


Tumeurs cardiaques



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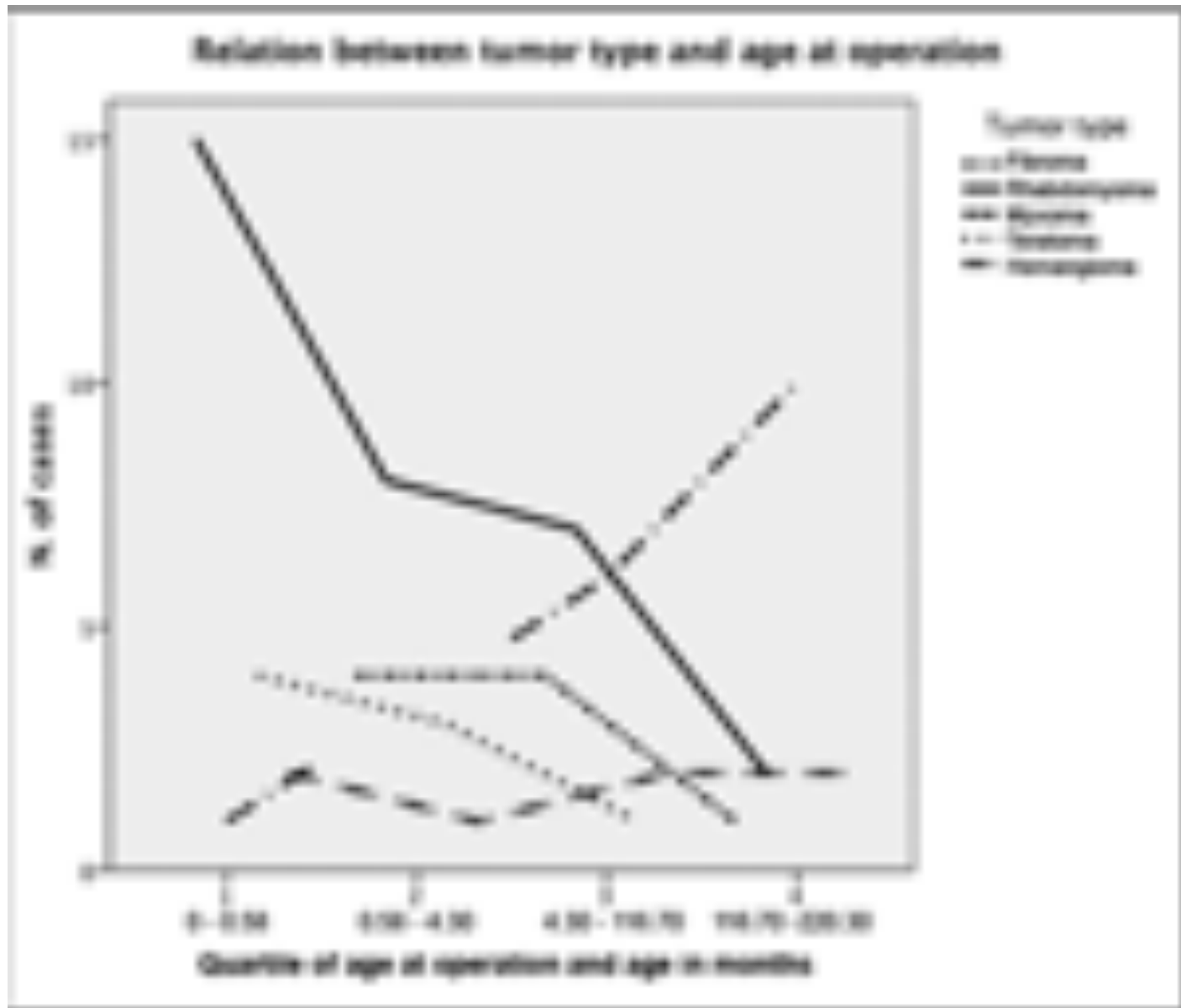
Circonstances de découverte: Dépend de la localisation!

- | | | |
|--------------------------------|--|-----------------------------|
| - Symptômes |  | - Souffle |
| - Diagnostic Prénatal | | - Cyanose |
| - Trouble du rythme/conduction | | - Douleur thoracique |
| - Cardiopathie sous jacente | | - Détresse respiratoire |
| - Exploration extracardiaque | | - Syncope |
| | | - Palpitation |
| | | - AVC embolique |
| | | - ACR |
| | | - Insuffisance circulatoire |
| | | - Tamponnade |
| | | - Ischémie |

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TUMOR	% of Group		
	Adults	Children	Infants
Myxoma	46	15	0
Lipoma	21	0	0
Papillary fibroelastoma	16	0	0
Rhabdomyoma	2	46	65
Fibroma	3	15	12
Hemangioma	5	5	4
Teratoma	1	13	18
Mesothelioma of AV node	3	4	2
Granular cell tumor	1	0	0
Neurofibroma	1	1	0
Lymphangioma	1	0	0
Hamartoma	0	1	0

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Tumeurs cardiaques chez l'enfant

• Tumeurs bénignes (90%)

Rhabdomyomes: 40-60%

Tératomes cardiaques et péricardiques: 15-20%

Fibromes: 12-16%

Hémangiomes: 5%

Myxomes: 2-4%

Lipomes, fibroélastomes, lymphangiomes

• Tumeurs malignes (10%)

Primitives :sarcomes

Secondaires: neuroblastome, néphroblastome, LNH

Tumeurs multiples: toujours rhabdomyomes
Tumeur unique: très souvent rhabdomyomes

Rhabdomyomes:

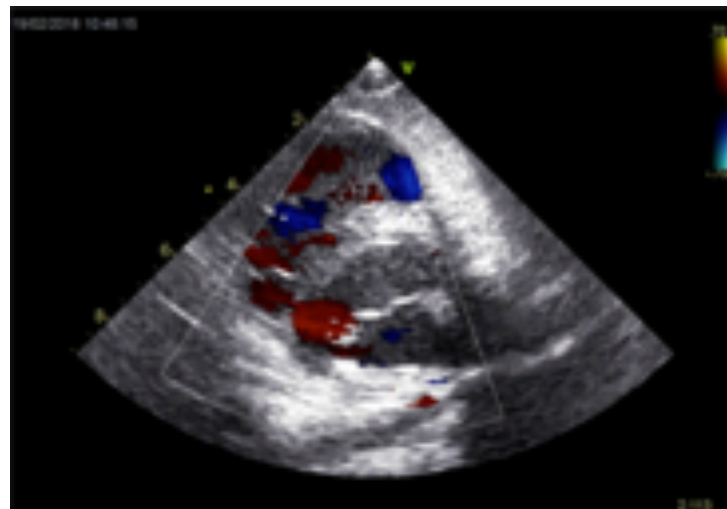
Tumeur la plus fréquente chez l'enfant

- Circonstance de découverte : DAN, Bilan extracardiaque, Signes cliniques
- Signes cliniques inconstants:
 - Souffle, cyanose: Obstruction hémodynamique à l'éjection ou au remplissage D et/ou G
 - Trouble du rythme (TJ, Syndrome de WPW, tachycardie atriale, TV ou exceptionnellement des troubles conductifs)
- Echographique: Tumeurs intra-myocardiques MULTIPLES homogènes, lisses, rondes, hyperéchogènes localisées parois VD, VG, SIV et valves AV
- 50 à 70% : sclérose Tubéreuse de Bourneville
- Régression avant 6 ans (première année de vie++)
- Indication chirurgicale en cas d'obstacle ou trouble du rythme réfractaire

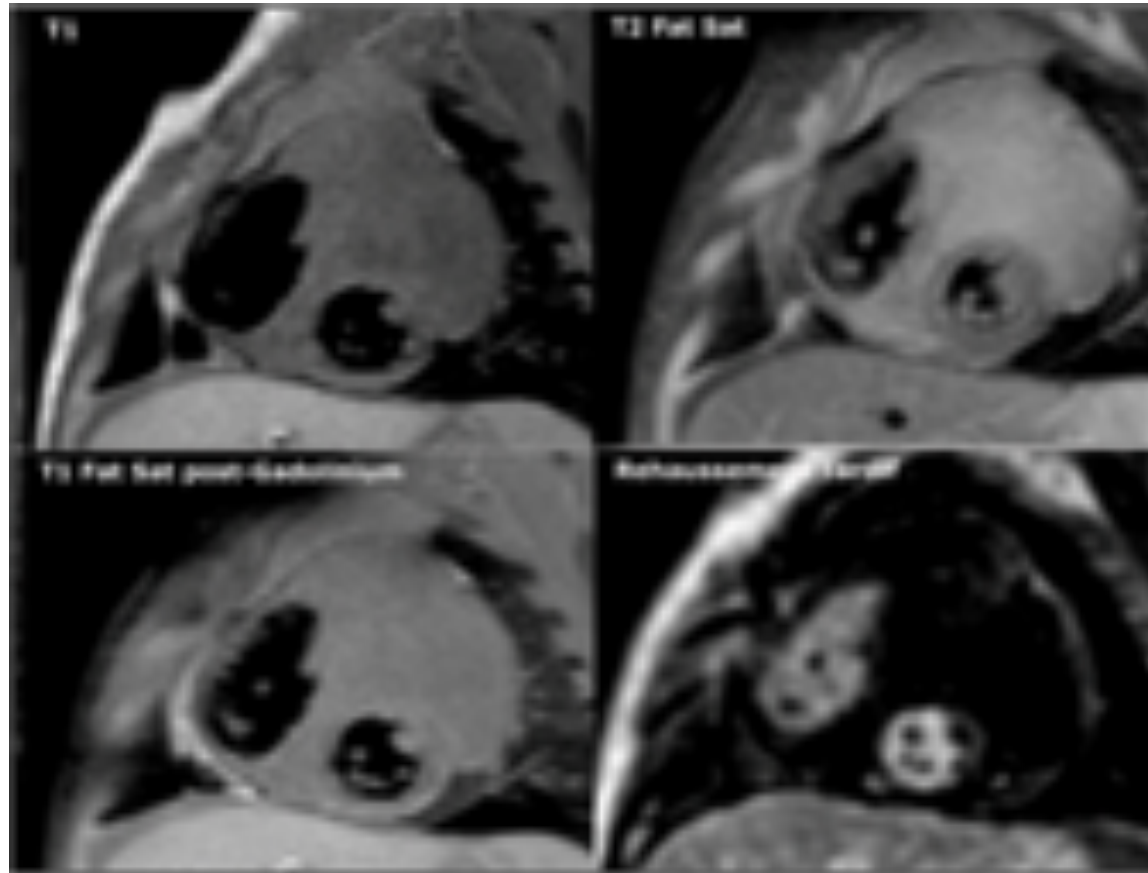
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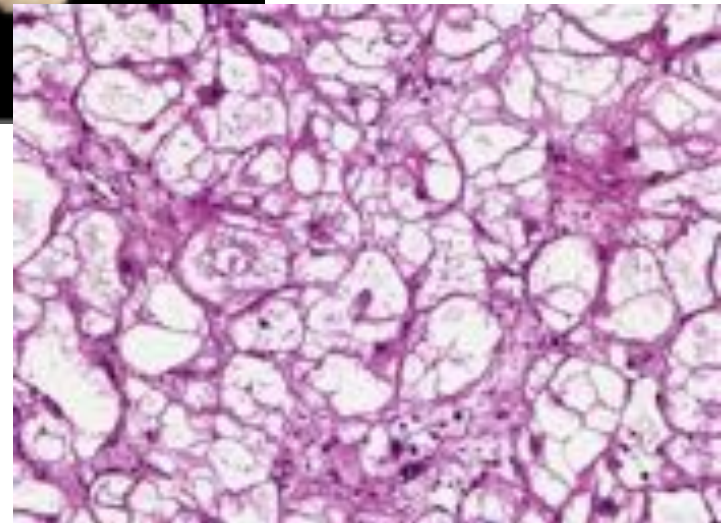


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- En IRM: Masses multiples, myocardiques ou intracavitaires hypodense avec un réhaussement post gadolinium

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Macroscopie: couleur jaune blanchâtre, ferme

Histologie: cellules rondes volumineuses avec fines travées radiaires et vacuoles de glycogènes (« spider cells »)

