Cardiac tumors in children

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Overview of cardiac tumors in children

Primary cardiac tumors

- Incidence 0.0017%-0.19% (100 to 1000 less than secondary cardiac tumors)
- Multiple histotypes
- Outcome usually favorable but can create mechanical complications (obstructions) or cause arrhythmias

Presenting symptoms of cardiac tumors

Clinical manifestation	Cases (N)	Percentage (%)
None	41	24.7
Heart murmur	54	32.5
Shortness of breath	13	7.8
Arrhythmia	11	6.6
Pericardial effusion	9	5.4
Twitch	6	3.6
Edema	3	1.8
Syncope	3	1.8
Embolism	2	1.2
Cyanosis	1	0.6
Others	23	13.9
Total	166	100.0

Prenatal diagnosis is frequent for rhabdomyomas, and teratoma.
Asymptomatic in 57% of cases
Sudden death or severe arrhythmias are rare

Shi L et al. Eur J Pediatr 2017;176:253-60

Pathological classification and frequency distribution of cardiac tumors in children

Primary Benign	*Frequency (%)*
Rhabdomyoma	40-60
Teratoma	15-19
Fibroma	12-16
Myxoma	2-4
Haemangioma	5
Lymphangioma	}
Haemangiopericytoma	} Very rate
Oncocytic tumours	}
Primary Malignant	
Rabdomyosarcomas	2
Fibrosarcoma	2
Secondary Metastatic tumours	
Neuroblastoma	}
Leukaemia	} very
Lymphoma	} rare
Melanoma	}

Mainly benign tumors
Mainly primary
30% operated
Mortality 3%

Uzun O et al. Orphanet J Rare Disease 2007;2:11
Shi L et al. Eur J Pediatr 2017;176:253-60

Age at diagnosis according to tumor histotype

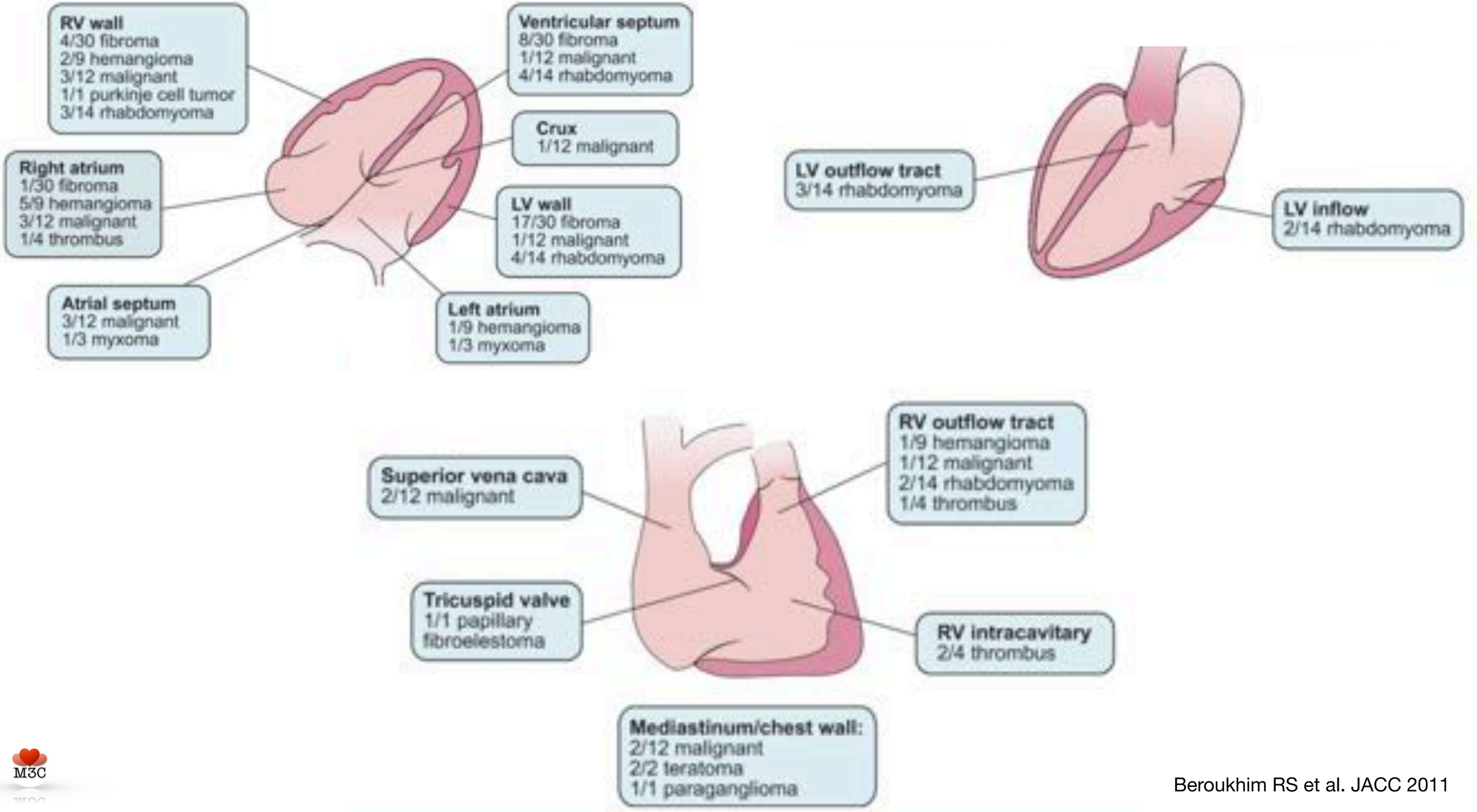
	% of Group		
TUMOR	Adults	Children	Infants
Myxoma	**46**	15	0
Lipoma	21	0	0
Papillary fibroelastoma	16	0	0
Rhabdomyoma	2	**46**	**65**
Fibroma	3	15	12
Hemangioma	5	5	4
Teratoma	1	13	18
Mesothelioma of AV node	3	4	2
Granular cell tumor	1	0	0
Neurofibroma	1	1	0
Lymphangioma	1	0	0
Hamartoma	0	1	0

Affected locations of cardiac tumors

Type	Cases (*N*)	Location (*N*)									
		LA	LV	RA	RV	VS	MV	TV	MPA	Pericardium	Multiple
Rhabdomyoma	100	1	15	1	21	6	–	1	–	–	55
Myxoma	15	8	1	1	3	–	1	–	–	–	1
Fibroma	21	–	10	2	5	3	–	–	1	–	–
Hemangioma	6	–	1	3	1	–	–	–	–	1	–
Lipoma	5	–	2	1	1	1	–	–	–	–	–
Papillary fibroma	2	–	–	–	–	–	2	–	–	–	–
Pericardial cyst	1	–	–	–	–	–	–	–	–	1	–
Fibrosarcoma	2	–	–	–	–	–	–	–	–	1	1
Malignant mesothelioma	2	–	–	–	1	–	–	–	–	1	–
Lymphoma	2	–	–	2	–	–	–	–	–	–	–
Rhabdomyosarcoma	1	–	–	–	–	–	–	–	–	1	–
Undifferentiated sarcoma	1	–	–	–	–	–	–	–	–	1	–
Adrenocortical carcinoma	1	–	–	1	–	–	–	–	–	–	–
Renal clear cell sarcoma	1	–	–	1	–	–	–	–	–	–	–
Wilms' tumor	1	–	–	1	–	–	–	–	–	–	–
Yolk sac tumor	1	–	–	1	–	–	–	–	–	–	–
Malignant epithelial cell carcinoma	1	–	1	–	–	–	–	–	–	–	–
Hepatoblastoma	1	1	–	–	–	–	–	–	–	–	–
Unknown	2	–	1	–	1	–	–	–	–	–	–
Total	166	10	31	14	33	10	3	1	1	6	57

Mainly ventricular walls
Followed by atrial walls
Unique except for rhabdomyomas

Shi L et al. Eur J Pediatr 2017;176:253-60



Beroukhim RS et al. JACC 2011

Tumor diagnosis prediction using MRI

Table 1 Tumor Diagnosis Prediction Table

Tumor Type	Location	SSFP	T1	T1 + Fat Sat	T2	FPP	MDE	Other
Fibroma	Intramyocardial, ventricular septum or free wall*	−	±	±	±	No*	++ (well-defined border ± dark core)*	Can be in an atypical location
Rhabdomyoma	Intramyocardial or intracavitary, attached to myocardium	±	±	±	+	No*	—	
Malignant	Infiltrative†		±		±	Variable	± (If − then heterogeneous appearance)	History of malignancy
Vascular‡	Variable	±	−	−	− (variable)	Strong*	+ (variable and heterogeneous)	Consider malignant tumor
Thrombus	Mural or intraluminal*	−	−	−	−	No*	—*	MDE sequence, long inversion time
Myxoma	Typically left atrium but can be in any chamber	±	±	±	+	No	±*	Irregular, pedunculated, mobile*
Fibroelastoma	Pedunculated, mobile endocardial or valvular mass	−	−	−	−	No		
Pleuropericardial cyst	Right cardiophrenic angle	+ +*	−	−	+ +*	No	−	Smooth-walled and well-defined
Purkinje cell tumor	Ventricular myocardium		+ −*	—*	−	No		Ventricular arrhythmia*
Teratoma	Intrapericardial (usually compressing SVC and/or RA)	±				No		Multilocular bosselated mass with solid and cystic areas
Lipoma§	Any chamber	−	+ −*	—*	±	No	−	

*Either strongly supportive of or necessary for diagnosis. †Infiltrative: 1) crossing an annular or tissue plane within the heart; 2) involving both cardiac and extracardiac structures; or 3) appearance of linear growth through a large vessel such as the superior or inferior vena cava. ‡Vascular refers to tumors with strong vascular supply, including hemangioma, malignant vascular tumors, and paraganglioma. §Lipoma was not tested, because no cases of biopsy-proven lipoma were included.

− = iso- or hypointense; ± = variable intensity; + = hyperintense; ++ = strongly hyperintense; fat sat = fat saturation; FPP = first pass myocardial perfusion; MDE = myocardial delayed enhancement; RA = right atrium; SSFP = steady state free precession; SVC = superior vena cava.

Beroukhim RS et al. JACC 2011

Risk of arrhythmia by histotype

	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.

Distribution of types, age at diagnosis, outcome of operated cardiac tumors in children

Table 1 Summary of 120 children with a cardiac tumor

Tumor type	Number of patients	% of total	Age range	Mean age (year)	No. (%) of deaths	Age of deaths (days)
Rhabdomyoma	42	35	1 day–6 years	0.6	3 (7)	3, 13, and 17
Myxoma	28	23	4 days–20 years	9	1 (4)	25
Fibroma	20	17	5 days–17 years	3.25	4 (20)	5, 15, 28, and 38
Teratoma	8	7	1 day–16 years	3	1 (12)	4
Pericardial	4	3.5	43 days–15 years	7.3	0 (0)	–
Rhabdomyosarcoma	3	2.5	20 days–8 years	–	0 (0)	–
Sarcoma	2	1.5	7 days–14 years	–	0 (0)	–
Hamartoma	1	1	1 year	1	0 (0)	–
Uncategorized	12	10	1 day–16 years	4.25	0 (0)	–
Total	120	100	1 day–20 years	3.6	9 (7)	Mean age 14

Bielefeld KJ et al. Pediatr Cardiol 2013;34-125-8

Indications for surgery according to tumor histotype

Tumor Histotype (No. of Patients)	Presence of Symptoms	Abnormal ECG	Echocardiographic Hemodynamical Impairment
Rhabdomyoma (32)	23 (71.8)	9 (28.1)	26 (81.2)
Myxoma (18)	10 (55.5)	0 (0.0)	14 (77.7)
Teratoma (12)	7 (58.3)	3 (25.0)	4 (33.3)
Fibroma (9)	5 (55.5)	1 (11.1)	9 (100.0)
Hemangioma (8)	8 (100.0)	2 (25.0)	3 (37.5)
Sarcoma (5)	5 (100.0)	3 (60.0)	3 (60.0)
Other* (5)	3 (60.0)	0 (0.0)	4 (80.0)
Total (89)	61 (68.5)	18 (20.2)	63 (70.8)

All data shown are number of patients (% of tumor histotype).
*Other includes pseudotumor (3), papilloma (1), and malignant teratoma (1).

Padalino M et al. Circulation 2012; 126:22-30

Primary surgical procedures and deaths rates according to tumor histotype

Histotype	No. of Patients	Complete Resection	Partial Resection	OHT	Other Surgery*	Early Death (Within 30 d From Operation)	Late Death (After 30 d From Operation)
Rhabdomyoma	32	15 (46.9)	14 (43.8)	1 (3.1)	2 (6.2)	1 (3.1)†	0 (0.0)
Myxoma	18	17 (94.4)	1 (5.6)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)
Teratoma	12	12 (100.0)	0 (0.0)	0 (0.0)	0 (0.0)	1 (8.3)	0 (0.0)
Fibroma	9	5 (55.6)	1 (11.1)	3 (33.3)	0 (0.0)	0 (0.0)	2 (22.2)‡
Hemangioma	8	7 (87.5)	1 (12.5)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)
Sarcoma§	5	2 (40.0)	3 (60.0)	0 (0.0)	0 (0.0)	1 (20.0)	2 (40.0)
Other‖	5	4 (80.0)	1 (20.0)	0 (0.0)	0 (0.0)	1 (20.0)¶	0 (0.0)
Total	89	62 (69.7)	21 (23.6)	4 (4.5)	2 (2.2)	4 (4.5)	4 (4.5)

All data are shown as number of patients (% of tumor histotype). OHT indicates orthotopic heart transplant.
*Other surgery includes cavopulmonary anastomosis (1), midline sternotomy and incisional biopsy (1).
†Postoperative brain hemorrhage.
‡Status post heart transplant.
§Two patients lost at follow up.
‖Other tumor histotypes include pseudotumor (3), papilloma (1), malignant teratoma (1).
¶Malignant teratoma.

Padalino M et al. Circulation 2012; 126:22-30

Postoperative complications according to tumor histotype

Histotype	No. of Patients	LCO	Postoperative Arrhythmia	PNX	Pleural and/or Pericardial Effusion	Phrenic Nerve Injury	Other Complications*	Total
Rhabdomyoma	32	1	1	1	3	1	3	10 (31)
Myxoma	18	0	1	1	1	0	1	4 (22)
Teratoma	12	2	1	0	0	0	1	4 (33)
Fibroma	9	0	1	0	0	0	2	3 (33)
Hemangioma	8	0	0	0	1	0	1	2 (25)
Sarcoma	5	1	0	0	0	0	0	1 (20)
Other†	5	1	0	0	1	0	0	2 (40)
Total	89	5 (5.6)	4 (5.4)	2 (2.2)	6 (6.7)	1 (1.1)	8 (9.0)	26 (29.2)

All data are shown as number of patients (% of tumor histotype). LCO indicates low cardiac output syndrome; PNX, pneumothorax.

*Other complications include undetermined minor secondary complications (3), acute cardiac transplant rejection (1), multiorgan failure (1), superior vena cava thrombosis after heart transplant (1), respiratory insufficiency requiring long-term mechanical ventilation (1), and cerebral hemorrhage on previous brain surgery site (1).

†Other tumor histotypes include pseudotumor (3), papilloma (1), and malignant teratoma (1).

Padalino M et al. Circulation 2012; 126:22-30

Rhabdomyoma(s)

Key points for Rhabdomyomas

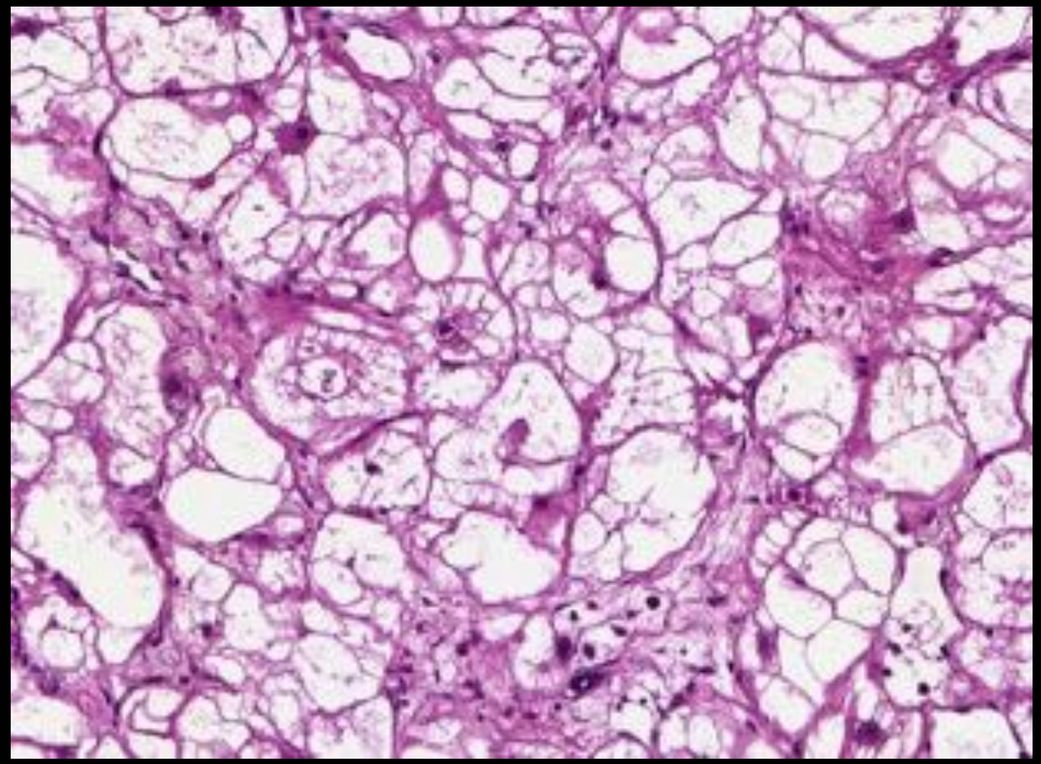
1. Prenatal diagnosis is frequent

2. Associated with TSC in a large proportion

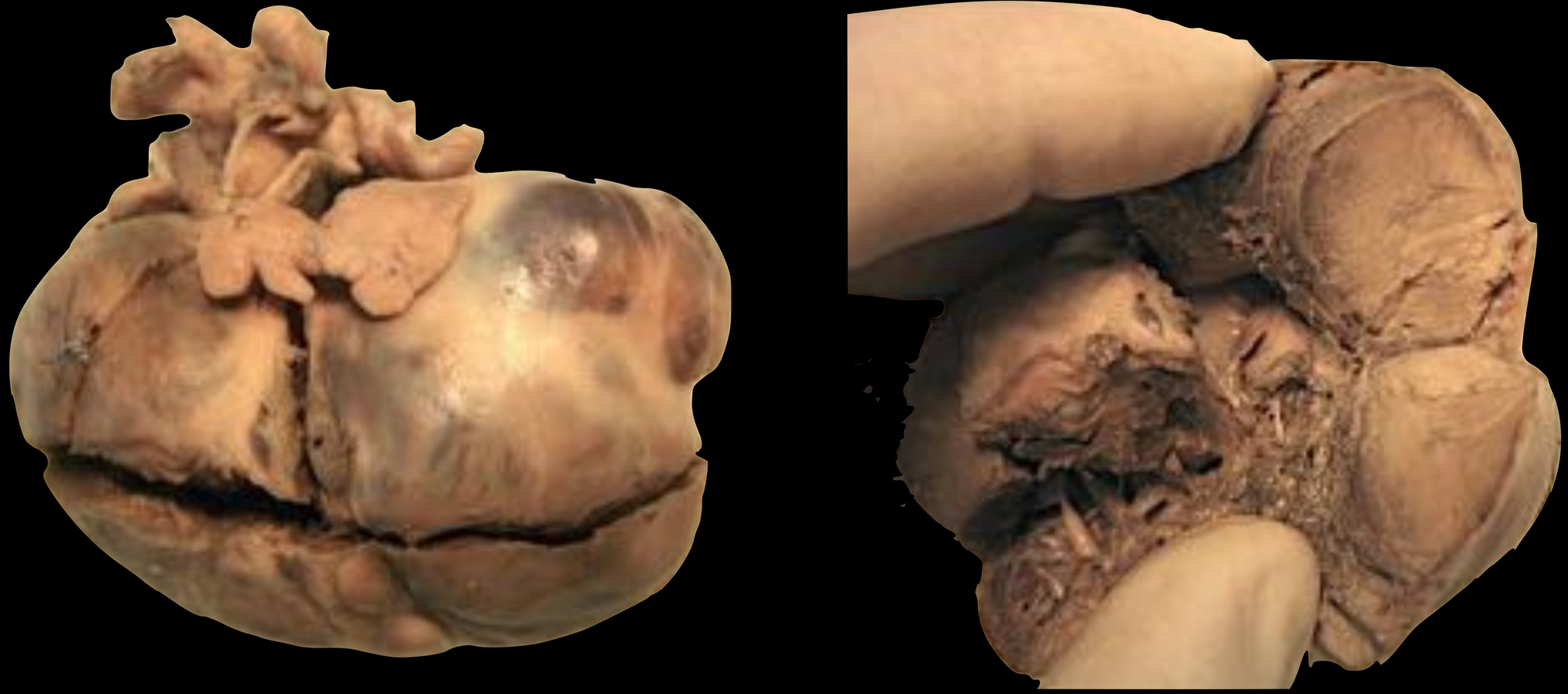
3. Cardiac outcome usually favorable with regression of tumors before 6 years of age

4. Global outcome is related to TSC (neurological outcome) and is difficult to predict

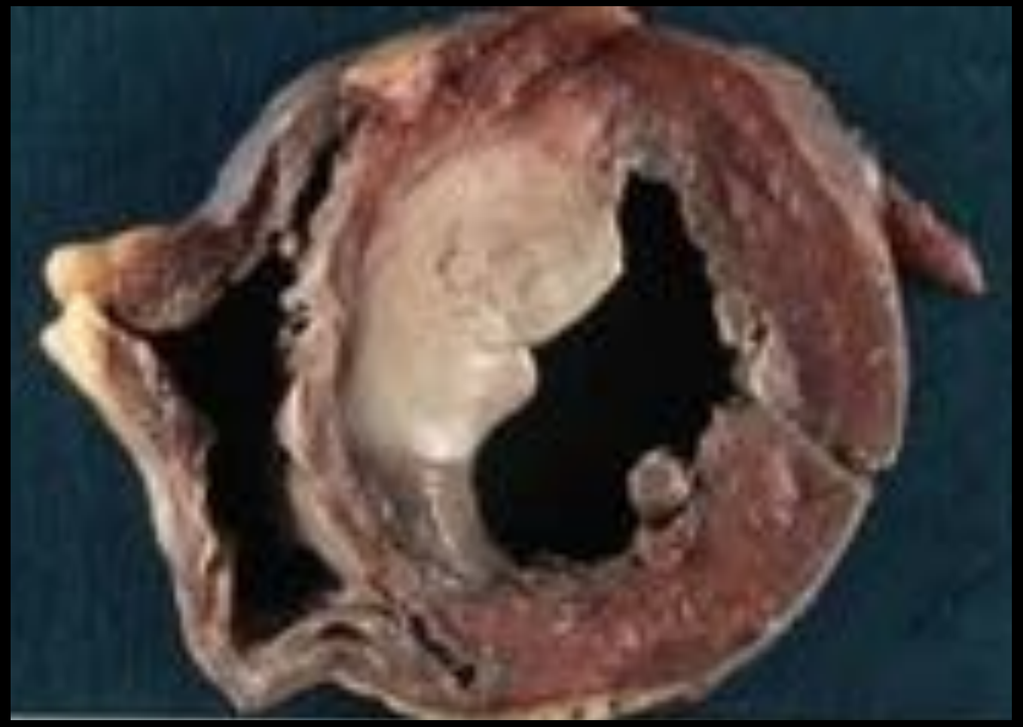
5. Surgical treatment is exceptional is case of obstruction of the in- or outflow tracts

