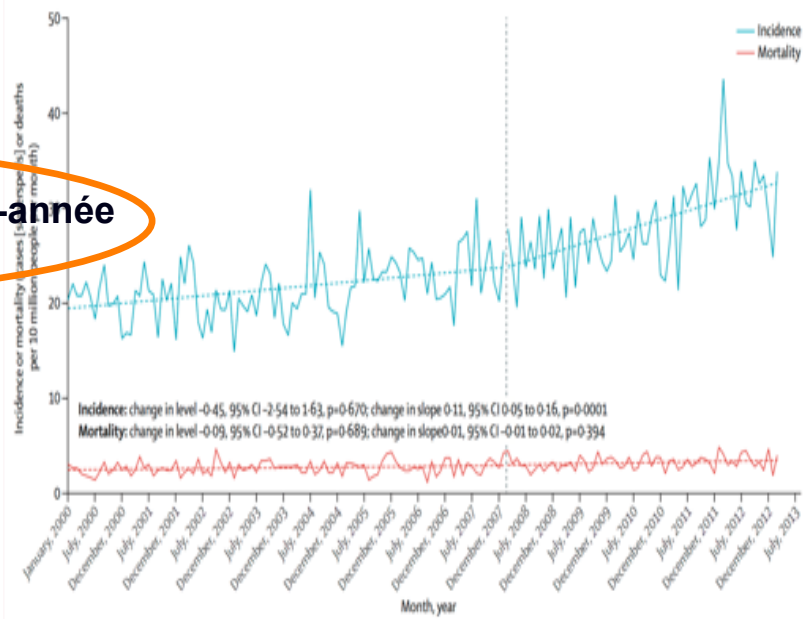


Valve Melody : **0.8 – 3% pt-année**

Valves/conduits pulmonaire chir : **0.5 - 3% pt-année**

TAVI: **0.67 – 2.1% pt-année**

Valves Ao/mitrale chir : **0.3 – 1.2% pt-année**



Dispositifs electroniques implantables : **1.9/1000 device-année**

Patients avec CC: **0.4 – 1.33 / 1000 pt-année**

Population générale : **30 -100/ million pt-année**

Miranda et al. Eur Heart Jour 2016
Wang et al. JAMA 2007
Rushani et al. Circulation 2013
Habib et al. Eur Heart Jour 2015
Dayer et al. Lancet 2015

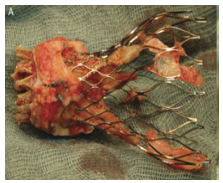
COMPARER CE QUI EST COMPARABLE

Table 2 Demographic characteristics.

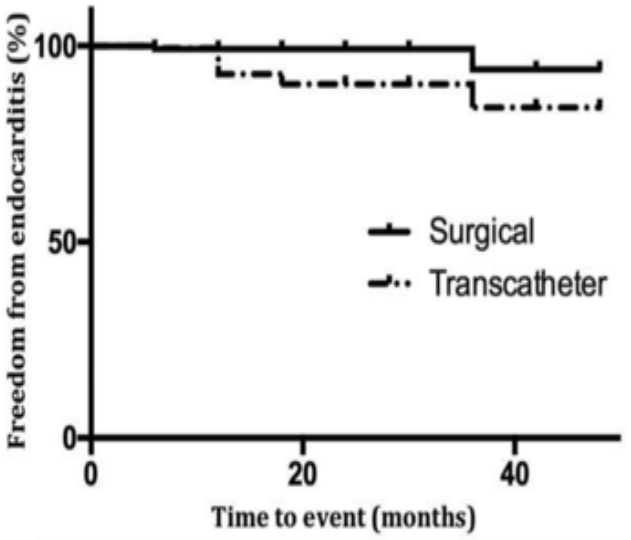
	Overall (*n* = 86)	IE (*n* = 5)
Age (years)	23.9 ± 10.5	29.8 ± 16.
Men/women	51/35	4/1
Body surface (m^2)	1.6 ± 0.3	1.8 ± 0.3
Type of CHD		
TOF	22 (25)	1
PAVSD	20 (23)	1
Truncus arteriosus	11 (13)	0
Aortic valve disease	13 (15)	2
PS	2 (2)	0
TGA-VSD-PS	2 (2)	1
DORV	4 (5)	0
Other	12 (14)	0
Co-morbidities	18	3
22q11 deletion	8	1
Noonan syndrome	1	0
Other syndrome	5	0
Trisomy 21	1	0
Beta thalassaemia	1	1
HCV	2	1
Portal cavernoma	1	1

Table 1. Baseline Characteristics and Procedural Details

Characteristics	Total, n=509	No TAVI-PVE, n=491	TAVI-PVE, n=18
Age, y (SD)	80 (6.9)	80 (6.9)	78 (6.9)
Men, n (%)	296 (58)	279 (57)	17 (94)
BMI, kg/m² (SD)	26.8 (5.1)	26.8 (5.2)	26.9 (3.5)
Arterial hypertension, n (%)	294 (58)	283 (58)	11 (61)
Diabetes mellitus, n (%)	103 (20)	99 (20)	4 (22)
Coronary artery disease, n (%)	259 (51)	249 (51)	10 (56)
Peripheral artery disease, n (%)	52 (10)	48 (10)	4 (22)
Chronic kidney disease†, n (%)	196 (39)	189 (38)	7 (39)
COPD, n (%)	73 (14)	72 (15)	1 (6)
Previous CVA, n (%)	72 (14)	69 (14)	3 (17)
Permanent pacemaker, n (%)	40 (8)	39 (8)	1 (6)
NYHA≥3, n (%)	364 (72)	352 (72)	12 (67)
Angina pectoris, n (%)	196 (39)	189 (38)	7 (39)
Syncope, n (%)	71 (14)	66 (13)	5 (28)
Previous endocarditis, n (%)	0 (0)	0 (0)	0 (0)



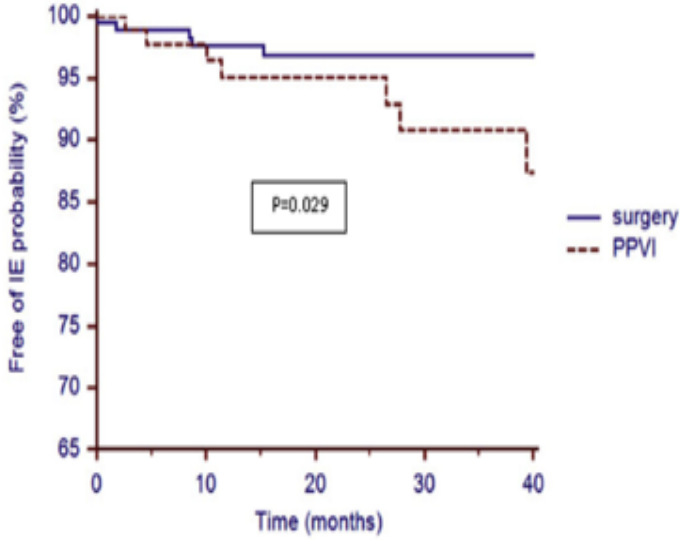
VALVES PERCUTANÉES VS CHIRURGICALES



134 chir et 208 percut (33 Sapien)

Incidence IE: 0.5 vs 1.5 %pt/années

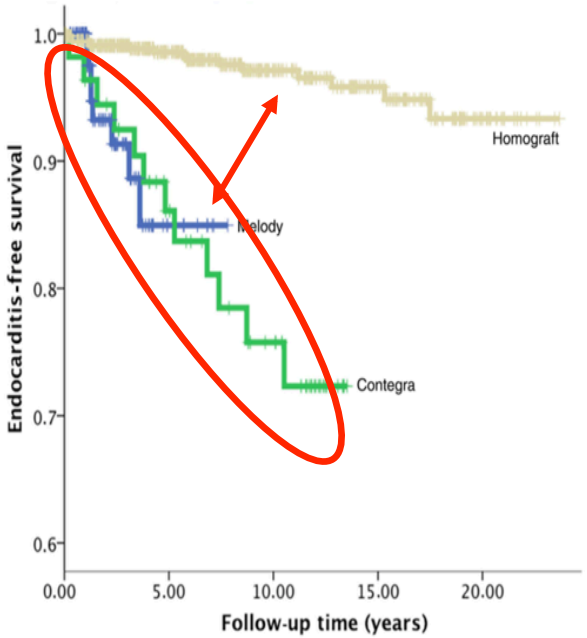
Lluri et al. CCI 2017



195 chir et 93 percut (0 Sapien)