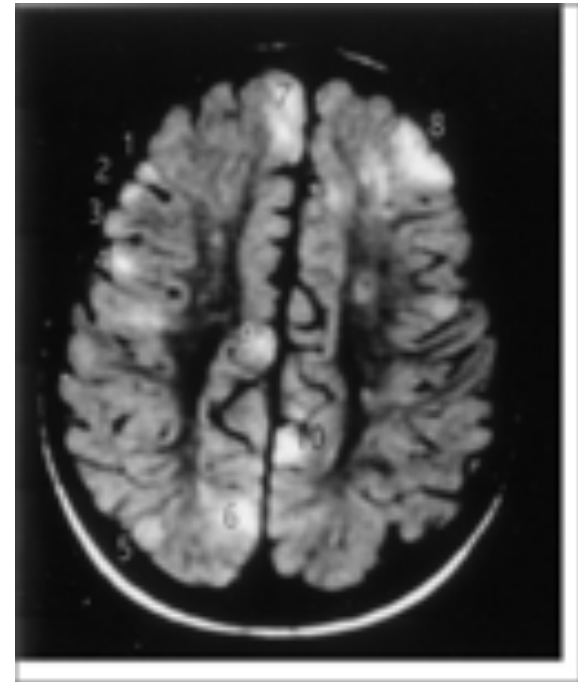
Slérose tubéreuse de Bourneville

1 cas sur 6000 naissances

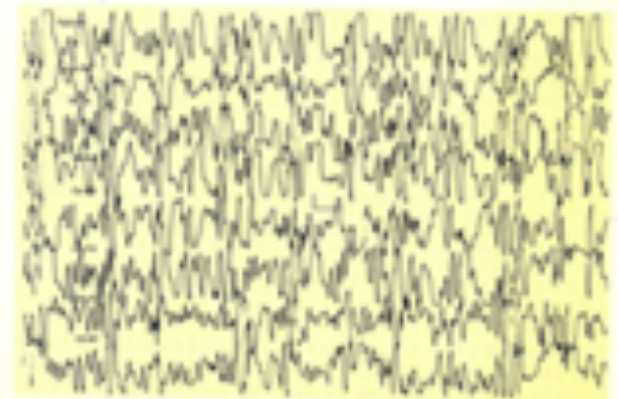
- Maladie autosomique dominante
- TSC1 (chromosome 9): code la protéine hamartine
- TSC2 (chromosome 16): code la protéine tubérine
- Atteinte cardiaque neurologique, dermatologique, néphrologique, ophtalmologique

STB: atteinte neurologique

- Epilepsie précoce (Syndrome de West)
80% des patients.
De tout type (spasme, focale, généralisée...)
Grande résistance aux traitements.
- Régression psychomotrice
- Troubles du comportement retard mental
- Psychose, troubles autistiques
- EEG: Hypsarythmie
- IRM: Tubers, HTIC, Anévrismes cérébraux



Hypsarythmie : activité de base désorganisée
Succession d'ondes lentes, de pointes et de pointes ondes de haut voltage



STB: atteinte dermatologique

- Taches hypomélaniques ou achromiques sur la peau (lumière de Wood)
- Angiofibromes de la face
- Aspect de peau de chagrin du bas du dos
- Tumeurs de Koënen des ongles



STB: atteinte néphrologique

- Angiomyolipome du rein: Tumeurs multiples et bilatérales très vascularisées, hémorragiques
- Kystes épithéliaux ou maladie poly-kystique, pouvant évoluer vers l'IR
- Cancers rénaux (sarcomes) rares mais précoces



Fibrome

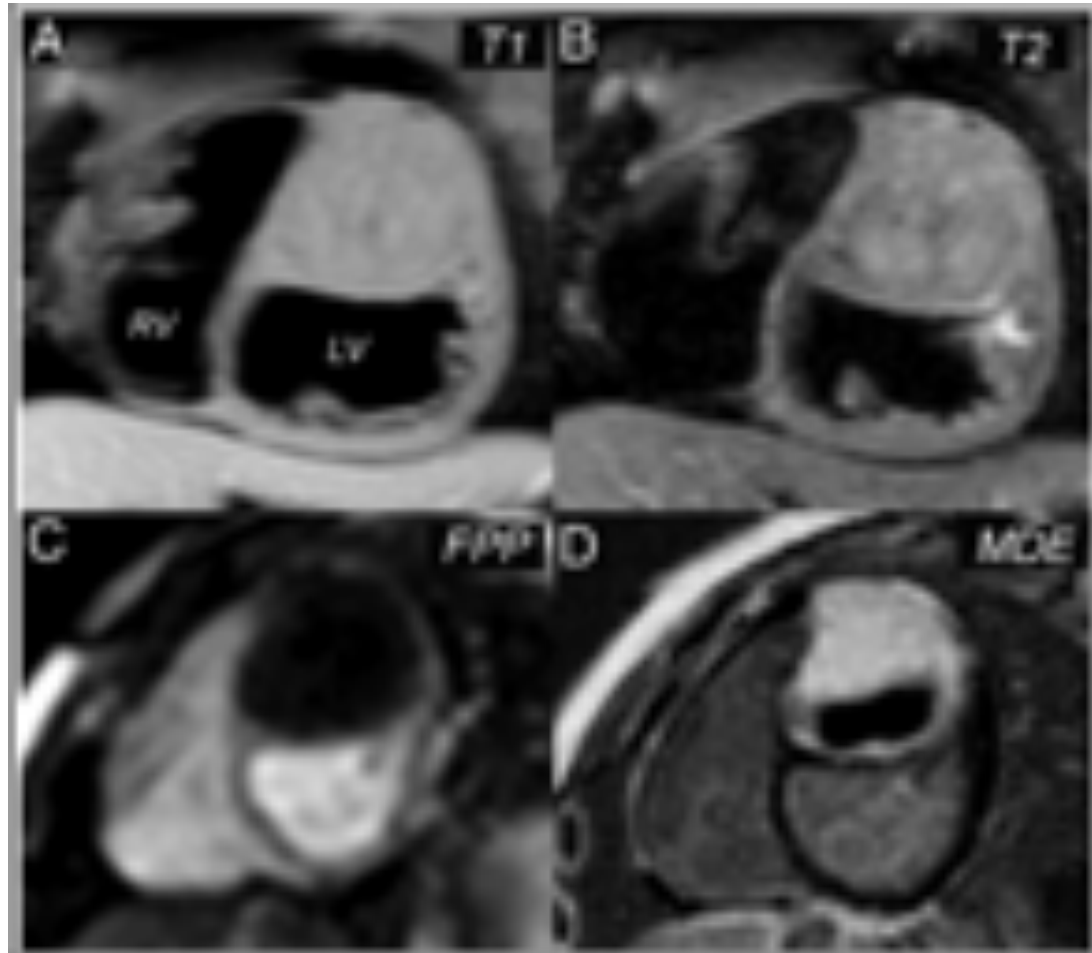
- Tumeur la plus fréquente après le rhabdomyome, 12-16%
- Tumeur unique envahissant le myocarde
- Circonstance de découverte souvent dans l'enfance
- Signes cliniques: Insuffisance cardiaque, troubles du rythme (+++) de conduction, ischémie par compression coronaire, souffle/cyanose obstructif
- **Echographie: localisation surtout ventriculaire**
- Pas de régression spontanée.
- Indication chirurgicale au diagnostic

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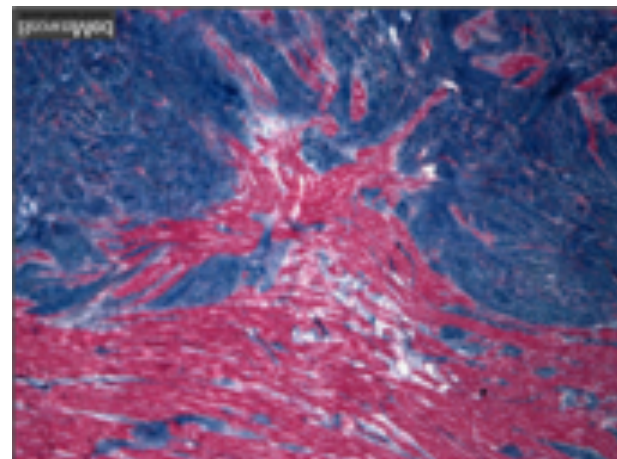
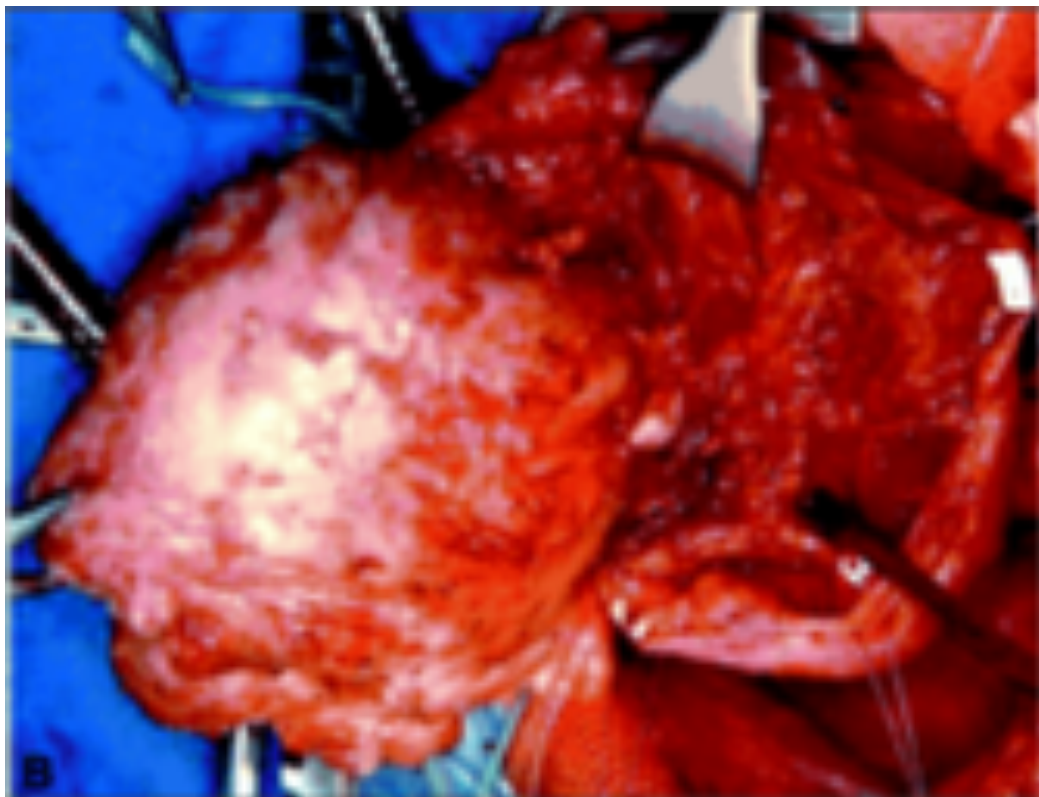
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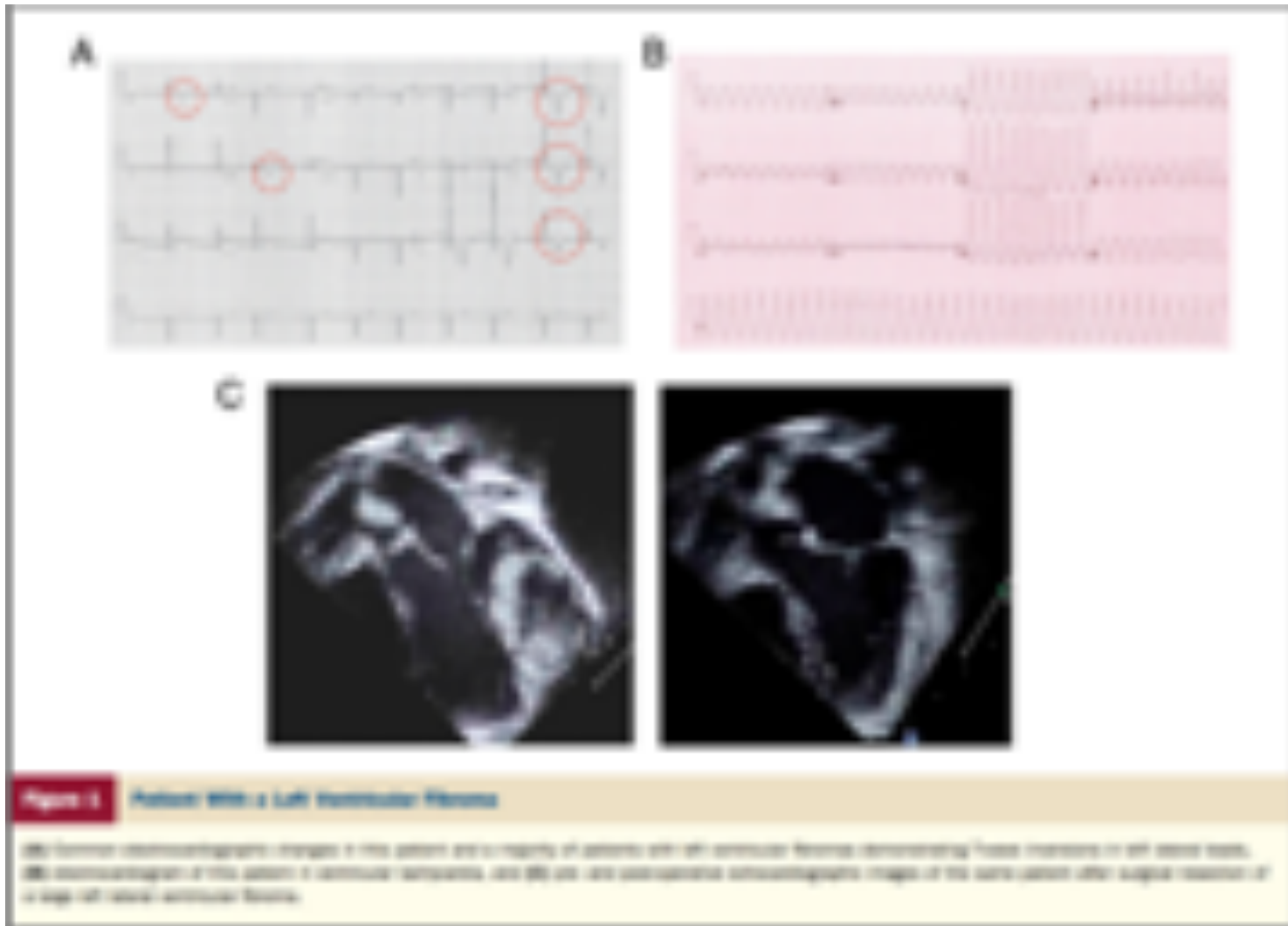
IRM: aspect tissulaire en iso signal T1. Leger hypersignal T2. Prise de contraste avec hypoperfusion centrale