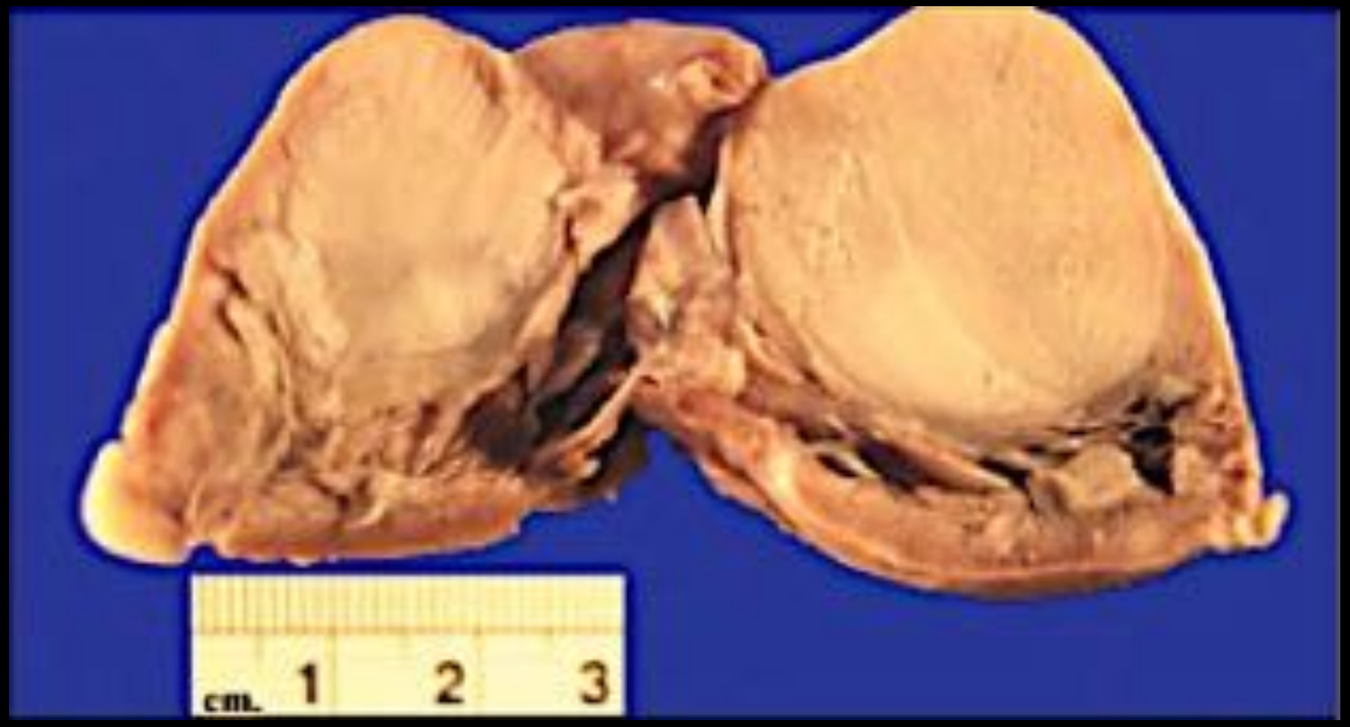


Rhabdomyoma

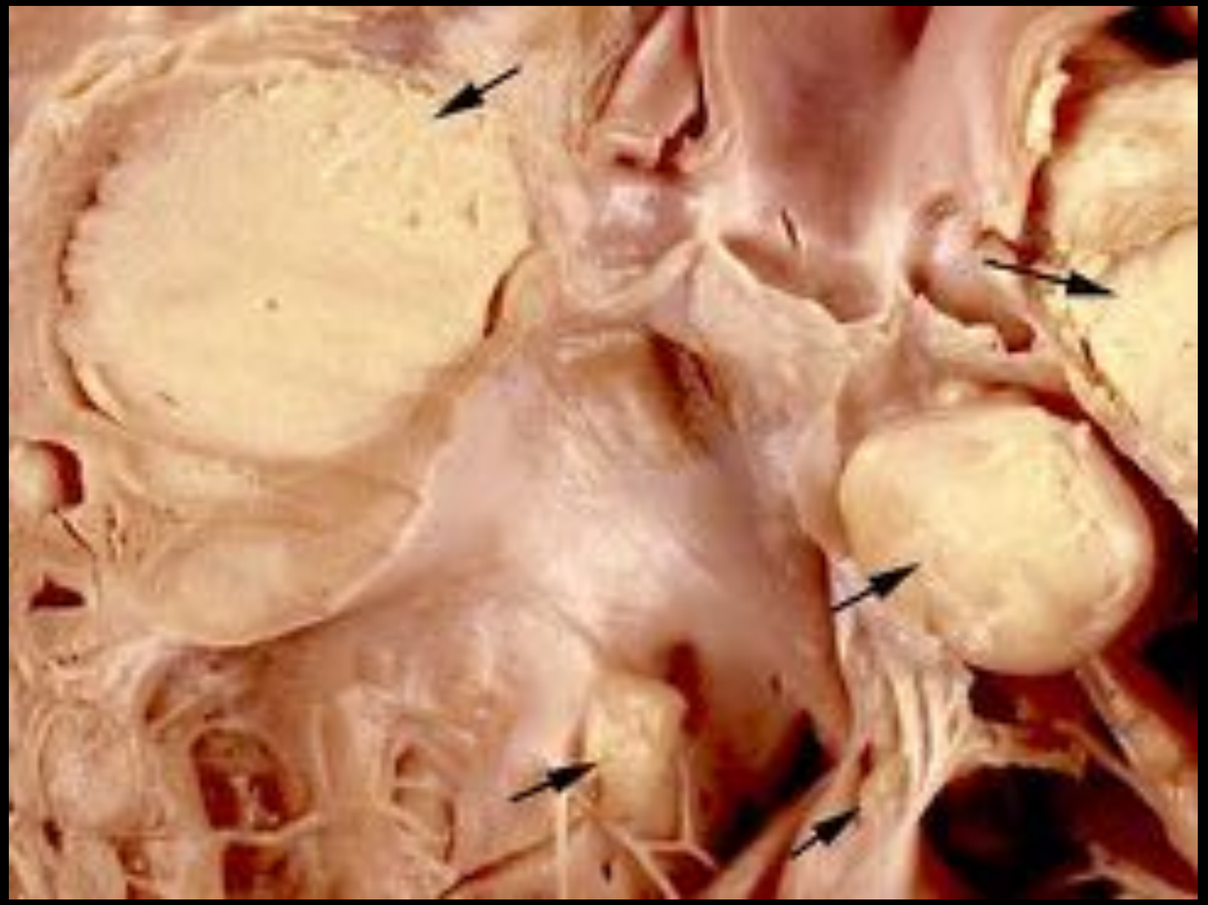


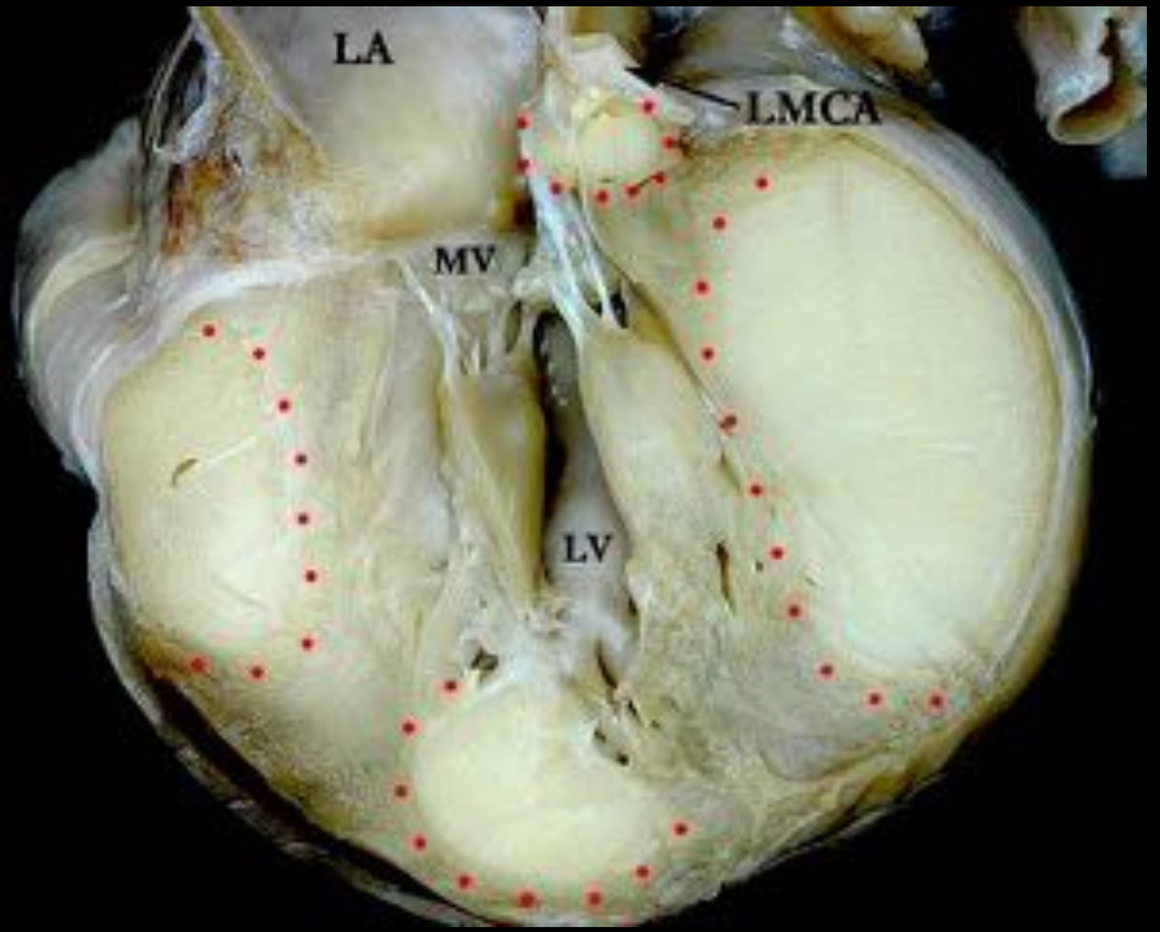
Rhabdomyoma





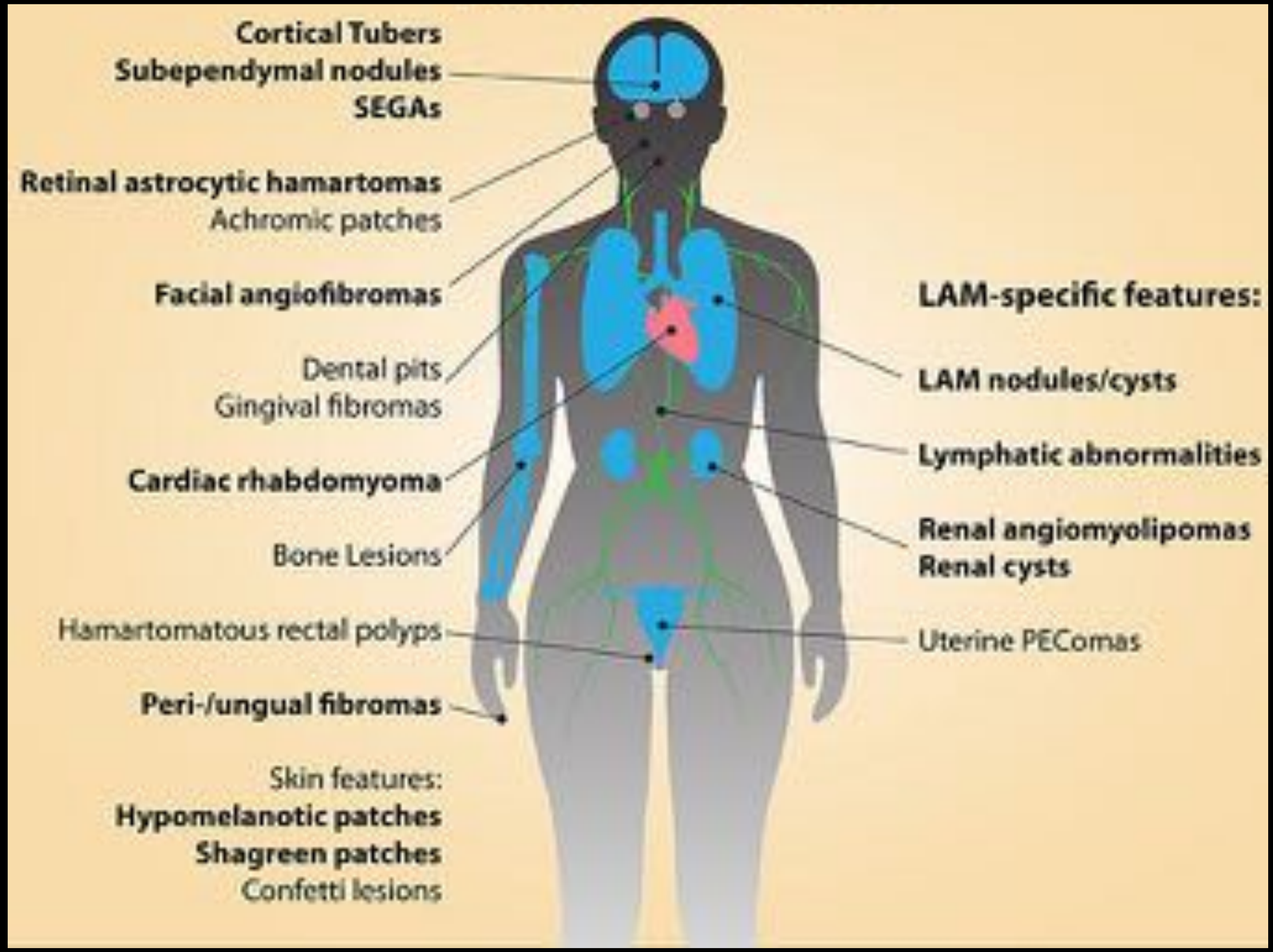
Rhabdomyoma



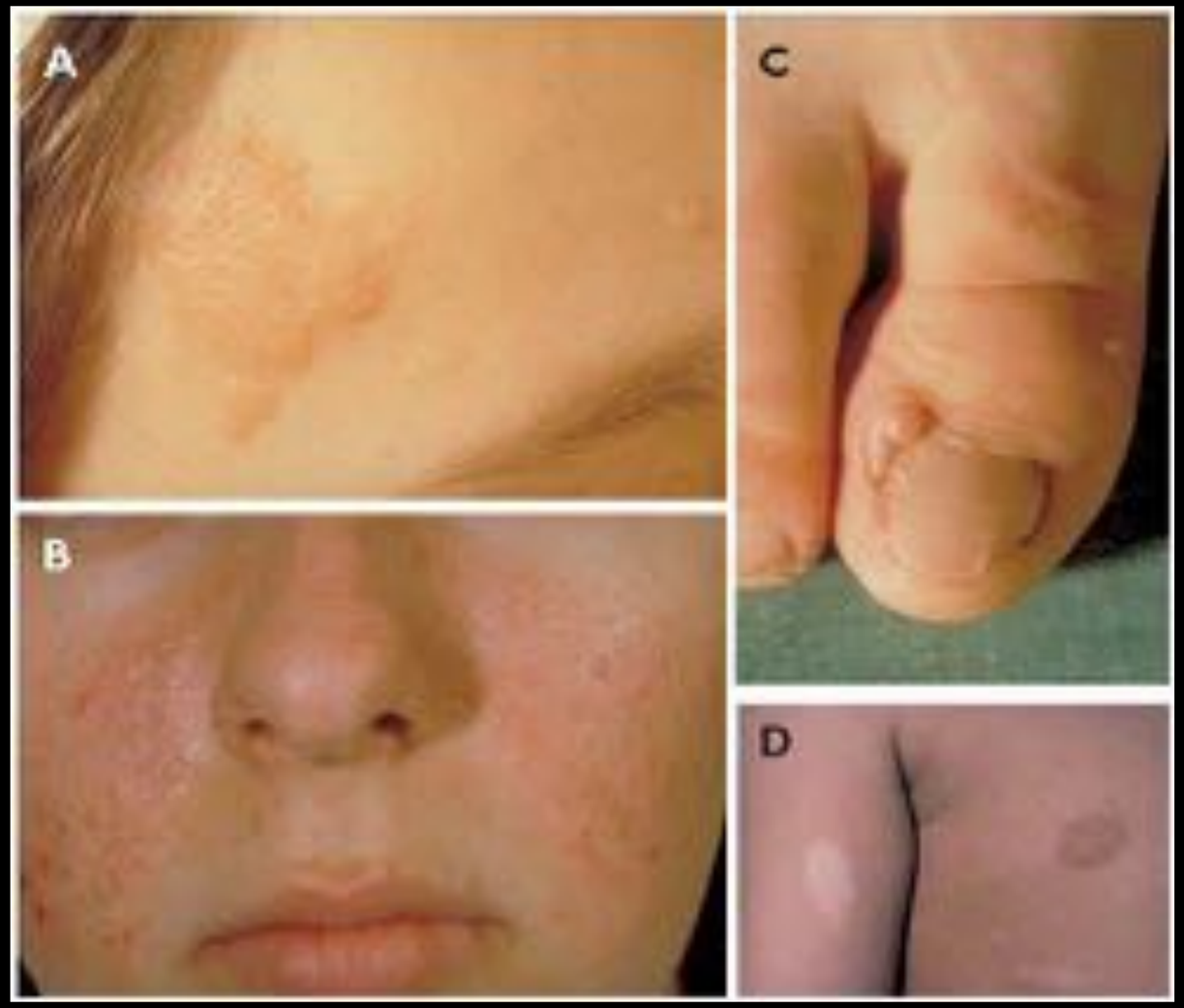


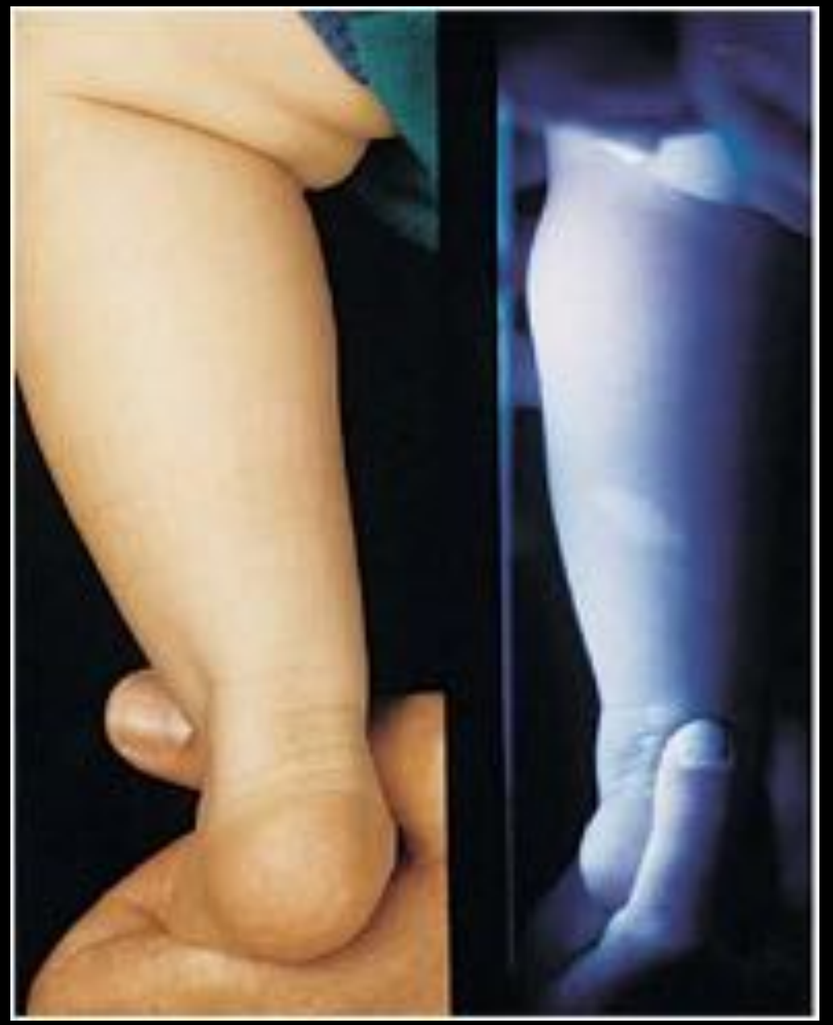
Rhabdomyoma

Tuberous Sclerosis Complex-Clinical features



Tuberous sclerosis - Cutaneous manifestations

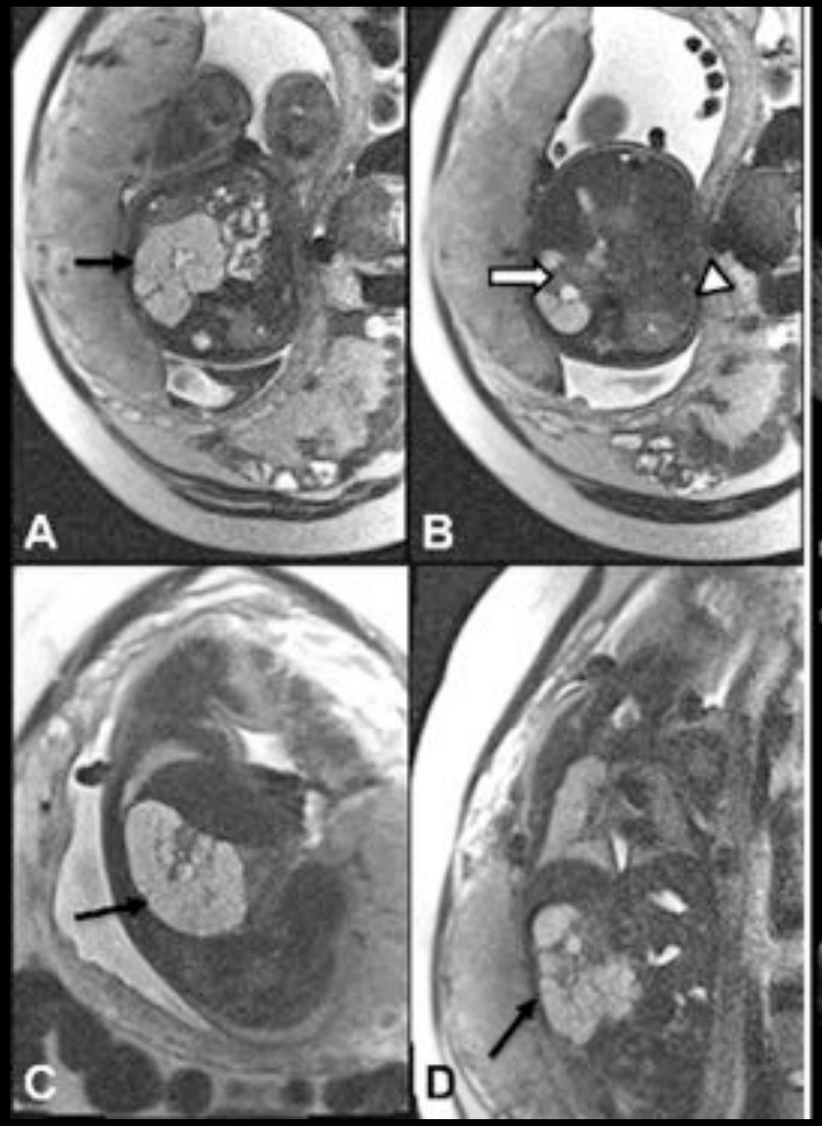


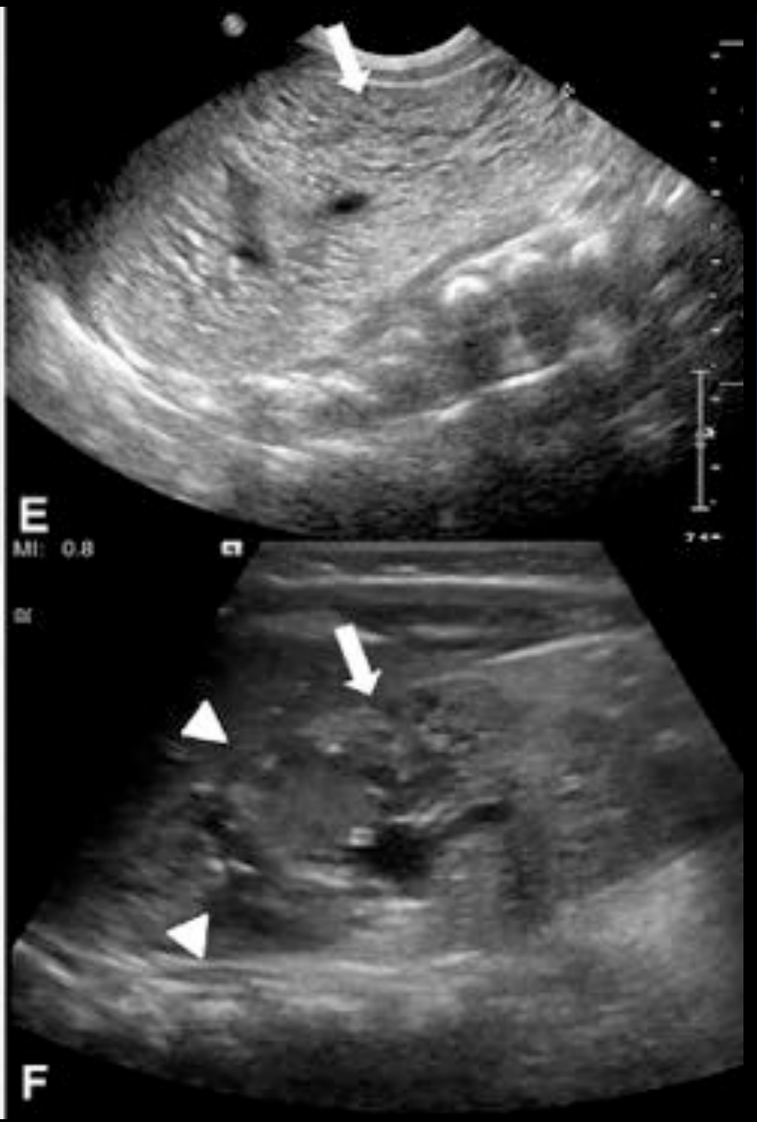


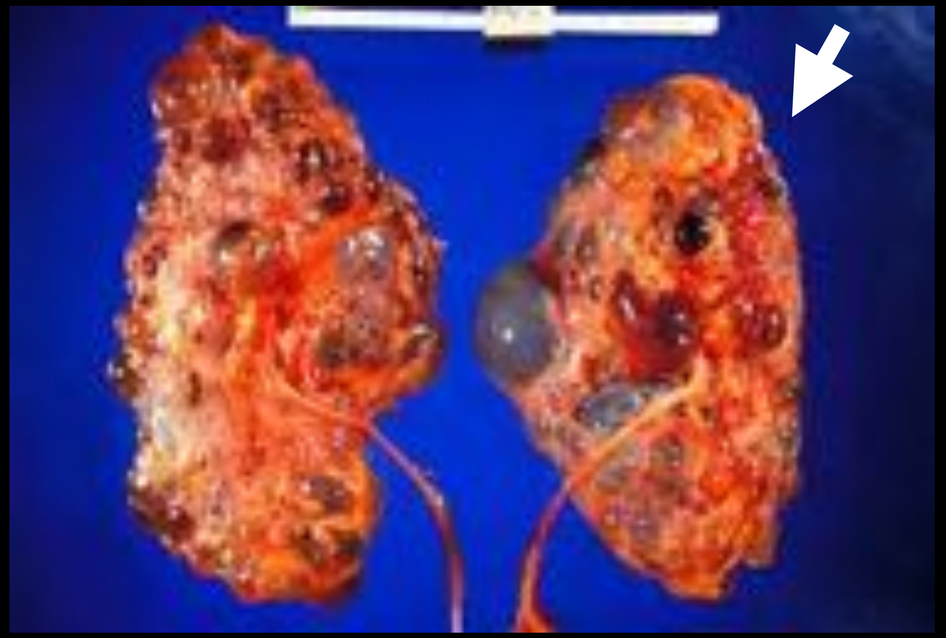
Wood Lamp

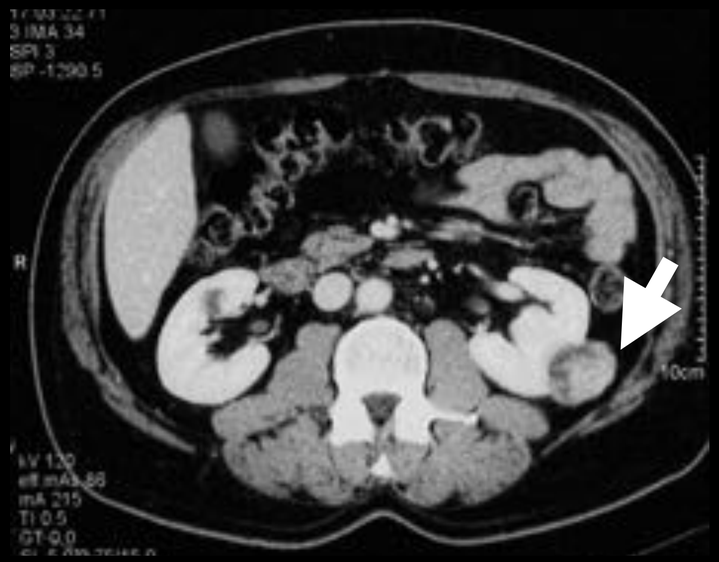
Rhabdomyoma

Tuberous sclerosis - Polycystic kidney disease



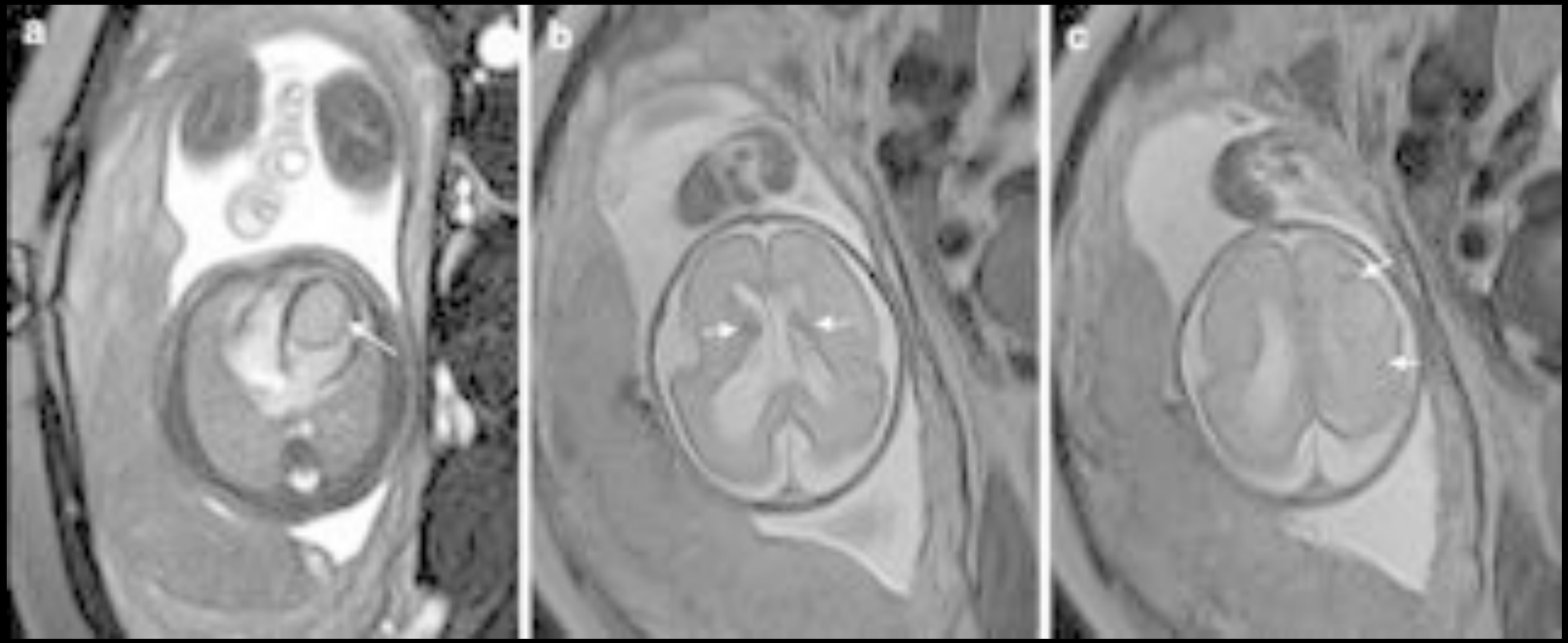






Rhabdomyoma

Tuberous sclerosis - Cerebral tuber



Rhabdomyoma

Tuber

Tuber

Tuberous Sclerosis Complex-Genetics

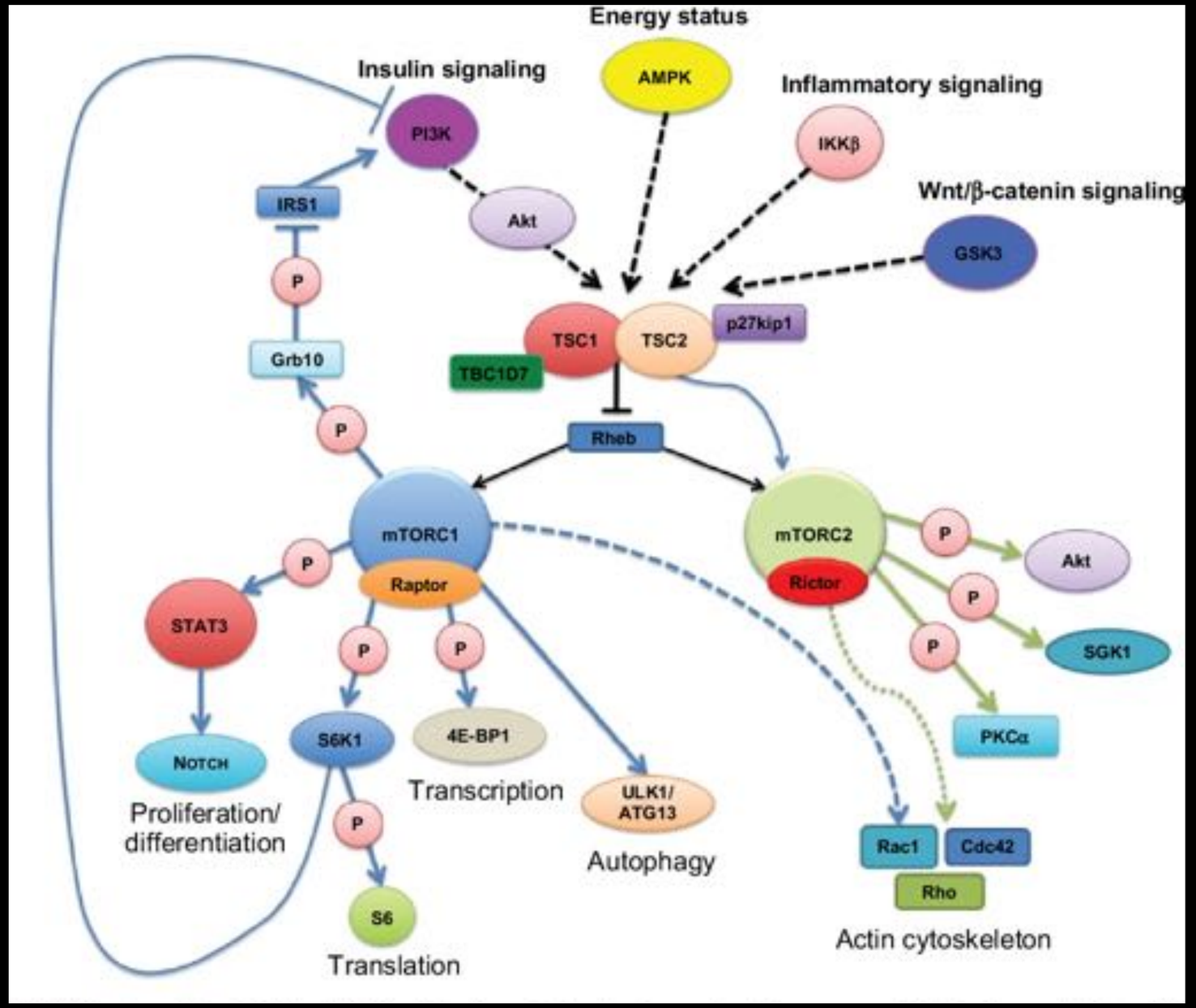
- The mutations in tuberous sclerosis complex may be in one of two tumor suppressor genes **TSC1** and **TSC2**. TSC1 on chromosome 9q34 codes for hamartin. TSC2 on 16p13 codes for tuberin.

- **Hamartin** and **tuberin** form a complex that activates GTPase-activating protein which in turn inhibits mTOR pathway. mTOR is a highly specific protein kinase that regulates protein synthesis, cell differentiation, growth and cell migration.

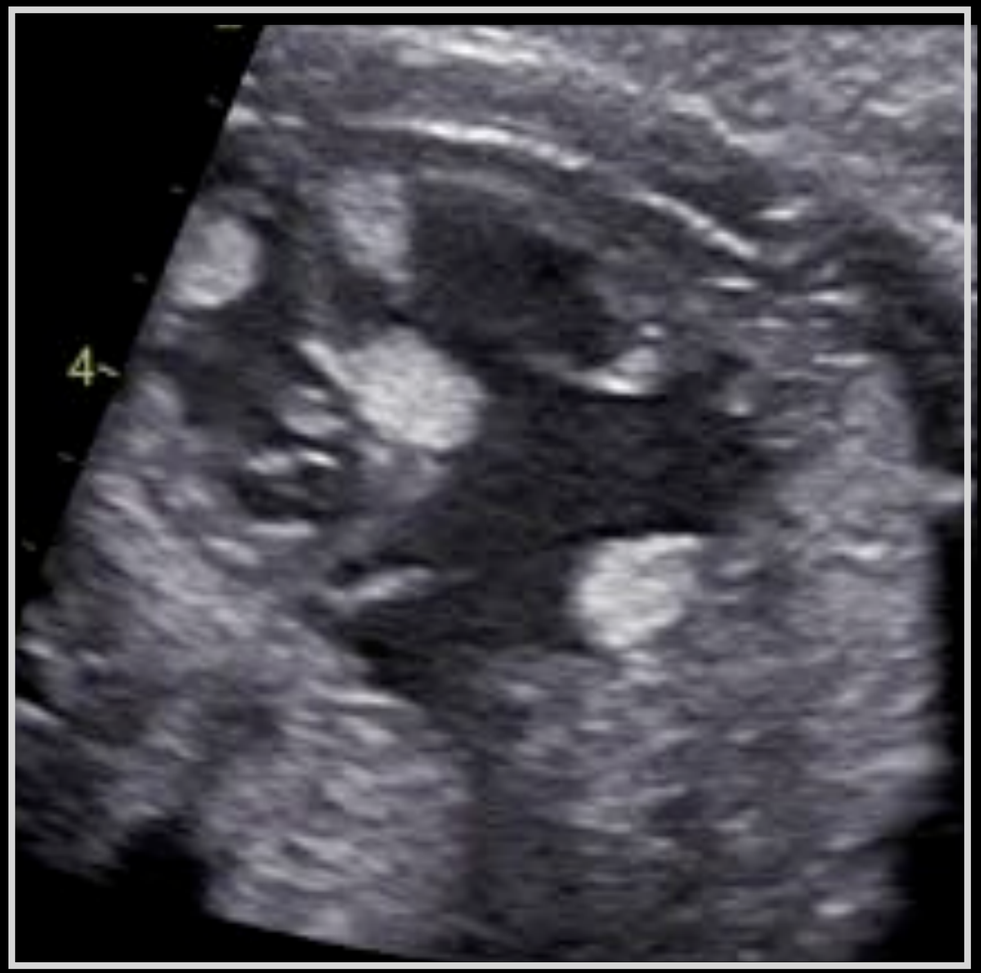
- Mutations in either TSC1 or TSC2 result in **constitutive activation of mTOR pathway** causing abnormal growth and tumor formation.

Tuberous Sclerosis Complex-Genetics

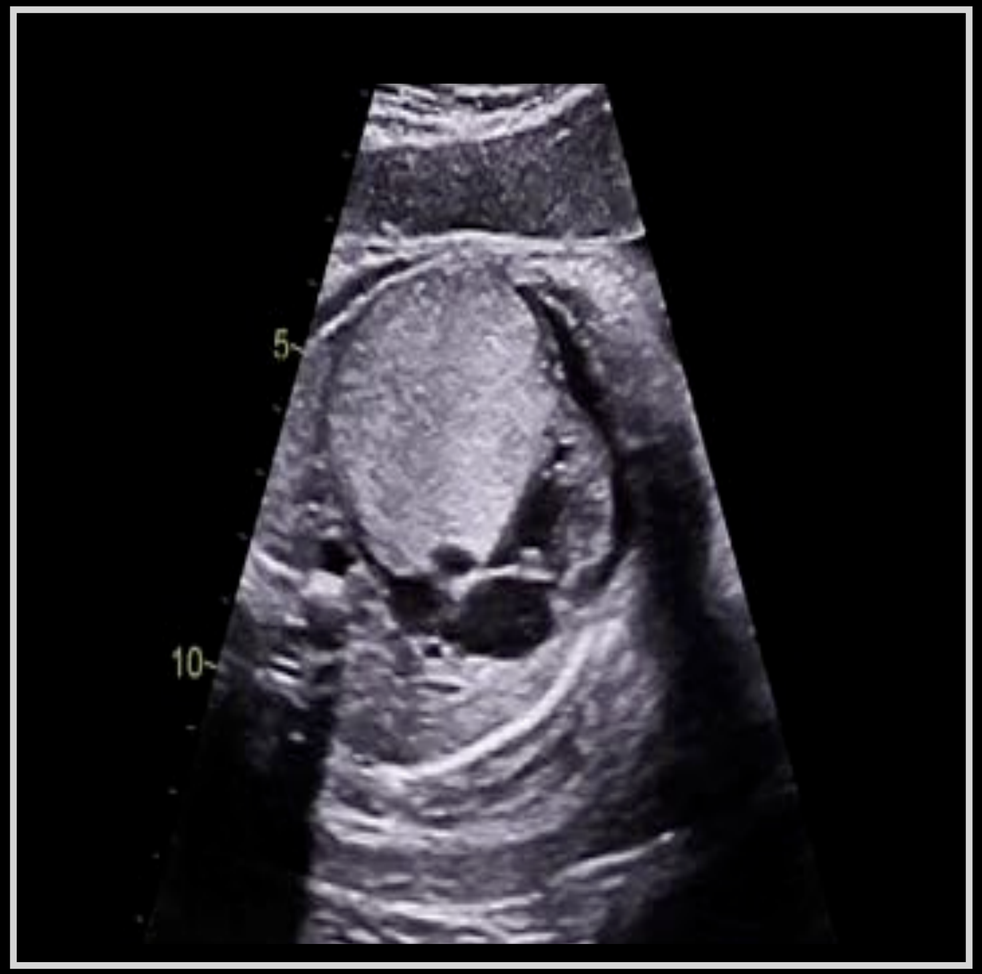
1/6000
60% of cases arise from de novo mutations