Incidence IE: 1.2 vs 3.9 %pt/années

Malekzadeh-Milani et al. JTCS 2014



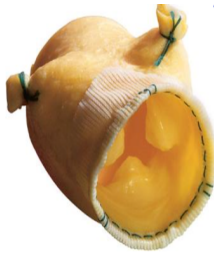
631 chir et 107 percut (0 Sapien)

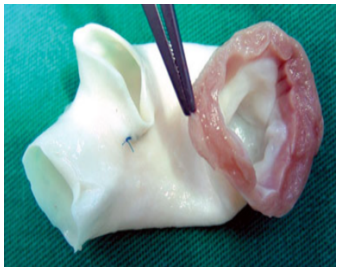
Incidence IE: 0.8 vs 2.7 vs 3% %pt/années

Van Dijck et al. Heart 2014

SUBSTRAT VALVULAIRE

Author	Year	n	Substrate	EI Cumulative incidence	EI Annualized Incidence (% pt-year)	Median Follow-up (years)
Albanesi	2014	12/106	Contegra	**11.3**		**7.6**
Malekzadeh	2014	5/190	Homografts Contegra	**2.6**	**1.2**	**2**
Ramanan	2015	6/115	Freestyle	**5.4**	**-**	**4.3**
Mery	2016	23/586	Homograft Contegra Porcine valve	**4**	**-**	**7**
Ugaki	2016	21/298	Contegra Homograft	**7**	**-**	**3.4**

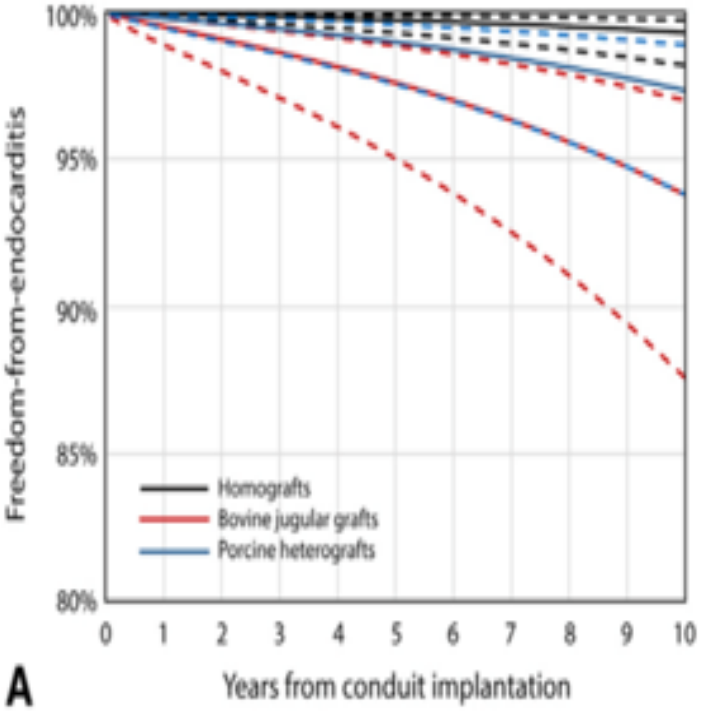


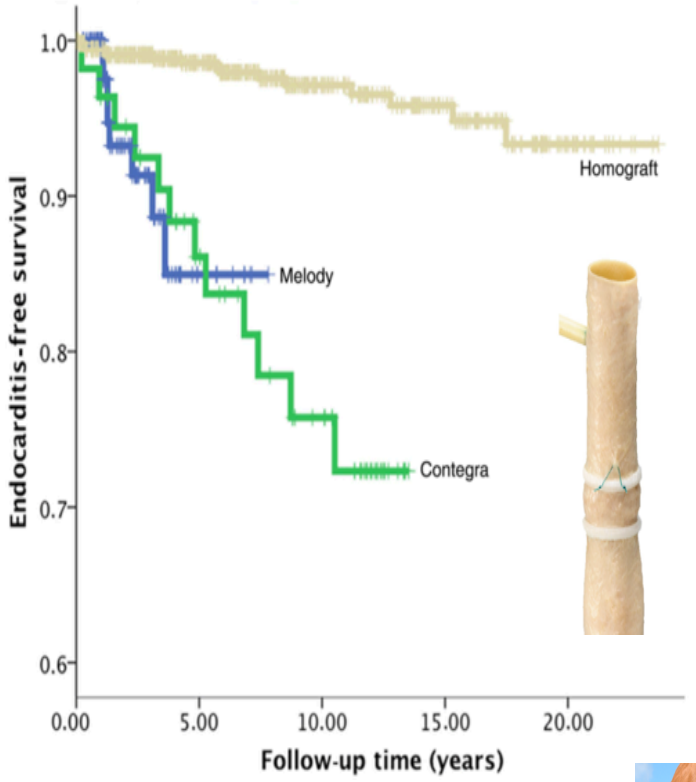


Tous les dispositifs valvulaires sont susceptibles d'être le siège d'une EI
Avec une incidence variable mais significative

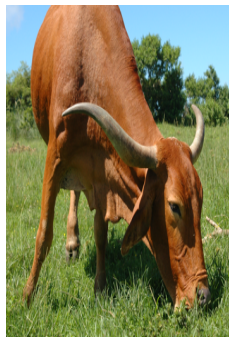
Albanesi et al. EJCTS 2014
Ramanan et al. Ann Thorac Surg 2015
Ugaki et al. Ann Thorac Surg 2016
Mery et al. JTCS 2016

SUBSTRAT VALVULAIRE





- EI plus fréquente chez les patients avec **VJB**
- **Quelle que soit la techniqued'implantation** (i.e. Contegra et Melody)
- Comparés aux homogreffes **RR=8.7 and 9.7** pour Melody et Contegra



Malekzadeh-Milani et al. JTCS 2014
Van Dijck et al. Heart 2014

Mery et al. JTCS 2016
Ugaki et al. Ann Thorac Surg 2015

SUBSTRAT VALVULAIRE

A Systematic Review of Infective Endocarditis in Patients With Bovine Jugular Vein Valves Compared With Other Valve Types

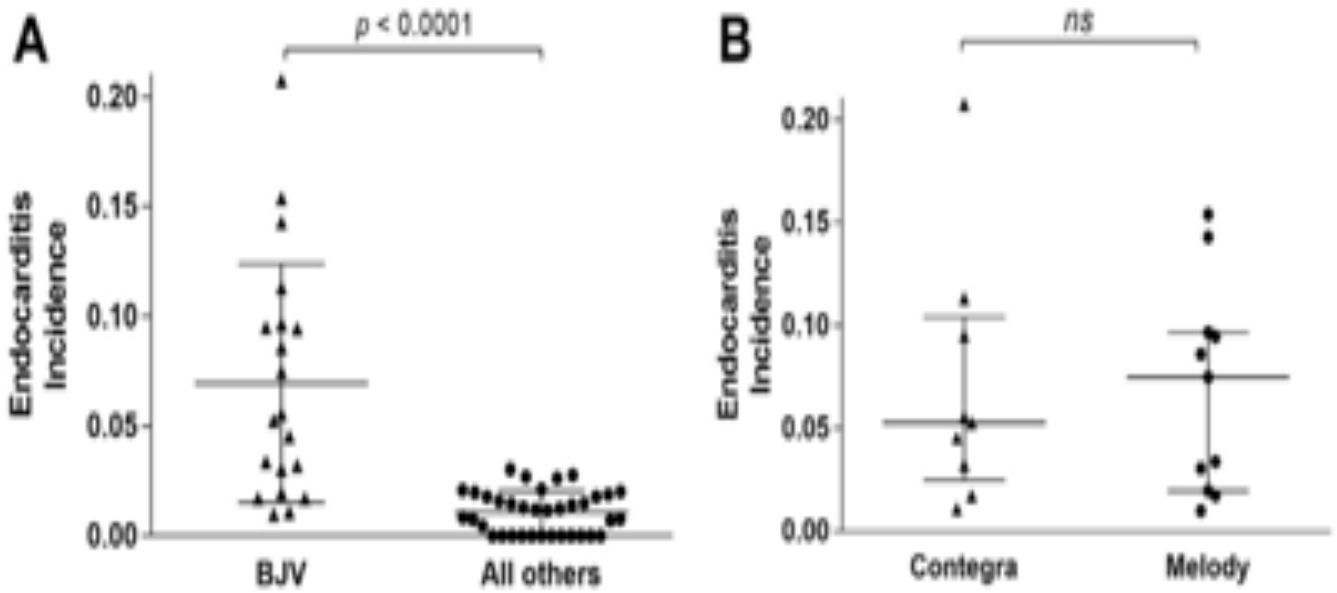
Méta-analyse sur IE chez les patients avec RVP chirurgical ou percutané