Macroscopie: Masses solides, larges, non contractiles, enchâssées dans l'épaisseur du myocarde sont. L'aspect est ferme, blanc, homogène sans foyer de nécrose ou d'hémorragie.

Histologie: fibroblastes siégeant dans un stroma de collagène. Calcification centrale typique.

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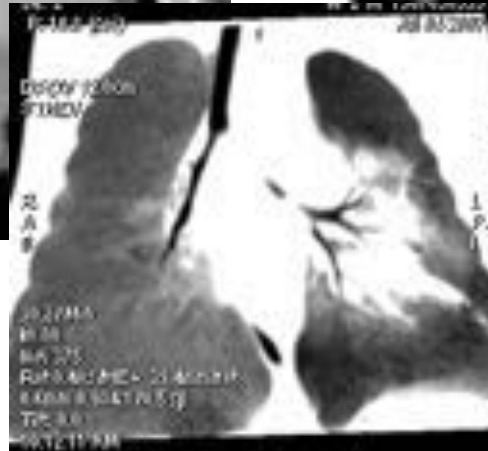
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Table 2 Arrhythmias by Tumor Type

	All Tumors	Rhabdomyoma	Fibroma	Myxoma	Vascular	Teratoma	Lipoma	Other
Patients, n	173	106	25	14	6	4	3	15
Clinically significant arrhythmia	42 (24%)	17 (16%)	16 (64%)	1 (7%)	1 (17%)	0	0	7 (47%)
Cardiac arrest/VF	4 (2%)	—	2 (10%)	—	1 (17%)	—	—	1 (6%)
VT	27 (16%)	6 (6%)	16 (64%)	1 (7%)	—	—	—	4 (27%)
WPW/sustained SVT	2 (1%)	2 (2%)	—	—	—	—	—	—
WPW/no SVT	9 (5%)	8 (8%)	—	—	—	—	—	1 (7%)
Non-WPW sustained SVT	9 (5%)	5 (5%)	—	—	—	—	—	5 (33%)
Low-grade arrhythmia	15 (9%)	13 (12%)	1 (4%)	1 (7%)	—	—	—	—
Any arrhythmia (low-grade + clinically significant)	57 (33%)	30 (28%)	17 (68%)	2 (14%)	1 (17%)	0	0	7 (47%)

Values are n or n (%). Some patients might have had more than 1 arrhythmia. Clinically significant arrhythmias are subdivided by type and defined in text. Low-grade arrhythmias included frequent premature atrial beats, ventricular ectopic beats or couplets, and brief nonsustained supraventricular tachycardia (SVT) in patients without manifest pre-excitation.

VF = ventricular fibrillation; VT = ventricular tachycardia; WPW = Wolff-Parkinson-White syndrome.



Compression arbre respiratoire

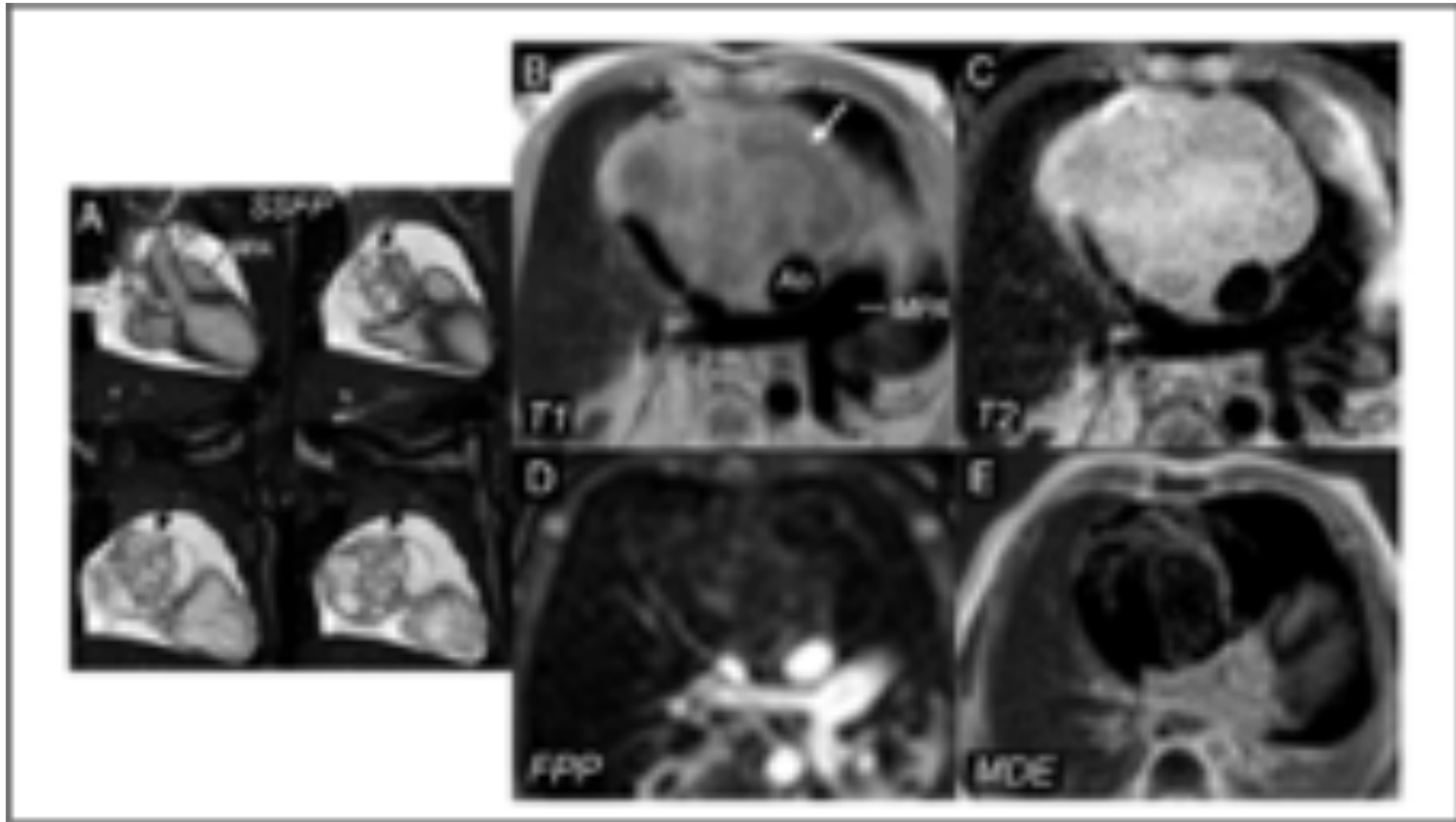
Tératome

- Circonstance de découverte: DAN ou précoce en postnatal
- Tumeur unique, volumineuse. Localisation Cardiaque ou péricardique
- Signes cliniques: Anasarque fœtal, Insuffisance cardiaque droite avec tamponnade post natale, détresse respiratoire par compression pulmonaire
- Echographie: épanchement péricardique, Tumeur unique hétérogène
- Pas de régression spontanée
- Risque rare de dégénérescence maligne
- Indication chirurgicale au diagnostic

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IRM: aspect tissulaire hétérogène en iso signal T1. Hypersignal T2. Hypo-intense après injection de gado