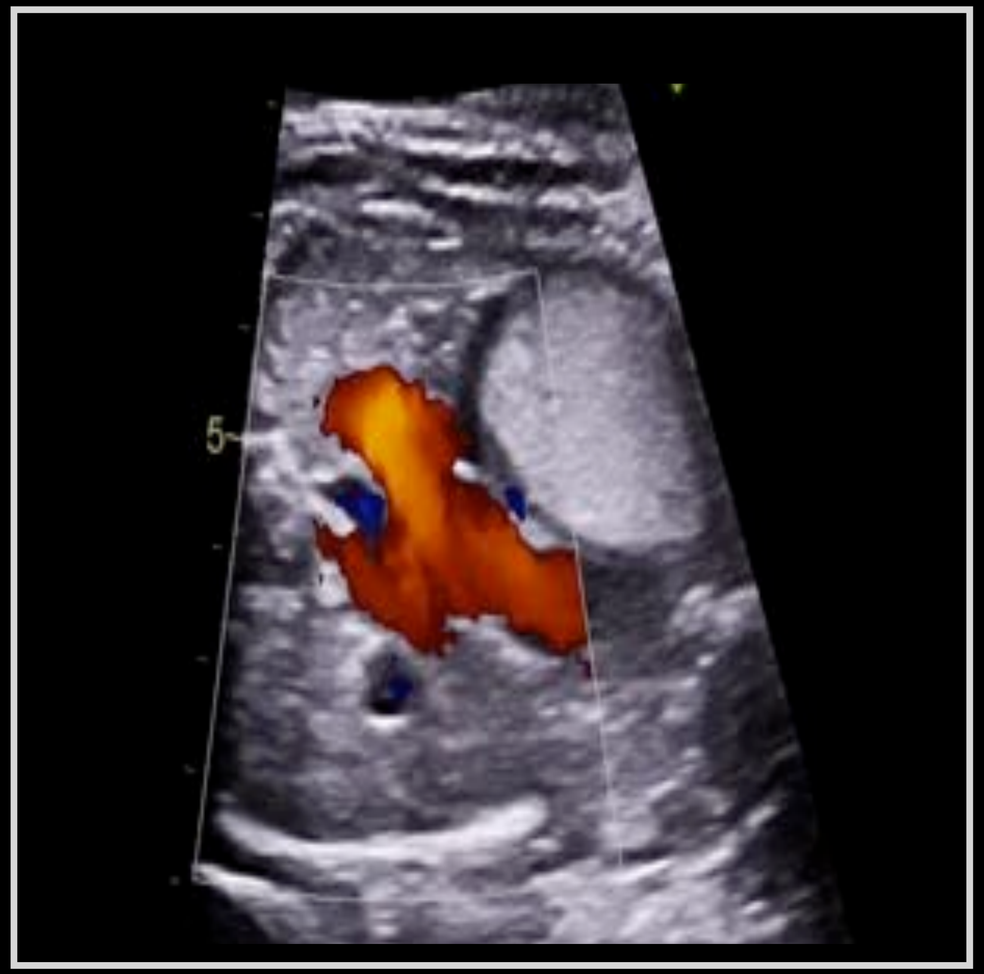


Fetal Rhabdomyomas

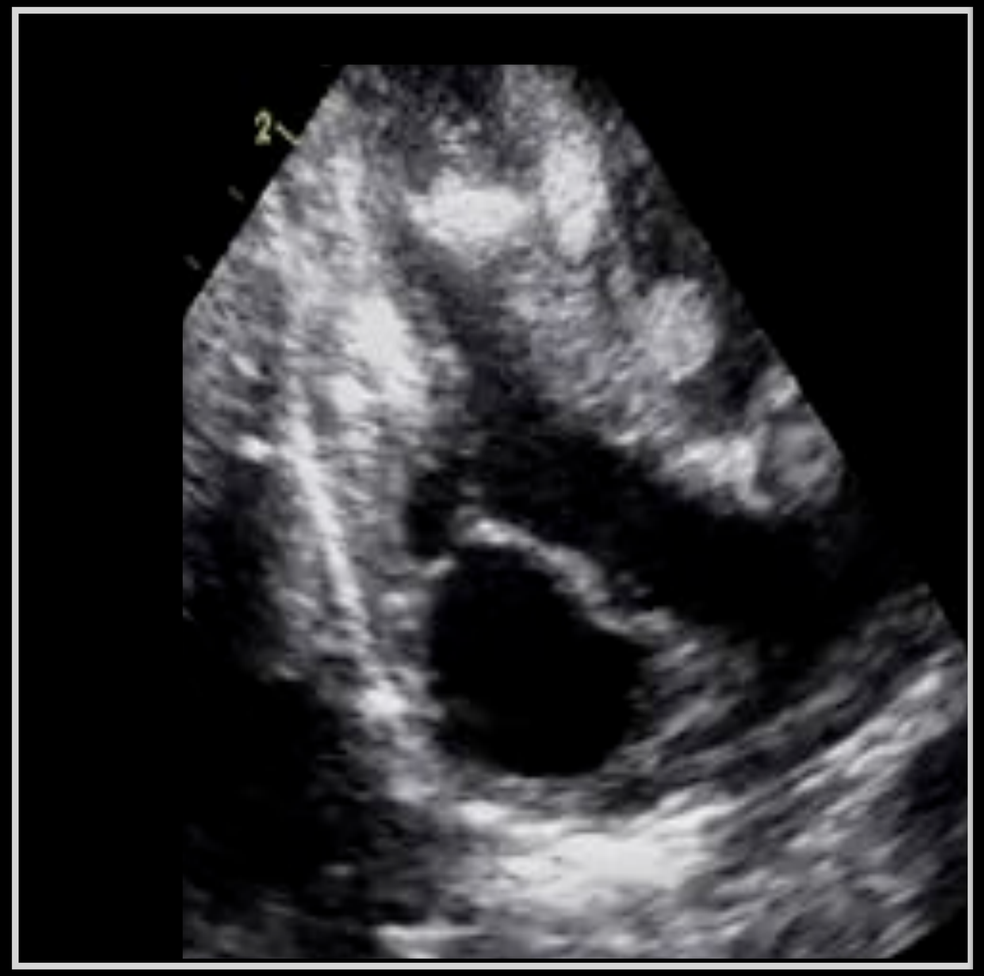


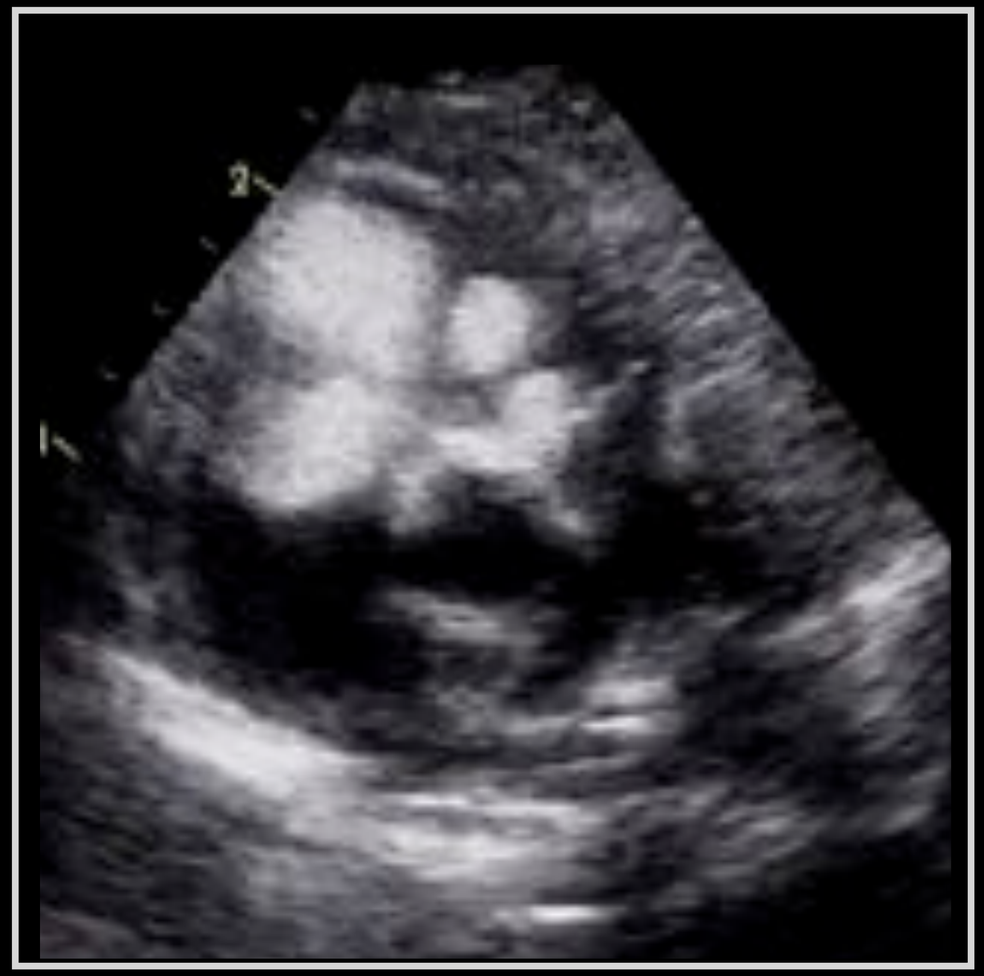
Fetal Rhabdomyomas



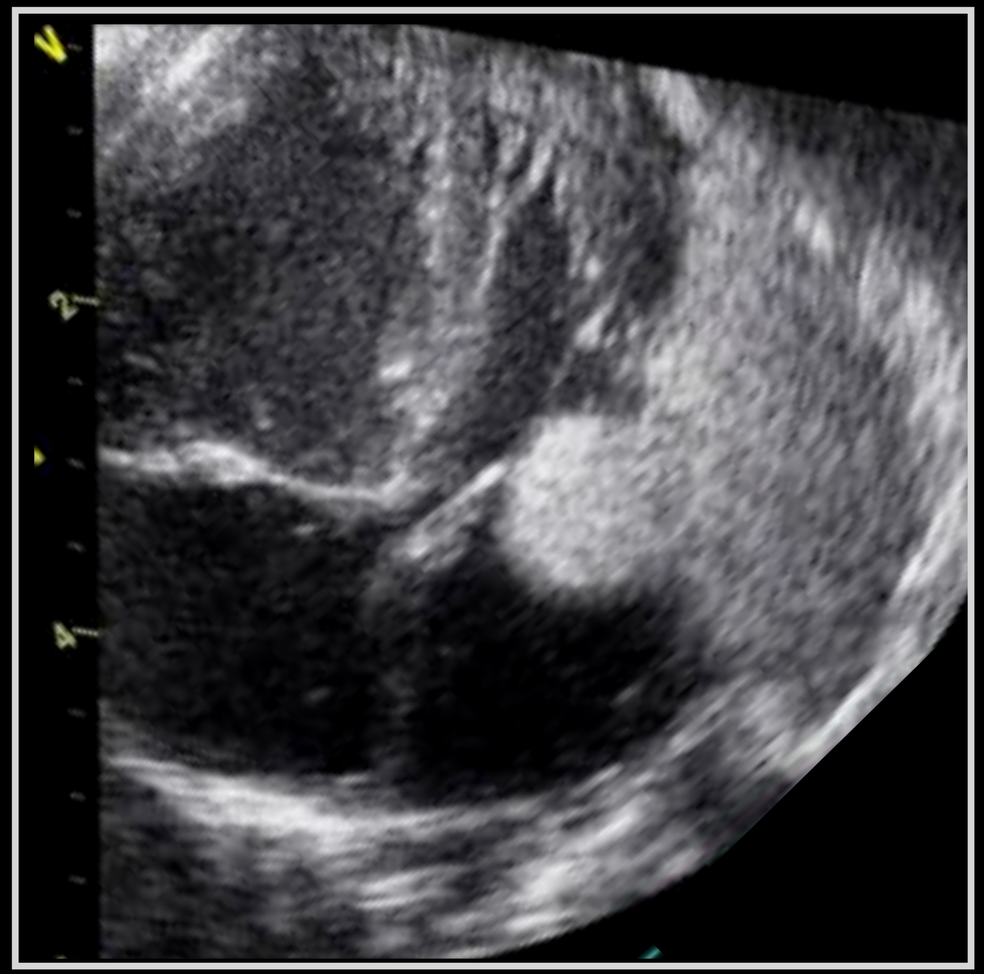


Rhabdomyomas - Multiple, hyperechogenic

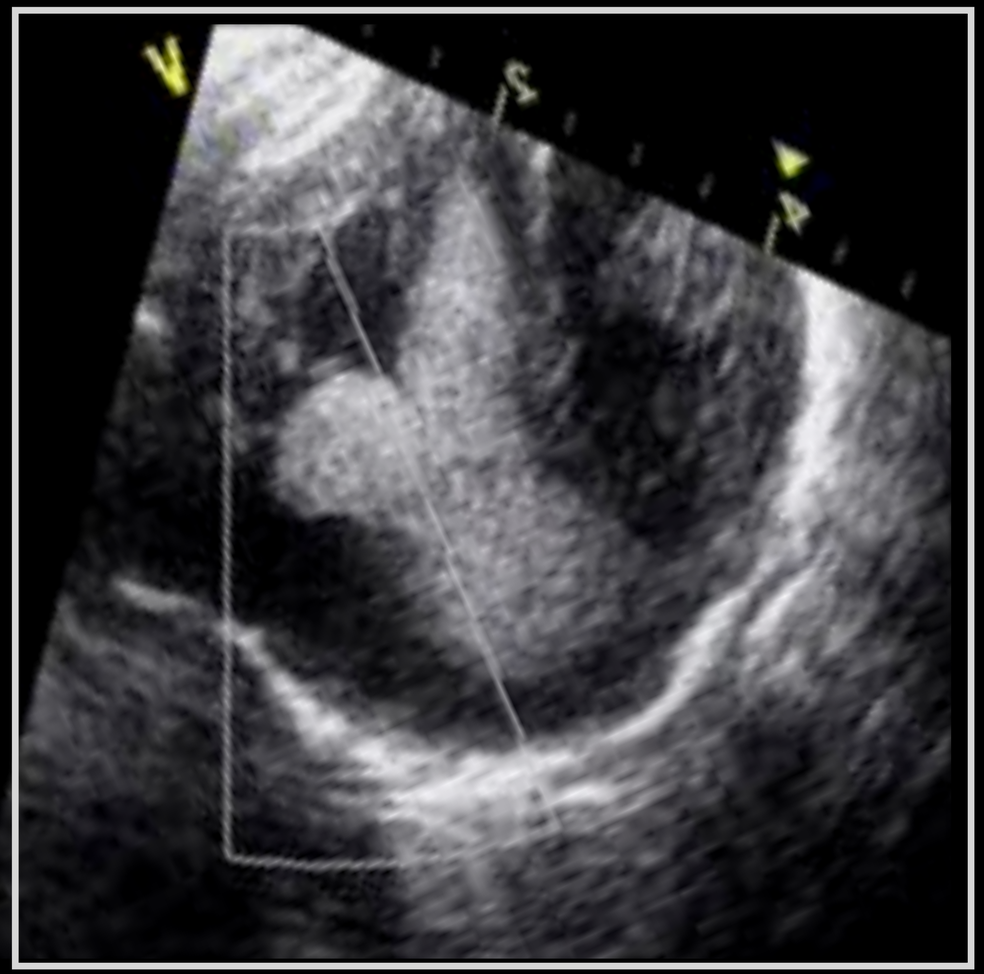




Rhabdomyomas - Inflow obstruction

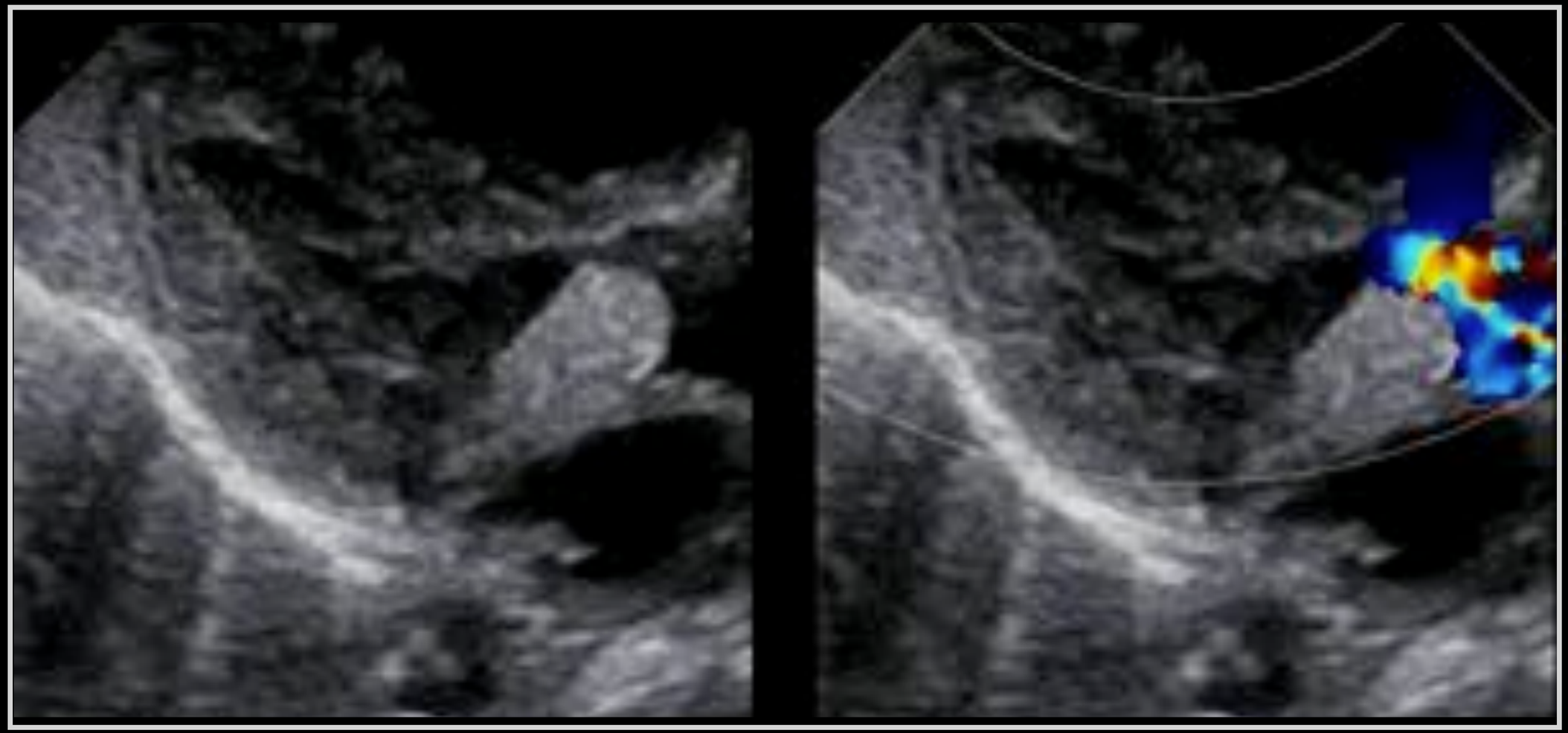


Mitral

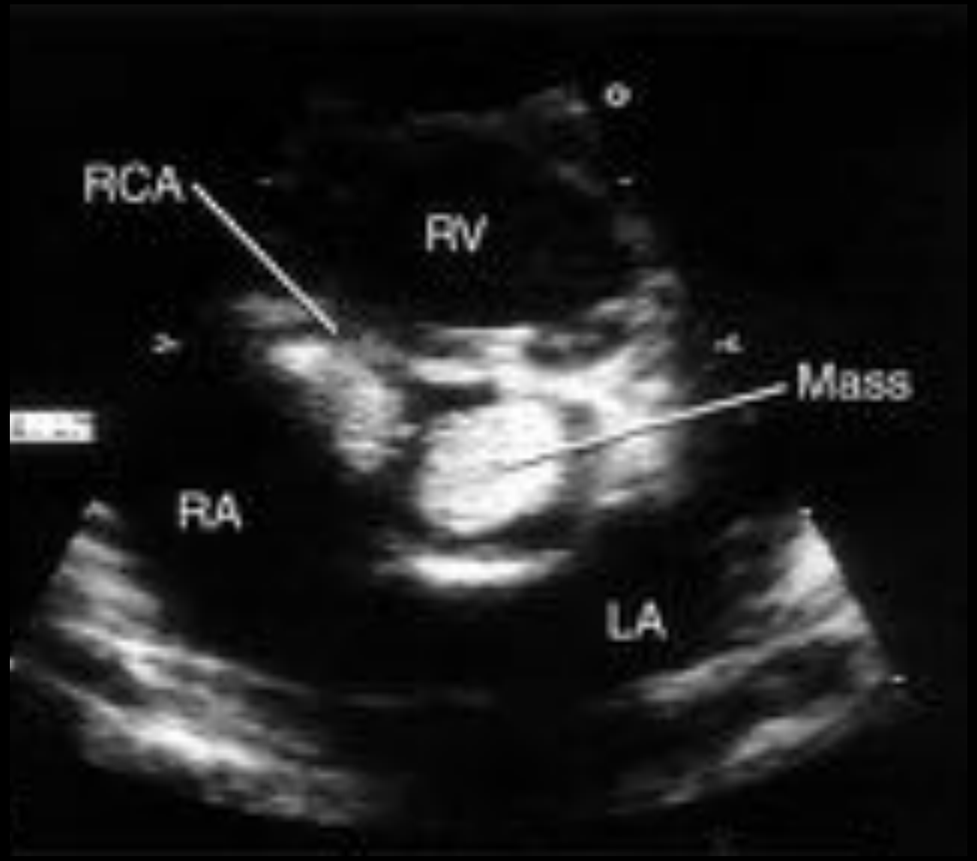


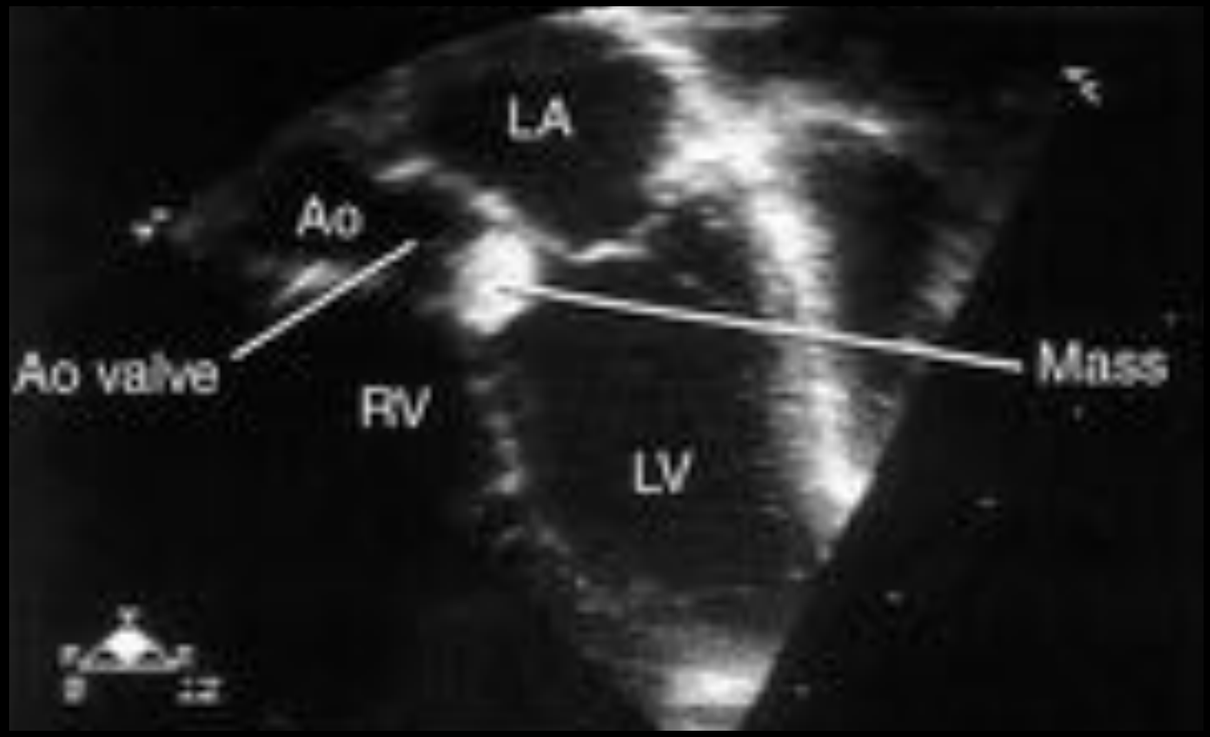
Tricuspid

Rhabdomyomas - Outflow obstruction



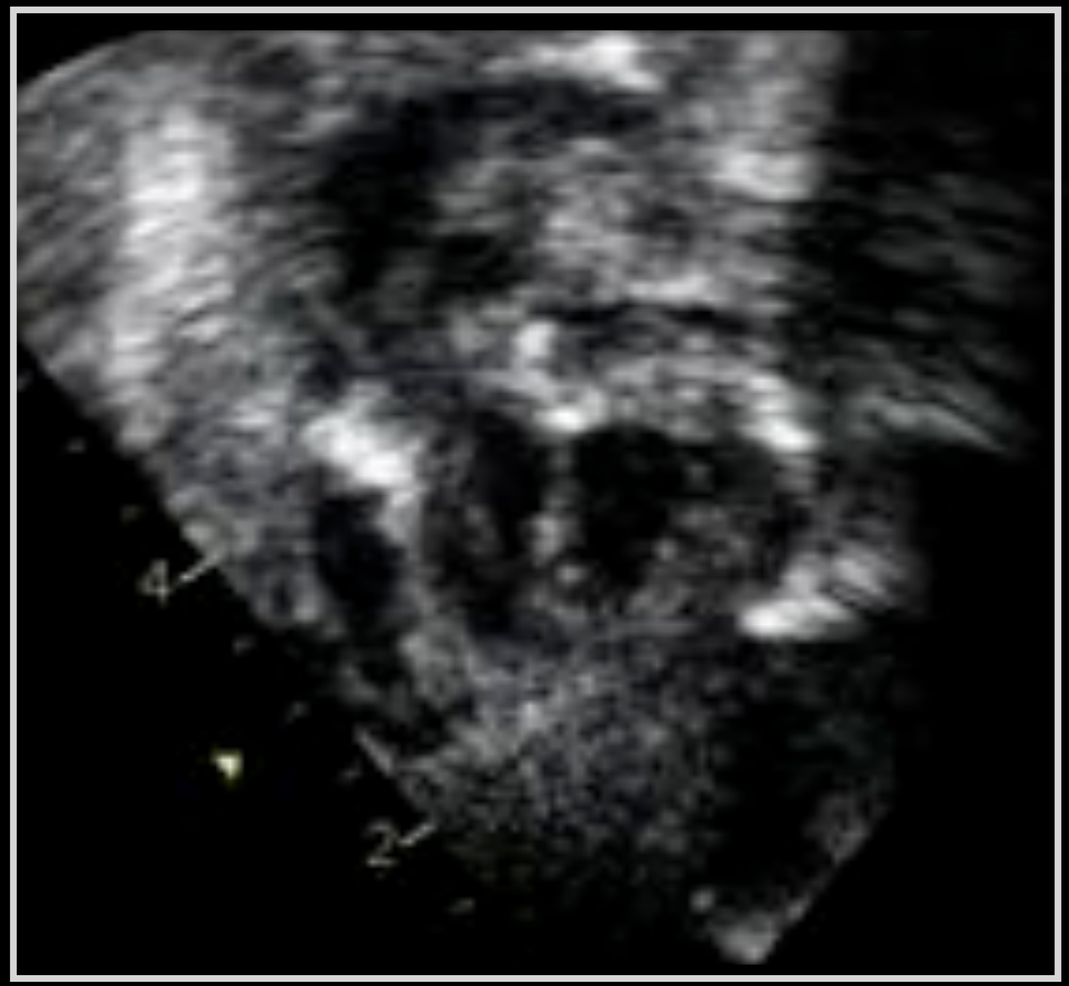
LVOT





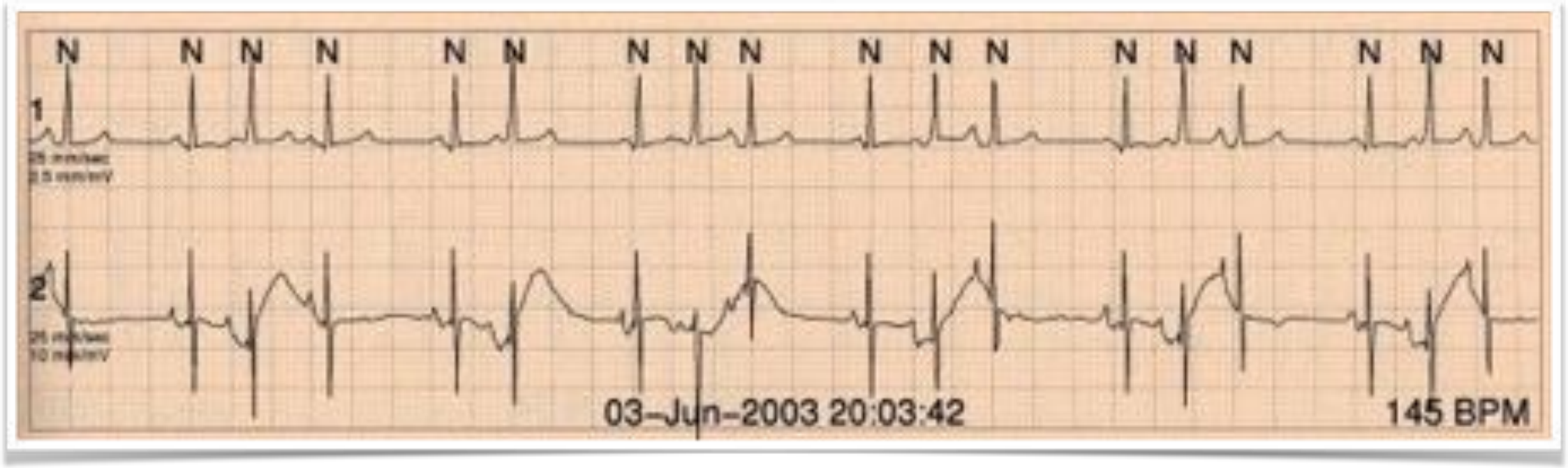
Rhabdomyoma - obstruction of the LVOT

Rhabdomyomas - Outflow obstruction

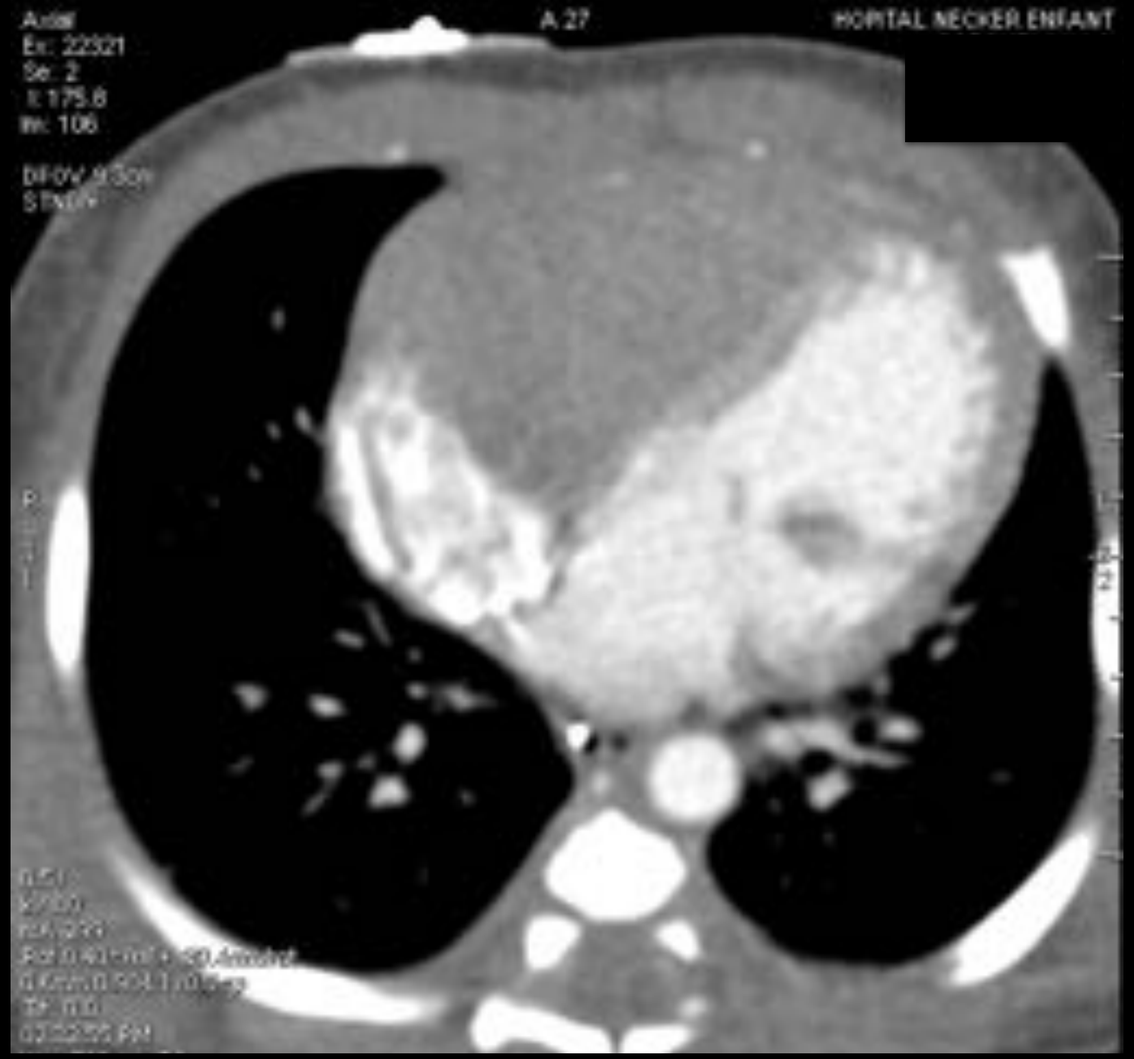


RVOT

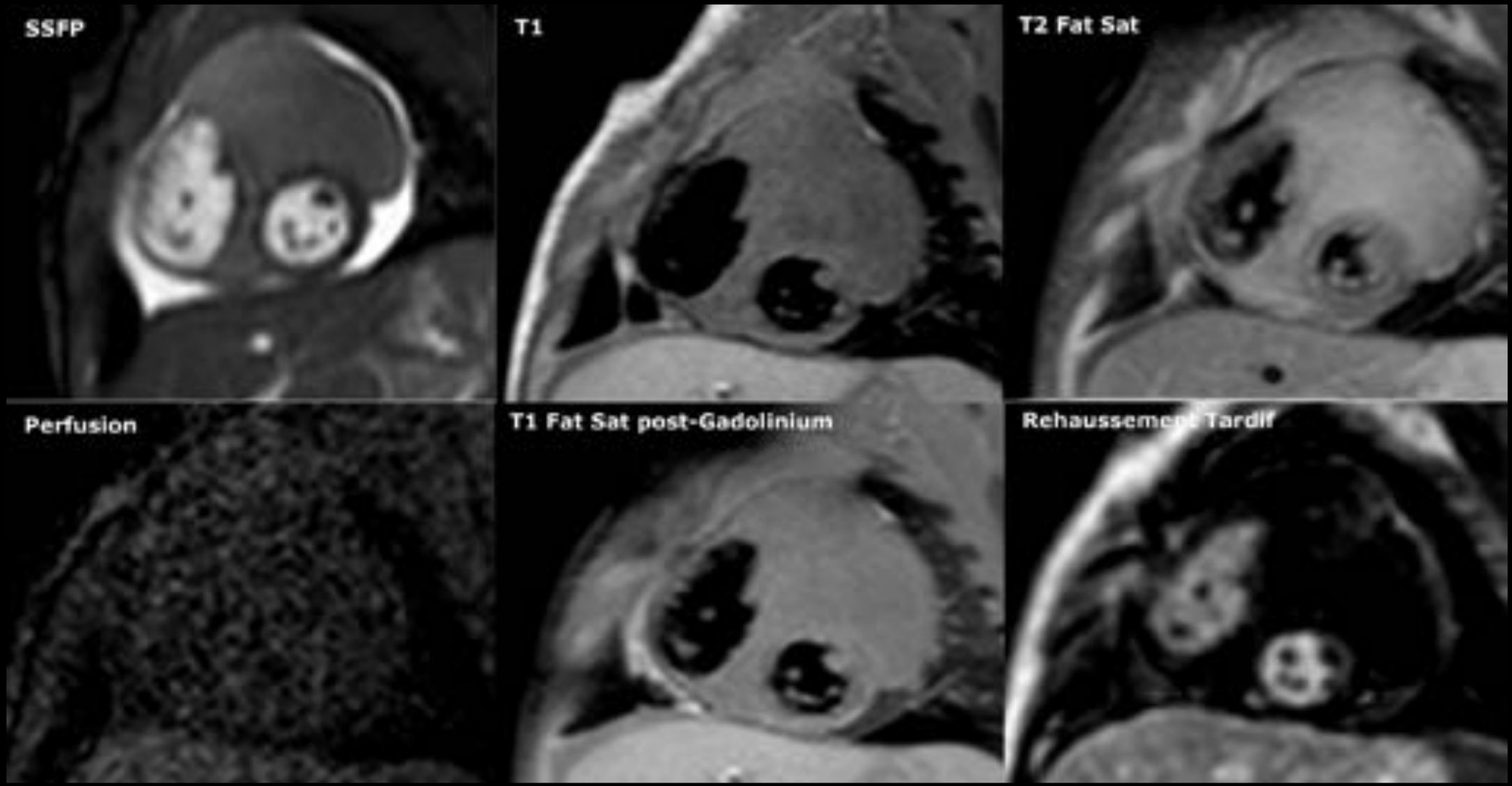
Rhabdomyomas - ECG



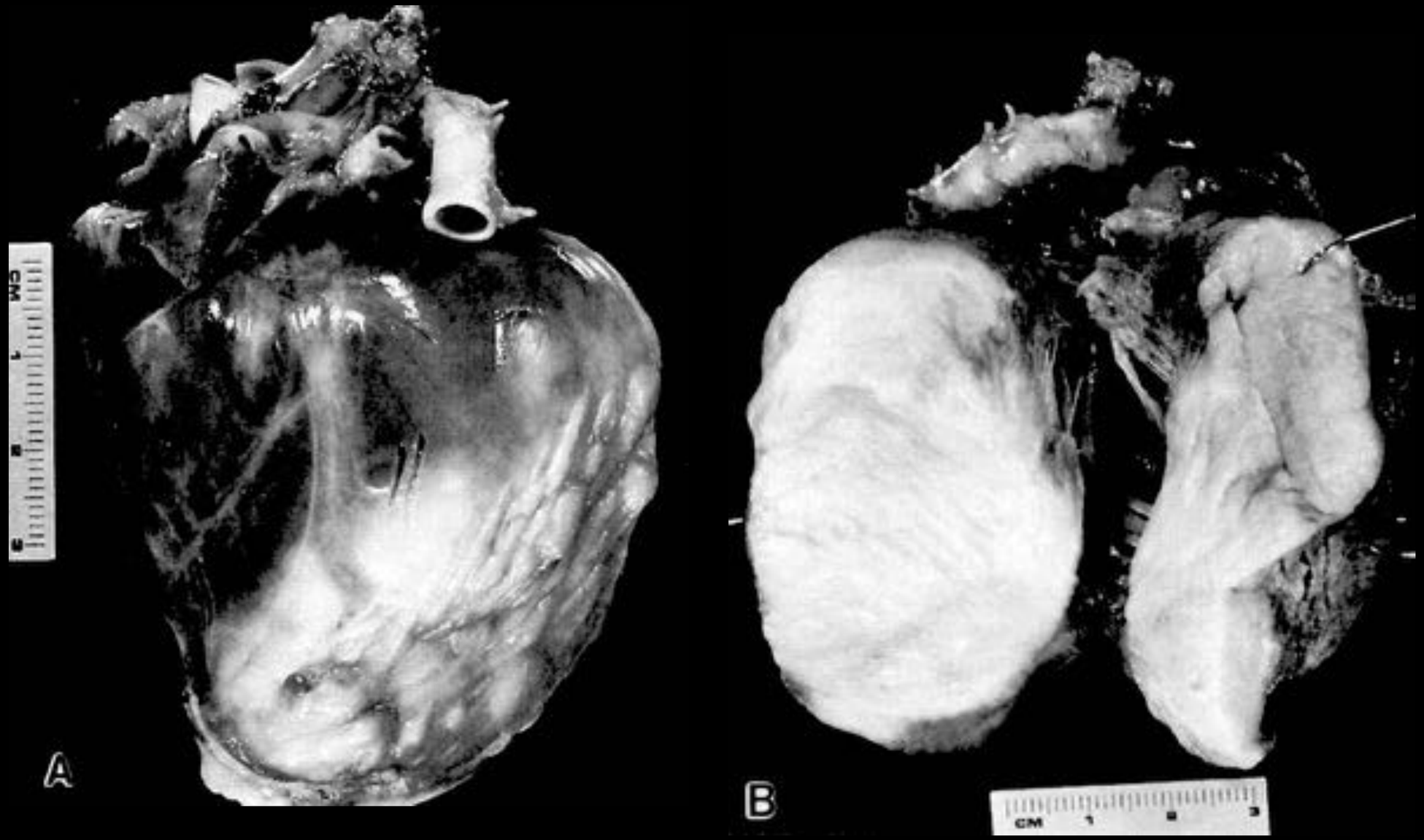
Rhabdomyomas - CT



Rhabdomyomas - MRI



Fibroma



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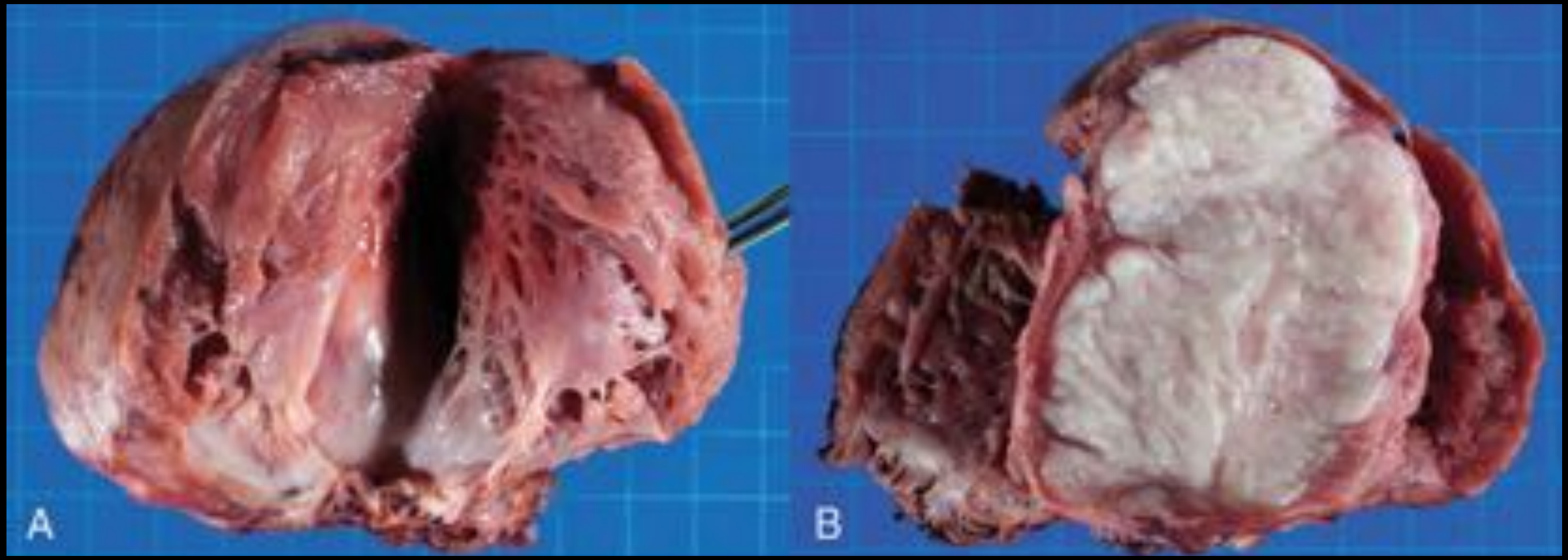
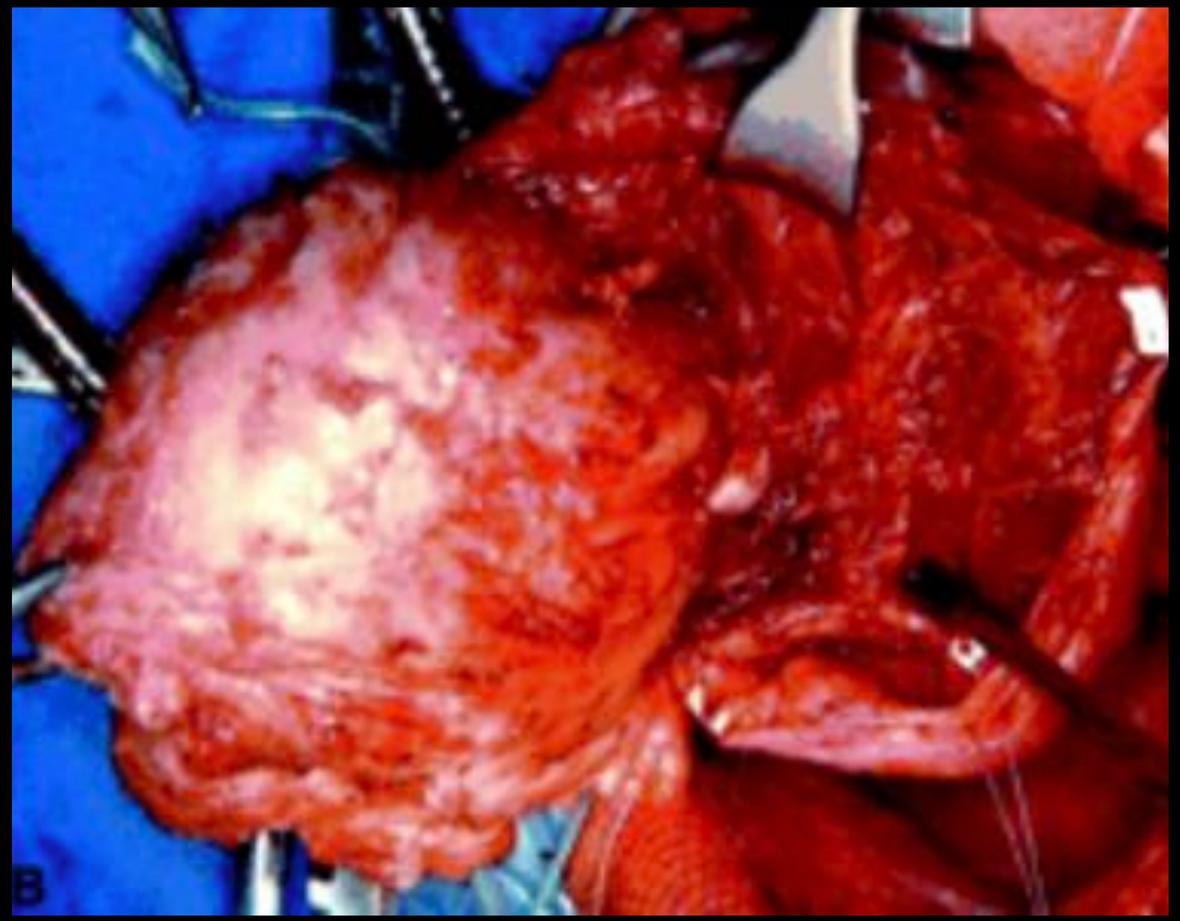
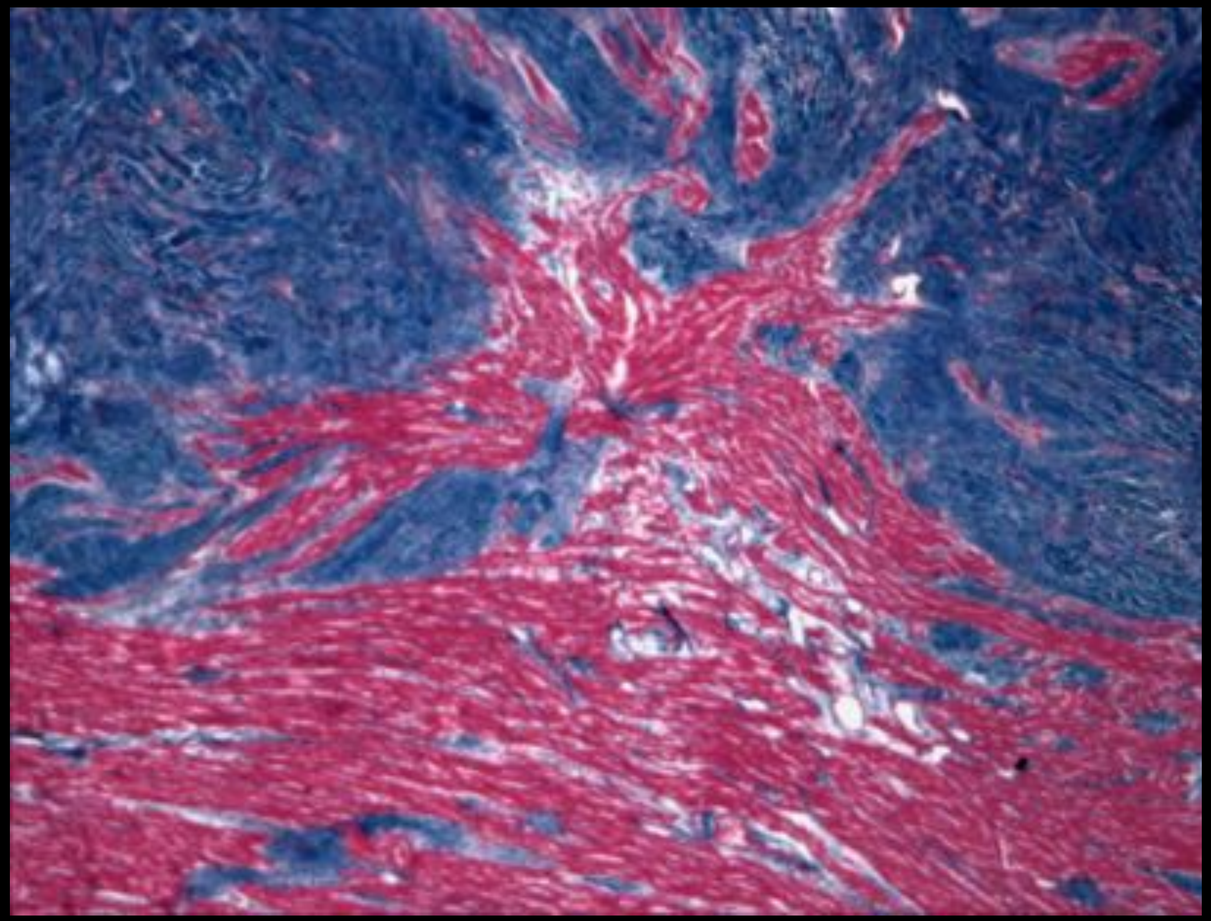