7063 patients

Incidence cumulative globale = 2.5%

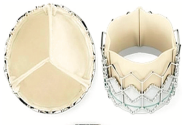
VJB vs autres substituts : 5.4% vs 1.2%; p < 0.0001

FIGURE 2 Incidence of Infective Endocarditis in BJV Compared to Other Valves



Sharma et al. JACC Int. 2017

VALVE SAPIEN



Infective Endocarditis Risk After Percutaneous Pulmonary Valve Implantation With the Melody and Sapien Valves

TABLE 1 Patient Demographics, Procedural Data, and Post-Procedural Outcomes

	PPVI With Melody Valve (n = 32)	PPVI With Sapien Valve (n = 47)	Standardized Difference
Age (yrs)	19.9 (15.8-28.9)	26.3 (18.9-39.9)	0.58*
Weight (kg)	56.5 ± 13.5	65.8 ± 17.6	0.59*
Male (%)	53.1	66.0	0.26
Genetic syndrome (%)	18.8	10.6	-0.23
History of severe infectious disease (%)	9.4	8.5	-0.03
History of endocarditis (%)	6.3	2.1	-0.20
Pacemaker/defibrillator (%)	6.3	10.6	0.16
Congenital heart diseases (%)			
Conotruncal malformation	81.3	68.1	
Ross procedure	9.4	21.3	
TGA	3.1	0.0	
PA-IVS/PVS	3.1	4.3	
DORV	3.1	6.4	
RVOT (%)			
Native RVOT	3.1	25.5	
Bioprosthesis	9.4	23.4	
Homograft	25.0	31.9	
Conduits	62.5	19.2	
RVOT lesion (%)			
Stenosis	84.4	50.0	
Regurgitation	0.0	35.7	
Mixed	15.6	14.3	

TABLE 1 Continued

	PPVI With Melody Valve (n = 32)	PPVI With Sapien Valve (n = 47)	Standardized Difference
Infective endocarditis during follow-up (%)	25.0	0.0	-0.80*
Pulmonary valve replacement during follow-up (%)	25.0	4.3	-0.59*
Percutaneous	3.1	2.1	-0.06
Surgical	21.9	2.1	-0.63*
Death during follow-up (%)	3.1	2.1	-0.06

Values are median [interquartile range] or %. Standardized difference computed as the difference in means or proportions divided by the SE. *Significant imbalance.

DORV = double-outlet right ventricle; PA-IVS = pulmonary atresia with intact ventricular septum; PPVI = percutaneous pulmonary valve implantation; PVS = pulmonary valve stenosis; RVOT = right ventricle outflow tract; TGA = transposition of the great arteries.

Hascoet et al. JACC Int. 2017

VALVE SAPIEN

- Monocentrique
- 2 cohortes non contemporaines: courbe d'apprentissage, prévention
- Populations peu comparables
  - Melody: conduits ou homogreffes (87% vs 51% Sapien)
  - Sapien: voies droites larges, pas de post dilatation
- Suivi plus court pour valves Sapien (1 an vers 4.9 ans)
- Pas de données sur les gradients résiduels (facteur prédisposant)

VALVE SAPIEN

COMPASSION TRIAL

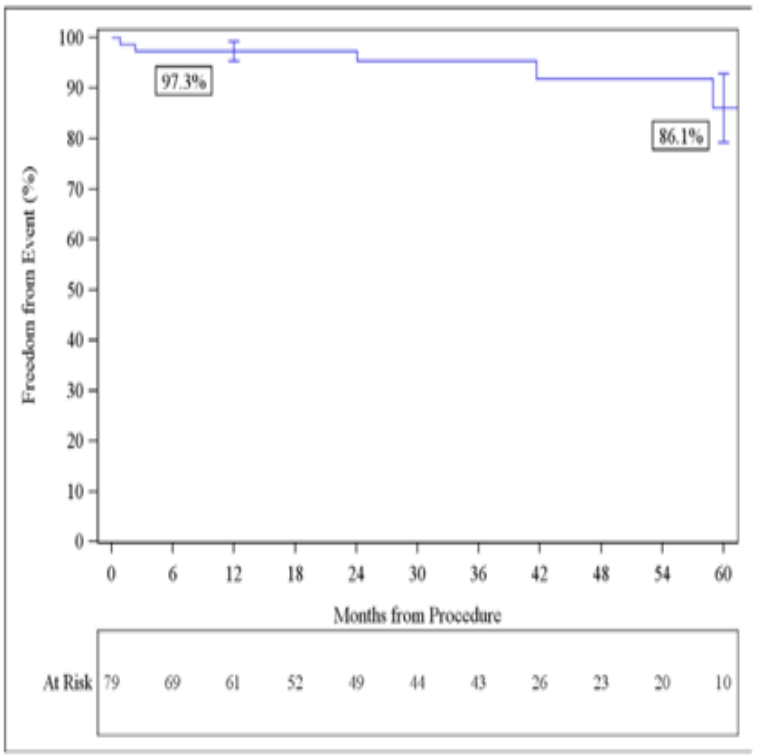
79 patients, Lésions mixtes

Suivi médian 3 ans

Table 9: Incidence of Site-Reported Serious Adverse Events by Study Visit (with CEC adjudication where available) in the Safety Population (N=79)

Adverse Event	≤ 30 Days		31 – 365 Days		All Events	
	Events	Patients with Event	Events	Patients with Event	Events	Patients with Event
Any Serious Adverse Event	29	3/ 79 (29.1%)	22	13/ 79 (16.5%)	132	38/ 79 (48.1%)
Other	2	2/ 79 (2.5%)	10	6/ 79 (7.6%)	37	13/ 79 (16.5%)
Infection (excluding endocarditis)	1	1/ 79 (1.3%)	3	3/ 79 (3.8%)	12	7/ 79 (8.9%)
CHF	1	1/ 79 (1.3%)	0	0/ 79 (0.0%)	11	4/ 79 (5.1%)
Electrolyte and/or CBC and platelet counts abnormal	1	1/ 79 (1.3%)	2	1/ 79 (1.3%)	11	2/ 79 (2.5%)
Valve stenosis	0	0/ 79 (0.0%)	1	1/ 79 (1.3%)	9	6/ 79 (7.6%)
Arrhythmia	2	2/ 79 (2.5%)	0	0/ 79 (0.0%)	8	6/ 79 (7.6%)
Endocarditis	1	1/ 79 (1.3%)	2	2/ 79 (2.5%)	5	4/ 79 (5.1%)

Clinical Figure 6: Freedom from Endocarditis at 5 Years (Safety Population, N=79)

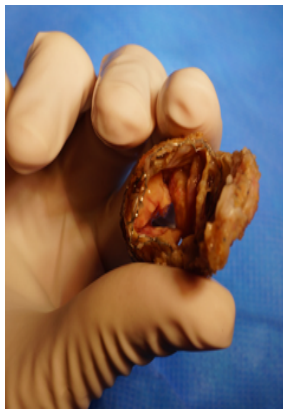


Edwards SAPIEN XT Transcatheter Heart Valve with the NovaFlex1Delivery System. Vol. 2016. https://www.accessdata.fda. gov/cdrh_docs/pdf13/p130009s037d.pdf

PREVENTION - EDUCATION

Table 3 **Cardiac conditions at highest risk of infective endocarditis for which prophylaxis should be considered when a high-risk procedure is performed**

Recommendations	Class[a]	Level[b]
Antibiotic prophylaxis should be considered for patients at highest risk for IE: (1) Patients with any prosthetic valve, including a transcatheter valve, or those in whom any prosthetic material was used for cardiac valve repair. (2) Patients with a previous episode of IE. (3) Patients with CHD: (a) Any type of cyanotic CHD. (b) Any type of CHD repaired with a prosthetic material, whether placed surgically or by percutaneous techniques, up to 6 months after the procedure or lifelong if residual shunt or valvular regurgitation remains.	IIa	C
Antibiotic prophylaxis is not recommended in other forms of valvular or CHD.	III	C