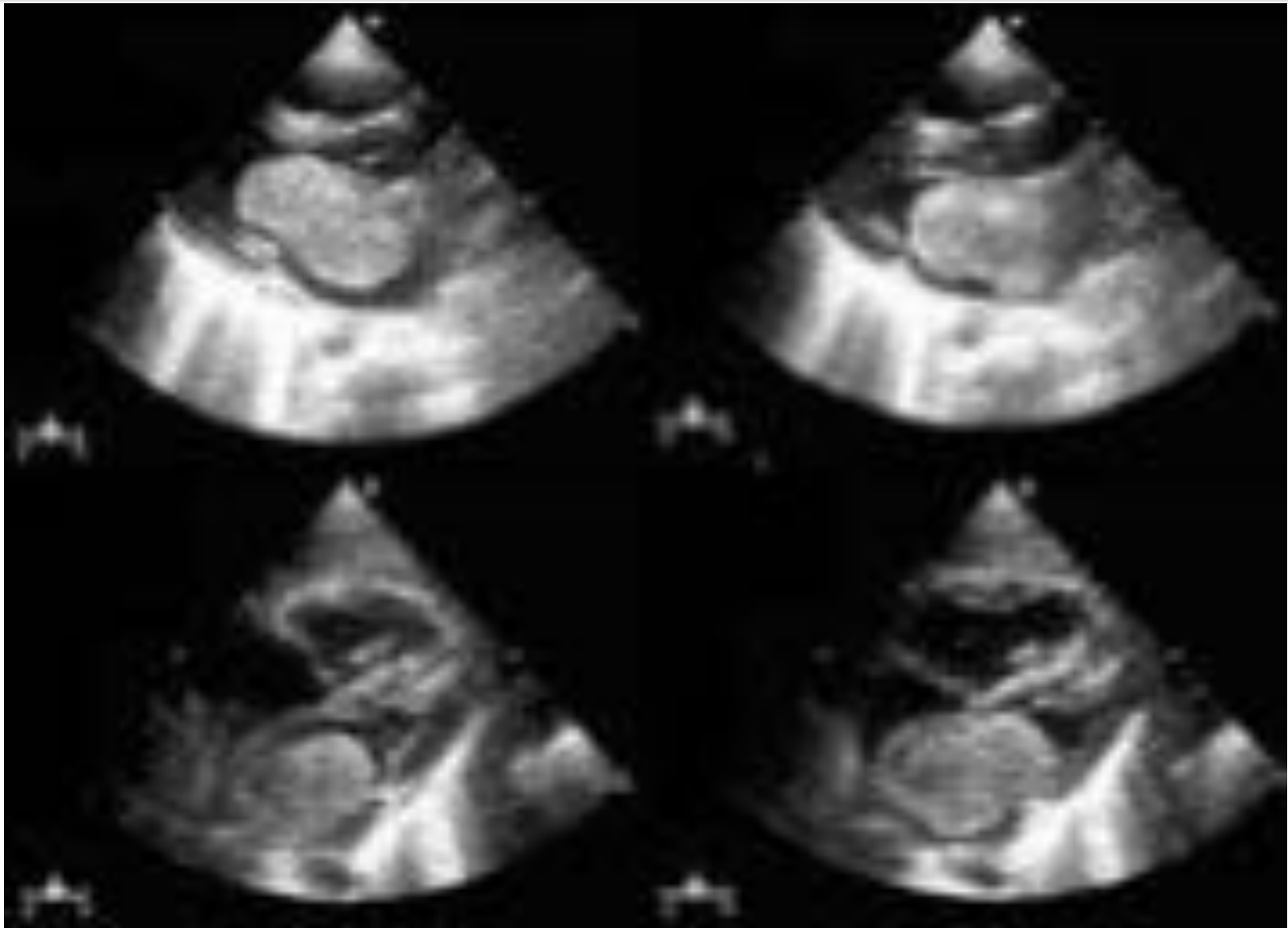


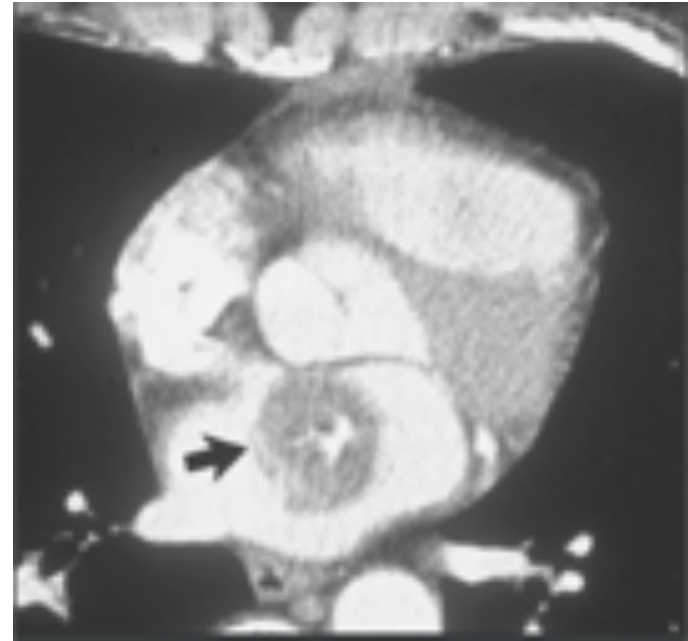
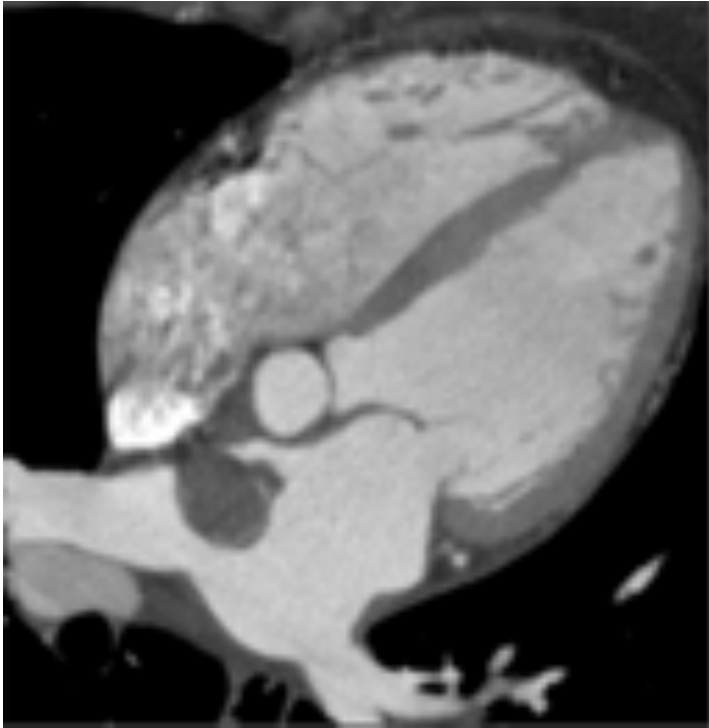
Tumeur germinale, formée par des cellules pluripotentes bien différenciées qui produisent un ou des tissus mûrs mais à des endroits inappropriés. Aspect multikystique et multilobulé associant les tissus des trois lignées embryonnaires

Myxome

- Tumeur rare chez l'enfant 2-4%, Tumeur cardiaque la plus fréquente chez l'adulte 40% avec une prédisposition féminine.
- Tumeur unique localisée dans 90 % des cas dans l'oreillette gauche. Multiples en cas de syndrome.
- Circonstance de découverte: Embolies artérielles (AVC), syndrome d'obstruction intermittente de la mitrale
- Echographie: aspect hétérogène près du foramen ovale. lobulée, pédiculée (pédicule court et large)
- Formes familiales (complexe de Carney: Anomalies pigmentaires de la peau, myxomes, tumeurs endocrines et schwannomes)
- Syndromes endocriniens Phacomatose, lentiginose
- Risque récurrence (5%)
- Résection chirurgicale (complète) au diagnostic

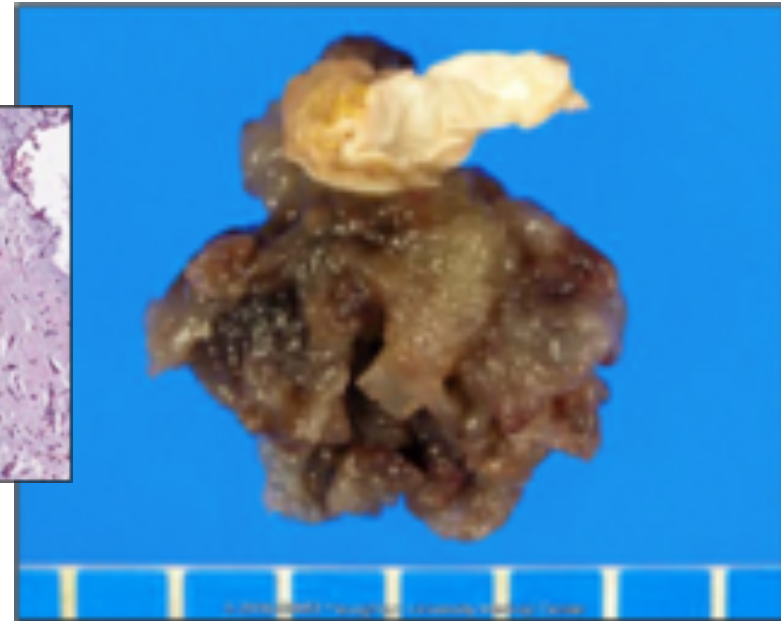
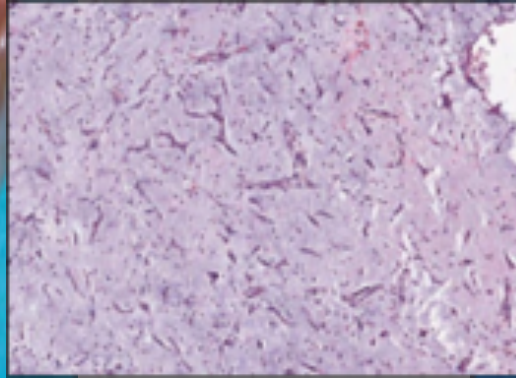
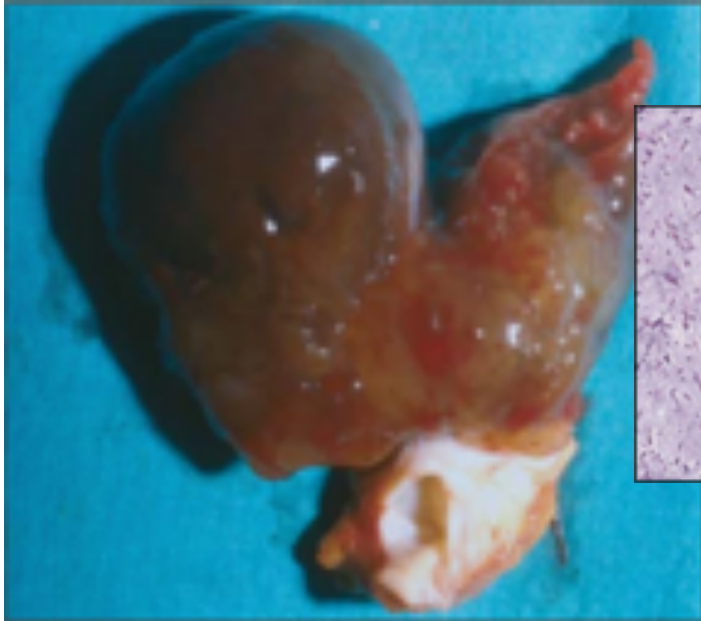
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IRM: Isosignal T1, Hyperintense T2, rehaussement tardif après injection.
Parfois hétérogène (nécrose, hémorragie)

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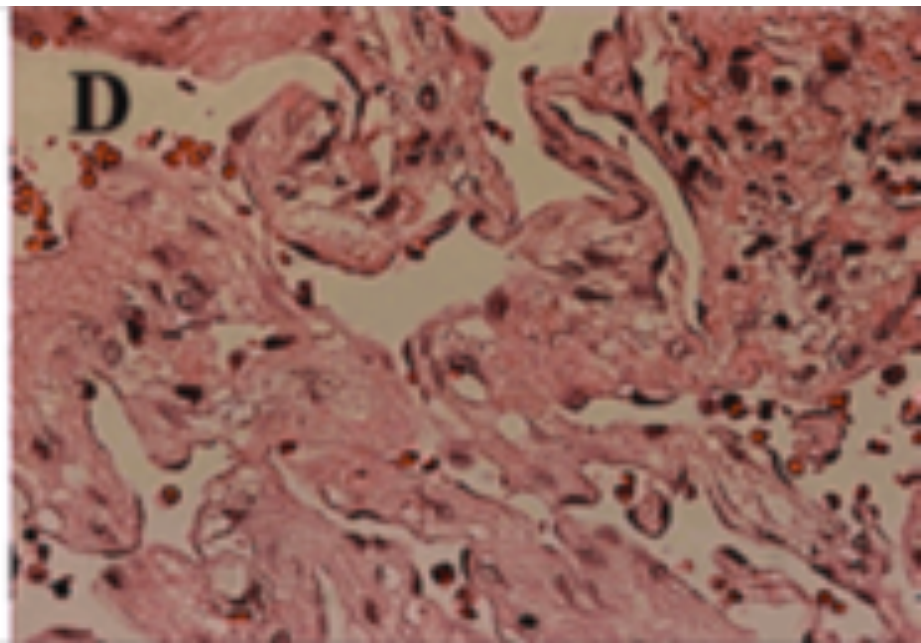
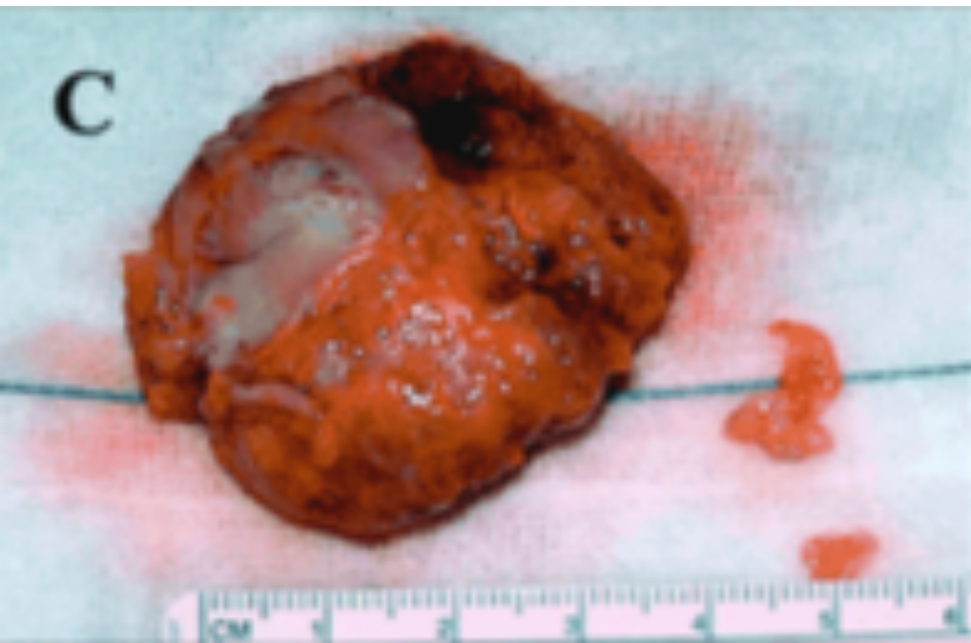