Fibroma

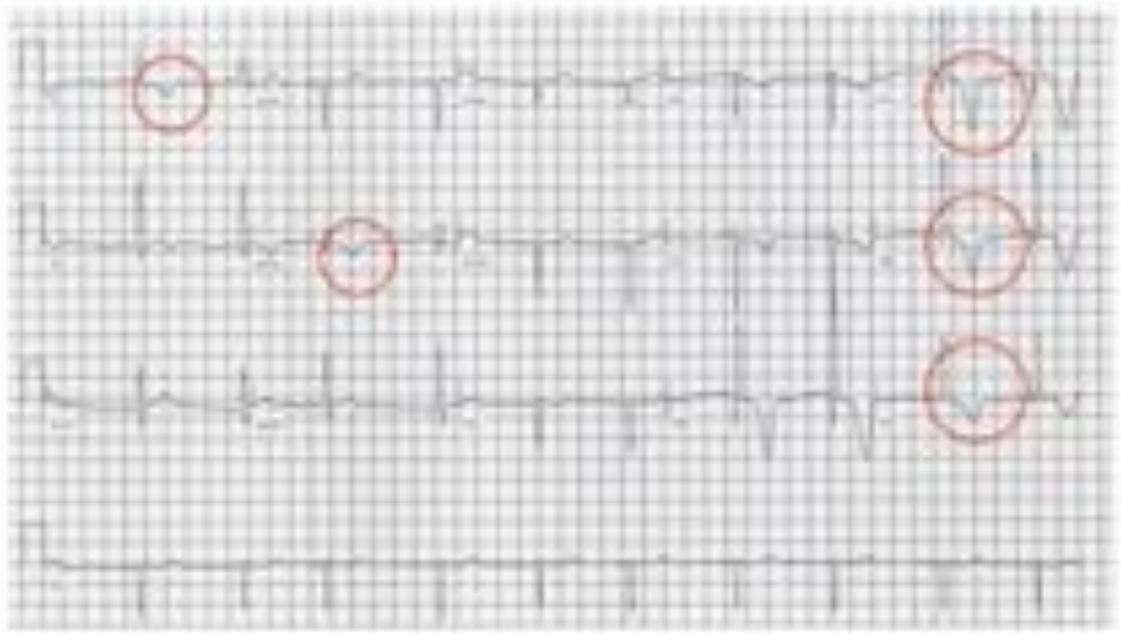




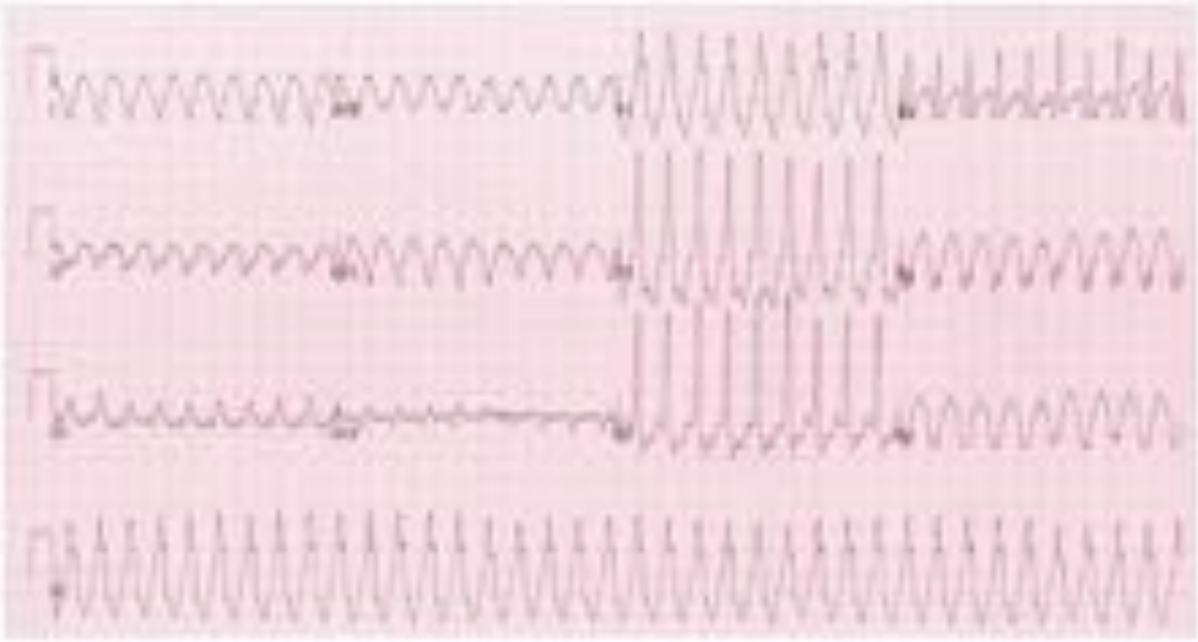
Fibroma

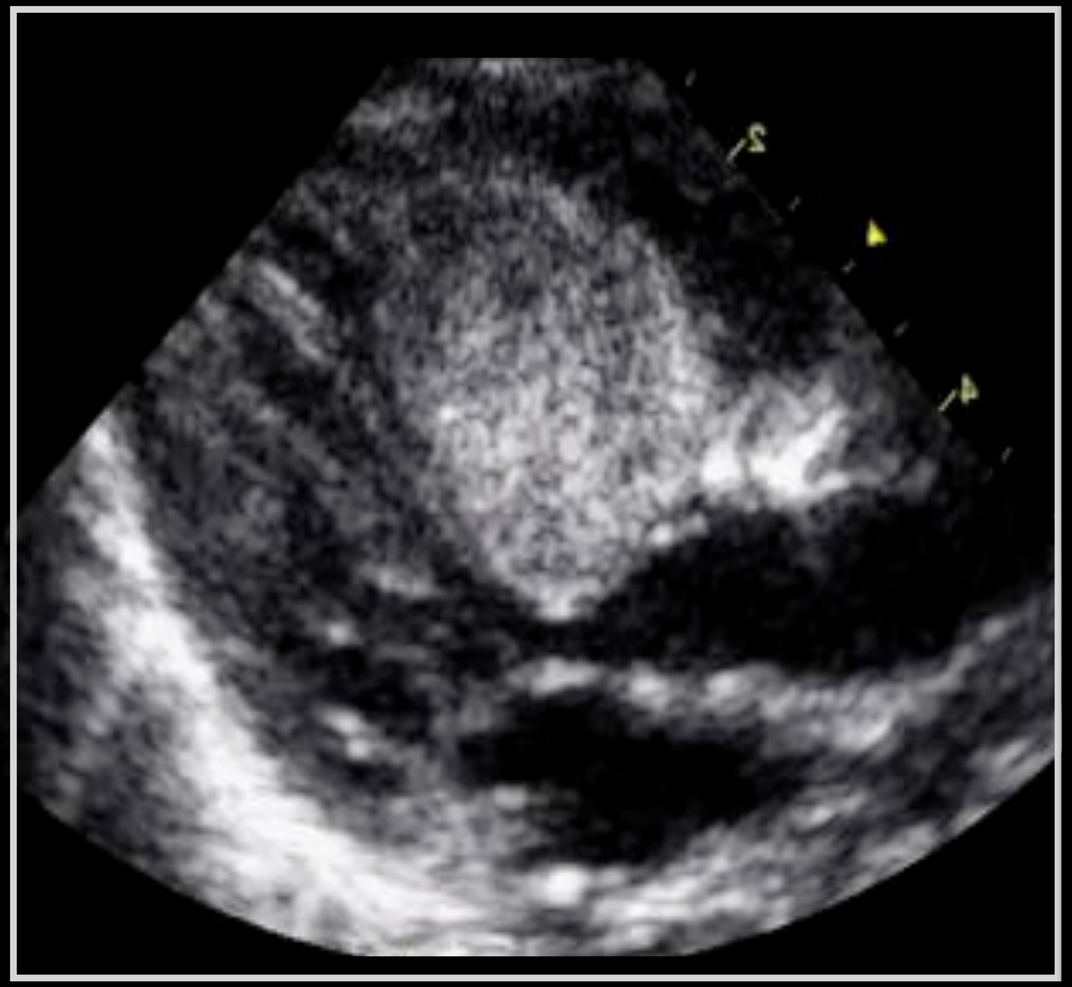
Fibroma - ECG

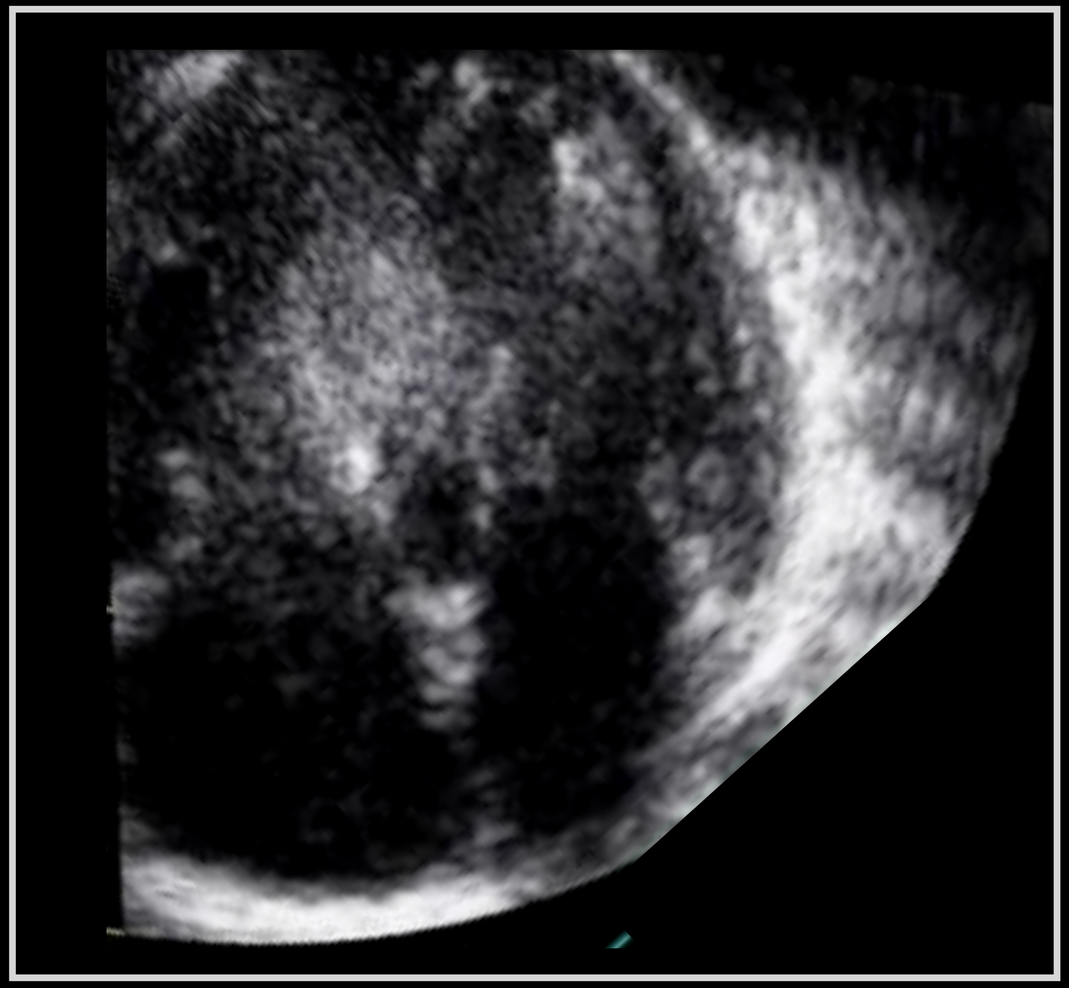
A



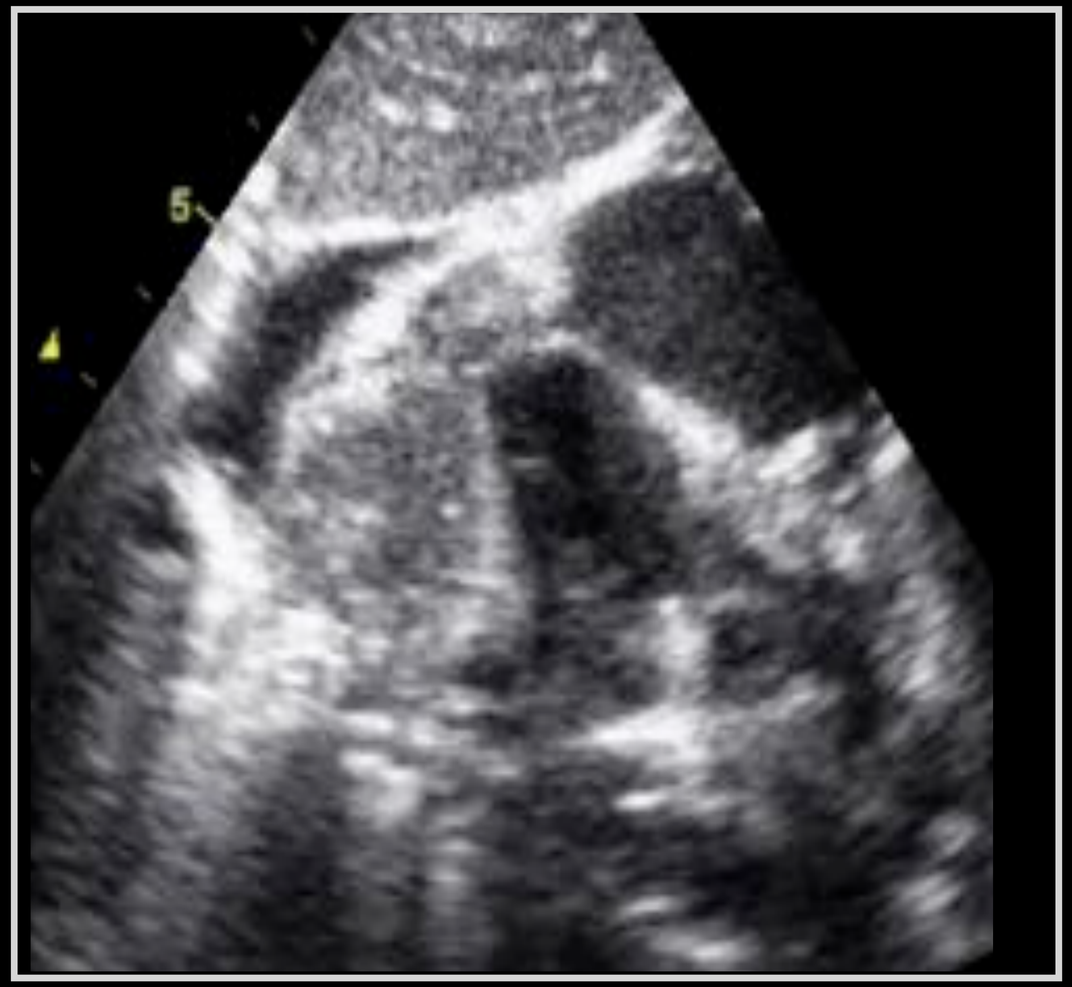
B

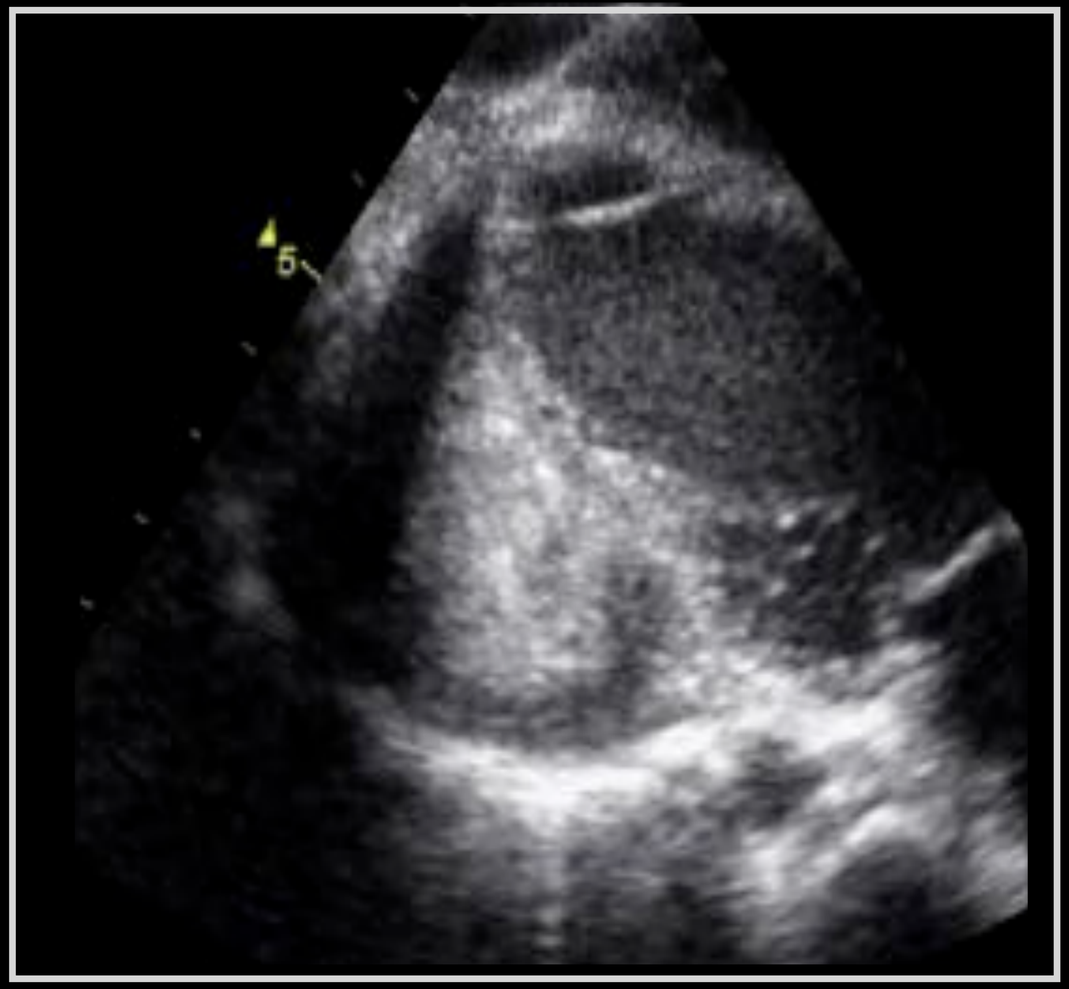




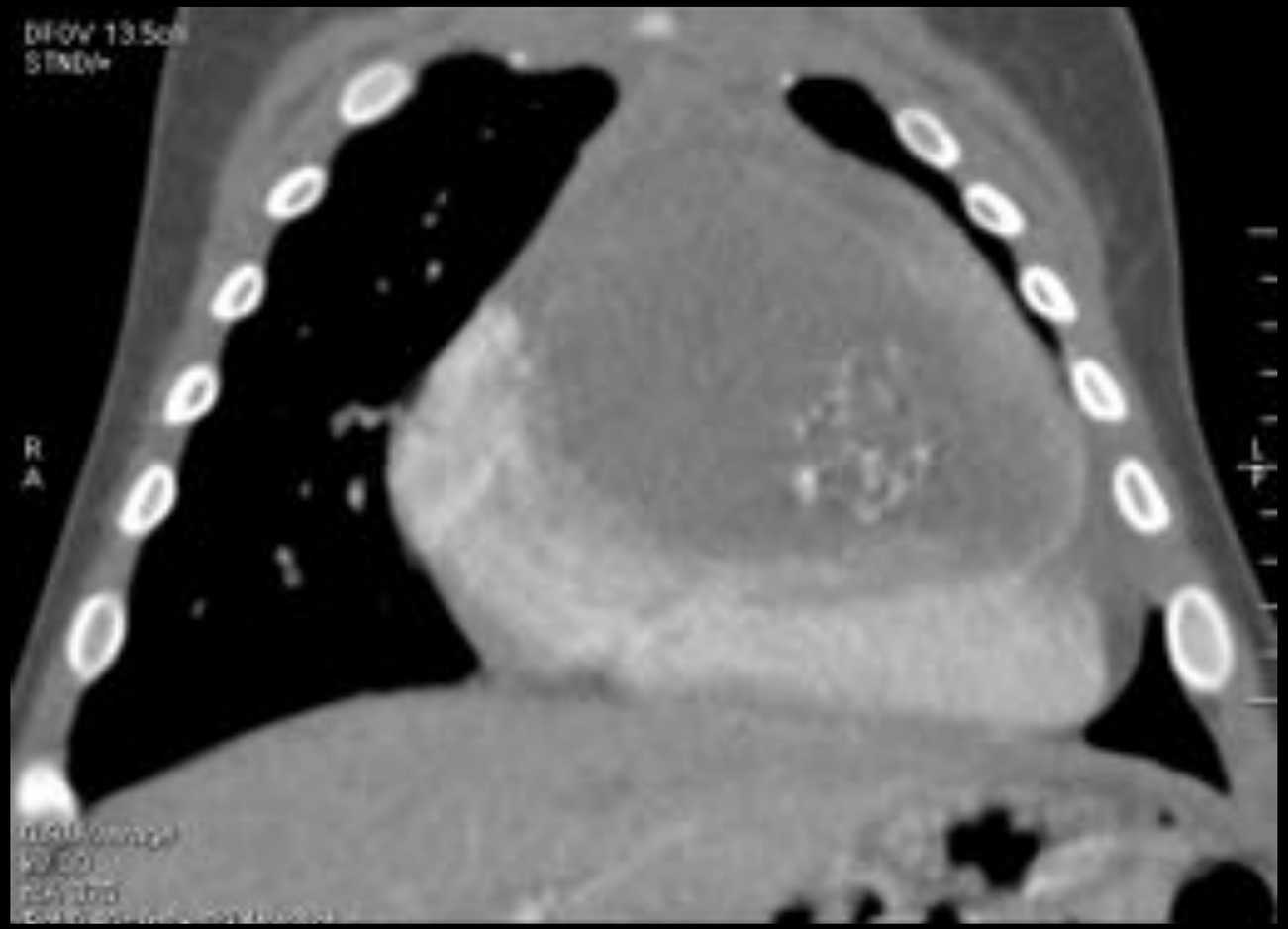


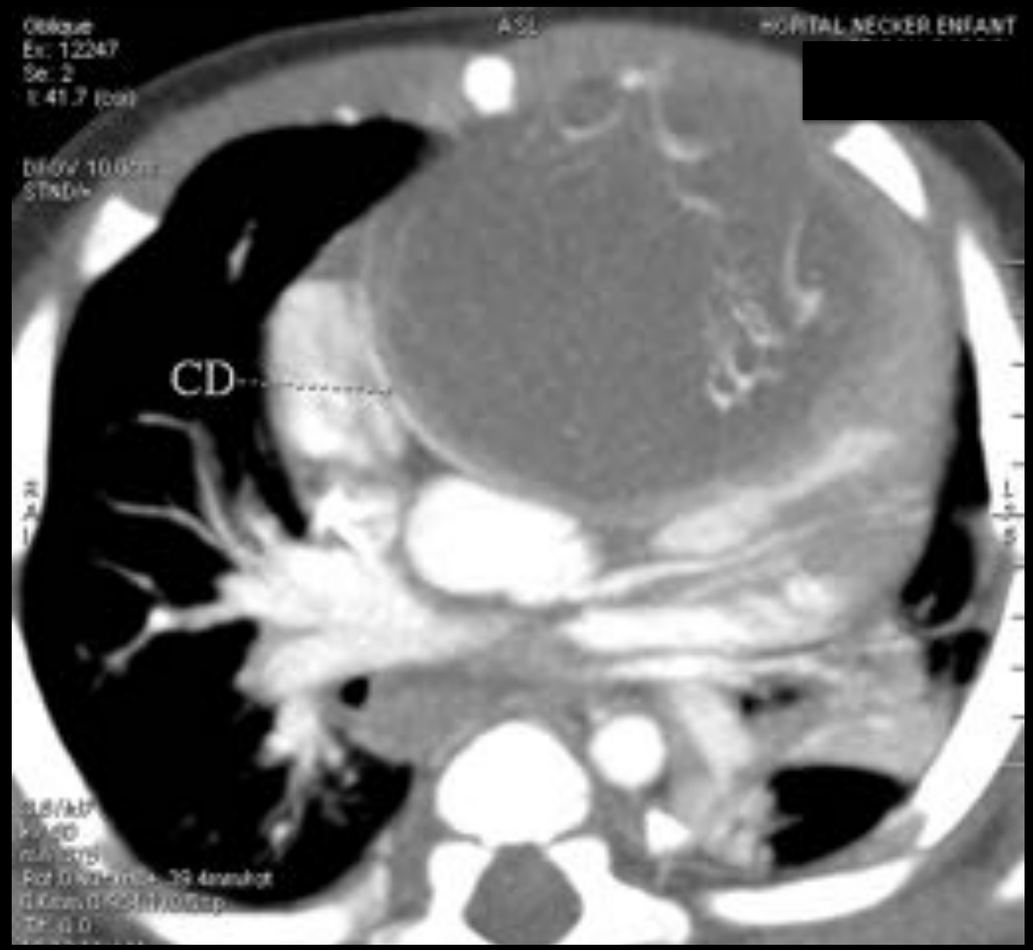
Fibroma - Unique, large, echogenicity close to that of adjacent myocardium