ST-IMPLANTATI

, parents, médeci

ggrégants au long

tibioprophylaxie à

Recommendations	Class[a]	Level[b]
A. Dental procedures		
• Antibiotic prophylaxis should only be considered for dental procedures requiring manipulation of the gingival or periapical region of the teeth or perforation of the oral mucosa	IIa	C
• Antibiotic prophylaxis is not recommended for local anaesthetic injections in non-infected tissues, treatment of superficial caries, removal of sutures, dental X-rays, placement or adjustment of removable prosthodontic or orthodontic appliances or braces or following the shedding of deciduous teeth or trauma to the lips and oral mucosa	III	C
B. Respiratory tract procedures[c]		
• Antibiotic prophylaxis is not recommended for respiratory tract procedures, including bronchoscopy or laryngoscopy, or transnasal or endotracheal intubation	III	C
C. Gastrointestinal or urogenital procedures or TOE[c]		
• Antibiotic prophylaxis is not recommended for gastroscopy, colonoscopy, cystoscopy, vaginal or caesarean delivery or TOE	III	C
D. Skin and soft tissue procedures[c]		
• Antibiotic prophylaxis is not recommended for any procedure	III	C

FACTEURS AGGRAVANTS

Portes d'entrée évitables

Manque observance

Déficiences mentales

Education - Prophylaxie EI

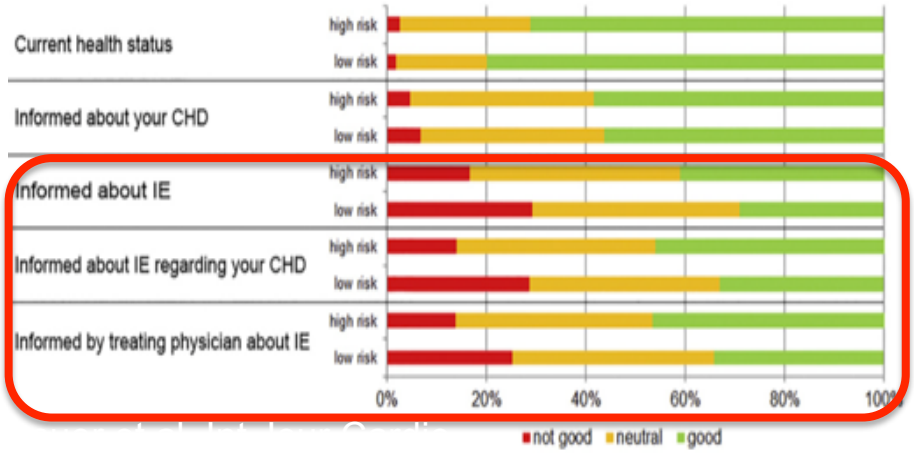
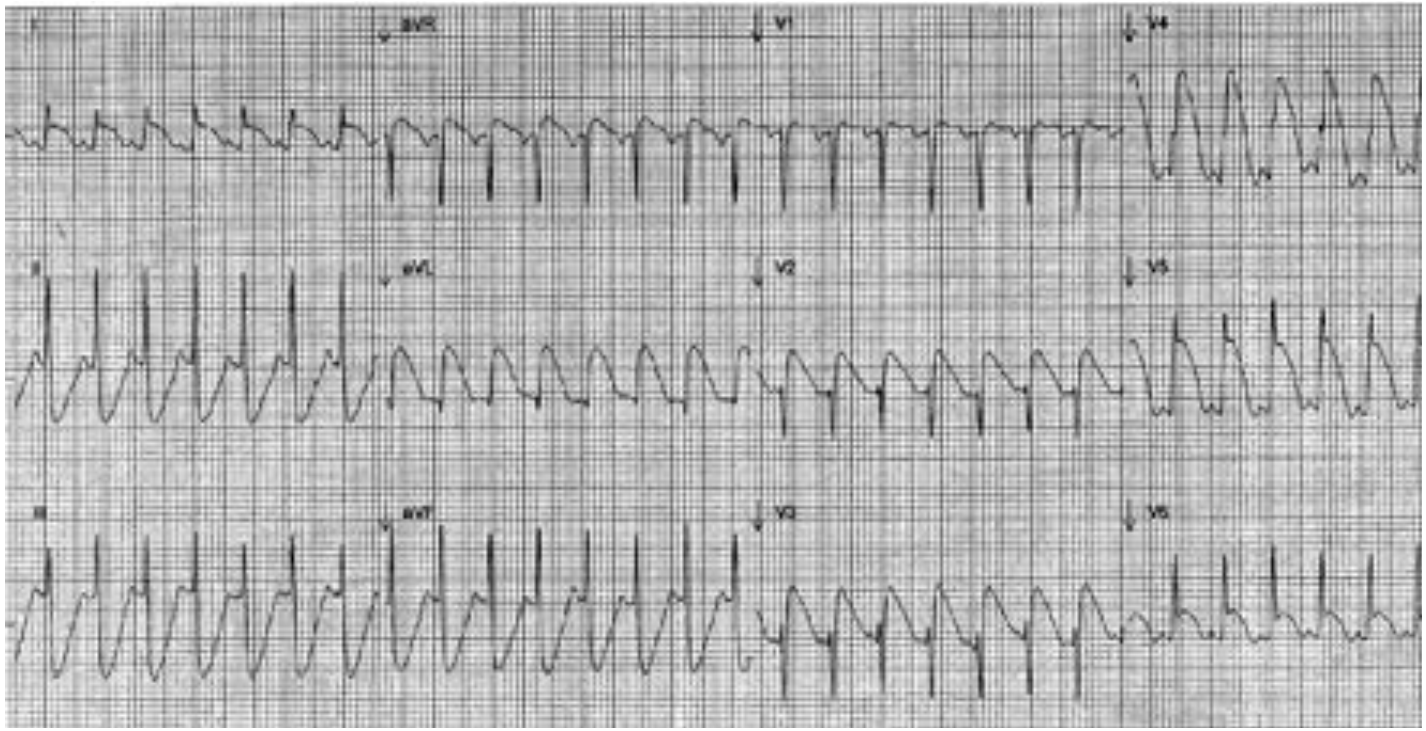


Table 1. Details of the 14 Index Bloodstream Bacterial Infection Cases

Case	Age, y	Sex	Time From Procedure to Systemic Infection (mo)	Underlying Cardiac Condition	Infecting Organism	Number of Positive Blood Cultures	Circumstances of Infection
1	4	Male	9	TOF/PA	*Streptococcus viridans*	3	Tooth extraction
2	28	Male	14	Congenital AS, S/P Ross procedure	*Streptococcus viridans*	4	None
3	41	Male	16	D-TGA	*Staphylococcus lugdunensis*	3	None
4	29	Male	56	D-TGA	*Streptococcus mitis*	3	Infection of oral ulcer
5	29	Male	53	TOF/PA	Coagulase negative *Staphylococcus*	2	Traumatic finger cut with subsequent cellulitis
6	25	Male	5	DORV	*Streptococcus anginosus* group	2	Pneumonia
7	42	Male	24	TOF/PA	Methicillin-resistant *Staphylococcus aureus*	2	Dental procedure
8	56	Male	10	TOF/PA	Methicillin-resistant *Staphylococcus aureus*	2	Sternal wound infection
9	14	Male	26	TOF/PA	*Streptococcus mitis*	3	Dental cleaning preceded
10	49	Male	20	TOF/PA	Methicillin-resistant *Staphylococcus aureus*	3	None
11	10	Male	18	TOF/PA	*Haemophilus parainfluenzae*	3	Bacterial gastroenteritis
12	21	Female	4	TOF	*Streptococcus viridans*	2	None
13	17	Male	30	Truncus Arteriosus	*Streptococcus mutans*	2	None
14	18	Male	1	TOF/PA	*Staphylococcus epidermidis*	2	Tracheostomy-associated infection

Buber et al. Circ Intrv 2013

Myocardites aiguës



Généralités

- Série autopsique: identification d'une myocardite dans 8,6% à 12% en cas de mort subite
- Evolution vers la CMD possible et non exceptionnelle