Macroscopie: Unique, lobulé, 2 aspect: gélatineuse, ou fibreuse friable

Histologie: révèle un stroma abondant mucoïde (Bleu alcian positif) au sein duquel se trouvent des cellules polygonales ou arrondies, allongées ou stellaires à cytoplasme peu abondant, éosinophile, regroupées en petits amas ou isolées. Le stroma peut être riche en fibres élastiques ou collagène

Hémangiome

- Tumeur rare (0.0010 à 0.0027%)
- Unique, épicardique ou intra-cavitaire. Développement parfois au dépend du réseau coronaire.
- Signes cliniques: Trouble du rythme ou obstacle ou ischémie sous épicardique en regard de la tumeur
- ETT: Masse échogène, homogène.
- Evolution: Possible involution, calcification, hémopéricarde
- Indication chirurgicale à discuter



Tumeur hamartomateuse

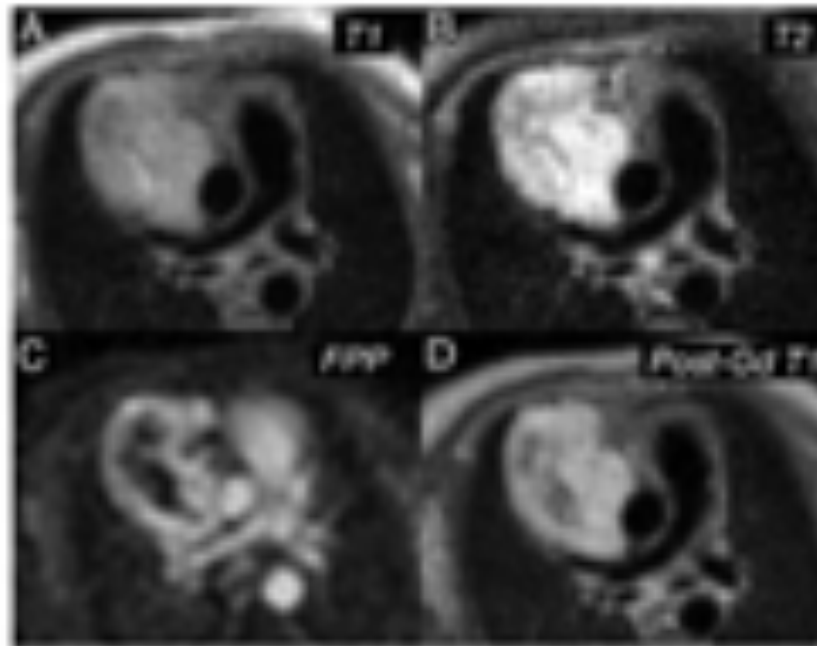


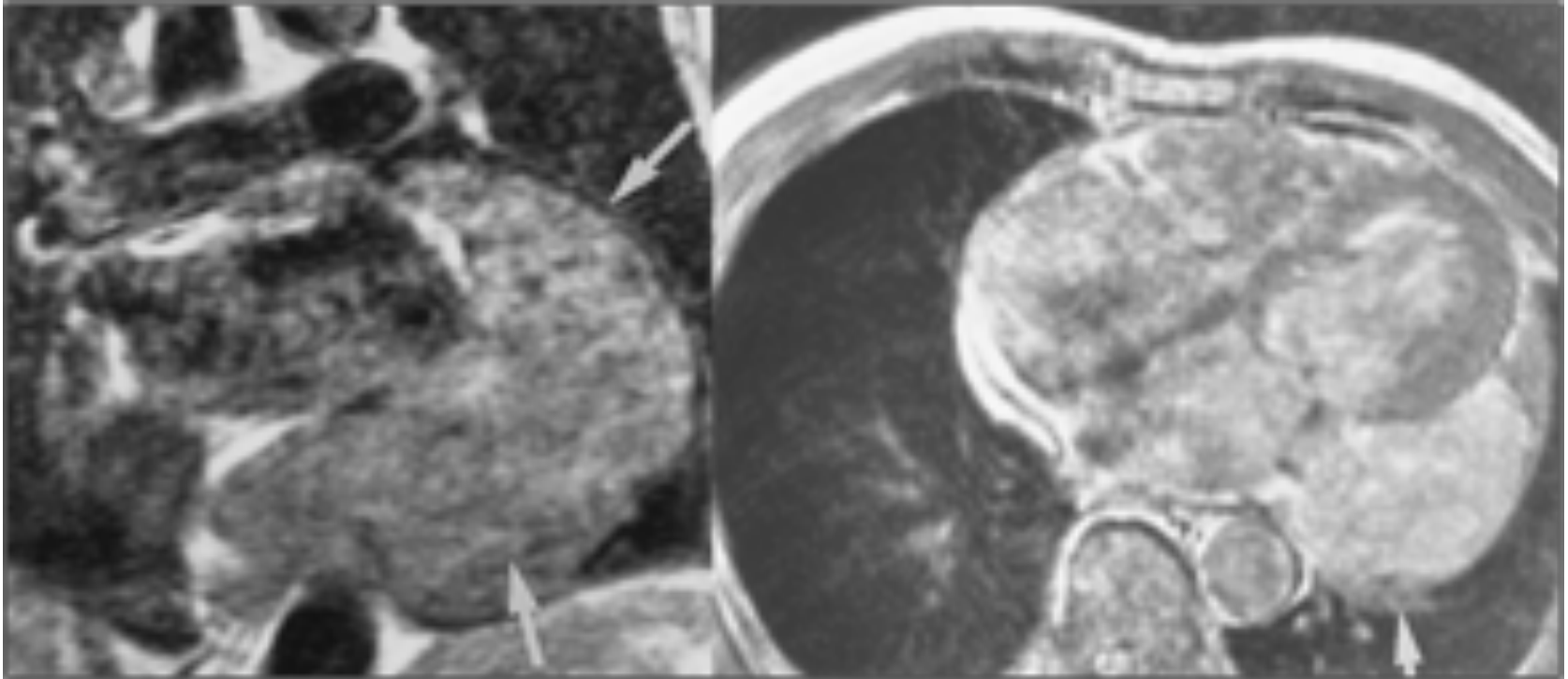
Figure 1 Right Atrial Hemangioma

(A) low to slightly hyperintense signal on T1/T2; (B) strongly hyperintense signal on T2/T2; (C) heterogeneous, strongly hyperintense signal on T1/T1 with avid perfusion of the tumor; and (D) hyperintense signal on post-contrast T1/T1. Abbreviations: A, Atrial; C, Contrast; T1, T1-weighted; T2, T2-weighted.

Hypersignal modéré T1, Hypersignal marqué T2, prise de contraste

Lipome

- Tumeur rare chez l'enfant
- Unique. Intracardiaque, rarement péricarde
- Echographie: Contours régulier hyper-échogène
- Histologie: masse de cellules graisseuses (adipocytes) encapsulées
- Indication chirurgicale en cas d'obstruction



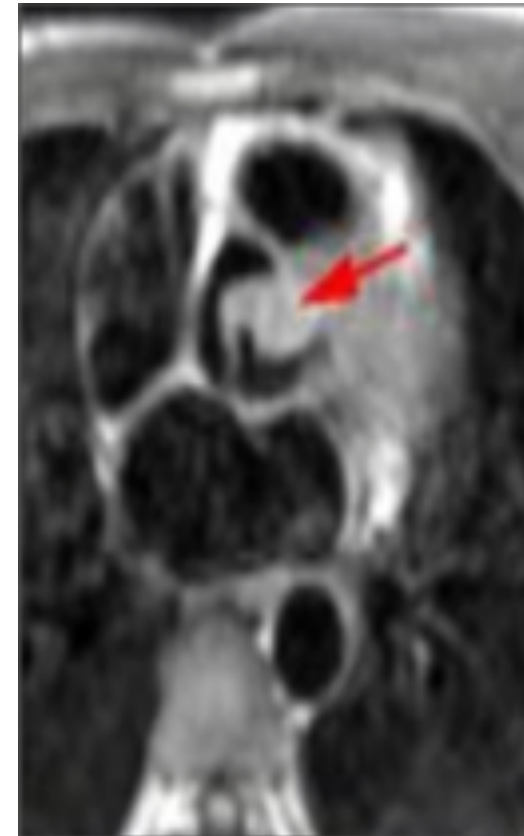
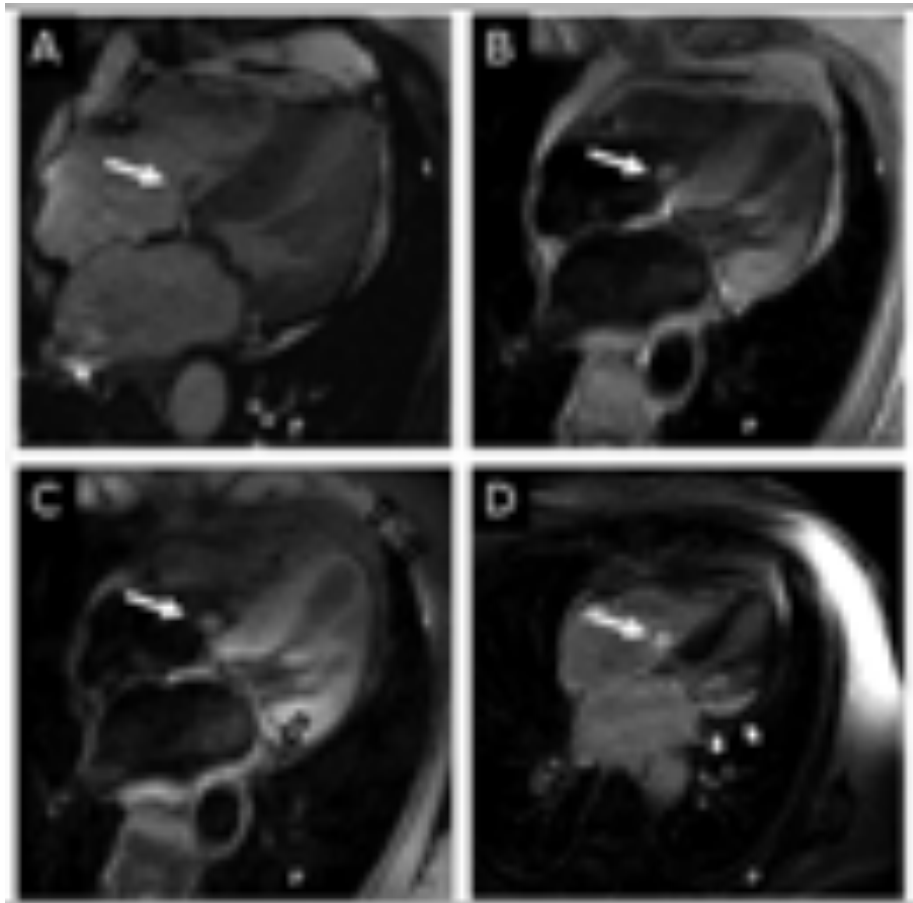
Masse hétérogène multilobulée en isosignal T1 et hypersignal T2

Fibro élastome

- Le fibroélastome est la 3ème variété de tumeur bénigne la plus fréquente chez l'adulte. Très rares chez l'enfant
- 85% sur les valves : aortiques > mitrales > pulmonaires > tricuspides (17%). Rarement multiples
- Echographie: Petite masses pédiculées, mobiles, vibratiles, hyperéchogène
- Tumeur bénigne mais risque obstructif ou embolique
- Indication chirurgicale au diagnostic