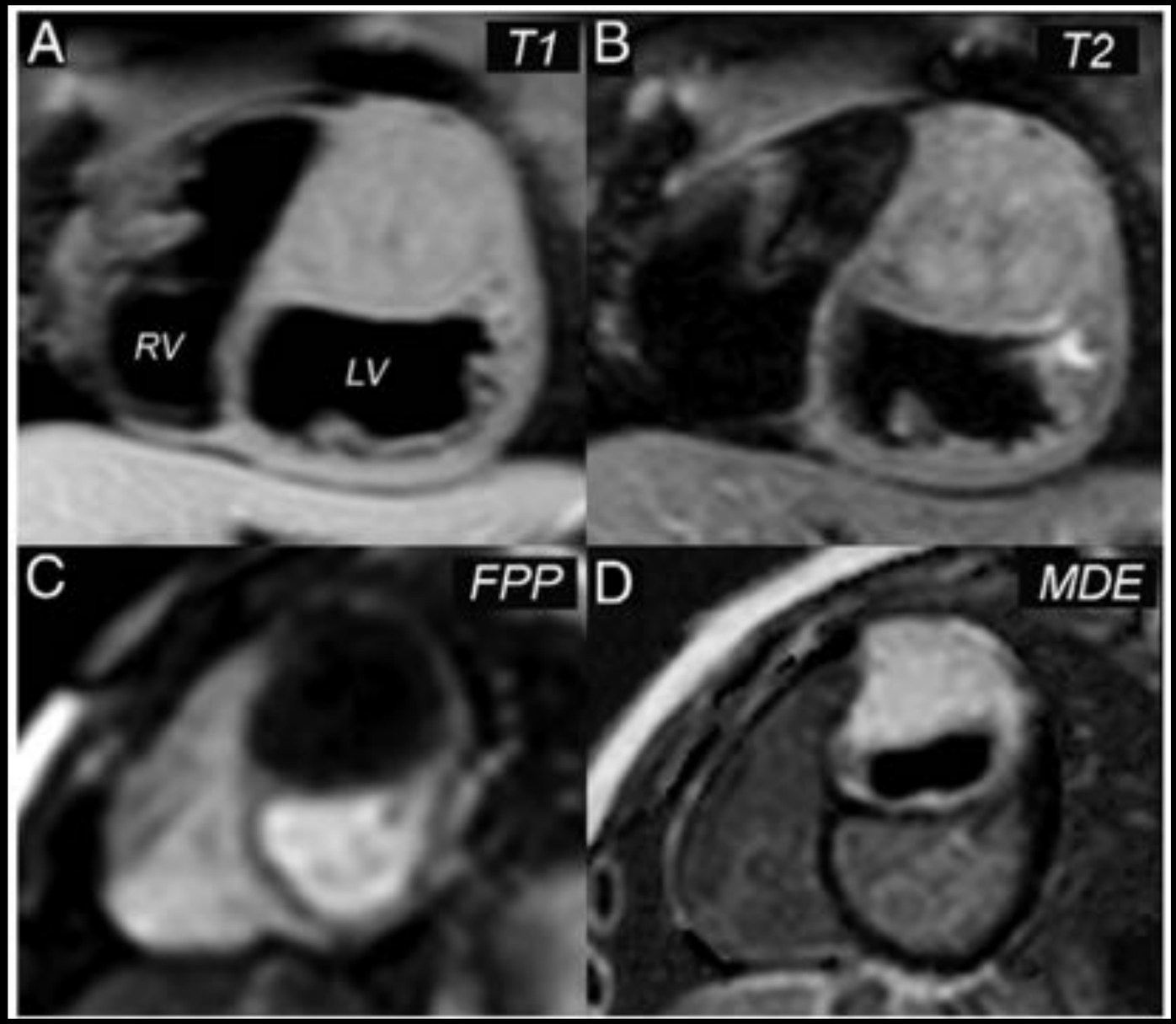


Fibroma



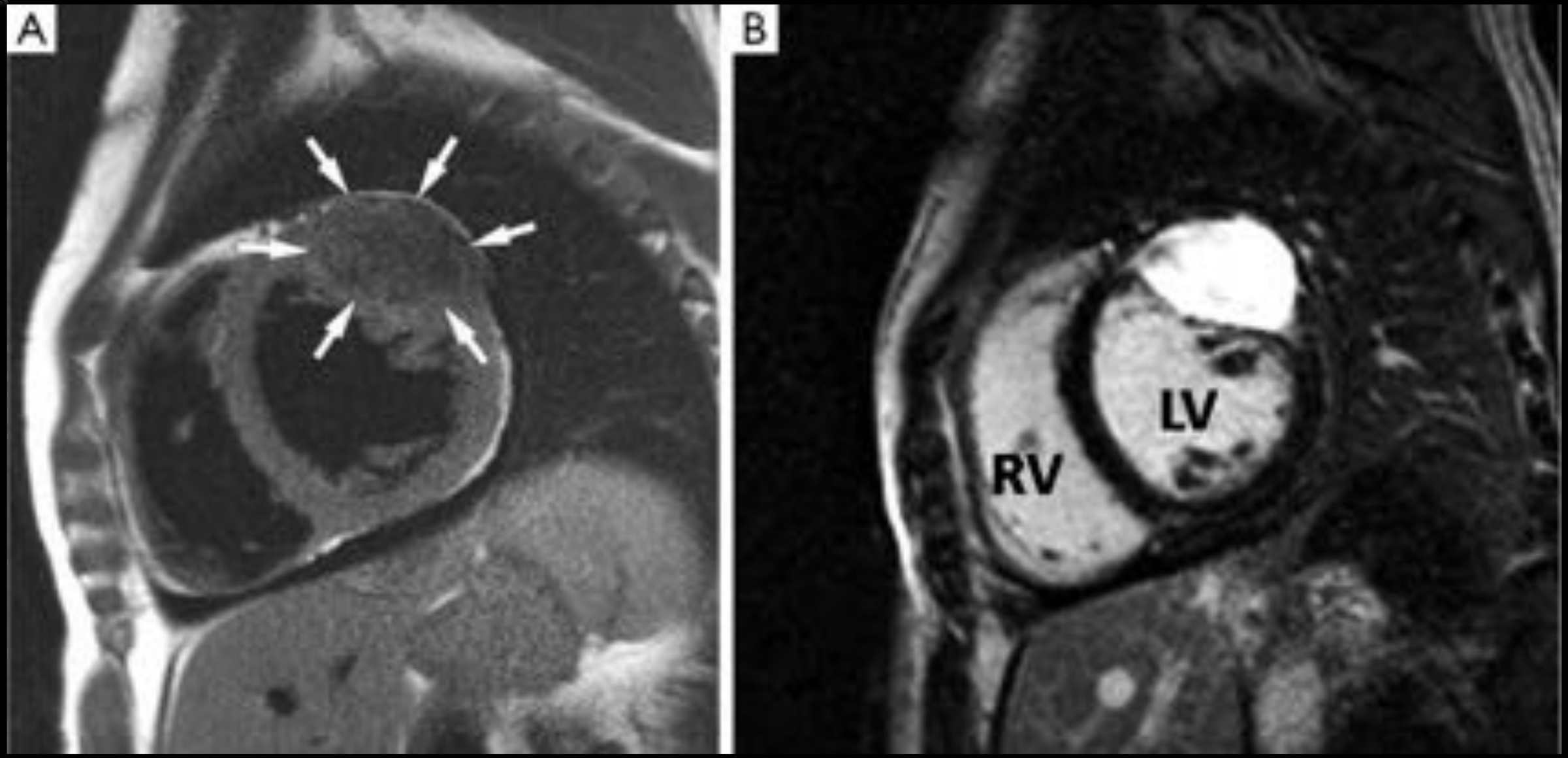


Fibroma - CT



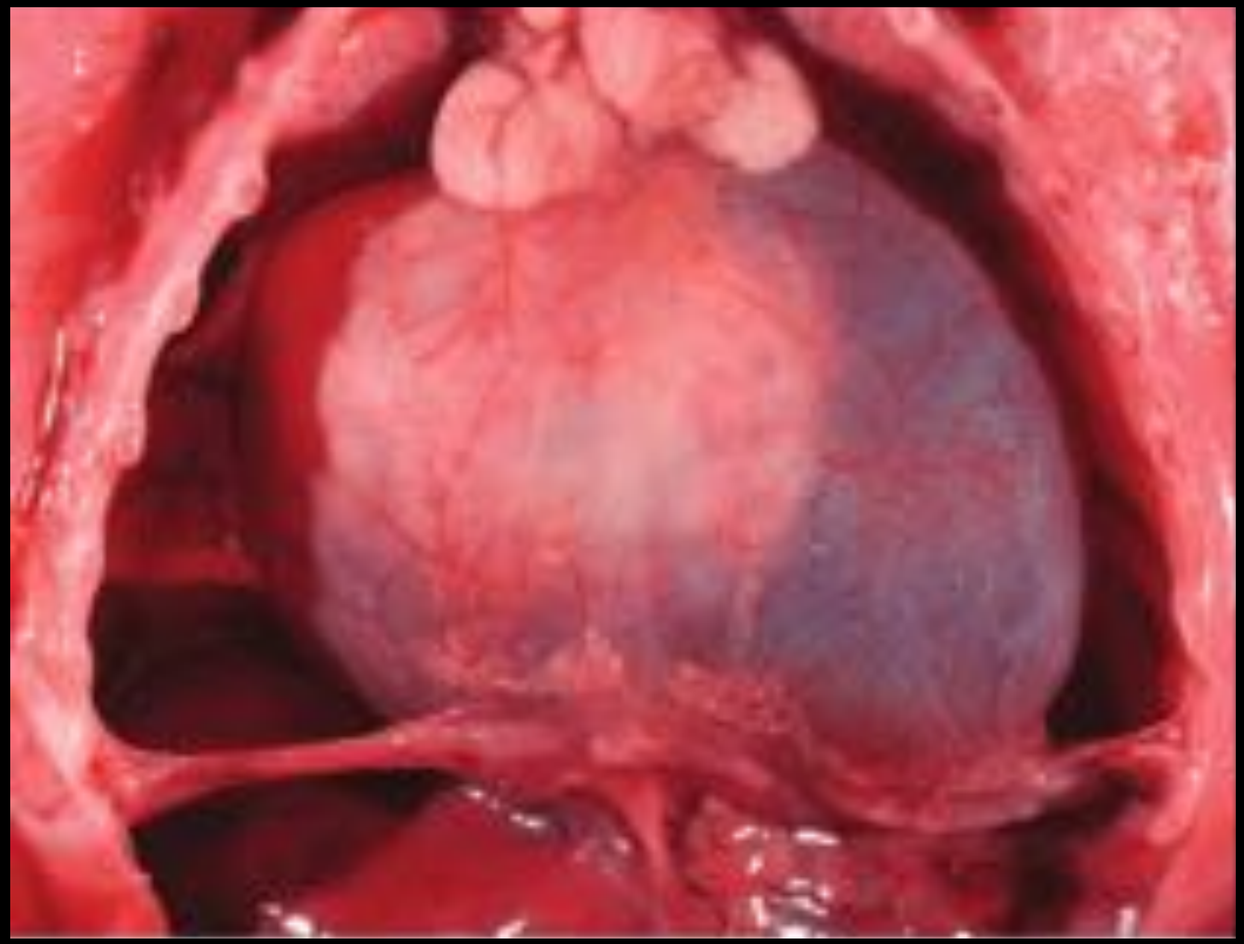
Fibroma - MRI

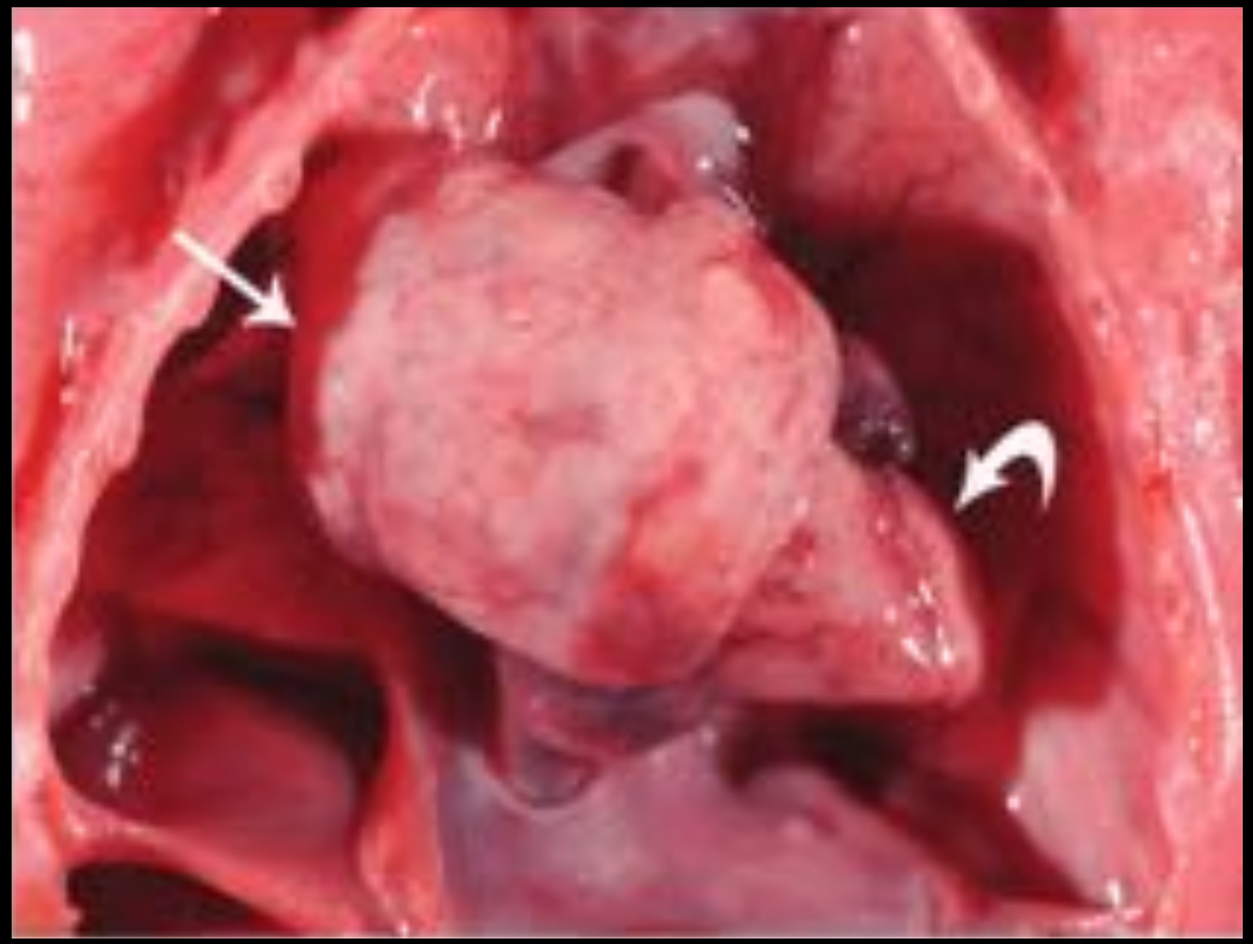
Beroukhim RS et al. JACC 2011



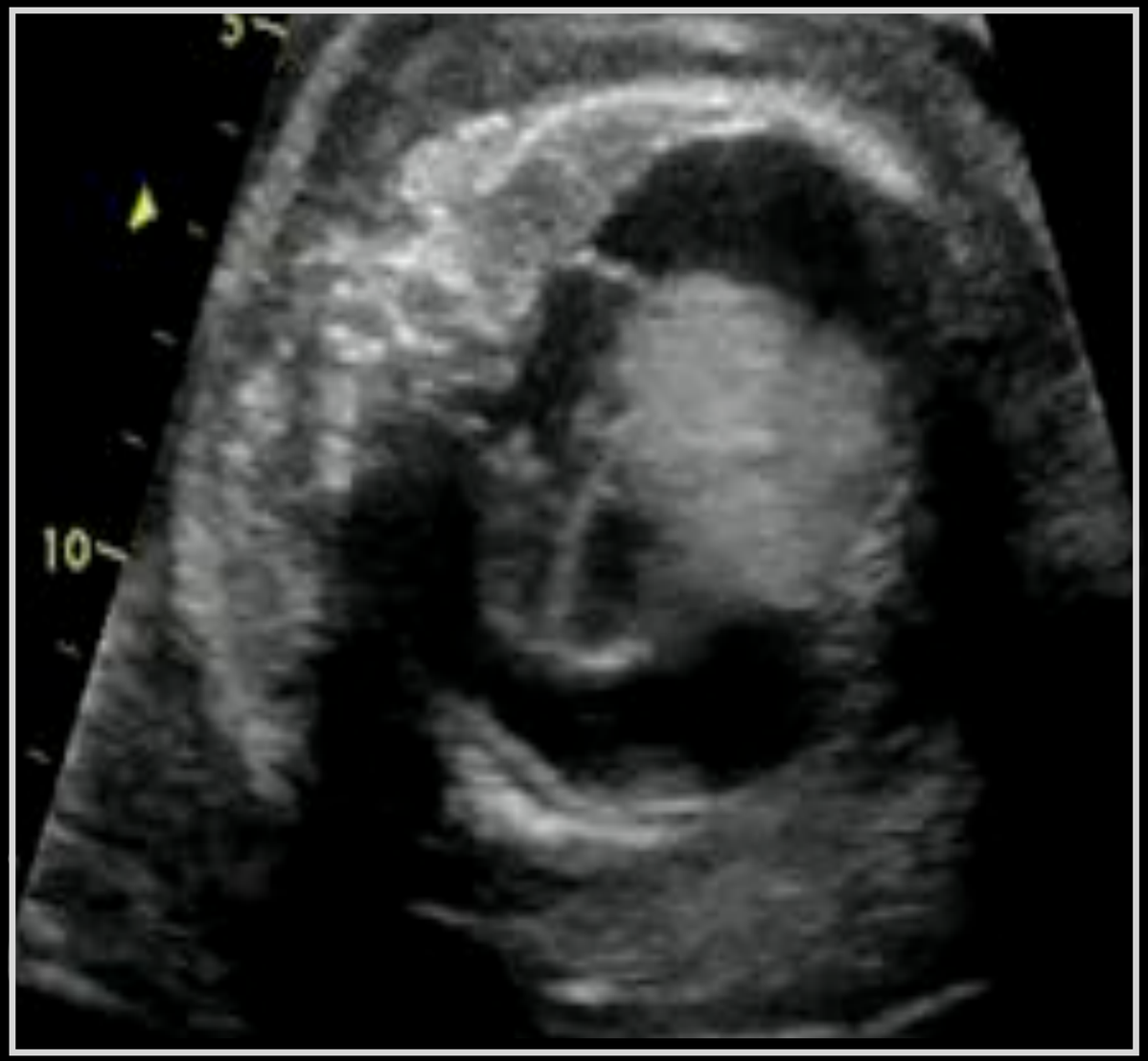
Fibroma - MRI

Teratoma

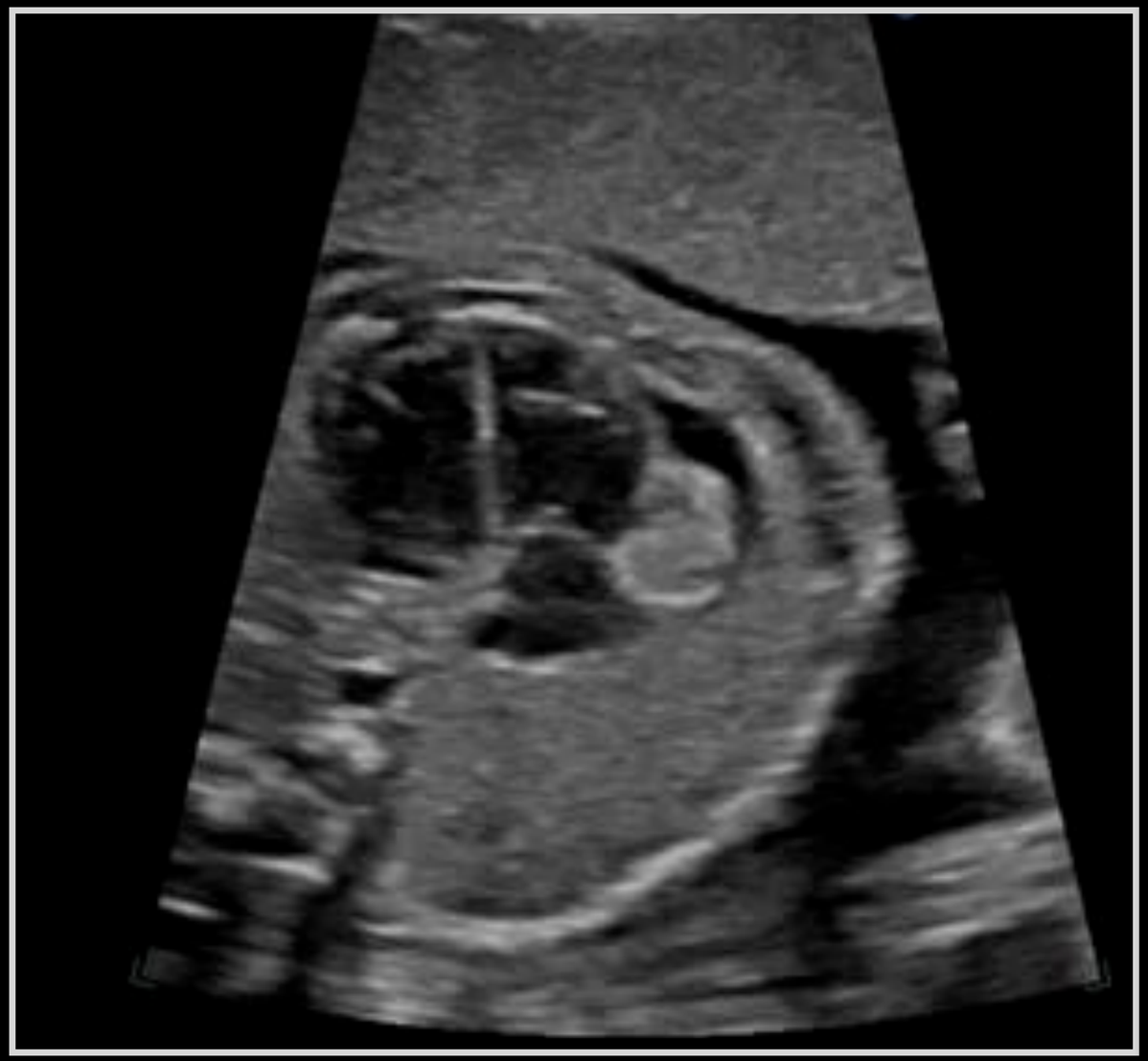




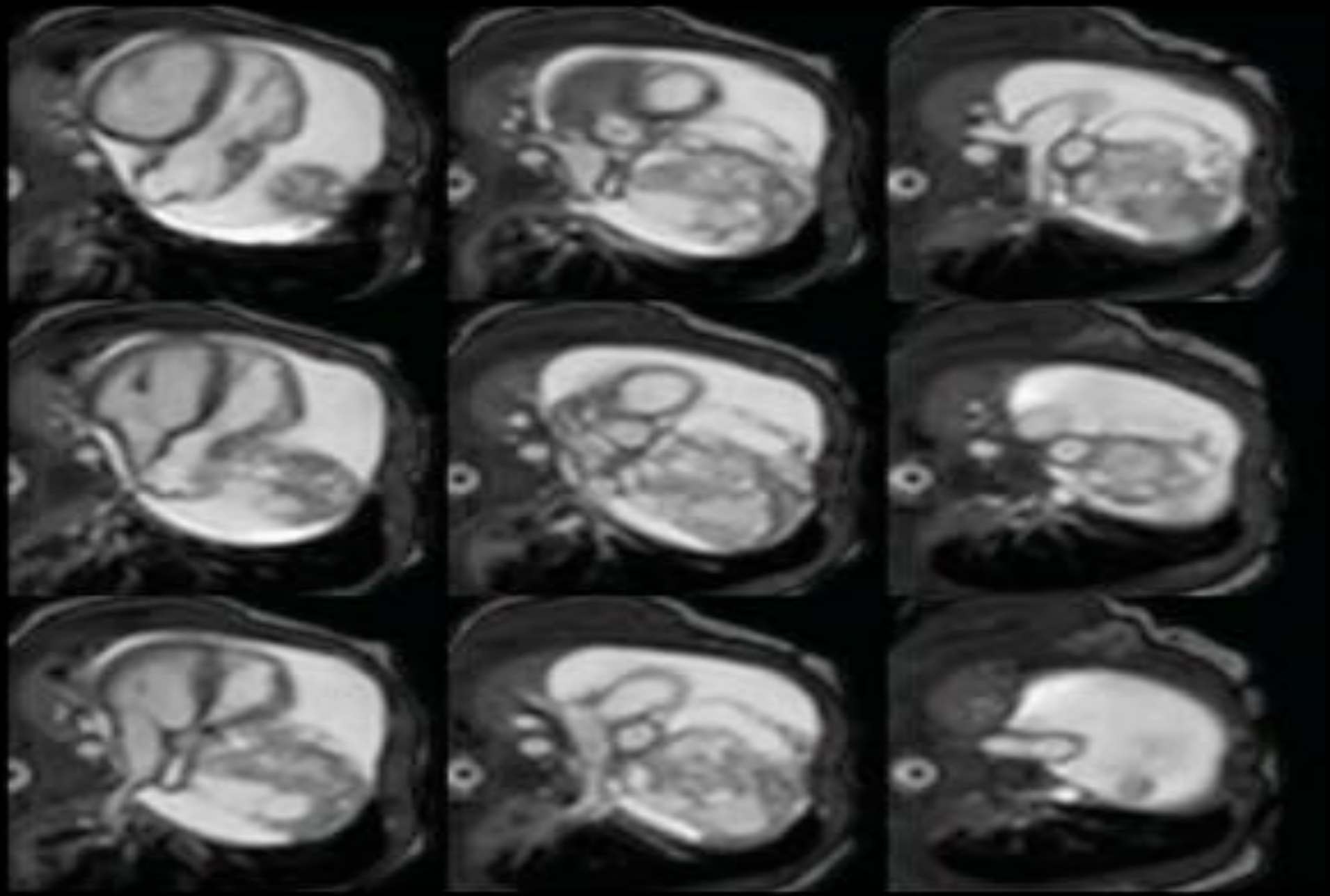
Intra-pericardial Teratoma



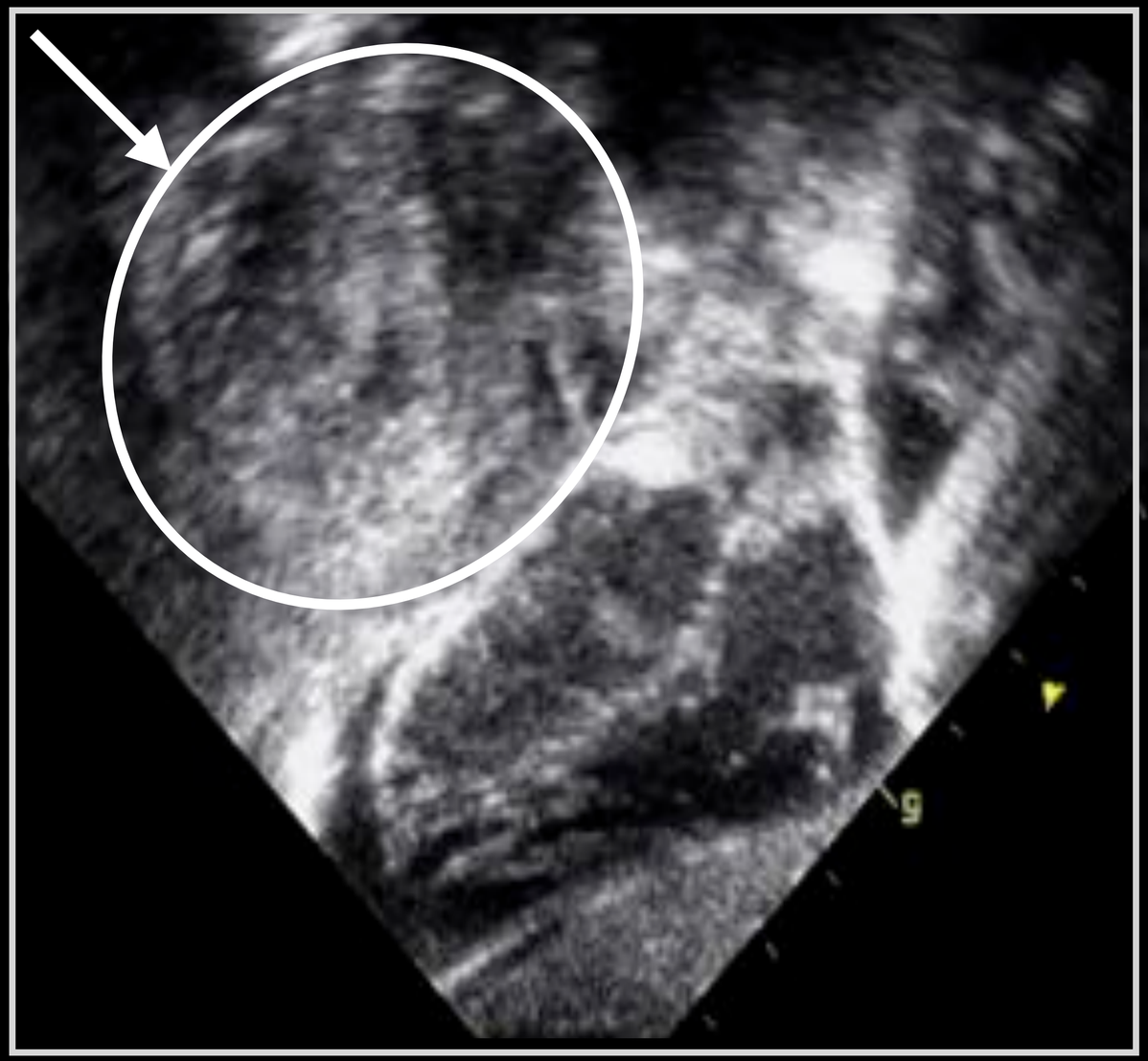
Intra-pericardial Teratoma



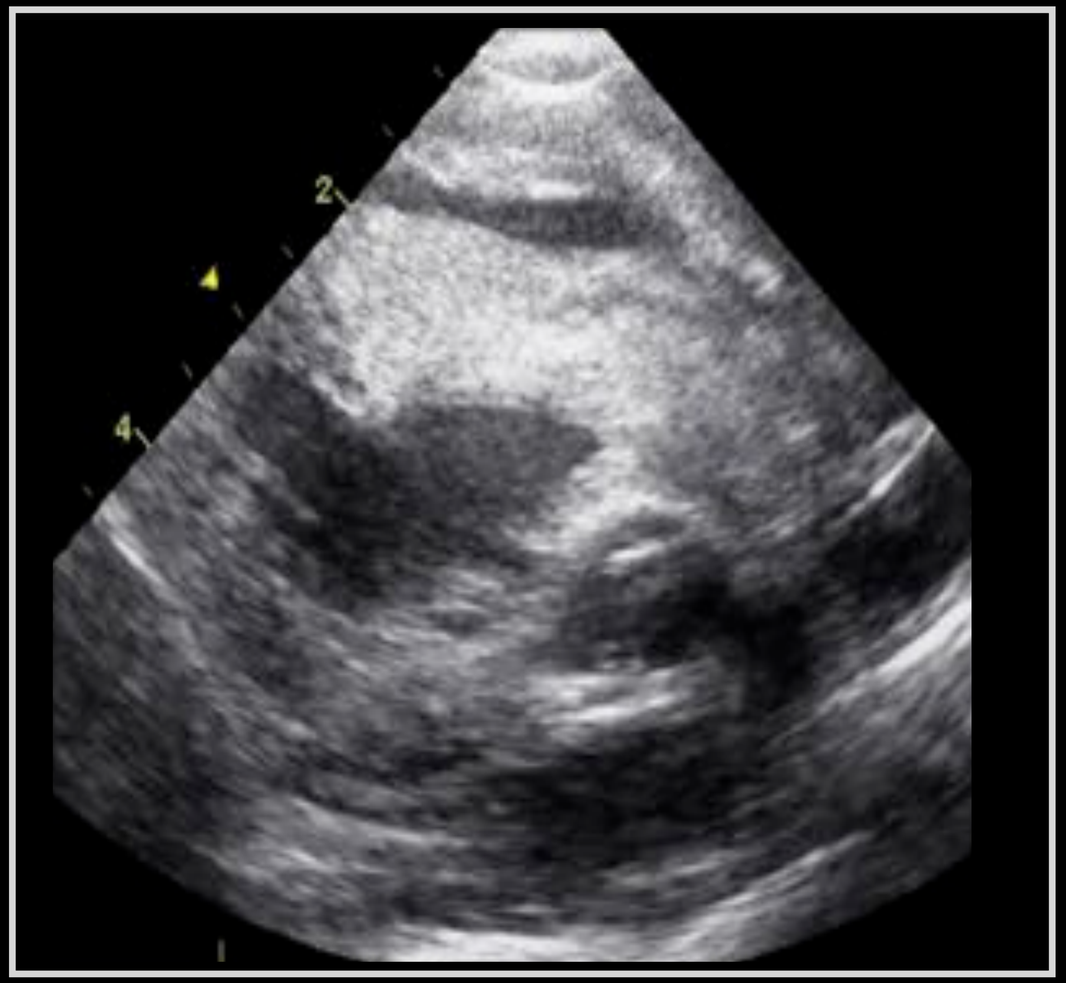
Intra-pericardial Teratoma

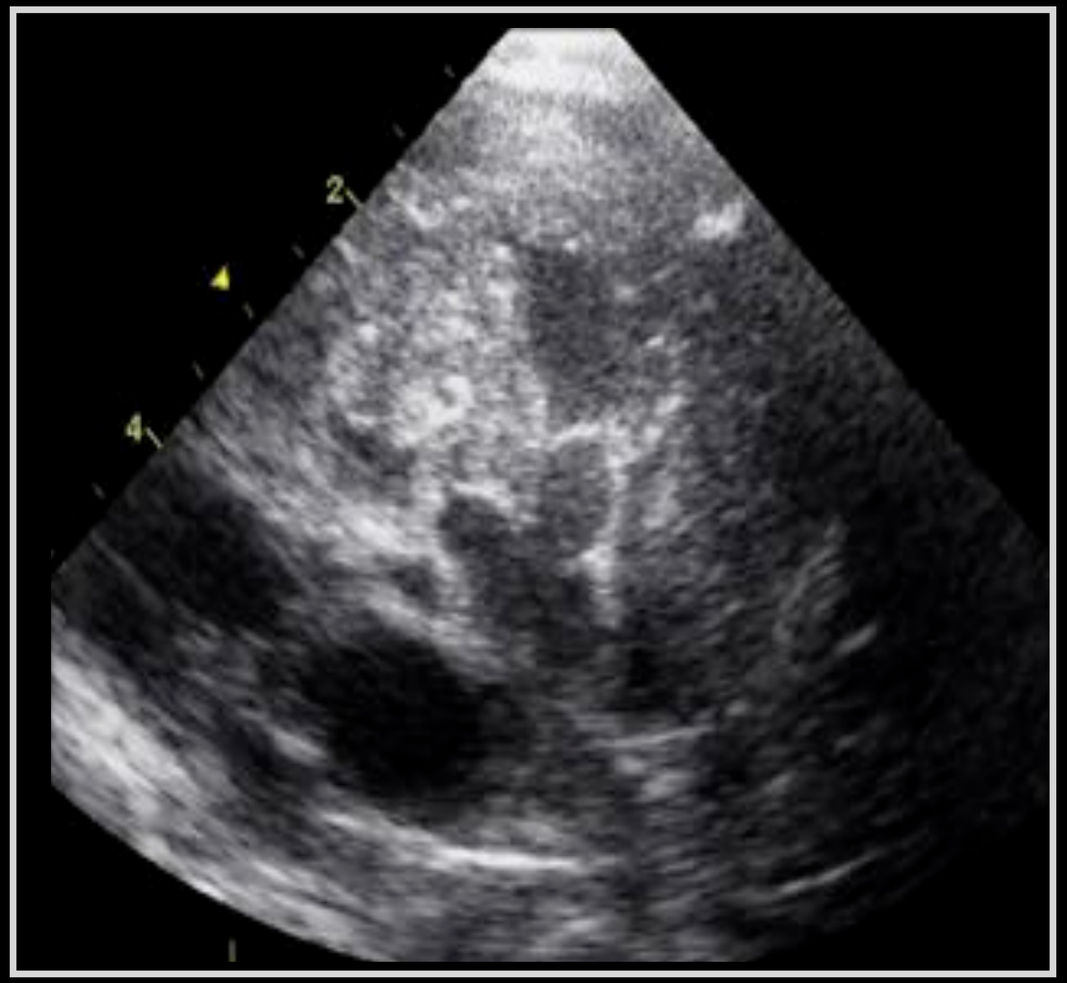


Intra-pericardial Teratoma

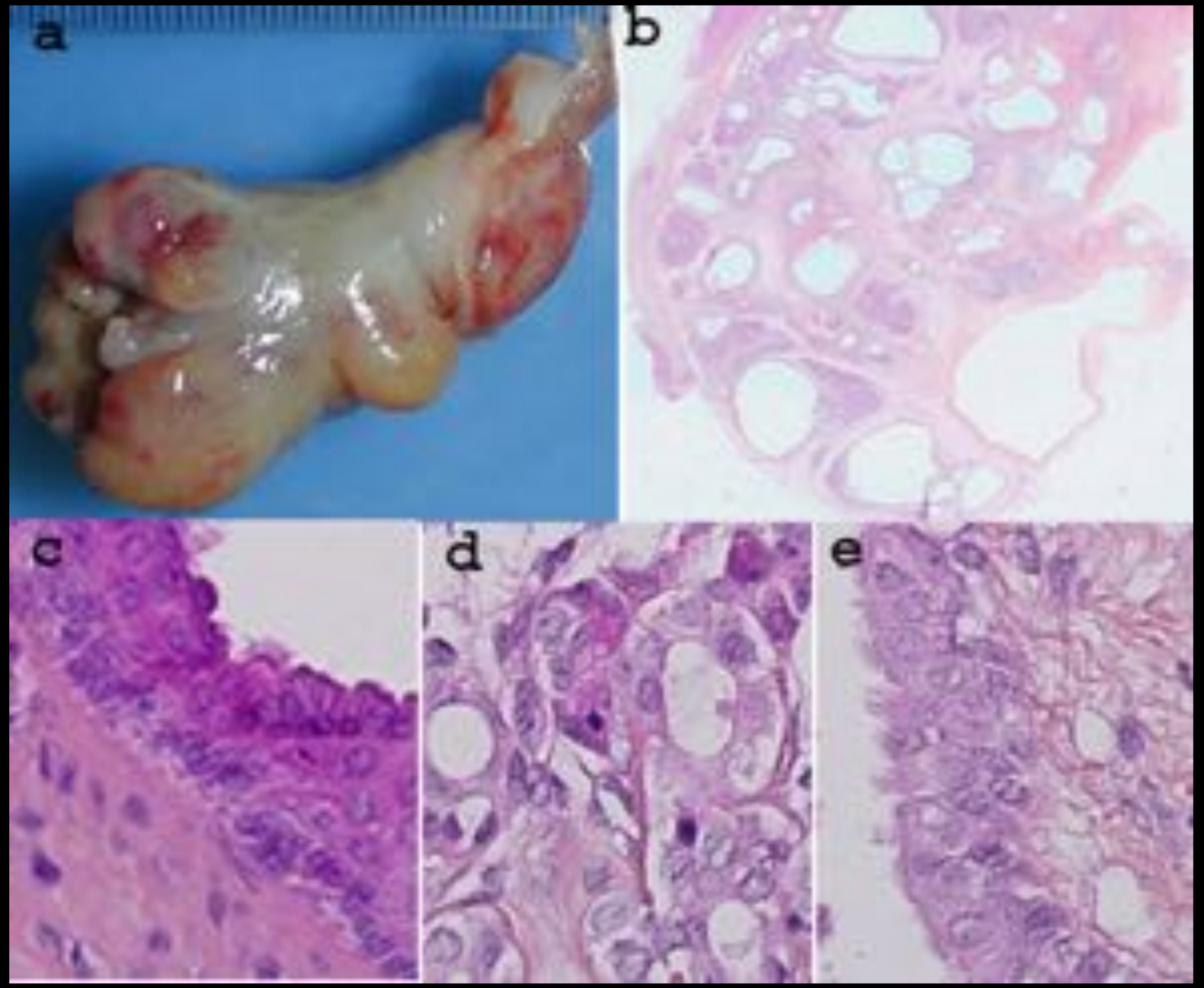


Intra-pericardial Teratoma - Neonatal



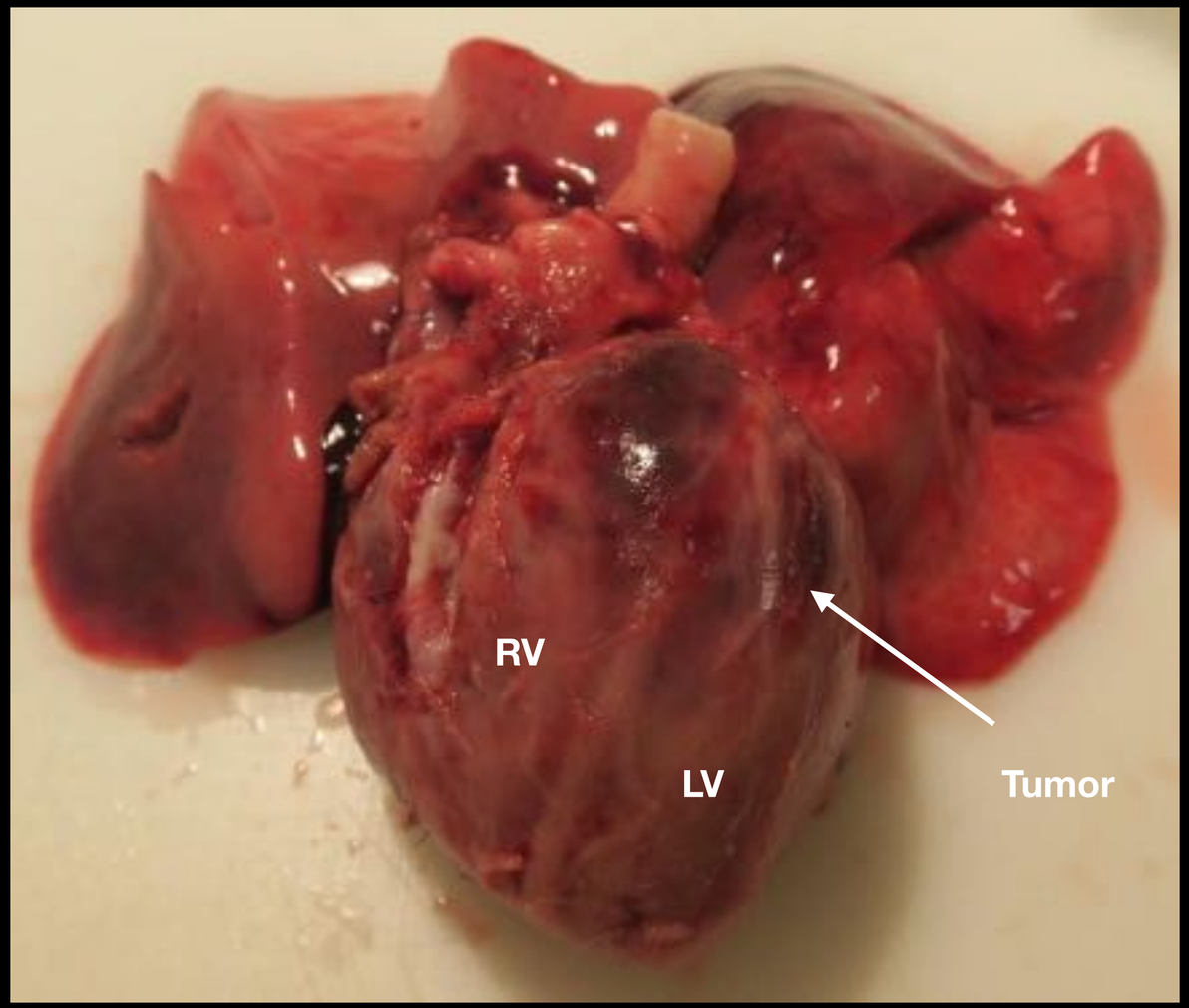


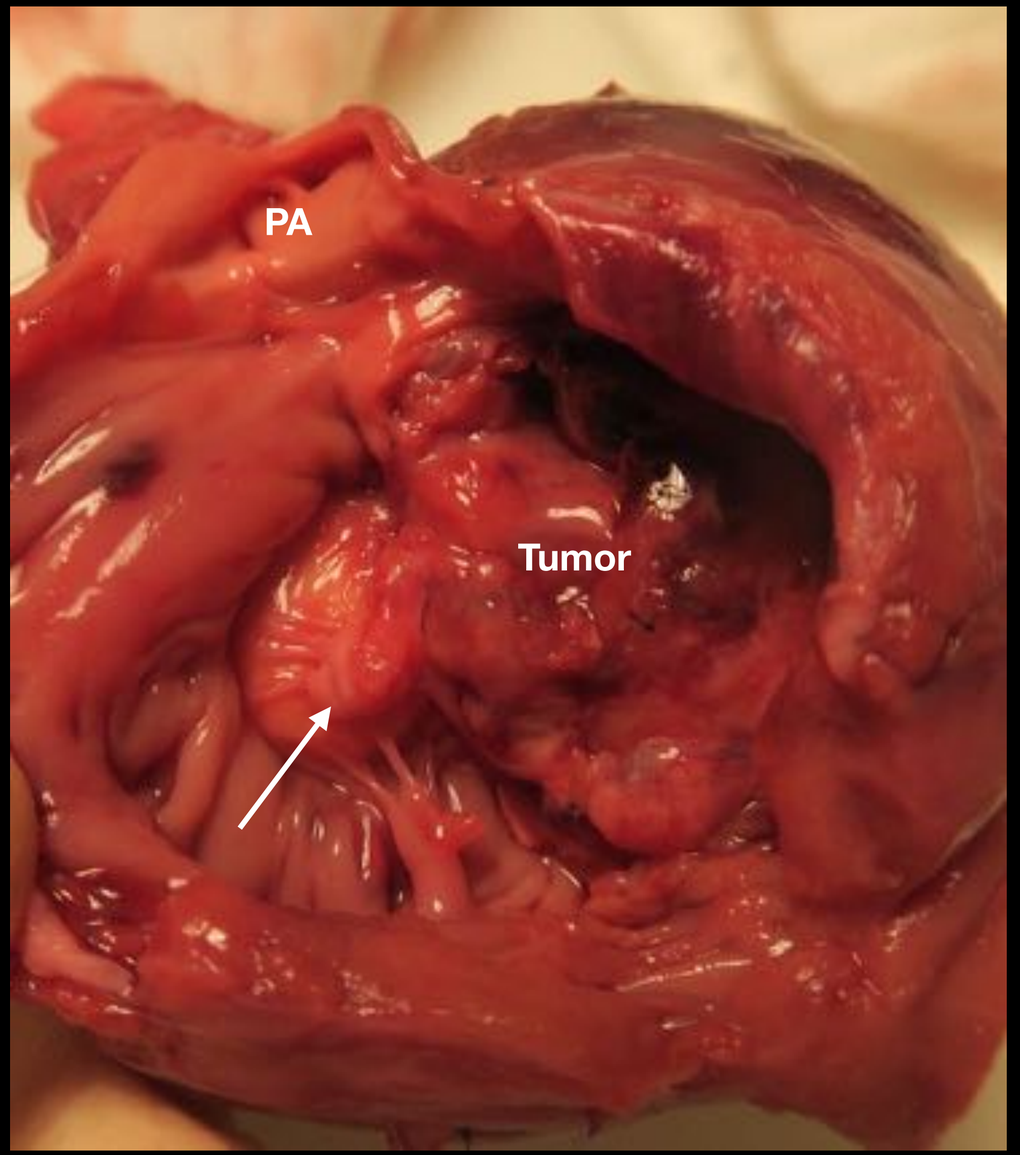
Cardiac Teratoma



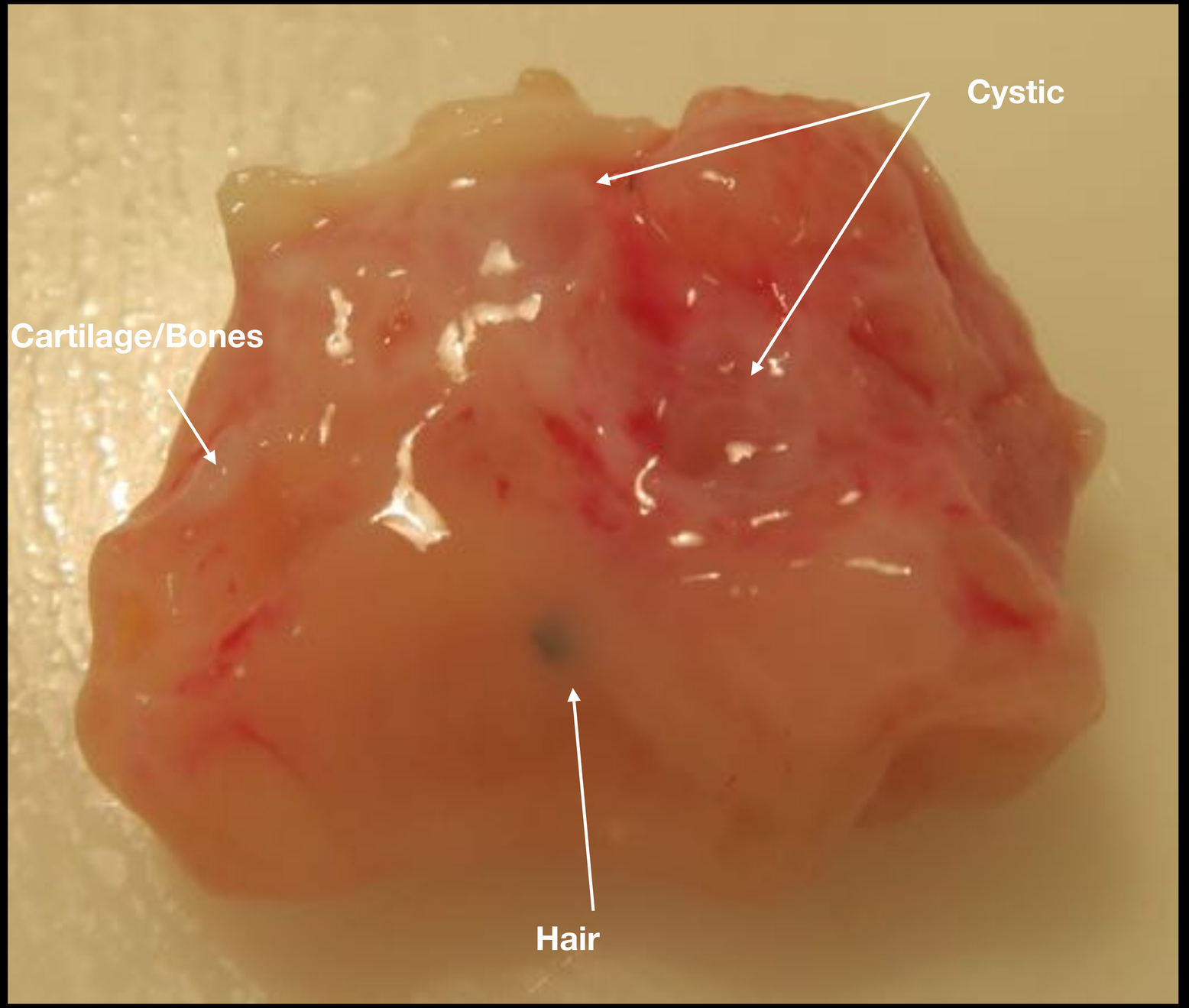
Cardiac Teratoma

Ou P et al. Circulation 2006;113:17-8

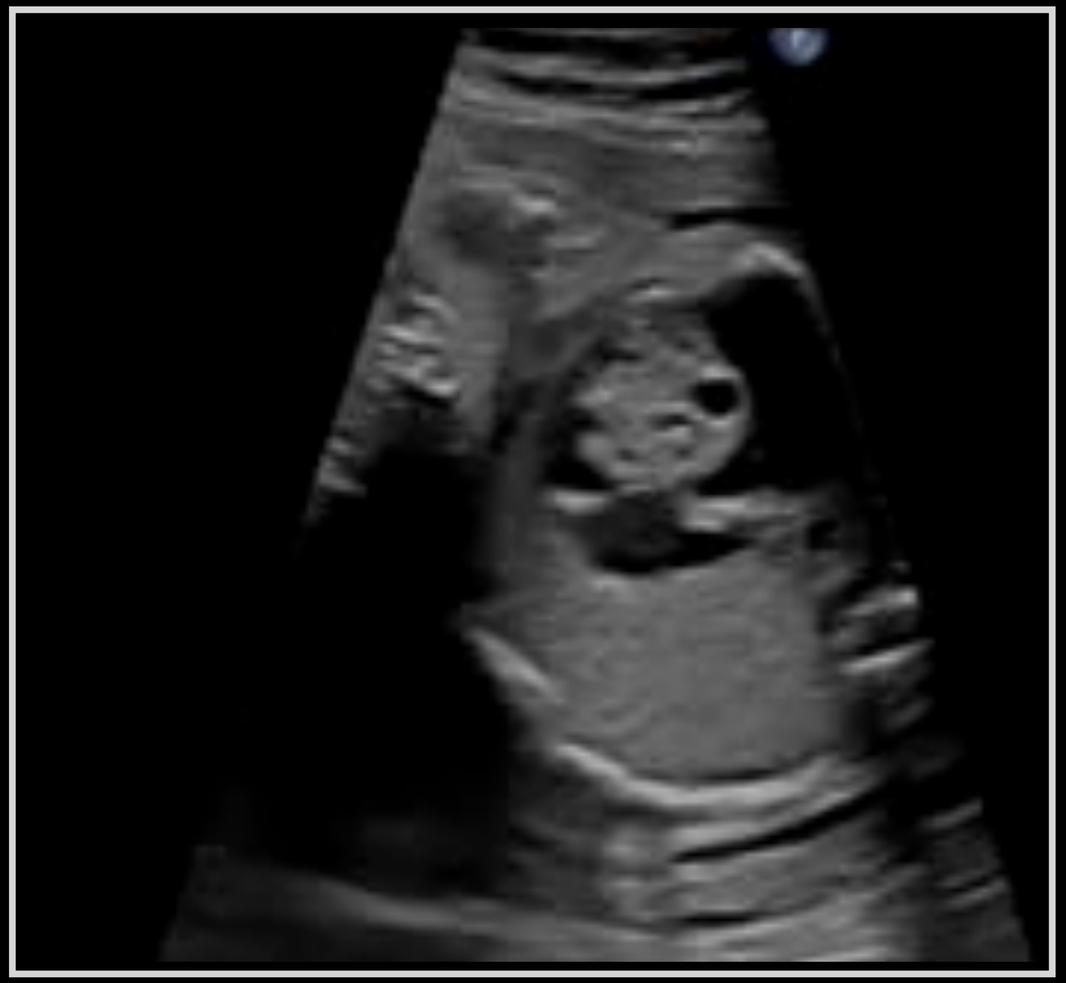


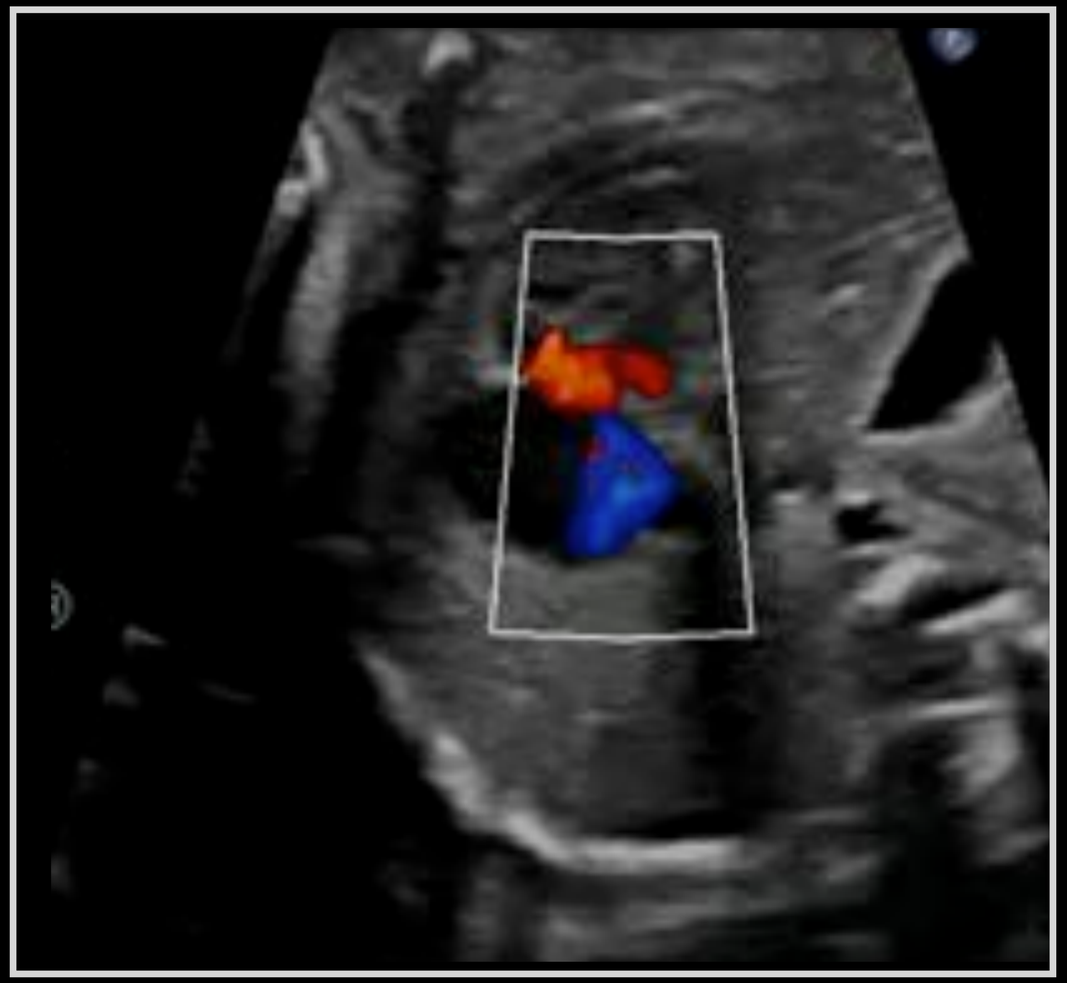


Cardiac Teratoma



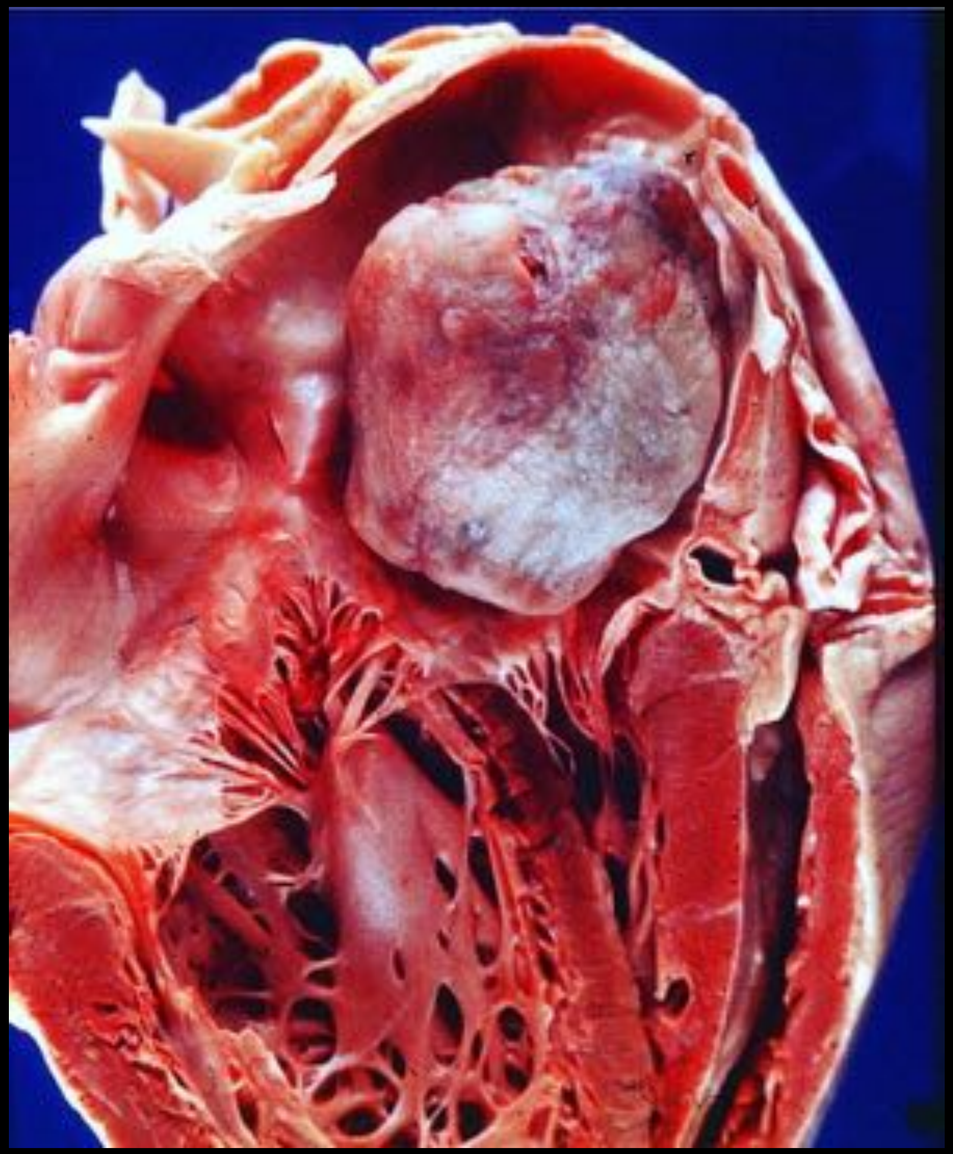
Cardiac Teratoma

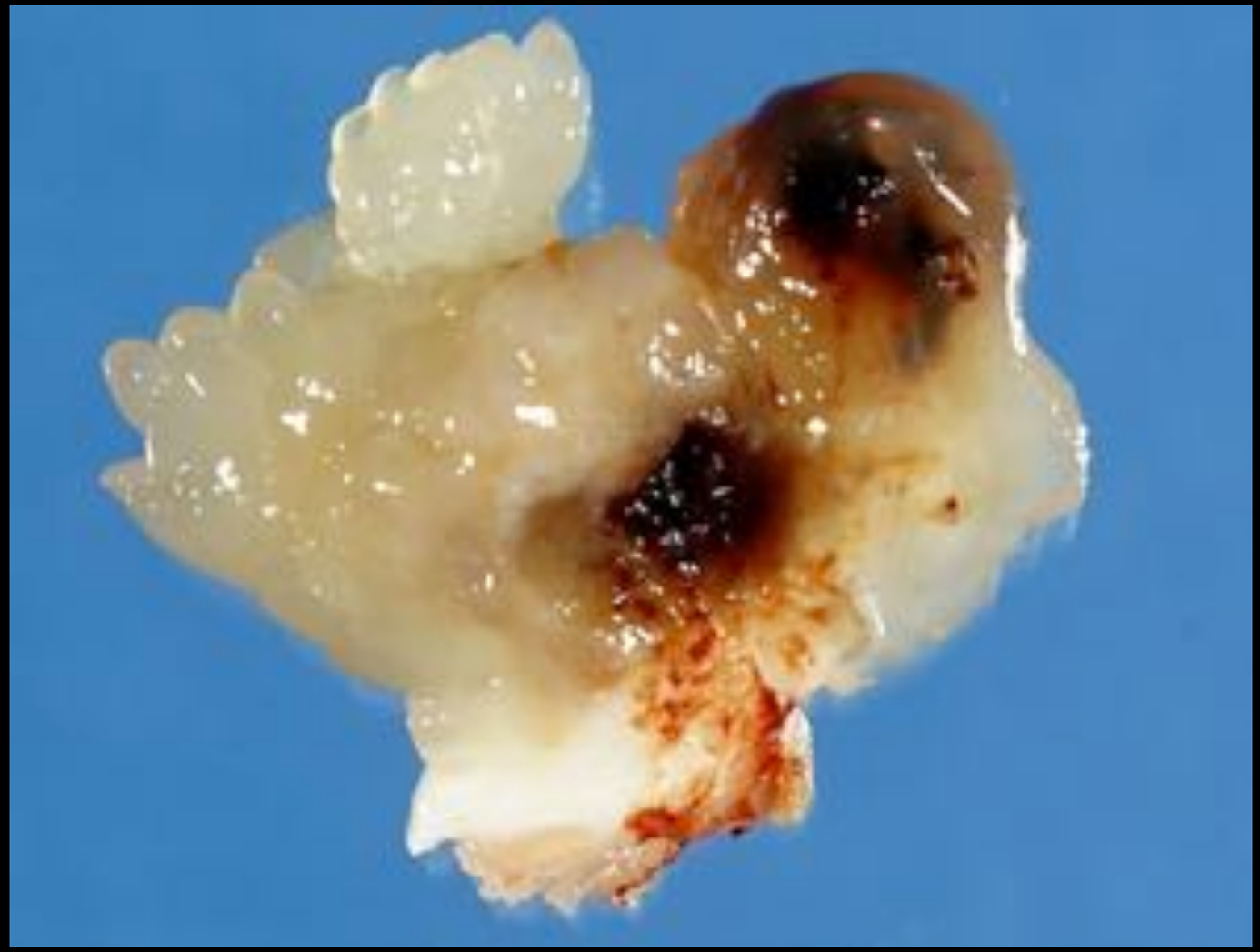




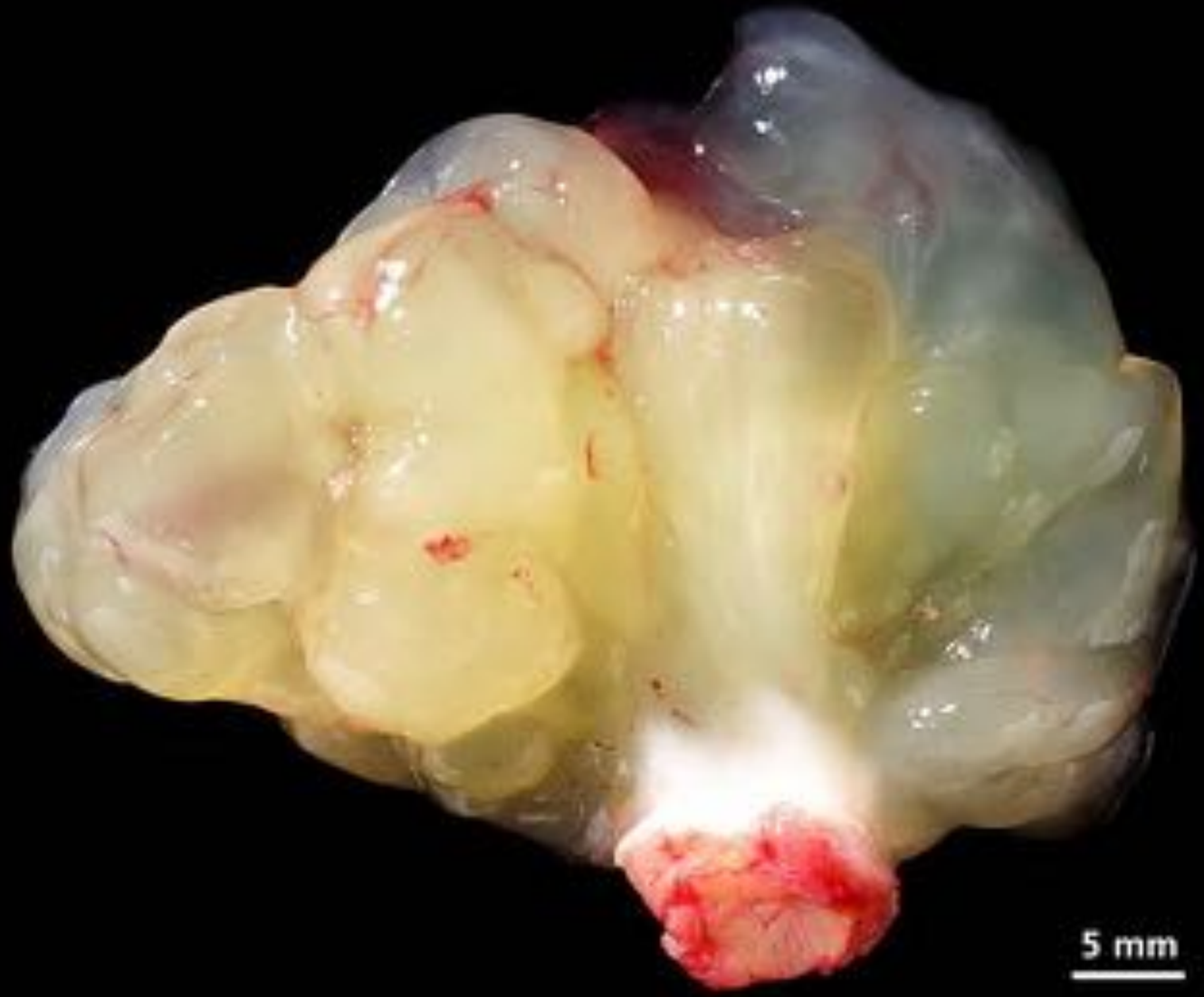
Cardiac Teratoma

Myxoma

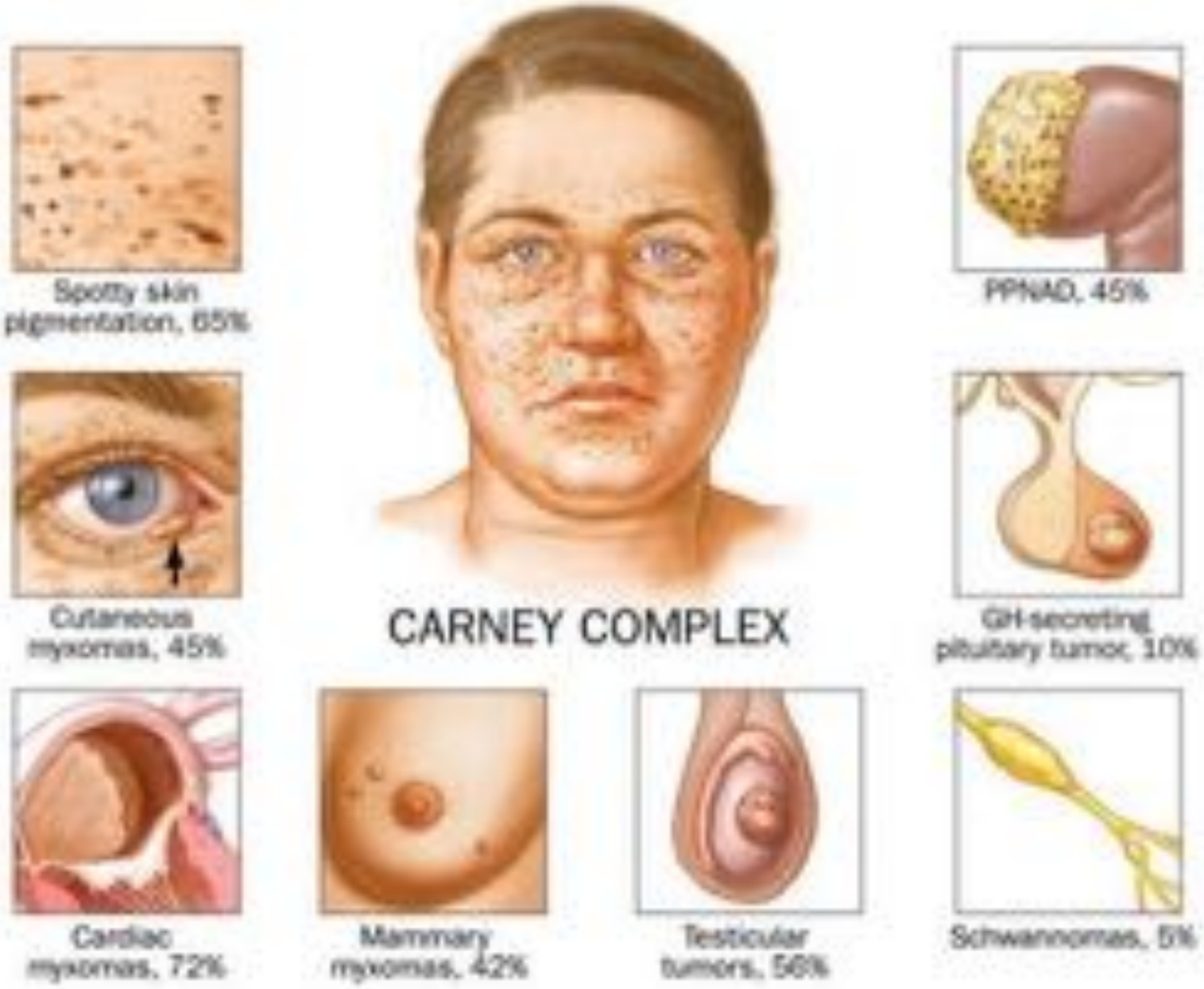