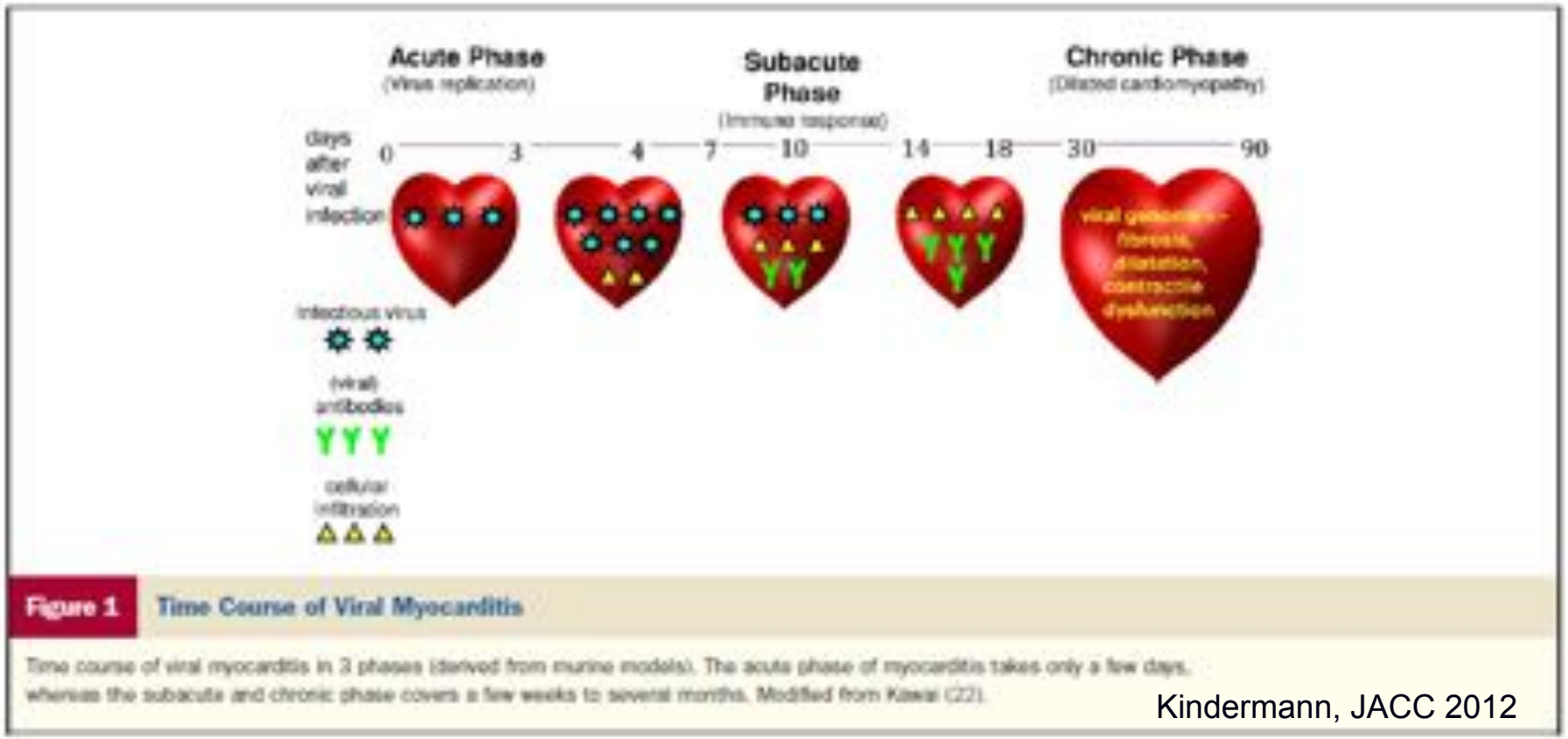
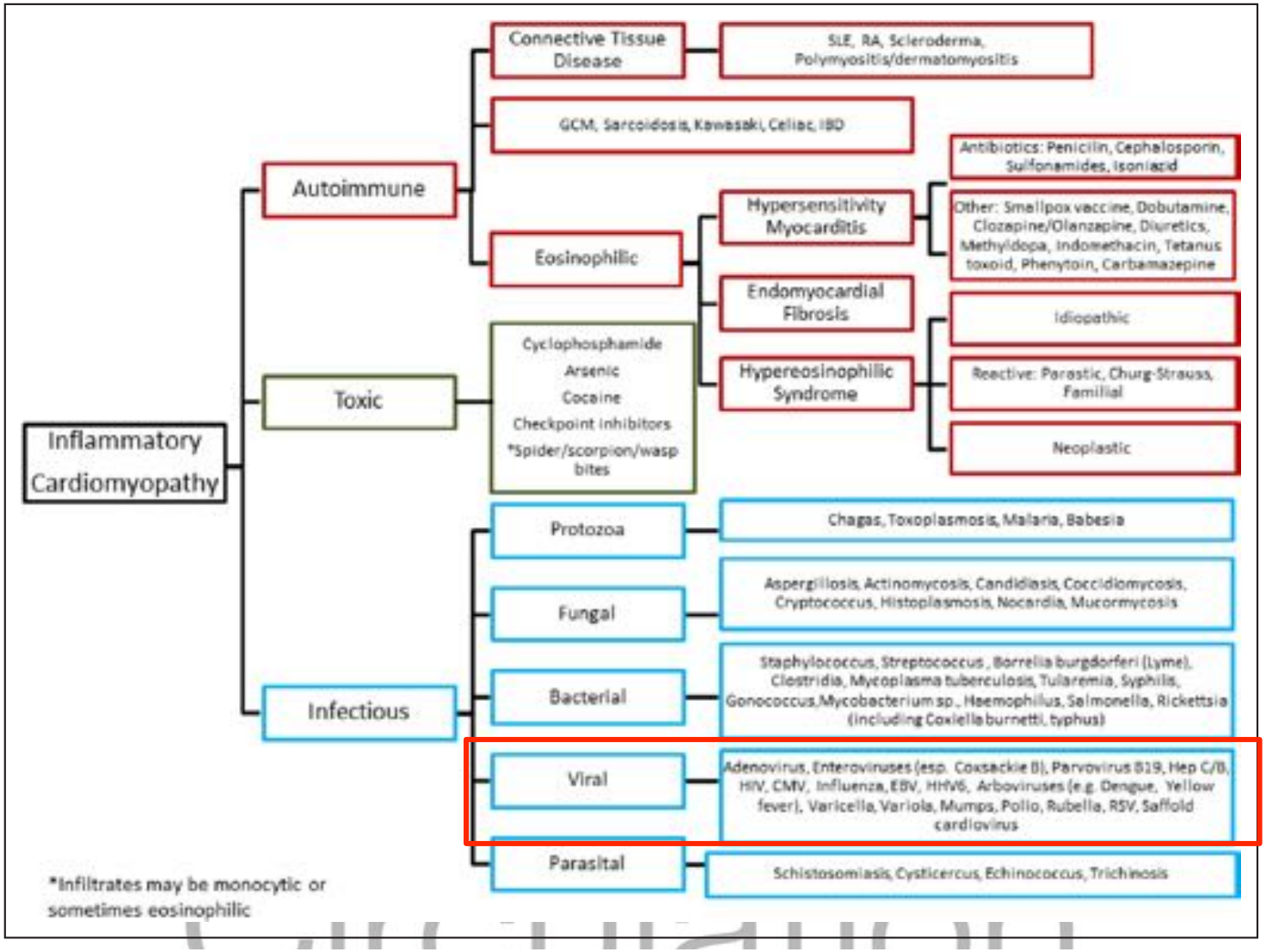


Figure 1 Time Course of Viral Myocarditis

Time course of viral myocarditis in 3 phases (derived from murine models). The acute phase of myocarditis takes only a few days, whereas the subacute and chronic phase covers a few weeks to several months. Modified from Kawai (22).