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Macroscopie: masse gélatineuse multilobée papillaire « anémone de mer »
Tissu conjonctif recouvert d'endothelium

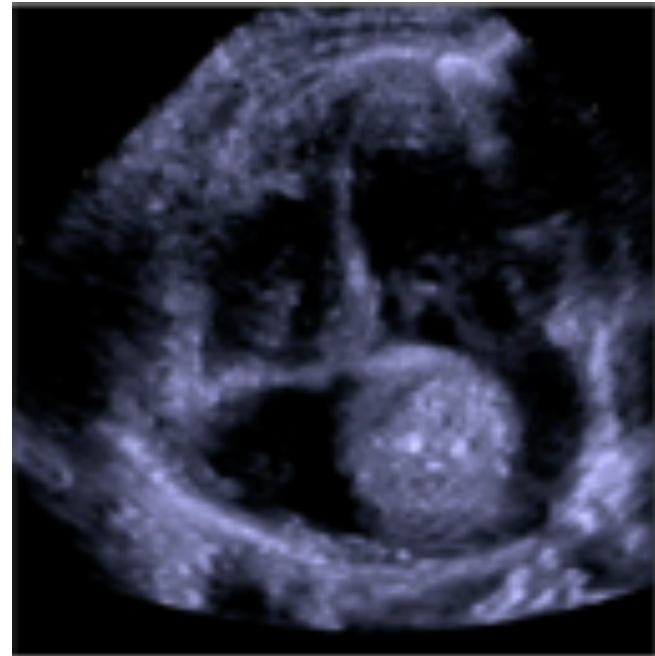
Myofibrome inflammatoire infantile

Syndrome rare 1/150000 naissance,
sporadique ou familial

Myofibromes localisées dans les tissus
sous cutanés et rarement dans les
viscères

Biopsie: fascicules enchevêtrés de
cellules fusiformes spinales
(myofibroblastes) à la périphérie
formant des nodules séparés par du
tissu de collagène sans atypie nucléaire

Résection car diagnostic étiologique non
invasif difficile



Tumeur histiocytoides (tumeur des cellules de Purkinje)

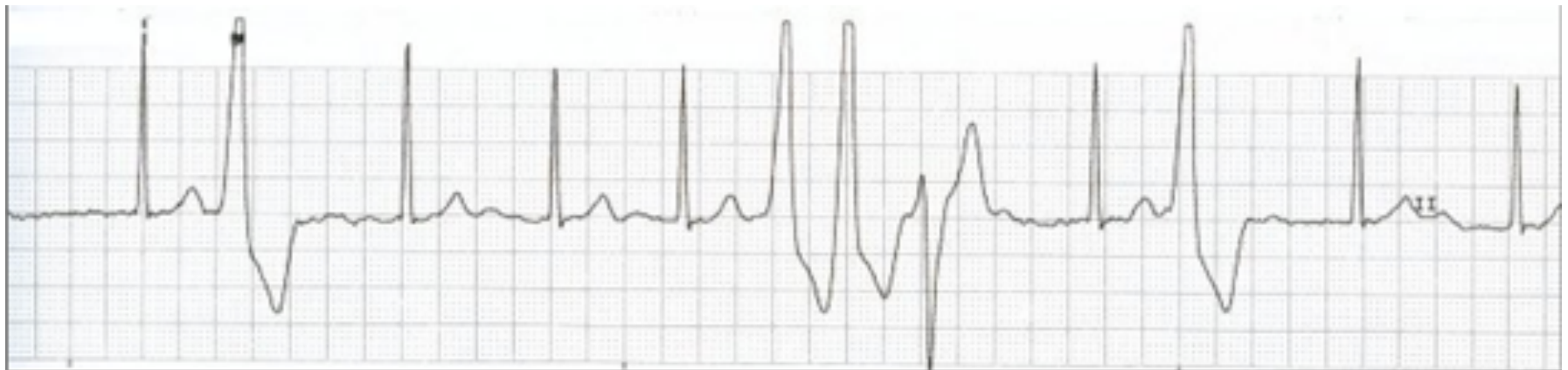
5% de cas familiaux (AR, ou lié à l'X)

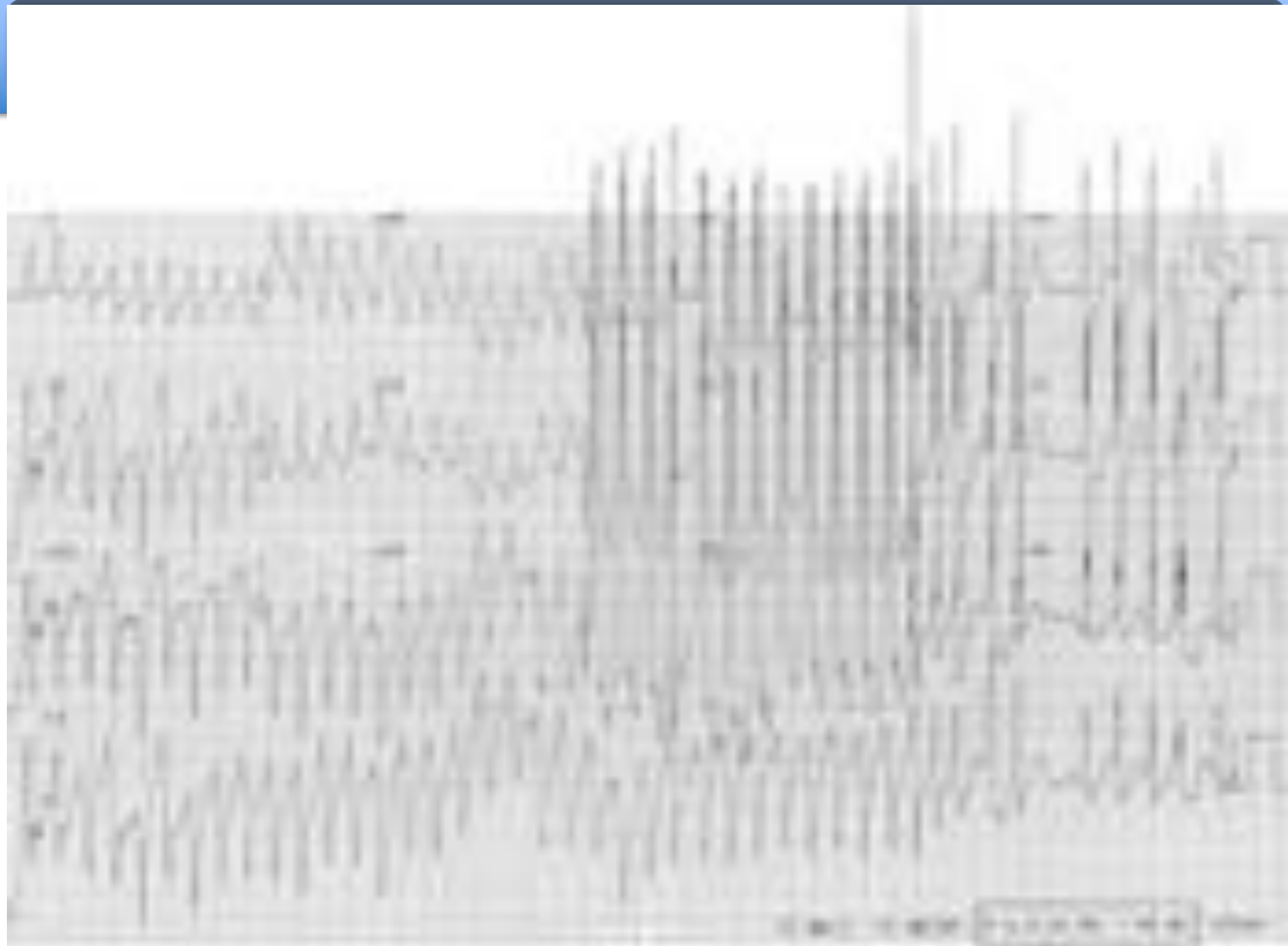
Prolifération hamartomateuse multifocale de cellules cardiaques.

Multiples nodules sous endocardiques jaunâtres mesurant moins de 5 mm, disséminés à tout le cœur

Troubles du rythme ventriculaires polymorphes et supraventriculaires résistants chez le nourrisson pouvant mener à des morts subites.