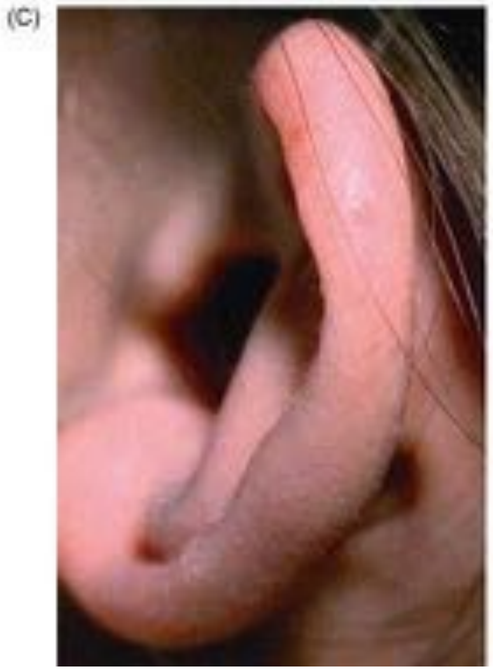
Myxoma

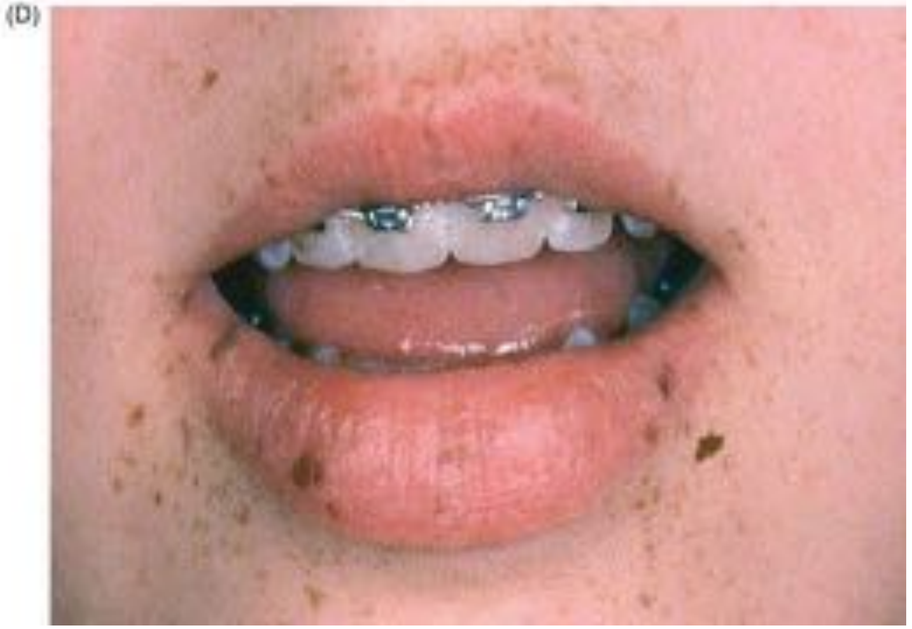


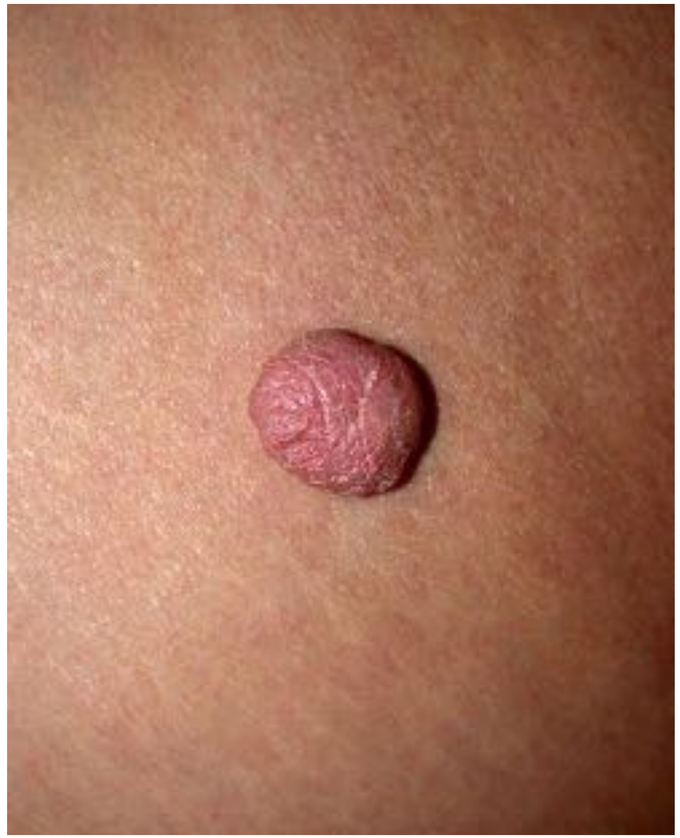
Myxoma


Spotty skin pigmentation, 65%
PPNAD, 45%
Cutaneous myxomas, 45%
CARNEY COMPLEX
GH-secreting pituitary tumor, 10%
Cardiac myxomas, 72%
Mammary myxomas, 42%
Testicular tumors, 56%
Schwannomas, 5%

Main features of Carney complex	(%)
Primary Pigmented Nodular Adrenocortical Disease (PPNAD)	25–60
Cardiac myxoma	30–60
Skin myxoma	20–63
Lentiginosis	60–70
Multiple blue nevus	
Breast ductal adenoma	25
Testicular tumors (LCCSCT: Large-Cell Calcifying Sertoli Cell Tumor) (in male)	33–56
Ovarian cyst (in female)	20–67
Acromegaly	10
Thyroid tumor	10–25
Melanotic schwannoma	8–18
Osteochondromyxoma	<10



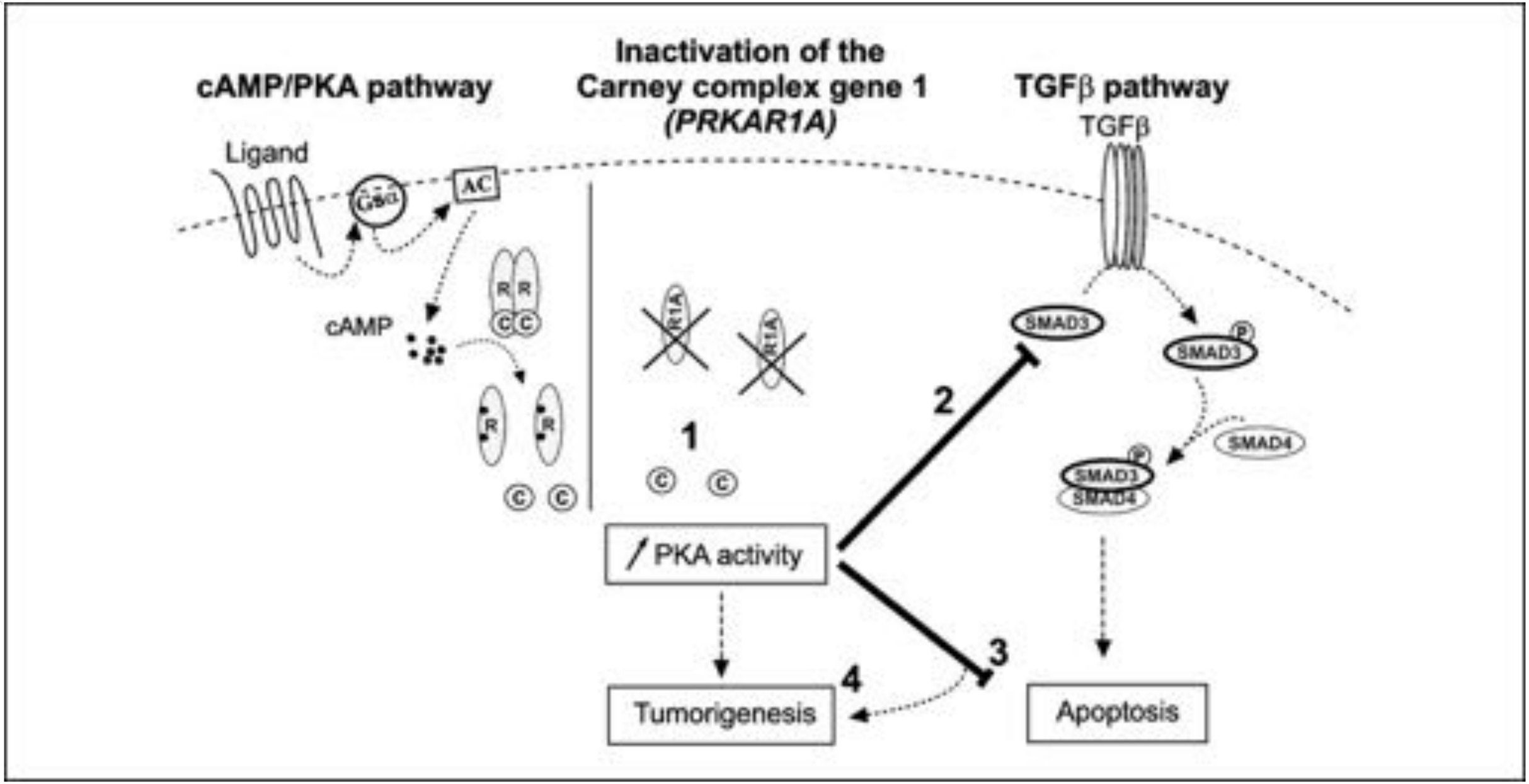




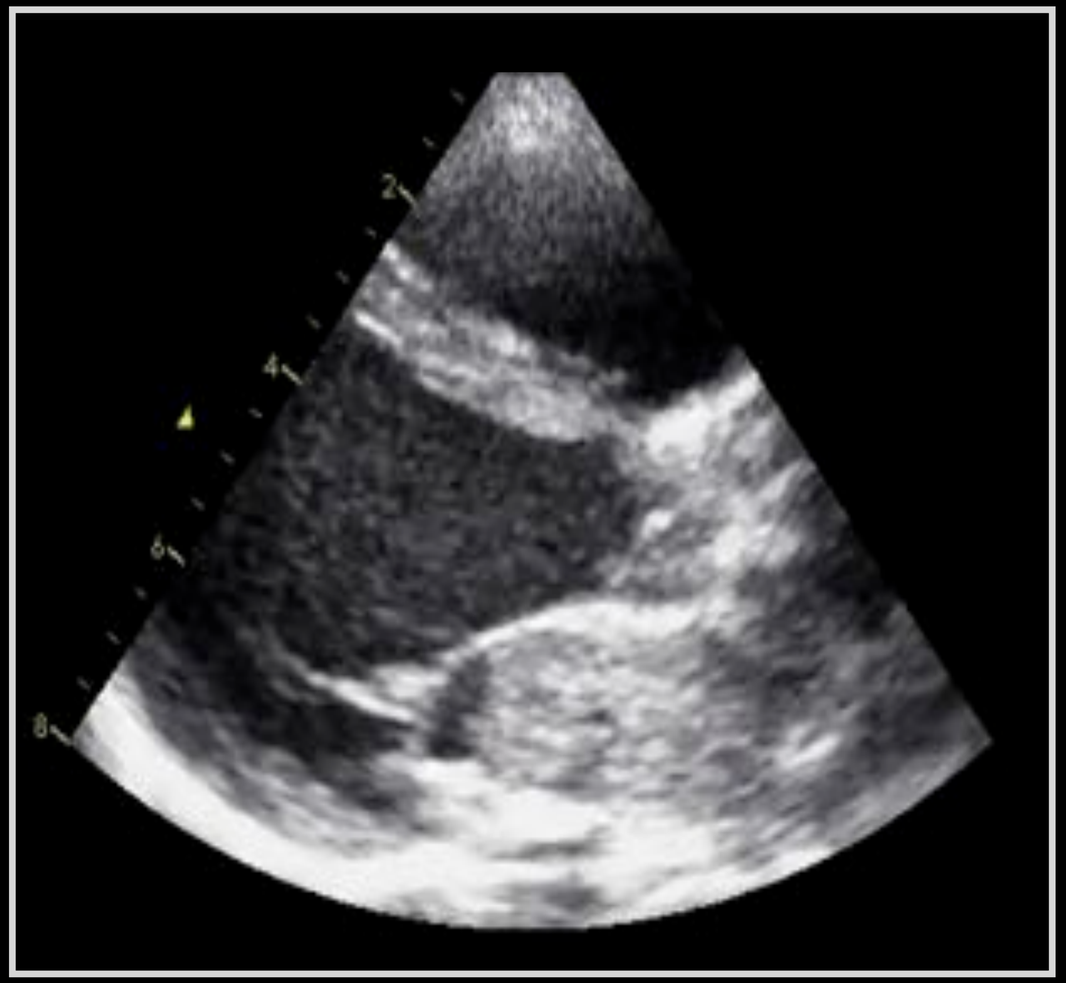
Carney Complex - Skin features

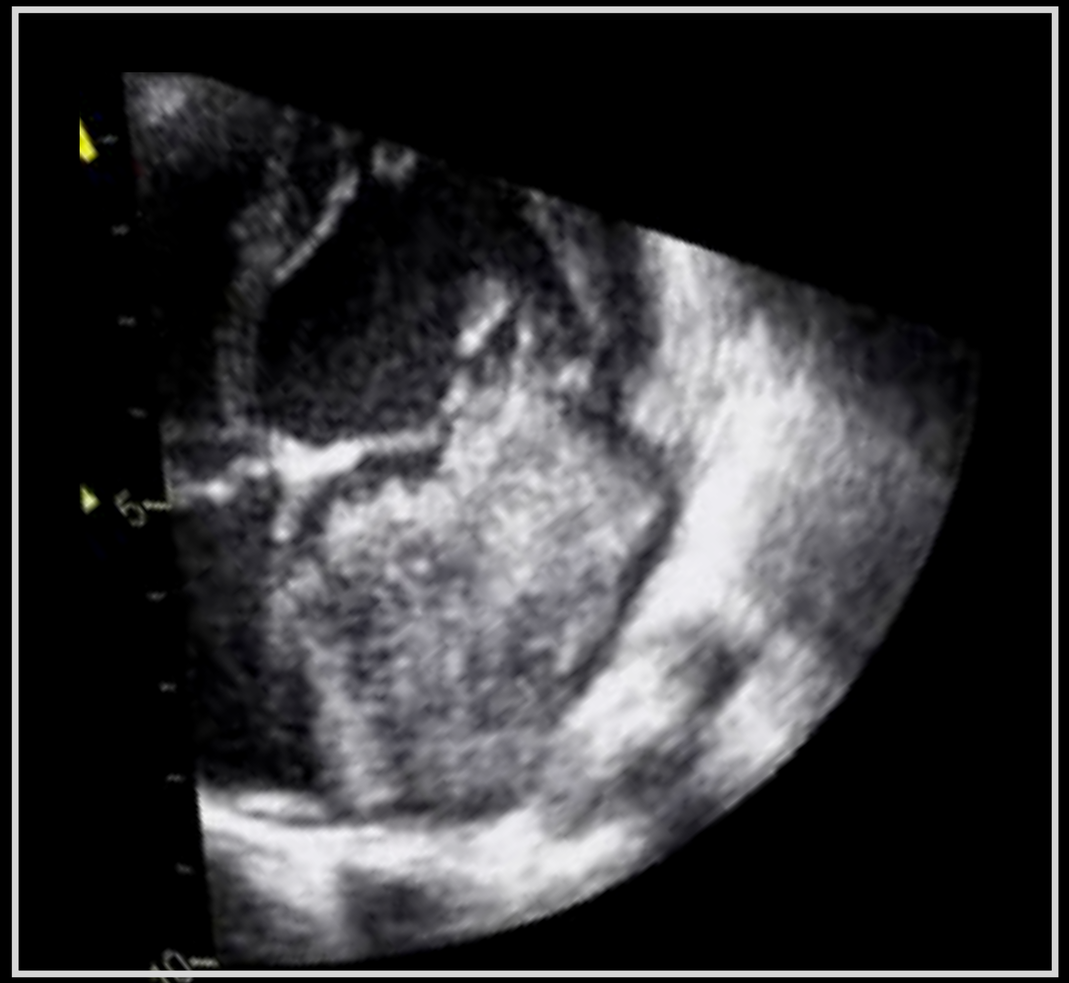
Carney complex and cardiac myxoma

- About **7% of cardiac myxomas** are associated with Carney Complex.
- The etiology of Carney Complex has been localized to **protein kinase A regulatory subunit gene PRKAR1A** on chromosome 17q23-24.
- **Germline inactivating mutations** in this gene have been found **in about 70% of individuals with Carney Complex**.