Kindermann, JACC 2012

Etiologies des myocardites



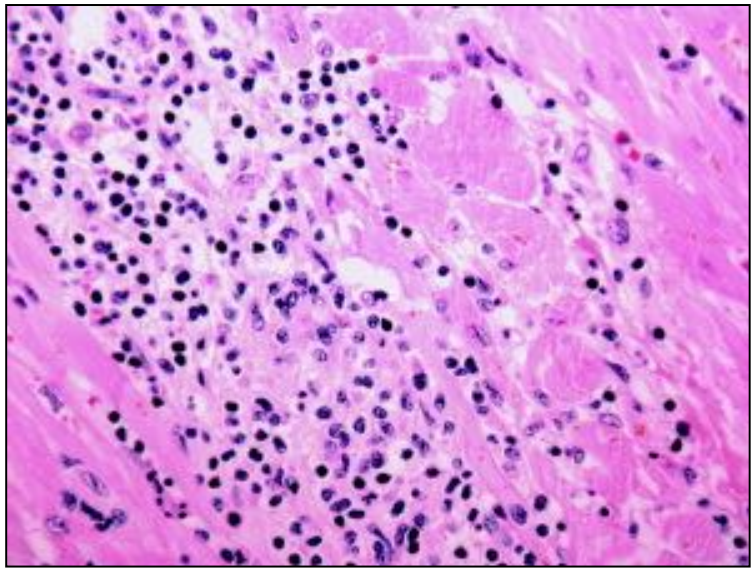
Kociol et al

Recognition and Management of Fulminant Myocarditis

Diagnostic positif

- **Clinique évocatrice:**
  - douleur thoracique, fièvre (30%), tachycardie (58%), dyspnée (68%)
  - Choc cardiogénique (Forme fulminante, 5-10/1 mill d'habitants/an)
  - Mort subite (TDR ou TDC)
- **Biologie:** Troponine, BNP ou N-proBNP
- **ECG:** infarctus du myocarde
- **Echocardiographie:** dysfonction modérée à sévère
- **IRM** et/ou **biopsie endomyocardique** (BEM)
- **Sérologies virales** peu utiles en pratique clinique

Critères de Dallas historiques (1986)



Infiltration lymphocytaire
Signe de nécrose non ischémique

Diagnostic	Image histologique
Myocardite aiguë	Infiltrat inflammatoire (lymphocytaire) Nécrose et/ou dégénérescence des myocytes en l'absence de maladie coronarienne
Myocardite subaiguë	Infiltrat inflammatoire sans nécrose apparente
Absence de myocardite	Image histologique normale

Tableau 2. Critères de Dallas.

Magnani JW et al. Circulation 2006 Cooper LT et al. Circulation 2007

Problèmes des critères de Dallas

- Myocardite avec atteinte hétérogène du myocarde -> Biopsies multiples > 5
- **Geste invasif:** mortalité 0,5%, complications 5%: perforation cardiaque, hémopéricarde, tamponnade
- Geste plus risqué chez le nourrisson
- Variabilité d'interprétation même entre experts
- « Goldstandard » mais discutée ++

Table 1. Risks Associated With Endomyocardial Biopsy in 546 Procedures

Overall 33 complications (6%)
Sheath insertion 15 (2.7%)
12 (2.0%) arterial puncture during local anesthesia
2 (0.4%) vasovagal reaction
1 (0.2%) prolonged venous oozing after sheath removal
Biopsy procedure 18 (3.3%)
6 (1.1%) arrhythmia
5 (1.0%) conduction abnormalities
4 (0.7%) possible perforation (pain)
3 (0.5%) definite perforation (pericardial fluid)
2 of 3 patients with definite perforation died

Data derived from Deckers et al (20).

Indication d'une BEM

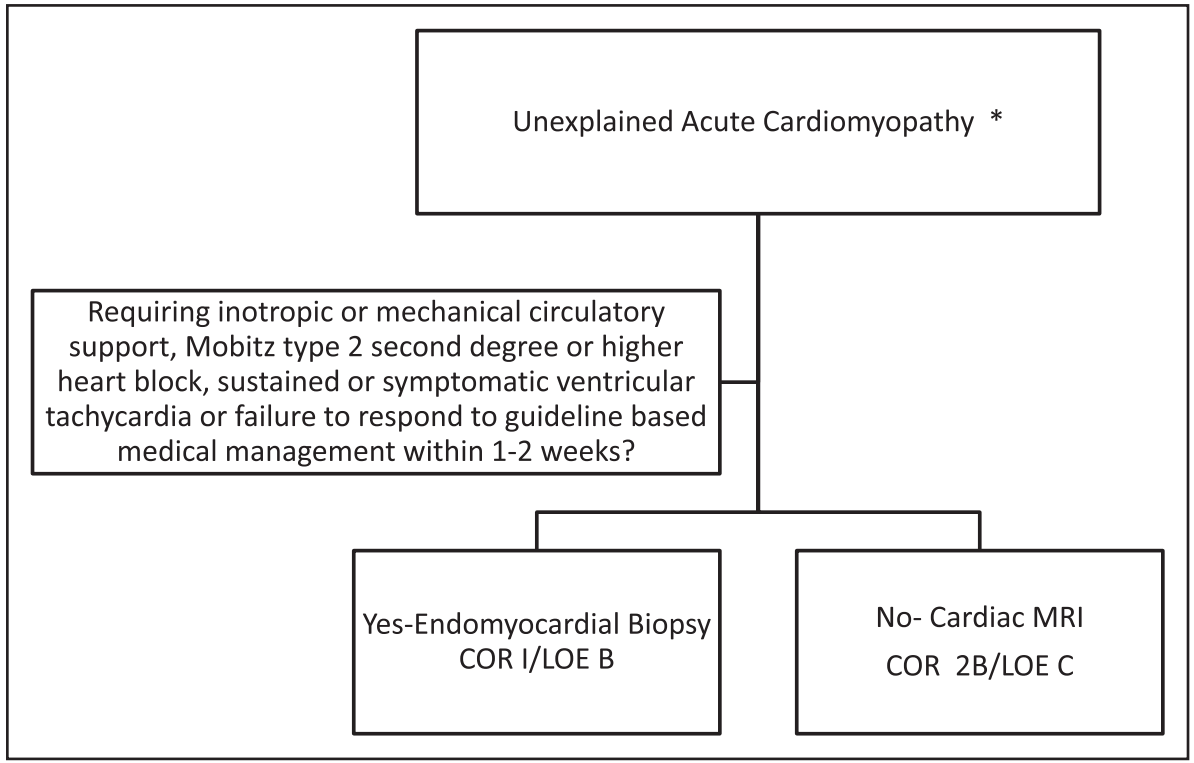


Figure 3. Indications for endomyocardial biopsy (EMB).
Guideline-based algorithm for whether EMB is indicated. COR indicates Class of Recommendation; LOE, Level of Evidence; and MRI, magnetic resonance imaging. *Usually a dilated cardiomyopathy. Fulminant myocarditis may have normal end-diastolic diameter with mildly thickened walls. Exclude ischemic, hemodynamic (valvular, hypertensive), metabolic, and toxic causes of cardiomyopathy as indicated clinically. Reprinted from Bozkurt et al.[3] Copyright © 2016, American Heart Association, Inc.

Lake Louise Criteria: IRM

Trois séquences IRM contributives:

- 1. Œdème en T2
- 2. rehaussement précoce du myocarde
- 3. rehaussement tardif du myocarde

- Diagnostic positif si > 2 critères :
  - Hypersignal T2
  - Ratio Signal myocarde / muscle périph augme
    Gadolinium
  - Hypersignal en rehaussement tardif

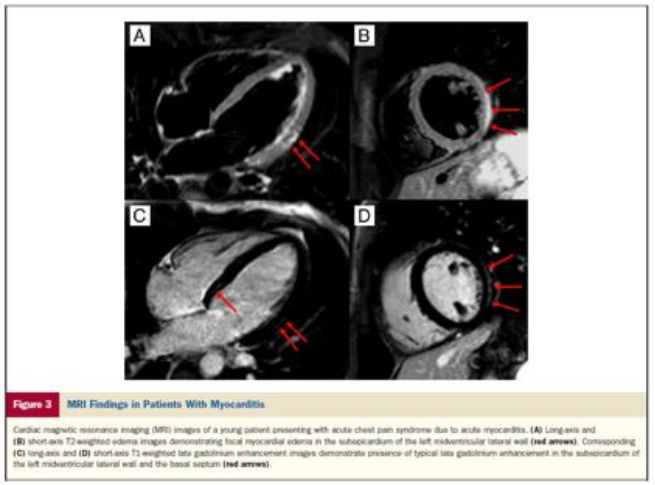


Figure 3 MRI Findings in Patients With Myocarditis

Cardiac magnetic resonance imaging (MRI) images of a young patient presenting with acute chest pain syndrome due to acute myocarditis. (A) Long-axis and (B) short-axis T2-weighted edema images demonstrating focal myocardial edema in the subepicardium of the left midventricular lateral wall (red arrows). Corresponding (C) long-axis and (D) short-axis T1-weighted late gadolinium enhancement images demonstrate presence of typical late gadolinium enhancement in the subepicardium of the left midventricular lateral wall and the basal septum (red arrows).