Pronostic très sévère

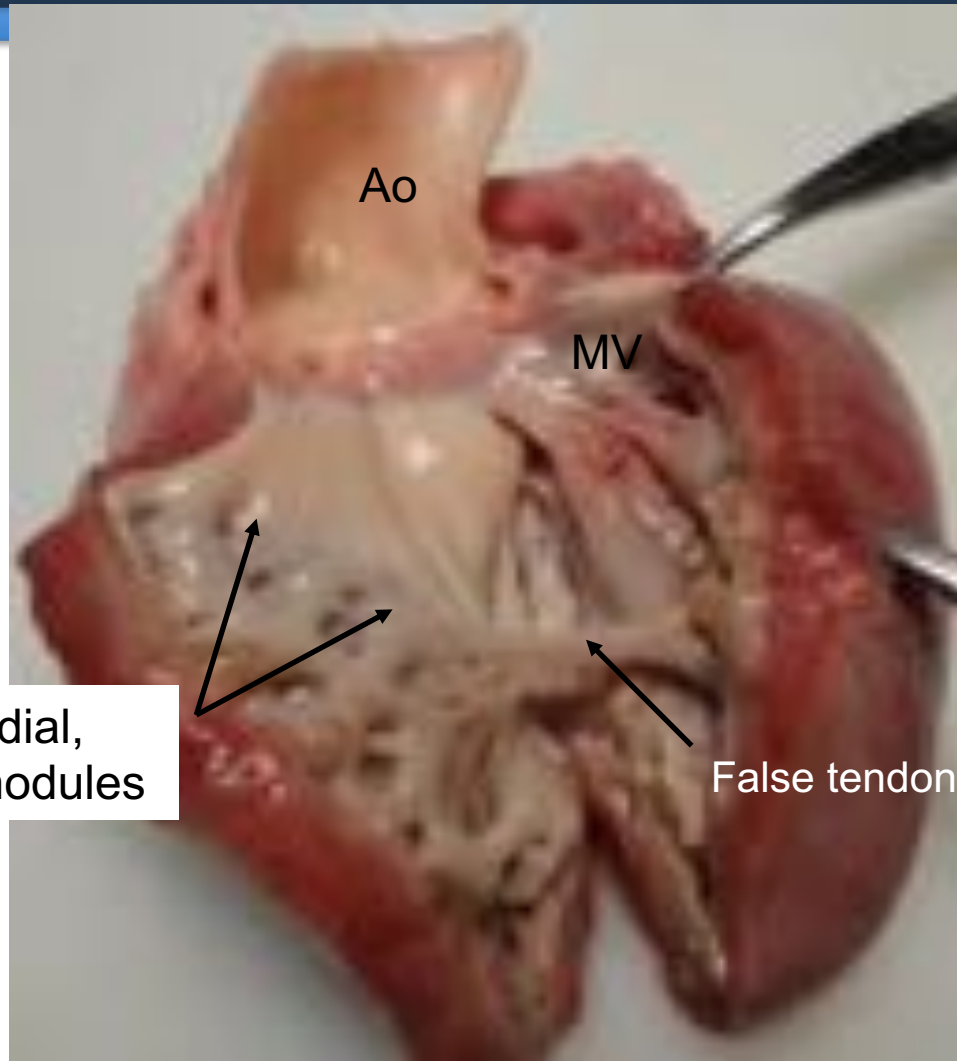






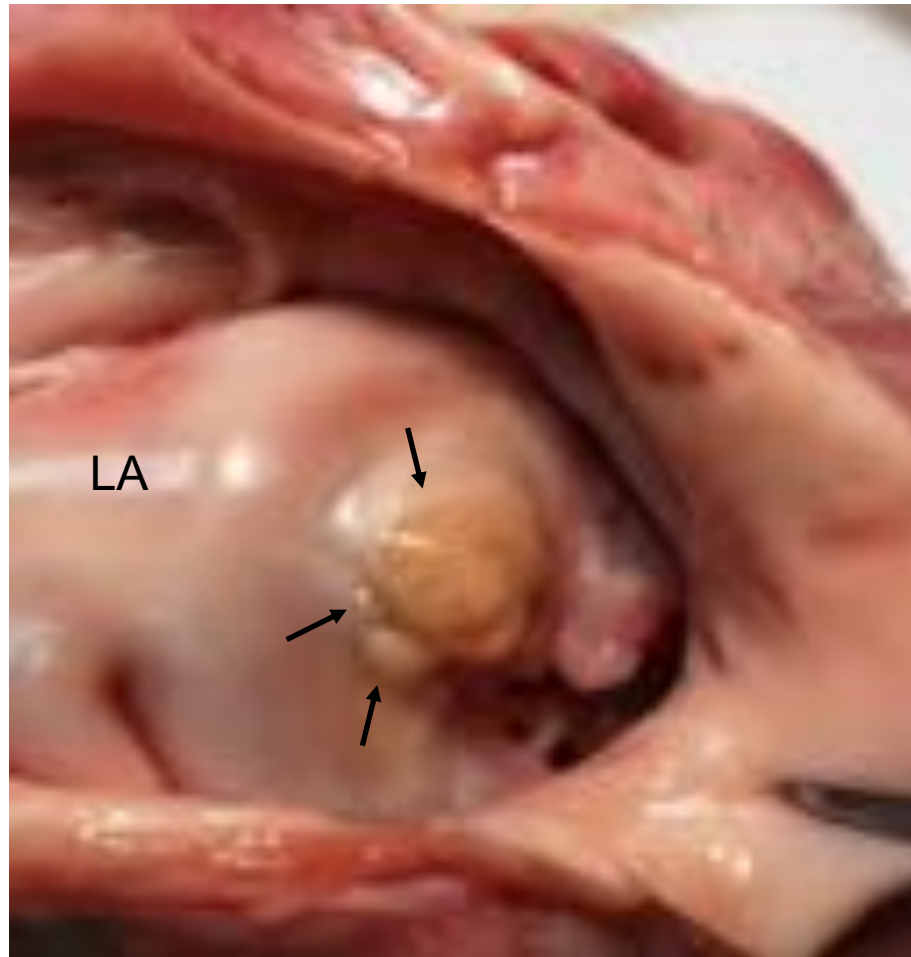
- Exemple de Lily : DAN de TSV non traitée
- A partir de 4 mois, TDR supra-ventriculaire et ventriculaire à répétition et torsades de pointe, avec arrêt cardiaque, hospitalisée en réanimation chirurgicale à 5 mois
- En échographie, elle a une myocardiopathie très modérée, avec une fonction ventriculaire gauche quasi-normale. On observe une image suspecte en échographie, descendant du septum vers l'anneau aortique. Cette image a été retrouvée en IRM. Il y a de nombreuses trabéculations à la pointe du ventricule gauche, et il n'y a aucune obstruction sous-aortique.
- BEM à 5 mois : arguments en faveur d'une cardiomyopathie histiocytoïde.
- Inscription sur liste de transplantation à 6 mois (5,5 kg) malgré réticence de l'équipe (pas d'assistance)

Décès 12 jours après (6 mois de vie) de TDR Ventriculaires réfractaires

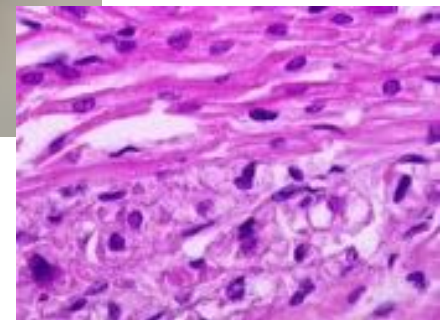
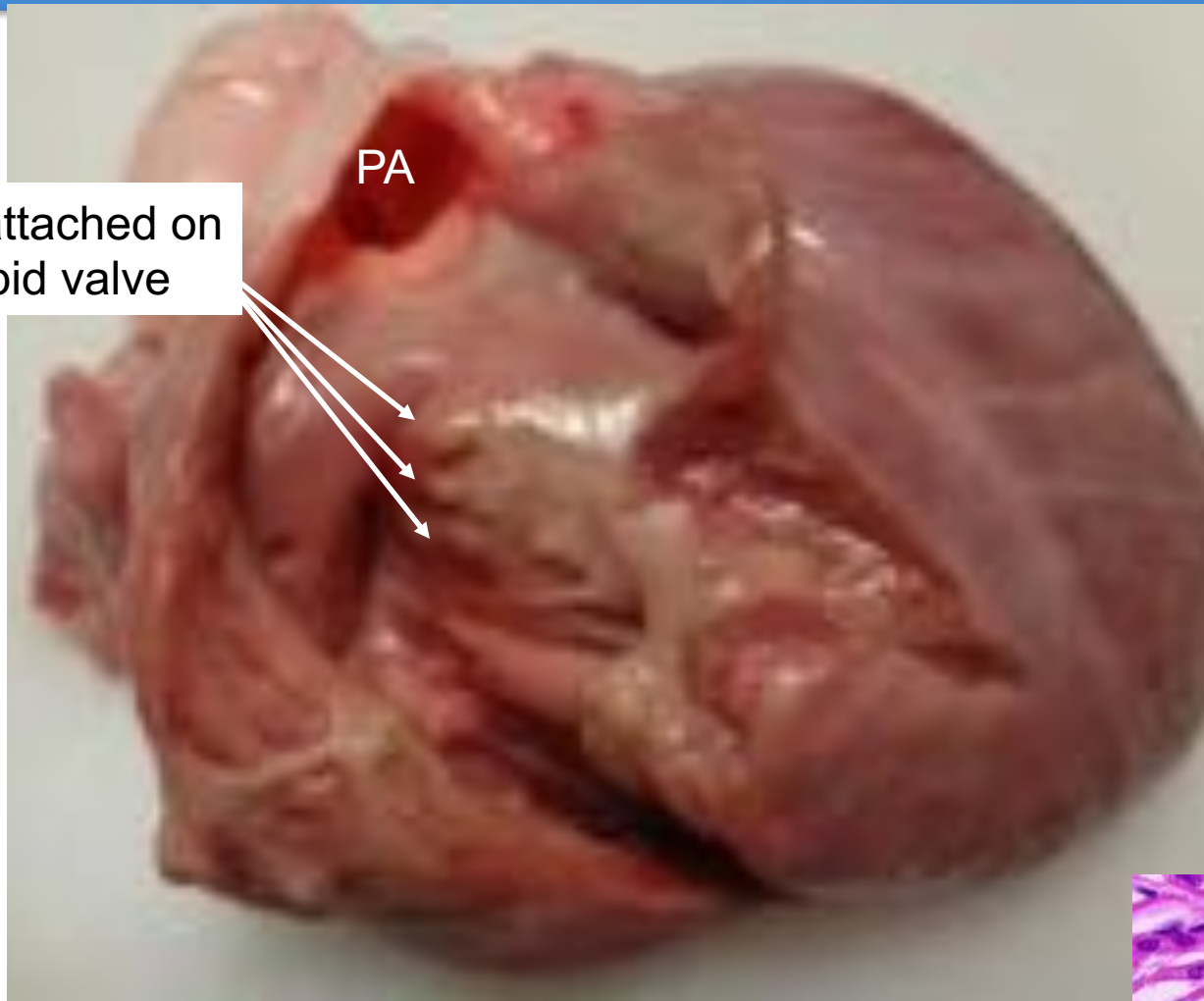


subendocardial, epicardial,
or valvular yellow-tan nodules

False tendon



3 tumors attached on
the tricuspid valve



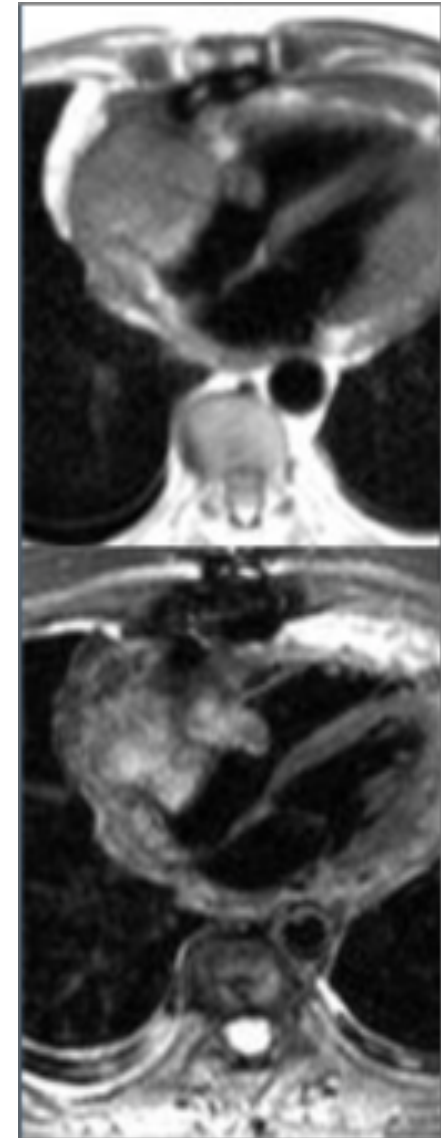
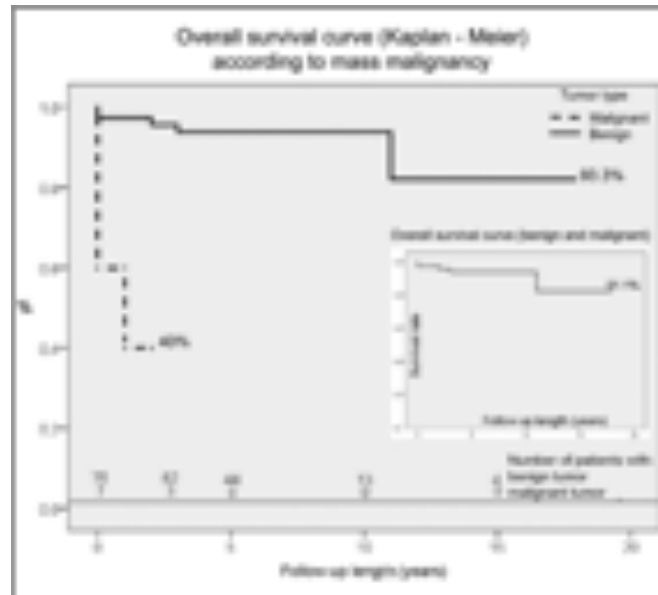
Tumeurs malignes

10%, rares

Rhabdomyosarcomes, fibrosarcome, angiosarcomes et lymphomes, métastases (mélanome).