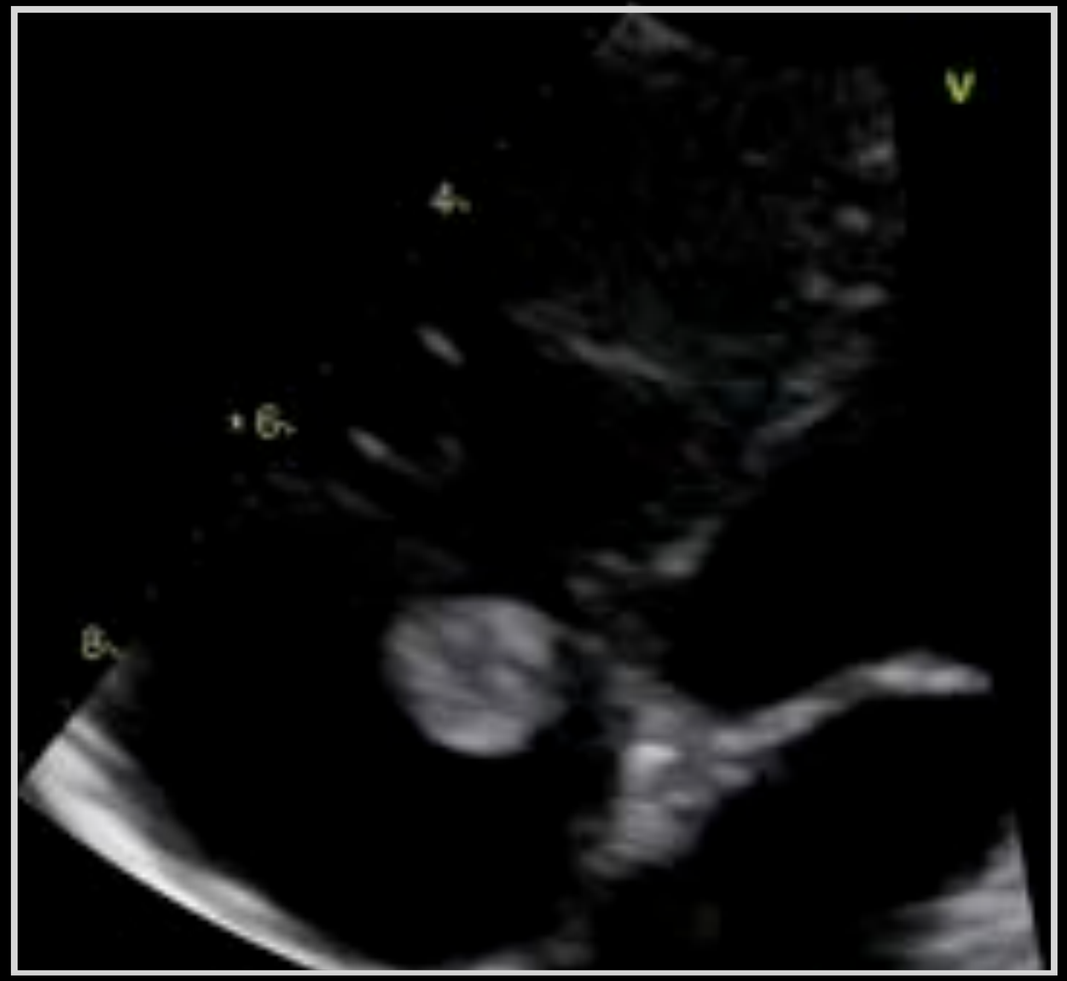


Carney Complex - Genetics

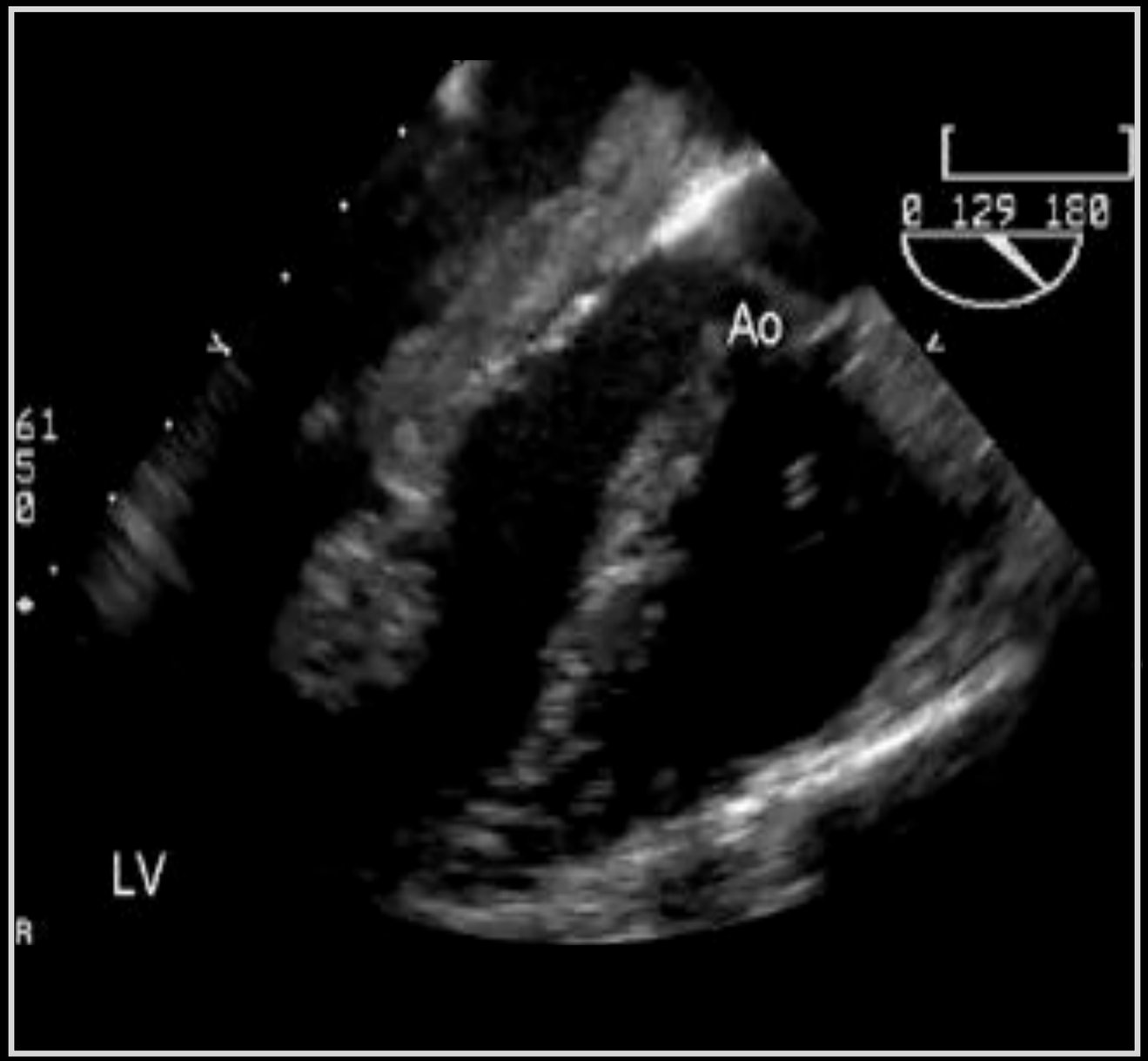




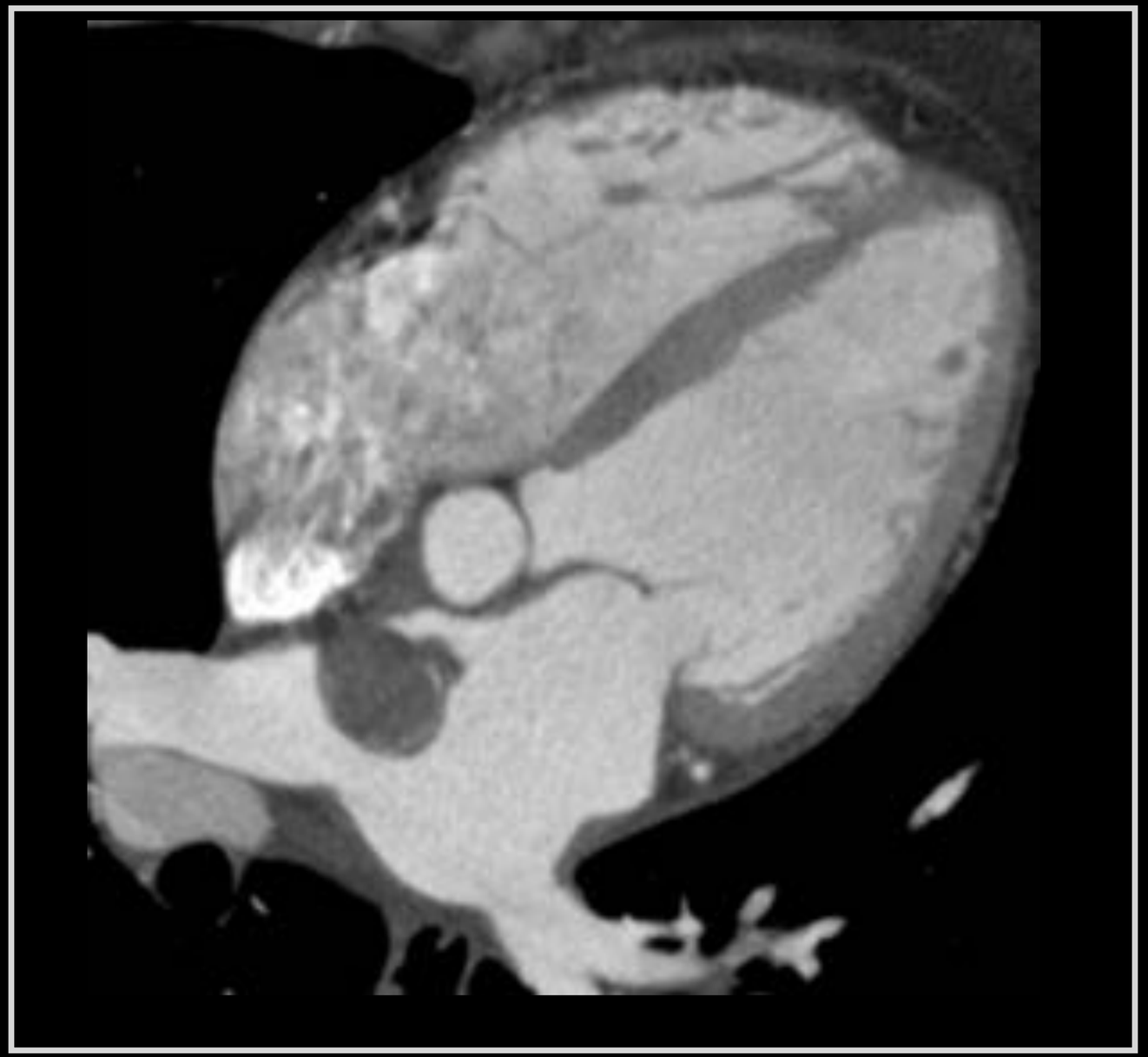
Myxoma



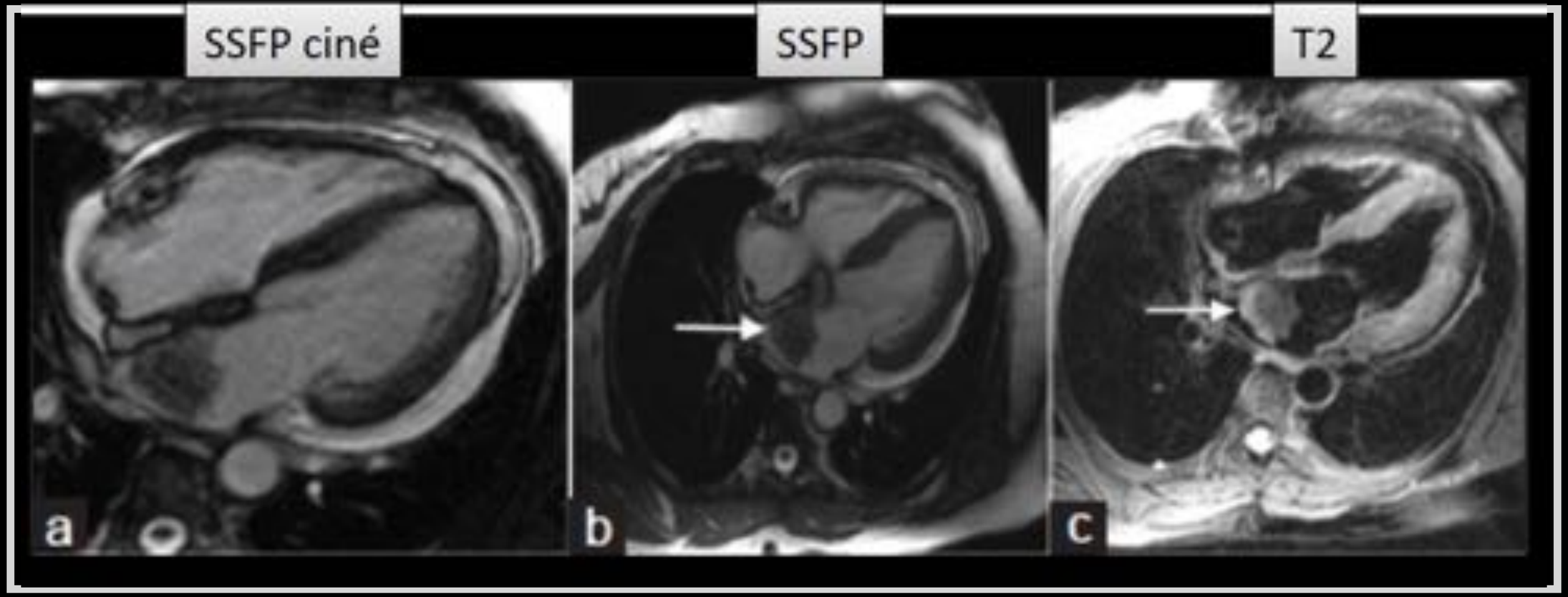
Myxoma - Right atrium



Myxoma - Left atrium

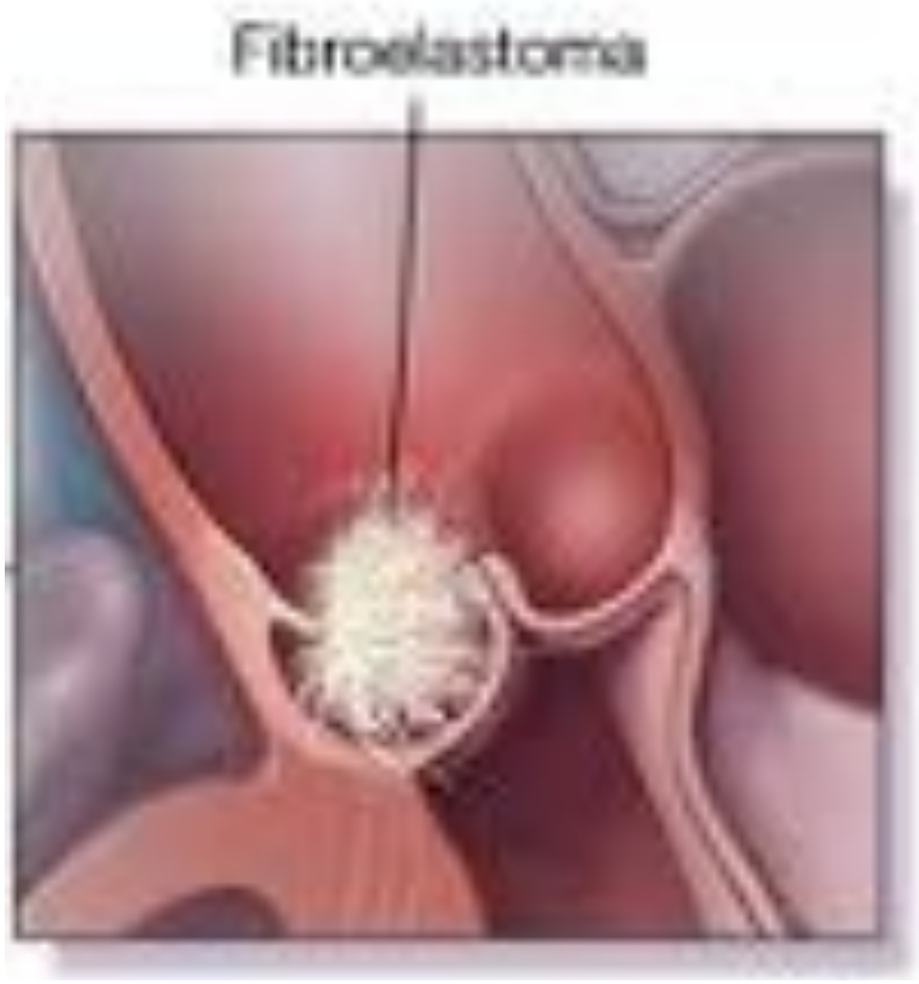


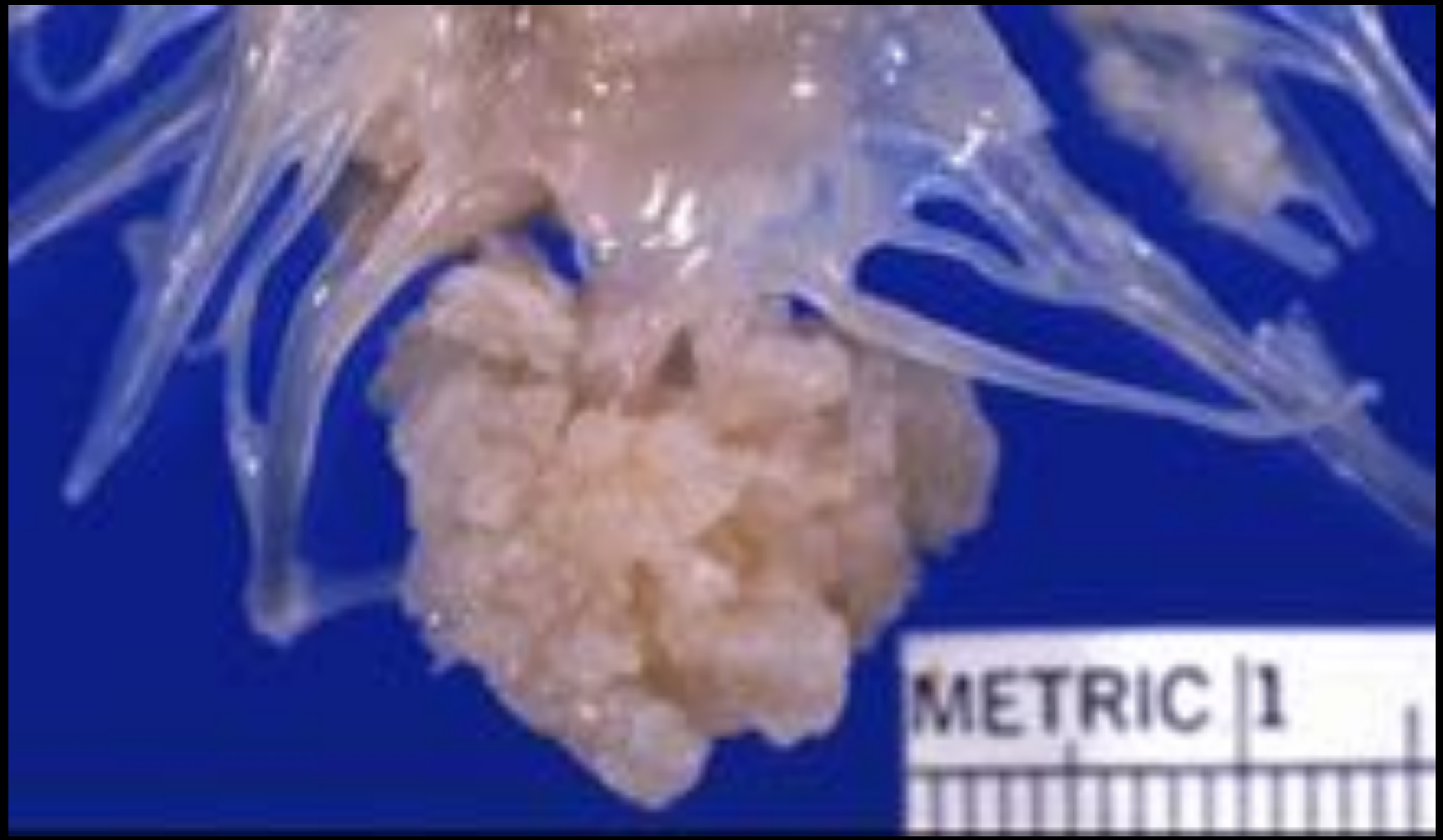
Myxoma - CT



Myxoma - MRI

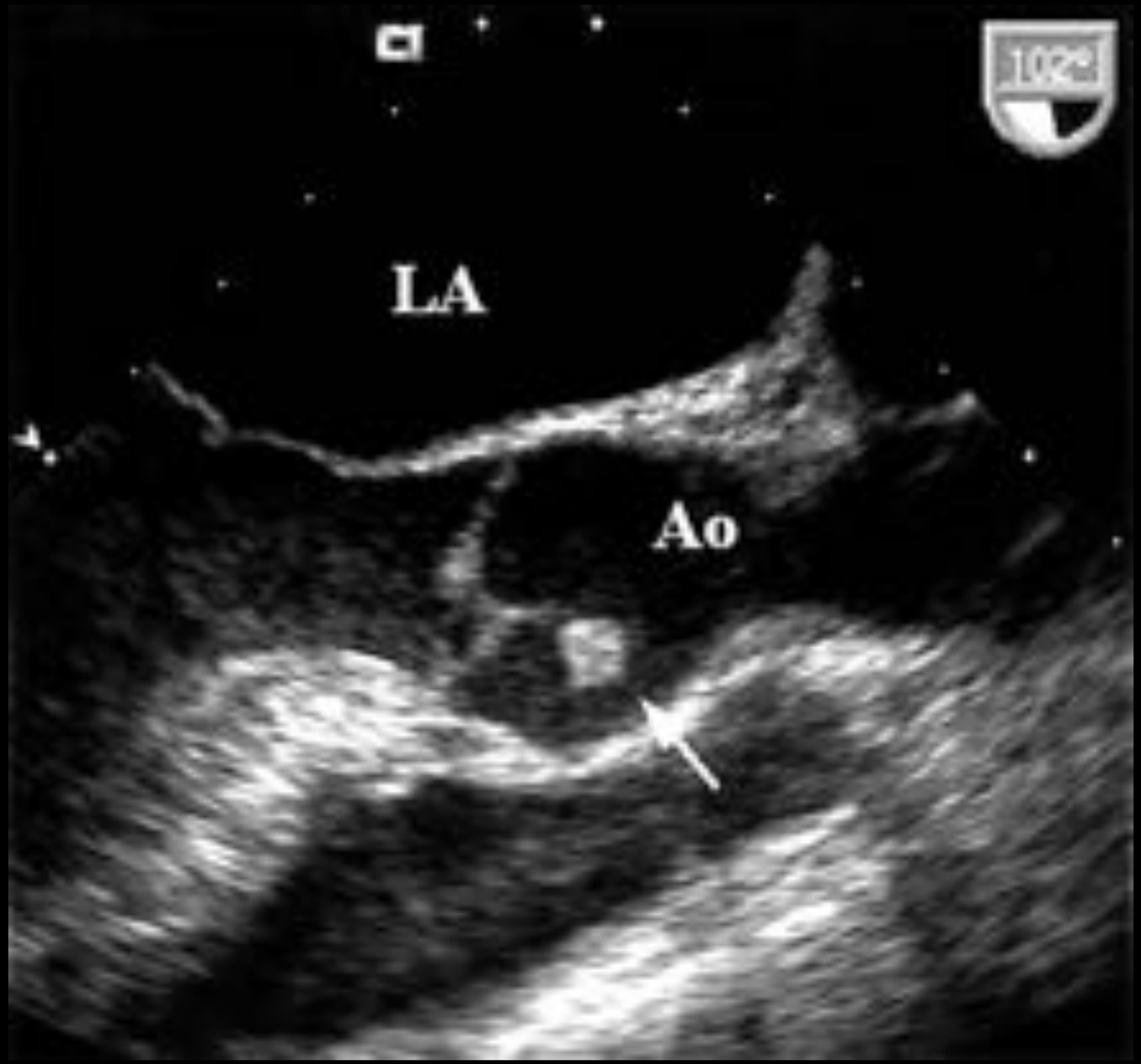
Papillary fibro-elastoma


Fibroelastoma

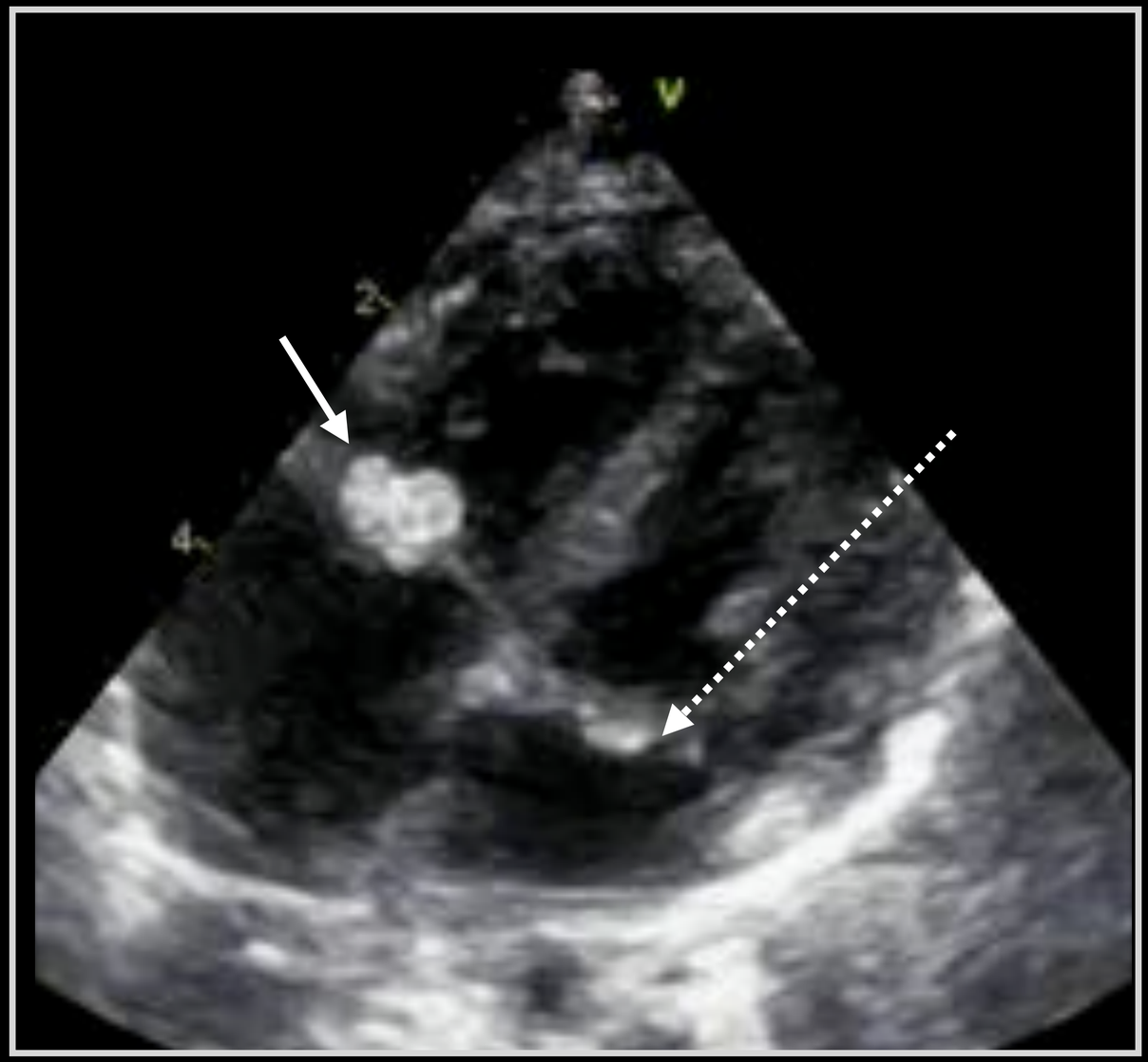


Fibroelastoma

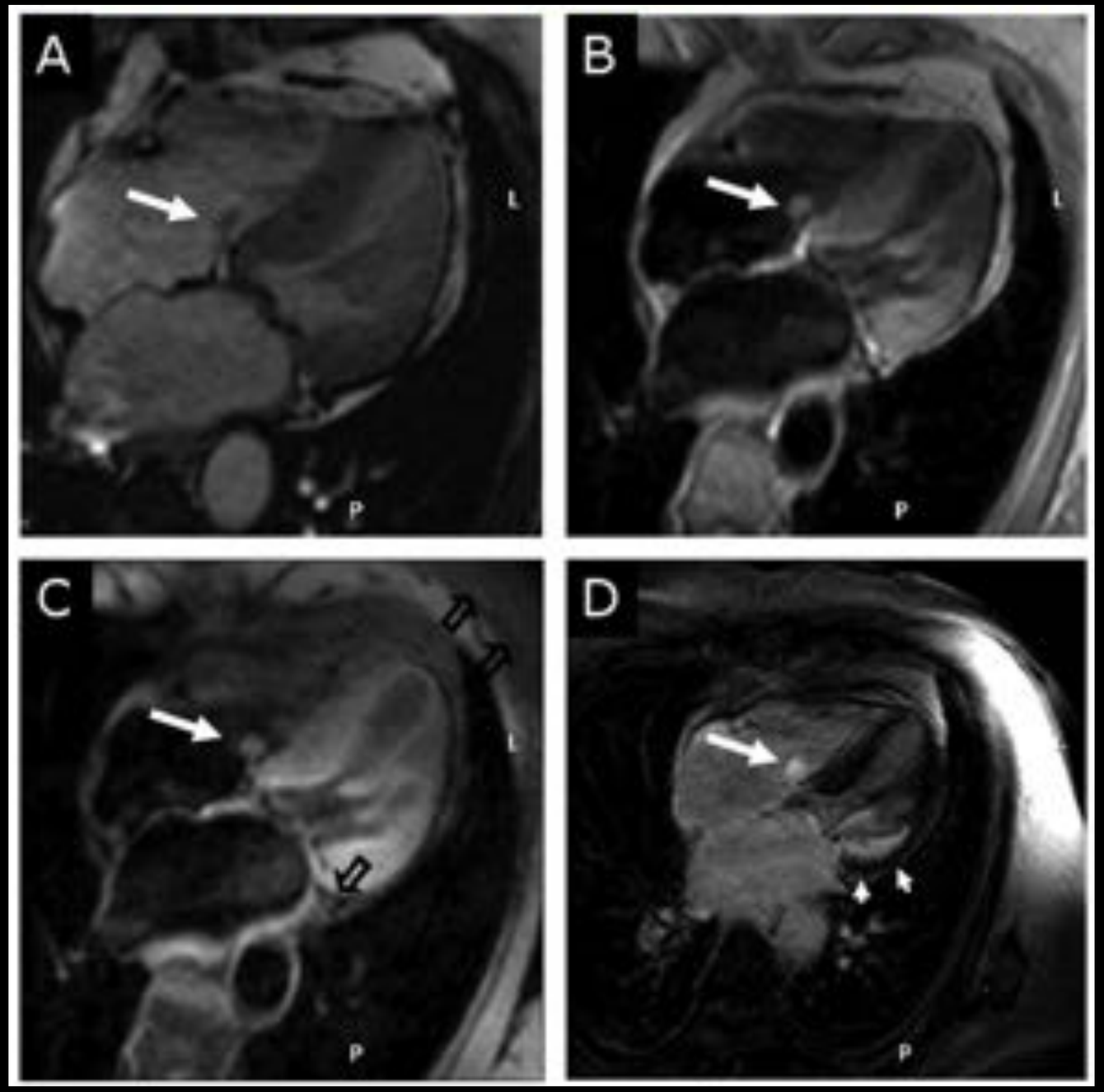
Fibroelastoma



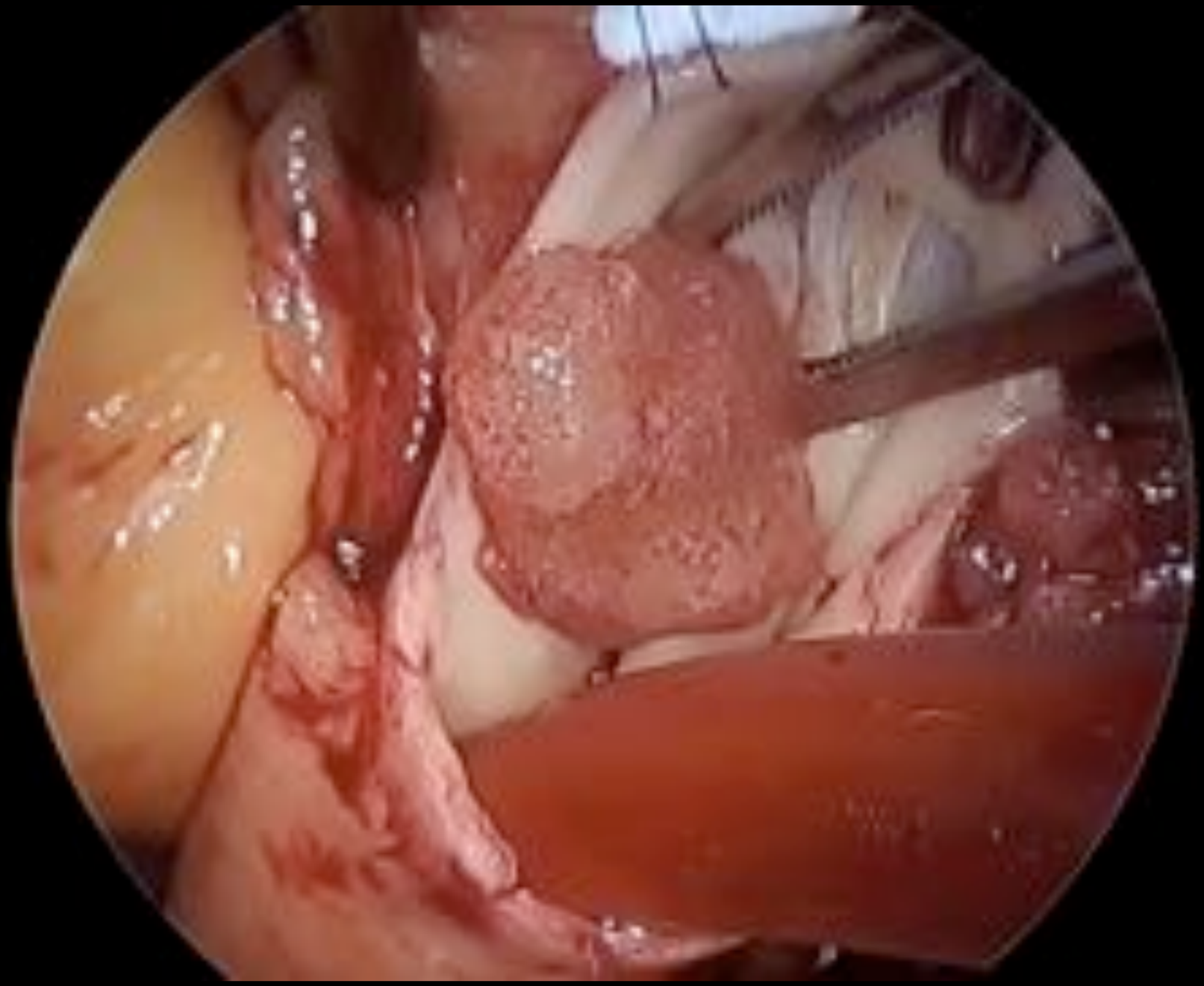
Fibroelastoma



Fibroelastoma

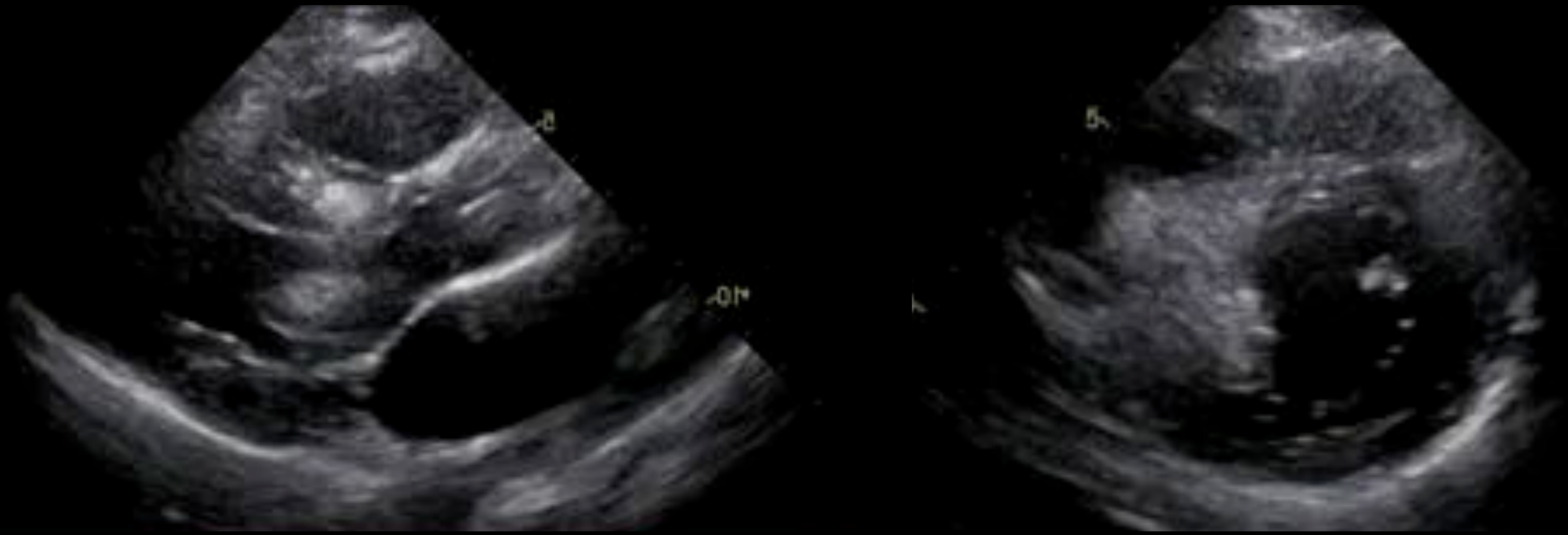


Fibroelastoma

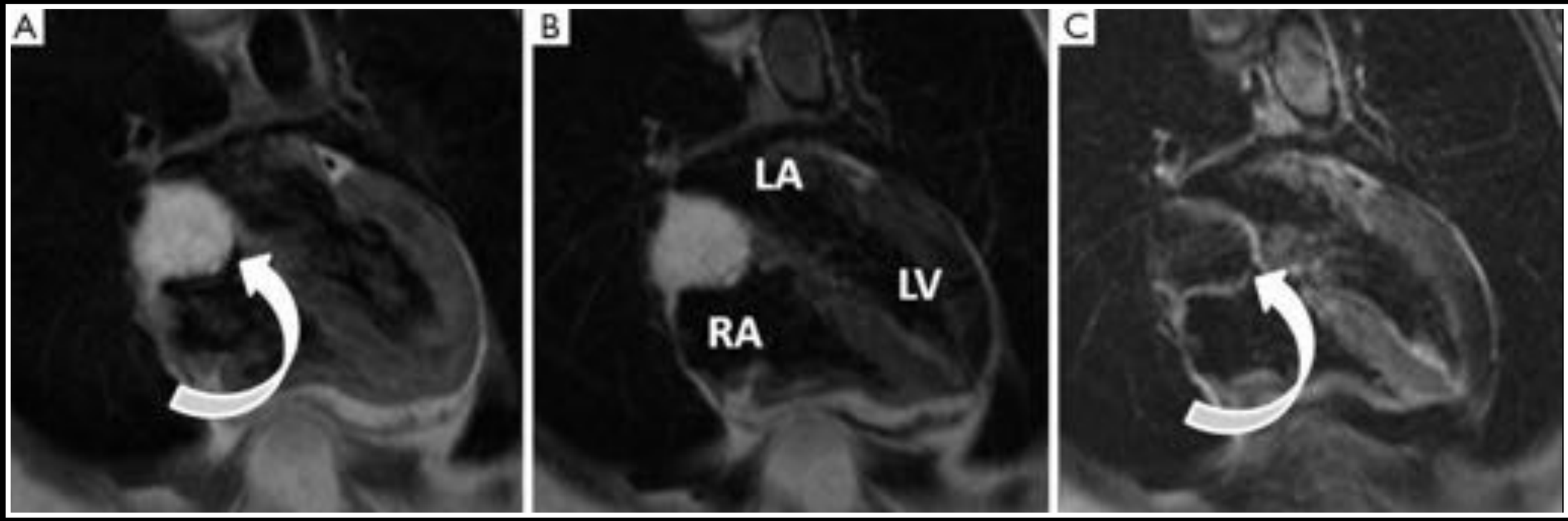


Fibroelastoma - Surgical removal - Pulmonary valve fibroelastoma

Lipoma



Lipoma



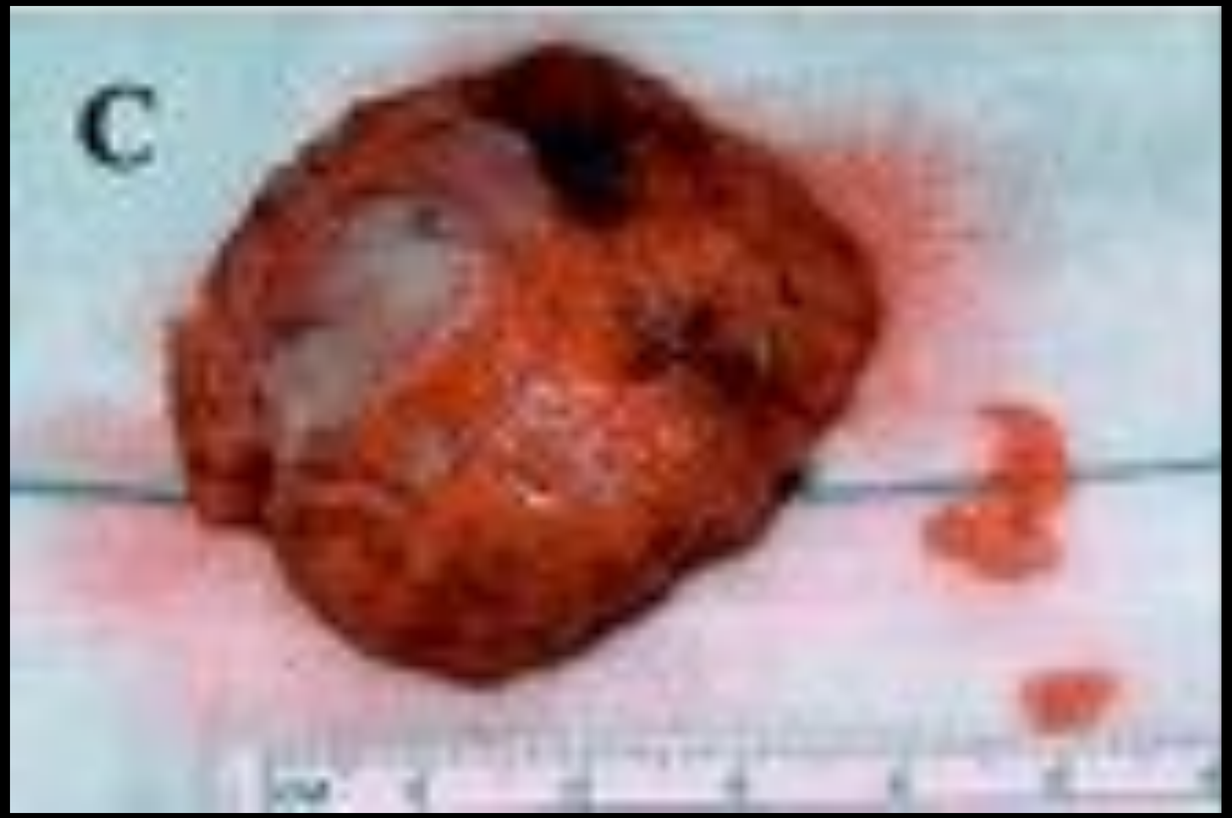
Atrial septal lipoma. (A) 4-Chamber T2-weighted black blood image showing a well-defined uniformly high signal lesion within the interatrial septum (curved arrow); (B) 4-Chamber T1-weighted black blood image again showing the lesion as having uniformly high signal in keeping with a fat composition; (C) 4-Chamber T2-weighted black blood image with fat suppression showing complete and uniform signal suppression (curved arrow). RA, right atrium; LA, left atrium; LV, left ventricle.

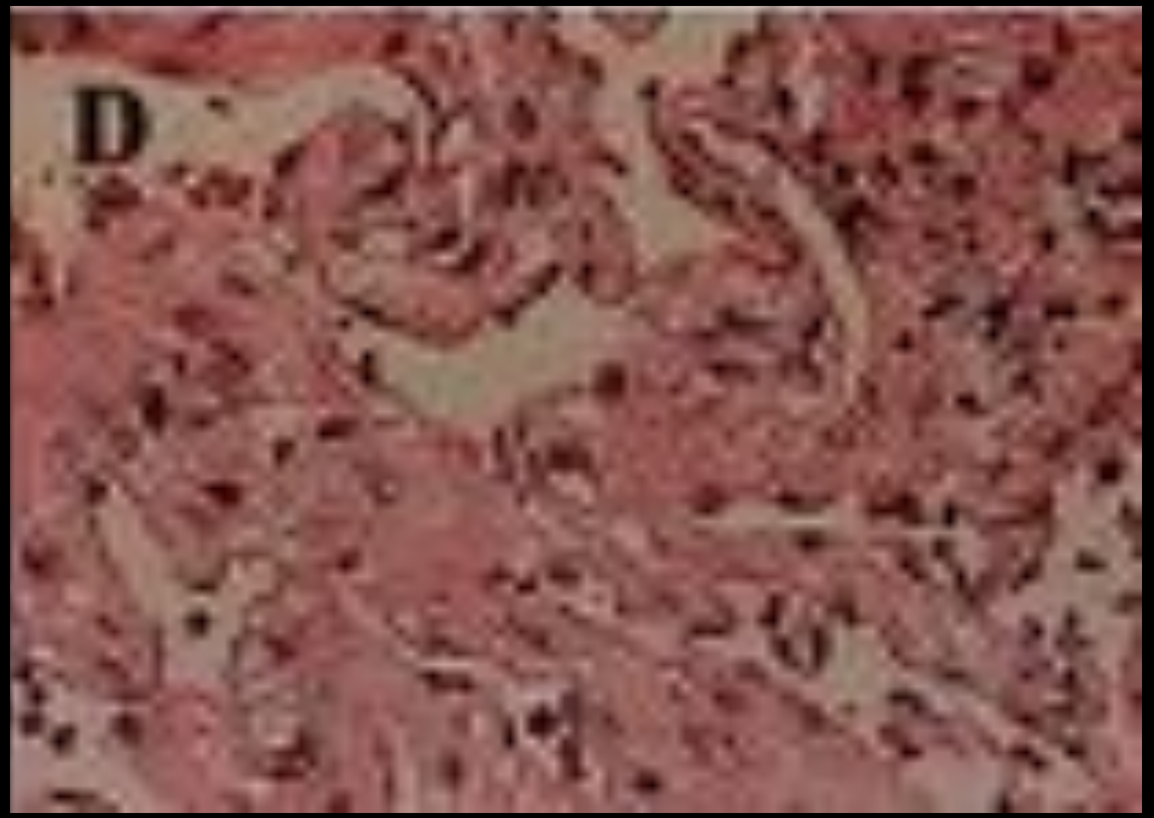