- Refaire IRM à 1-2 semaines si:
  - 0 critère mais symptômes trop récents, forte suspicion clinique
  - 1 seul critère présent

Friedrich MG et al. JACC 2009

Traitement en fonction de la forme clinique

- **Myocardite segmentaire focale:** Repos
- **Myocardite aiguë diffuse chez l'enfant**
  - Surveillance +/- assistance circulatoire (HNF)
  - Traitement d'attaque:
    - Immunomodulateurs, immunosuppresseurs, Anti-inflammatoire, immunoadsorption
- **Myocardite fulminante**
  - PEC du choc cardiogénique
  - (Traitement spécifique en fonction du type histologique)
- **Myocardite chronique active**
  - Discuter immunosupresseurs

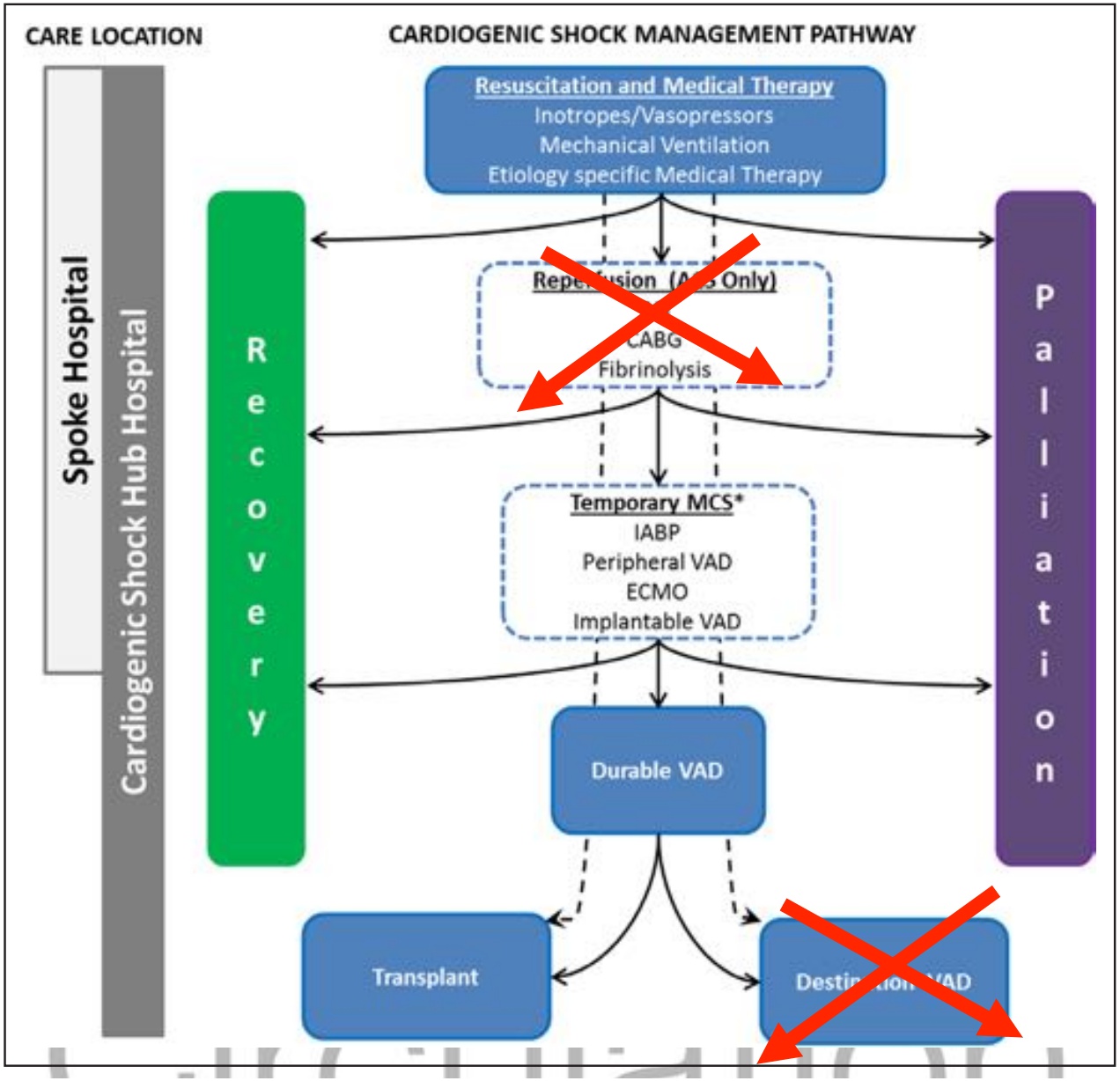
Traitement
ce qui est admis …

Traitement de l'insuffisance cardiaque

- **Selon les guidelines**
- **Selon la classe fonctionnelle NYHA**
  - IEC
  - Diurétiques
  - B-bloquant
  - ARA II
- **Formes sévères: Prise en charge en réanimation**
  - **Traitement « agressif »**
  - **Assistance circulatoire** (> 60 à 80 % survivants et récupération ad integrum possible)
  - **Drogues Inotropes positives et héparine!**

Kindermann, et al., JACC 2012
Amabile et al. Heart 2006

Traitement de la myocardite fulminante



Circ 2020

Traitement
ce qui est discuté …

Immunoglobulines
Anti inflammatoires
Antiviraux
Immunosuppresseurs

Immunosuppressive Treatment for Myocarditis in the Pediatric Population: A Meta-Analysis

Study	N	Age	Study methodology	IMSA	IMSA dosage, time of IMSA start	Follow-up	Observed variables	Inclusion criteria
Camargo et al. (9)	50	5 months–15 years	PNCT	P, CyA	P & A: 2.5 mg/kg/d, 1 week; 2.0 mg/kg/d, 3 weeks; 1.5 mg/kg/d, 4 weeks Cy: 1.5 mg/kg/d, 1 week; 1.0 mg/kg/d, 7 weeks; 0.5 mg/kg/d, 1 week	8.4±1.2 months	LVEDD, LVEF, PWP, CI, HR	Active myocarditis based on EMB findings
Aziz et al. (6)	68	3.7 ± 2.9 years	RCT	P	2 mg/kg/d, 1 month	15.1±9.2 months	LVEDD, LVESD, LVEF	Duration of symptoms for<3 months and continued LV failure and reduced EF
Drucker et al. (7)	46	–	CCT	IVIG	2,000 mg/kg 24 h; 1,000 mg/kg/d, 1 weeks	10.5±2.1 months	LVFS, LVEDD, death	Acute (<3 months) onset of congestive heart failure and echocardiographic documentation of diminished LV function and EMB
Bhatt et al. (8)	83	4.4 ± 3.2 years	PNCT	IVIG	400 mg/kg/d, 5 days	-	LVEF, death	Had viral infection with fever of < 2 weeks' duration; developed acute and severe heart failure after this illness; evidence of LV dysfunction on echocardiography EF< 40%; no previous or family history of cardiomyopathy
Gagliardi et al. (10)	114	36.6 ± 42.8 months	CCT	P, Cy	P: 2 mg/kg/d, 1 month; 0.5 mg/kg/d, 6 months; Cy: 6–8 mg/kg/d until blood concentration reached 170–210 ng/cm^3	13 years	LVEF, LVEDV, death	Congestive heart failure patients received right cardiac characterization and EMB
Camargo et al. (11)	10	42.1 ± 18.9 months	CCT	P, A	2.5 mg/kg, 4 weeks; 1.5 mg/kg, 4 weeks (both drugs)	9 months	LVEF, CI, death	Patients presenting with dilated cardiomyopathy who were clinically stable, under ambulatory care, with LVEF between 15 and 30%

PNCT, prospective non-controlled trial; RCT, randomized controlled trial; CCT, case–control study (including historical controls); IMSA, immunosuppressive agent; P, prednisolone; CyA, cyclosporine; A, azathioprine; IVIG, intravenous immunoglobulin G; LVEF, left ventricular ejection fraction; LVEDD, left ventricular diastolic dimension diameter; LVESD, left ventricular systolic dimension diameter; PWP, pulmonary wedge pressure; HR, heart rate; LVFS, left ventricular fractional shortening; CI, cardiac index, EMB, endomyocardial biopsy.