Tumeurs agressives, infiltrantes, métastasent à distance, comportant des foyers de nécrose, entraînant des épanchement ou des hémorragies péricardique,

Mauvais pronostic

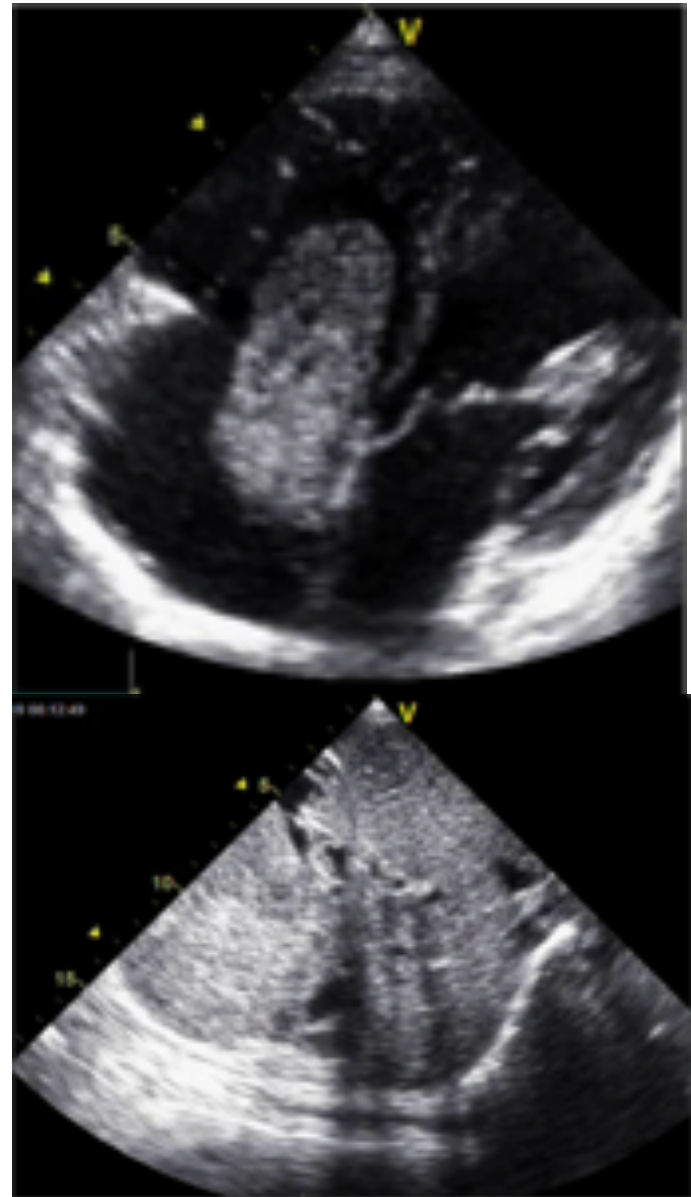


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Tumeur secondaire: Extension cardiaque du néphroblastome

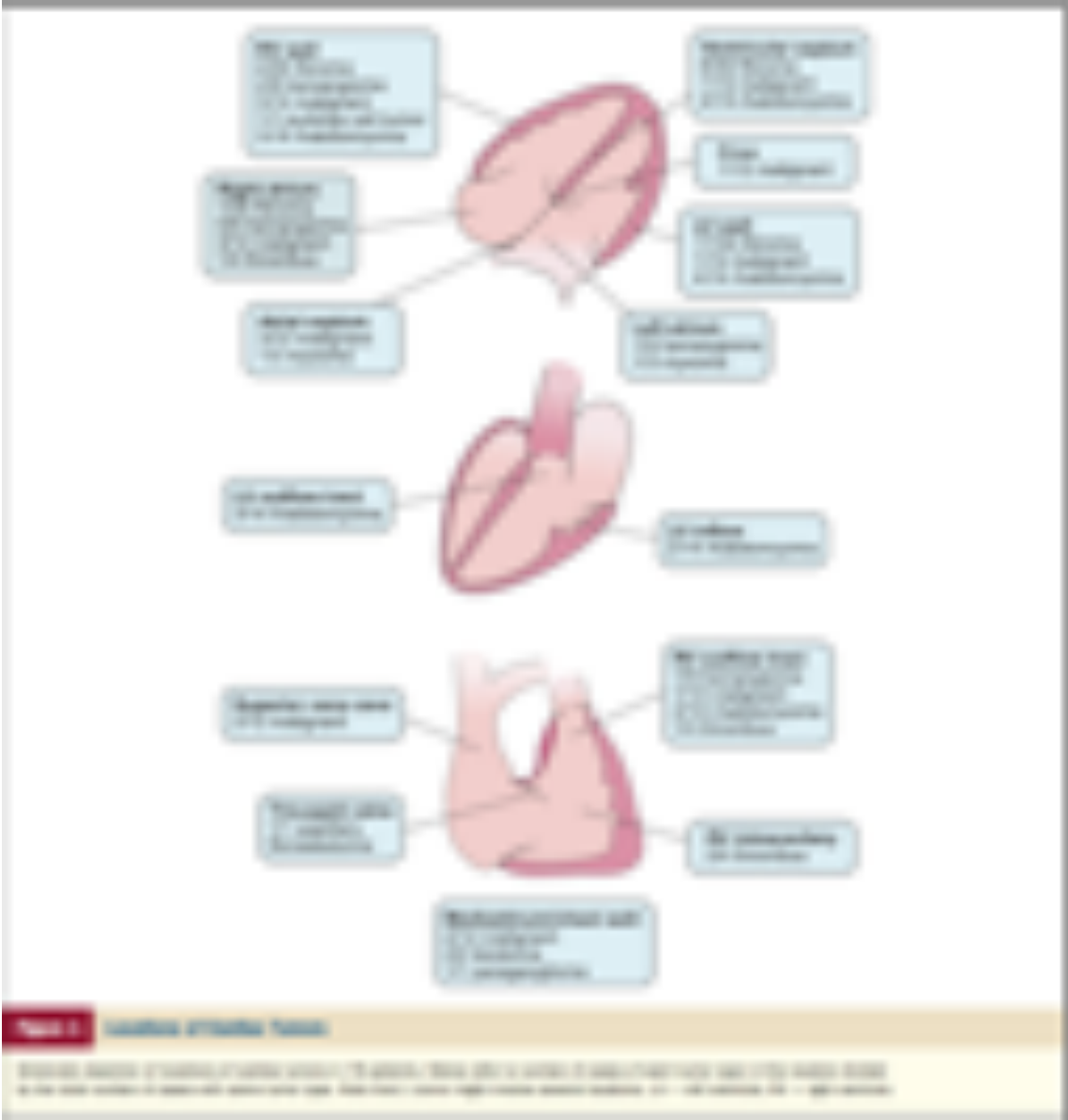
- Extension tumorale jusque dans l'OD
- Risque d'enclavement dans la tricuspide
- Chimiothérapie de réduction en urgence à + CEC en urgence



Comment faire le diagnostic sans anapath?

- Nombre de tumeur
- Localisation
- Aspect échographique
- Complications
- Aspect en IRM
- Atteinte extracardiaque

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IRM

Table 1. Tumor Diagnostic Predictive Table

Tumor Type	Location	SSFP	T2	T2 + Fat Sat	T2*	SSFP	MRV	Other
Fibroma	Intracardiac, ventricular septum or free wall	++	+	+	+	Ref ¹	++ (well-defined border) or both absent ²	Can be in an atypical location
Rhabdomyoma	Intracardiac or intramural, attached to septum/wall	++	+	+	+	Ref ¹	---	
Myxoma	Subvalvular		+		+	Variable	++ or ++ (large heterogeneous appearance)	History of myxomas
Teratoma	Variable	++	---	---	++ (enhanced)	Strong ²	++ variable and heterogeneous	Consider malignant tumor
Leiomyoma	Mural or intraluminal ³	++	---	---	---	Ref ¹	---	MRV negative, long duration flow
Myosarcoma	Typically left atrium but can be in any chamber	++	+	+	+	Ref ¹	++	Improve, unenhanced, subtle ²
Angiosarcoma	Pericardial, mural, subendocardial or subvalvular mass	++	+	+	+	Ref ¹		
Thrombus/mural clot	Right ventricular angle	++ or +	+	+	++ or +	Ref ¹	+	Smooth surface and well-defined
Myxoid wall tumor	Subvalvular septum/wall	++ or +	---	---	+	Ref ¹		Subvalvular septum/wall ²
Leiomyoma	Intramural (usually compressing RV and/or LV)	++				Ref ¹		Multifocally distributed mass with wall and septal areas
Coronary	Any chamber	++	++ or +	---	+	Ref ¹	---	

Notes: strongly suggestive if ++ response for diagnosis. Indeterminate if imaging is equivocal or demonstrates features with the least 2 imaging with cardiac and subcardiac structures or 3 components of flow present through a large vessel such as the aorta or inferior vena cava. Indeterminate refers to normal wall among discrete masses, including thrombus, malignant masses, tumors, and congenital lesions. (Sarcoidosis can not occur, however, in cases of myocardial disease and included.)

++ = definite hyperintense; + = variable intensity; - = hypointense; ++ = strong hyperintense; Ref¹ = Ref. 1; Ref² = Ref. 2; Ref³ = Ref. 3; Ref⁴ = Ref. 4; Ref⁵ = Ref. 5; Ref⁶ = Ref. 6; Ref⁷ = Ref. 7; Ref⁸ = Ref. 8; Ref⁹ = Ref. 9; Ref¹⁰ = Ref. 10; Ref¹¹ = Ref. 11; Ref¹² = Ref. 12; Ref¹³ = Ref. 13; Ref¹⁴ = Ref. 14; Ref¹⁵ = Ref. 15; Ref¹⁶ = Ref. 16; Ref¹⁷ = Ref. 17; Ref¹⁸ = Ref. 18; Ref¹⁹ = Ref. 19; Ref²⁰ = Ref. 20; Ref²¹ = Ref. 21; Ref²² = Ref. 22; Ref²³ = Ref. 23; Ref²⁴ = Ref. 24; Ref²⁵ = Ref. 25; Ref²⁶ = Ref. 26; Ref²⁷ = Ref. 27; Ref²⁸ = Ref. 28; Ref²⁹ = Ref. 29; Ref³⁰ = Ref. 30; Ref³¹ = Ref. 31; Ref³² = Ref. 32; Ref³³ = Ref. 33; Ref³⁴ = Ref. 34; Ref³⁵ = Ref. 35; Ref³⁶ = Ref. 36; Ref³⁷ = Ref. 37; Ref³⁸ = Ref. 38; Ref³⁹ = Ref. 39; Ref⁴⁰ = Ref. 40; Ref⁴¹ = Ref. 41; Ref⁴² = Ref. 42; Ref⁴³ = Ref. 43; Ref⁴⁴ = Ref. 44; Ref⁴⁵ = Ref. 45; Ref⁴⁶ = Ref. 46; Ref⁴⁷ = Ref. 47; Ref⁴⁸ = Ref. 48; Ref⁴⁹ = Ref. 49; Ref⁵⁰ = Ref. 50; Ref⁵¹ = Ref. 51; Ref⁵² = Ref. 52; Ref⁵³ = Ref. 53; Ref⁵⁴ = Ref. 54; Ref⁵⁵ = Ref. 55; 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Types	Complications
Rhabdomyome	Obstacle hémodynamique Trouble du rythme
Fibromes	Obstacle hémodynamique Trouble du rythme
Myxomes <small>Rare chez l'enfant, possible caractère génétique</small>	Dysfonction valvulaire Embolies
Tératomes intra péricardique	Epanchement péricardique

Prise en charge

Abstention thérapeutique: En cas de tumeur à potentiel régressif, ou absence de complication, tumeur bénigne non évolutive

Indication chirurgicale:

- Hémodynamique: Obstacle, fuite valvulaire, épanchement péricardique, insuffisance ventriculaire
- Rythmique: Trouble du rythme réfractaire ou trouble conducteur de haut degré
- Compression: Médiastinale, bronchique, coronaire
- Embolie artérielle
- Prophylactique: Risque d'embolie, Risque de dégénérescence

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Table 3. Surgical indication for surgery according to histological type

Indication	Total	Rhabdomyoma	Fibroma	Teratoma	Myxoma	Inflammatory myofibroblastic tumor	Lipoma	Fibrosarcoma	Hemangioma
Hemodynamic impairment	10	12	6	6	4	2	0	1	1
Obstruction	20	11	2	2	2	2	0	1	0
Valve impairment	3	1	2	0	2	0	0	0	0
Myocardial dysfunction	1	0	1	0	0	0	0	0	0
Pericardial effusion	6	1	1	2	0	0	0	0	1
Prophylactic	7	1	0	1	1	2	0	0	0
Rhythm or conductive disorders	5	2	2	0	0	0	1	0	0
Bronchial compression	2	1	1	0	0	0	0	0	0
Coronary compression	1	1	0	0	0	0	0	0	0
Embolism	2	0	0	0	2	0	0	0	0

Quelle chirurgie?

Exérèse complète:

Tumeur limitée, sans envahissement massif du myocarde ou des structures cardiaque.
Pas de potentiel évolutif. Risque de récurrence.

Exérèse partielle:

En cas de risque de lésion d'une structure cardiaque (valve, coronaire) ou en cas de risque de chirurgie délabrante (paroi latérale VG)

Chirurgie sans Exérèse:

Chirurgie palliative type blalock en cas de tumeur obstructive mais potentiel régressif (rhabdomyome +++)

Pace maker: En cas de trouble conducteur et tumeur bénigne sans indication d'exérèse

Transplantation:

Tumeur volumineuse non réséquable, Insuffisance cardiaque terminale ou trouble du rythme cardiaque réfractaire malgré résection ou traitement médical