Immunosuppressive Treatment for Myocarditis in the Pediatric Population: A Meta-Analysis

- Groupe d'enfants avec immunosuppresseurs amélioration significative:
  - Fraction d'éjection VG
  - Diamètre télédiastolique VG
  - Diminution décès et transplantation

- **MAIS:** 1 seule étude RCT, effectifs faibles, follow-up court

Conclusions: There may be a possible benefit, in the short term, to the addition of immunosuppressive therapy in the management of myocarditis in the pediatric population. However, further prospective investigation is warranted to validate this finding.

Bing He, Xiaoou Li and Dan Li*

November 2019 | Volume 7 | Article 430

Traitement de la myocardite fulminante

Table 7. Major Myocarditis Subtypes Resulting in a Fulminant Presentation

Subtype	H&E Findings	Clinical Manifestations	Treatment
Fulminant lymphocytic myocarditis	Extensive dense lymphocytic infiltrate with associated myonecrosis. May have occasional isolated multinucleated giant cells or eosinophils.	Acute heart failure rapidly leading to cardiogenic shock, conduction abnormalities, or ventricular arrhythmias/SCD. Chest pain.	Treatment is primarily supportive; circulatory support as needed to prevent MOSF. Some evidence that in the absence of cardiotropic viral genome by PCR, steroids may be helpful
GCM	Extensive mixed inflammatory infiltrate characterized by the presence of several multinucleated giant cells (usually present after 1–2 wk), eosinophils, monocytes, and macrophages in the absence of noncaseating granulomas. Edema and extensive myonecrosis often present.	Acute heart failure caused by systolic dysfunction, myocardial restriction, or both. Conduction abnormalities, including CHB and EMD; ventricular arrhythmias, including sustained VT/VF and SCD. Tends to comigrate with other autoimmune diseases.	Treatments consists of multimodality therapy and should be implemented after a tissue diagnosis has been confirmed. Usual therapy includes a combination of a high-dose steroids, a calcineurin inhibitor (such as cyclosporine), and an antimetabolite such as azathioprine. Cytolytic therapy (purified rabbit-derived polyclonal IgG directed at human thymocytes) used for suppression of life-threatening GCM has been reported.
Acute NEM	Extensive inflammatory infiltration of the myocardium with mononuclear cells and eosinophils. Associated myonecrosis or fibrosis. On EM, may see eosinophil degranulation and deposition of major basic protein.	Acute heart failure/cardiogenic shock. May present with a restrictive cardiomyopathy. Prothrombotic intracardiac state. Peripheral eosinophilia may or may not be present. Recent viral infection or new medication.	Identify potential precipitant, especially if a drug hypersensitivity (Table 5). High-dose steroids. Anticoagulation. Often presents with ST-segment elevations and chest pain mimicking an ST-segment–elevation myocardial infarction. Rapid angiography, EMB with subsequent circulatory support, and initiation of high-dose intravenous corticosteroids can be lifesaving.
ICI myocarditis	Newly identified lymphocytic myocarditis resulting from the introduction of novel chemotherapeutic agents that unleash inhibited antitumor T cells, which also may infiltrate and attack the myocardium. Histopathology consistent with lymphocytic infiltrate and myocardial necrosis.	Acute heart failure, cardiogenic shock, and atrial fibrillation developing soon after ICI therapy is started and generally more severe with combination ICI therapy. Typically occurs early in treatment and has a fulminant course.	Treatment includes immediate cessation of therapy, high-dose corticosteroids (1 g solumedrol intravenously daily for 3 d and then 2 mg/kg prednisone daily to start, followed by a slow wean) and initiation of an angiotensin receptor blocker or sacubitril/valsartan. May initially need MCS.

Circ 2020

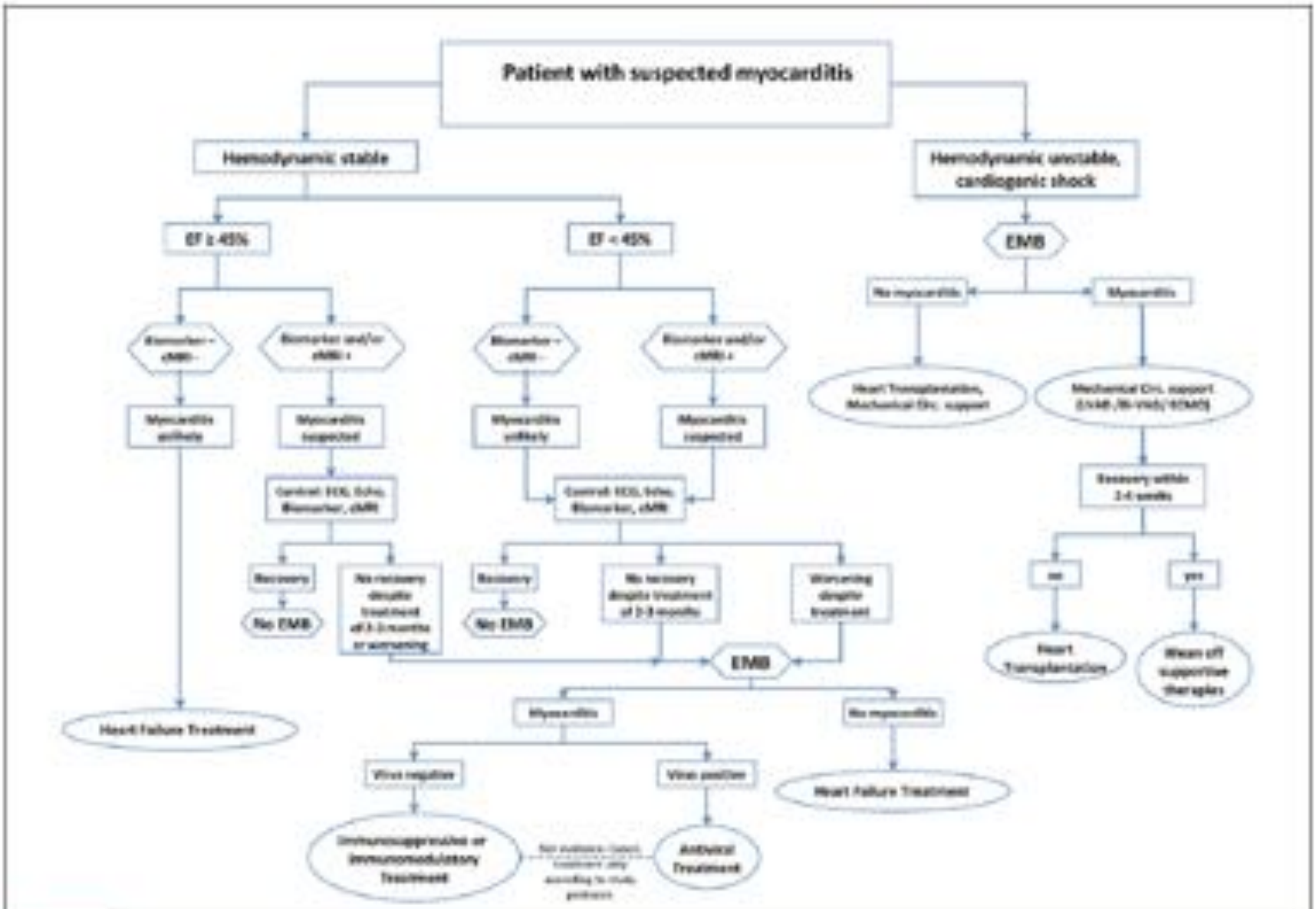


Figure 5 Proposed Diagnostic and Therapeutic Algorithm for Suspected Myocarditis

Proposed diagnostic and therapeutic algorithm for patients with suspected acute myocarditis considering biomarkers, cardiac magnetic resonance imaging (CMRI), and endomyocardial biopsy (EMB). BiVAD = biventricular assist device; Circ. = circulatory; ECMO = extracorporeal membrane oxygenation; LV = left ventricular; LVAD = left ventricular assist device.

Kindermann et al. JACC 2012

Pronostic

- Bon pronostic
  - Myocardite active
  - Fonction VG préservée
- Myocardite fulminante
  - Très bon pronostic à long terme si on passe la phase aigue vivante
  - Complète récupération possible
- Mauvais pronostic
  - Dysfonction VD, PAP élevée, syncope, PA basse, Fc élevée, QRS > 120 msec…