Lipoma

Hemangioma

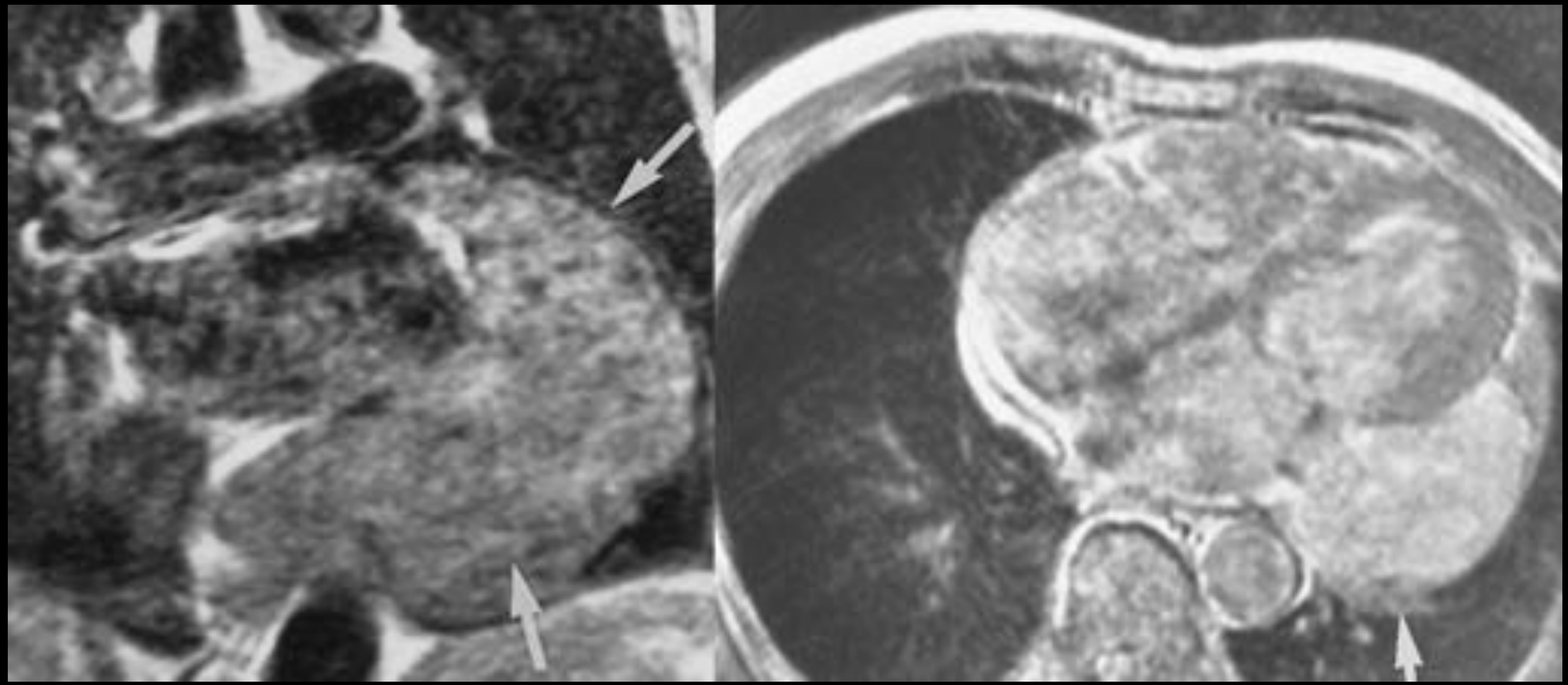
Cardiac hemangiomas

- Cardiac Hemangiomas are rare tumors that are observed in both children and adults.
- Hemangioma is generally a mix of capillary, cavernous, and arteriovenous (AV) hemangioma type
- There are no known risk factors for sporadic Cardiac Hemangiomas; however, in some individuals, the tumor is seen to be associated with Kasabach-Merritt syndrome.
- Many small hemangiomas are often undiagnosed and do not cause any signs and symptoms. In some cases, Cardiac Hemangiomas may present with chest pain, breathing difficulties, arrhythmias, and pericardial effusion
- Complications could include heart failure, severe obstruction of heart function, and even sudden death due to severe arrhythmias
- Surgical removal of the hemangioma is typically the best treatment option for Cardiac Hemangioma. The prognosis of the tumor with suitable treatment (surgical excision and removal) is reported to be good

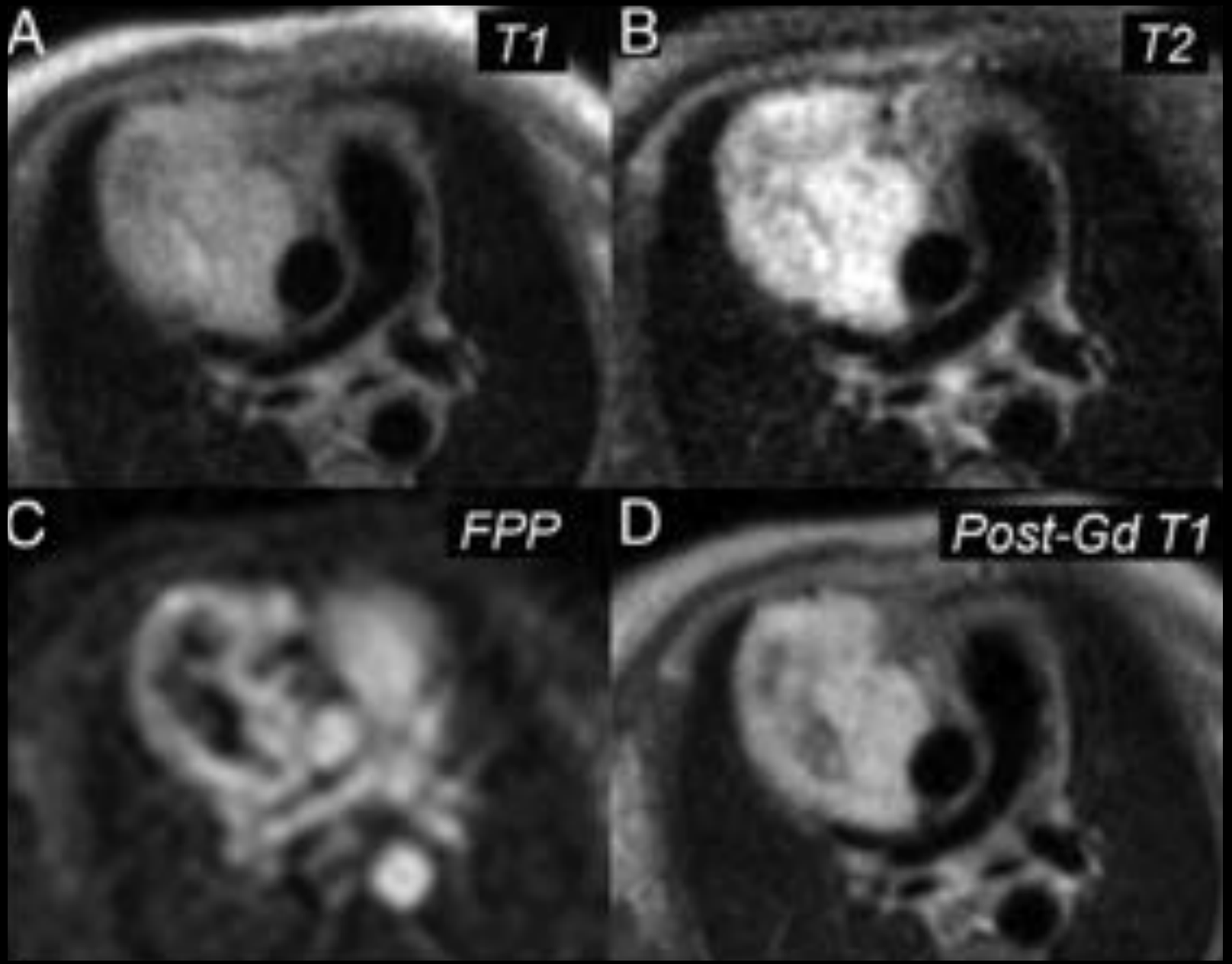




Hemangioma



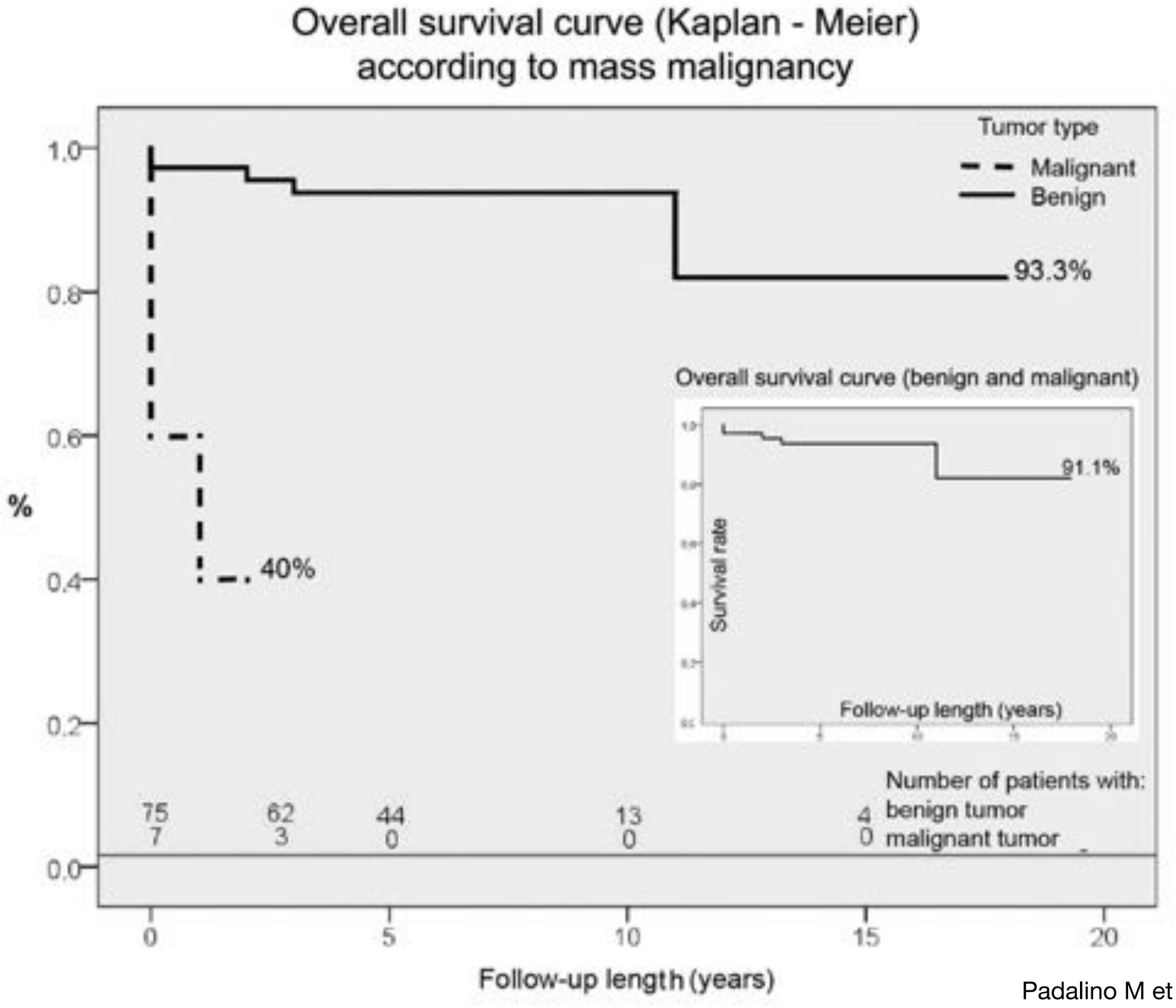
Hemangioma



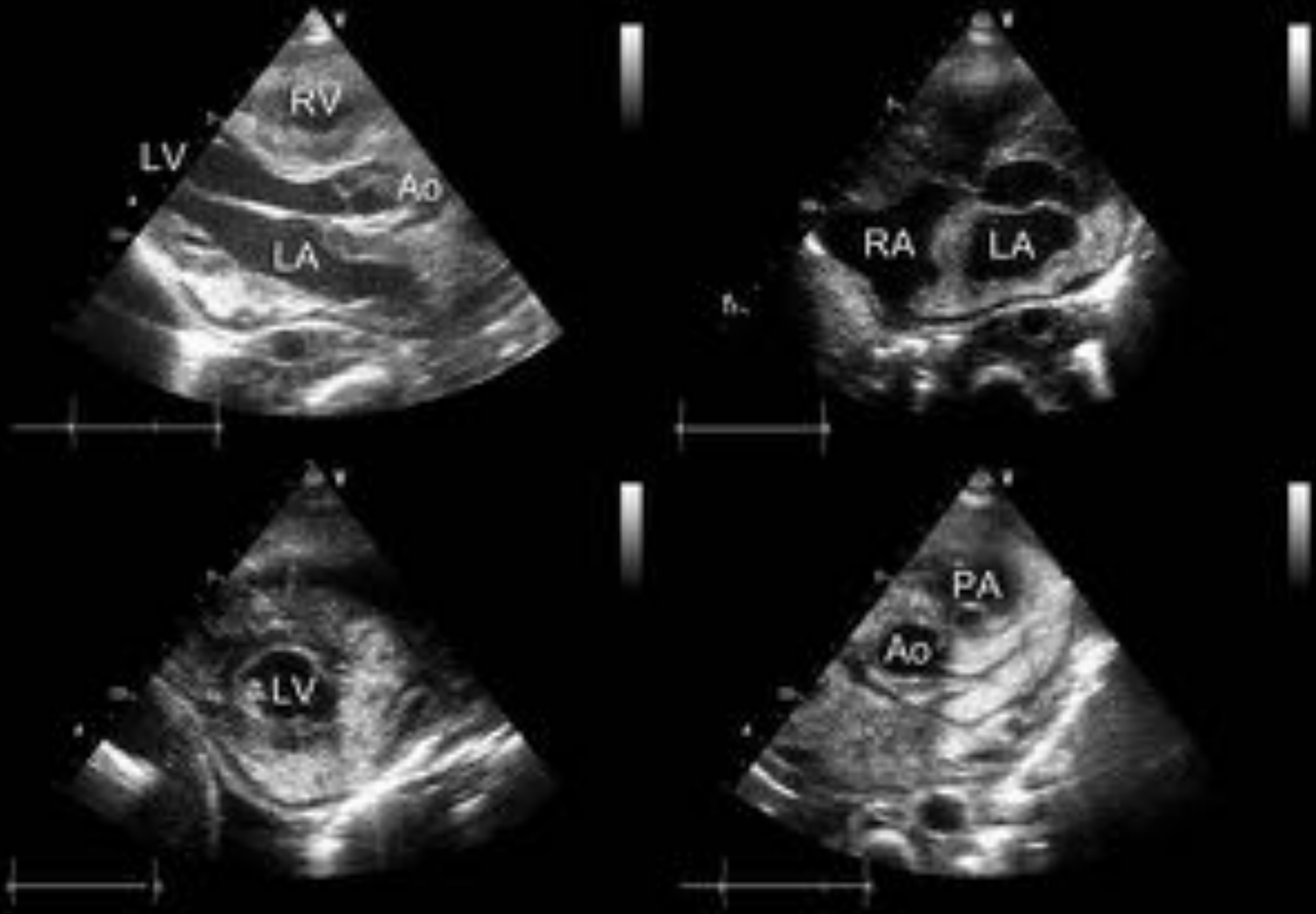
Hemangioma

Beroukhim RS et al. JACC 2011

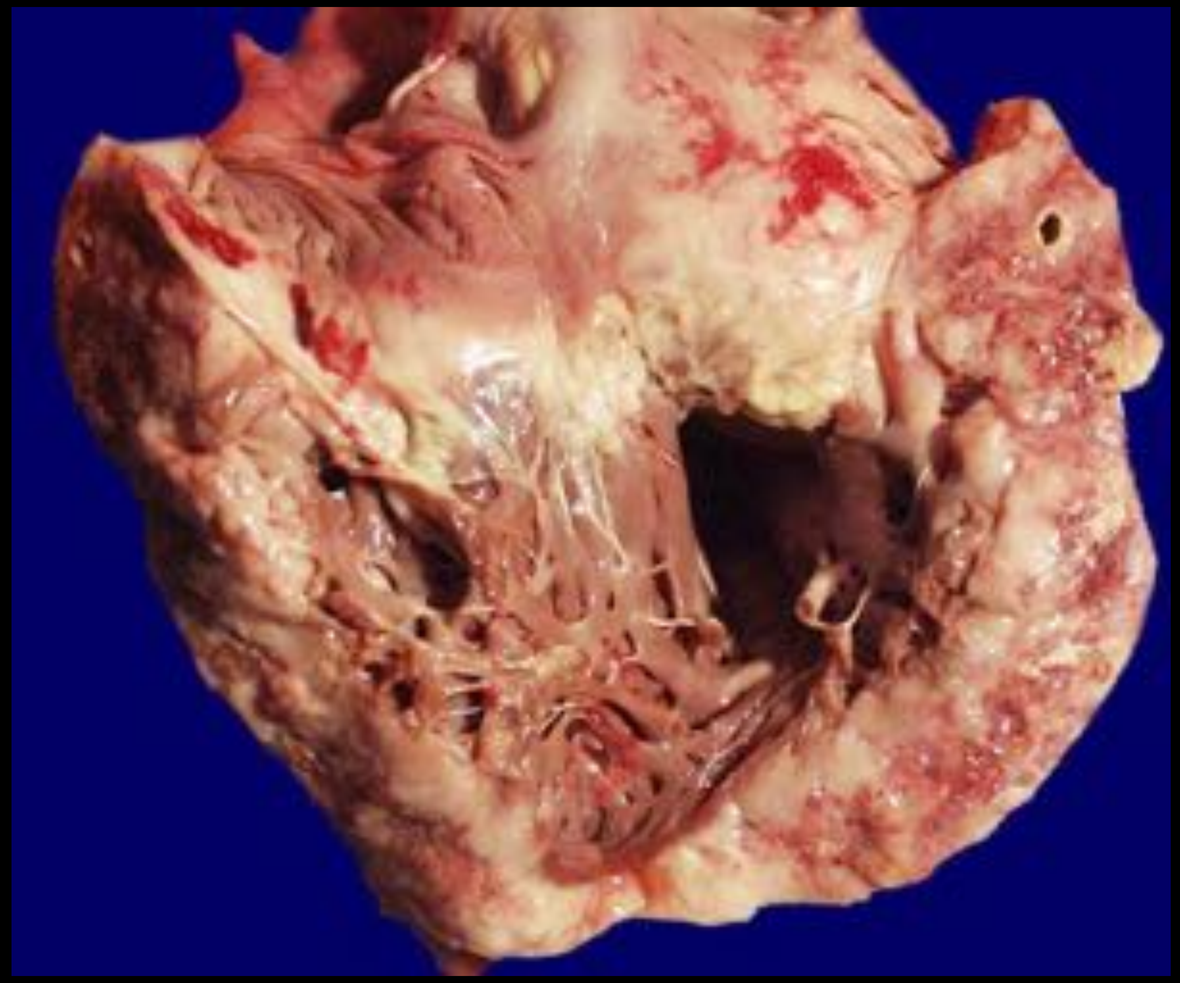
Malignant tumors

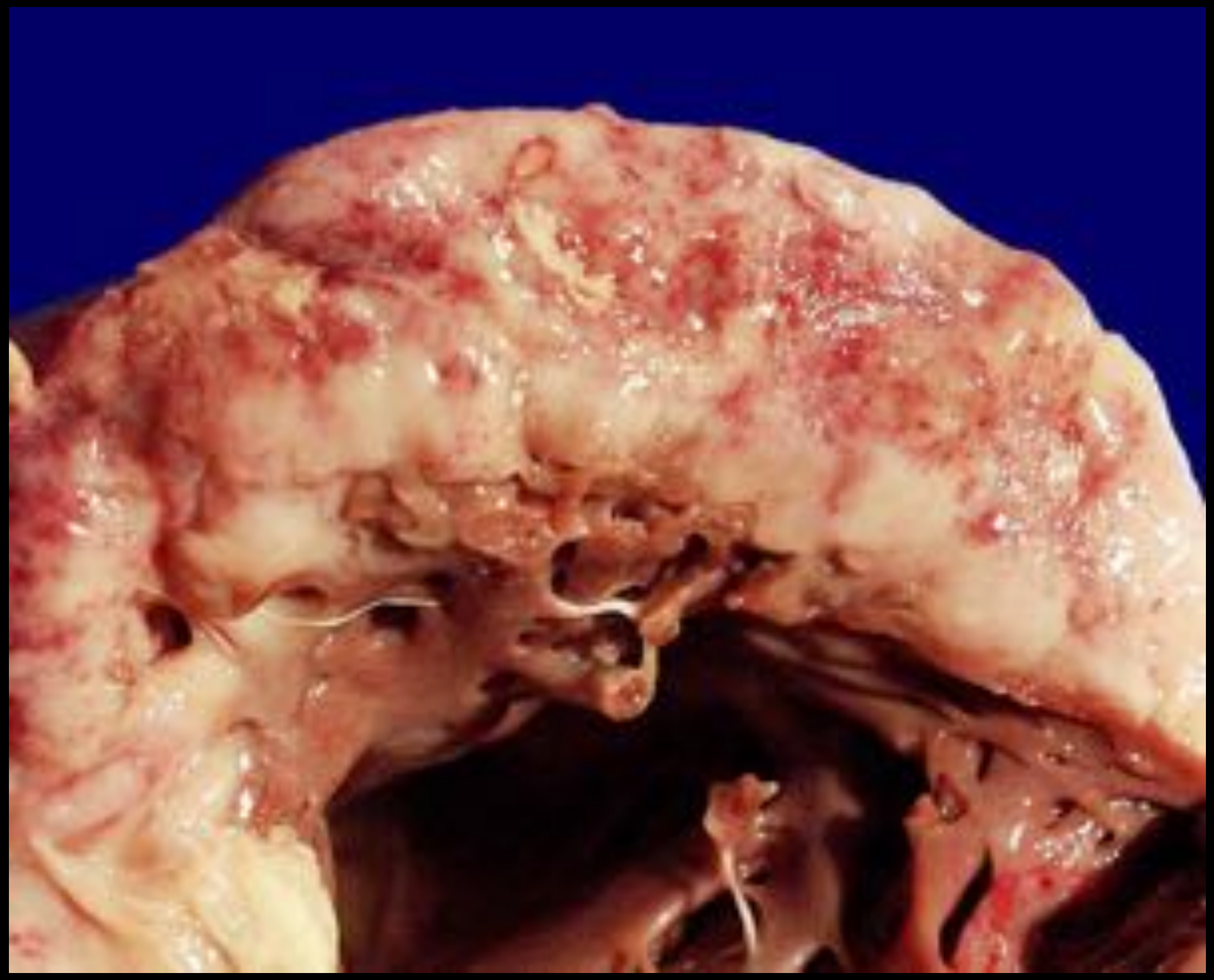


Padalino M et al. Circulation 2012; 126:22-30

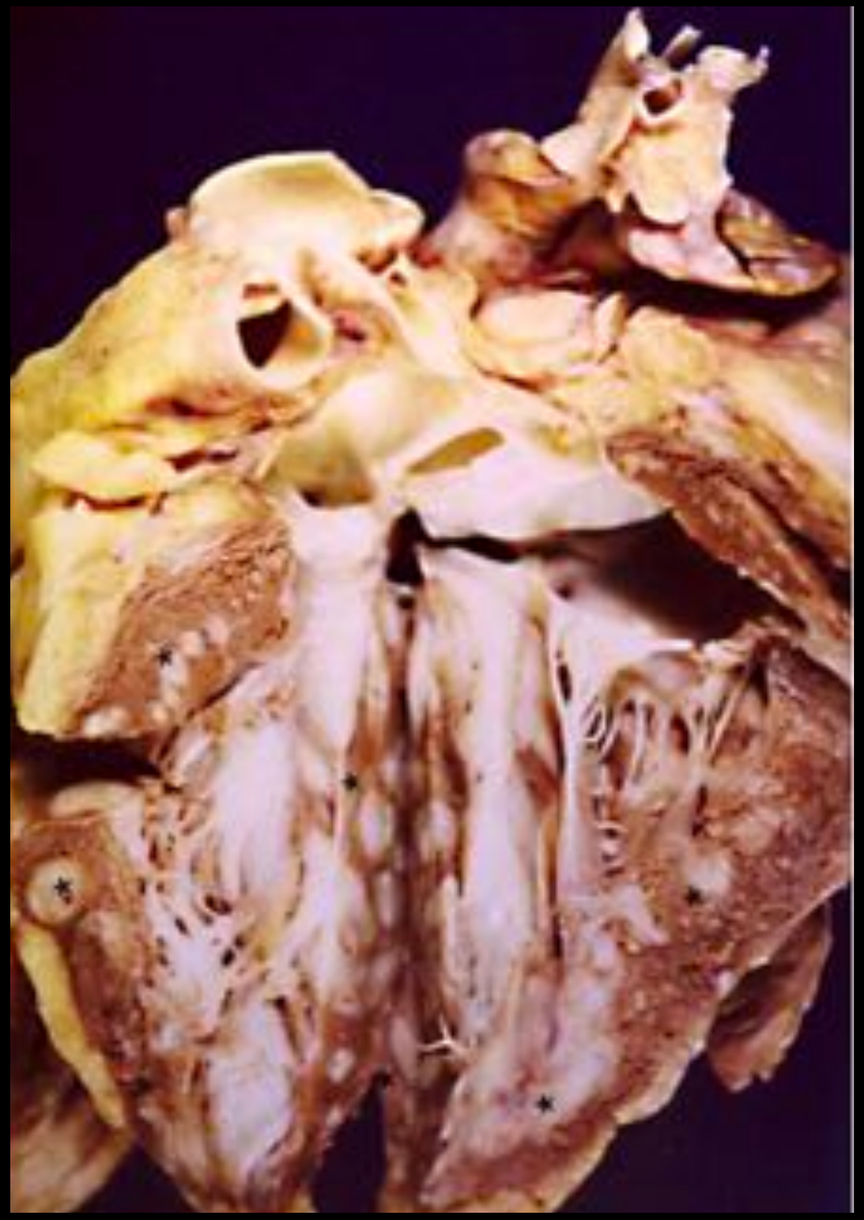


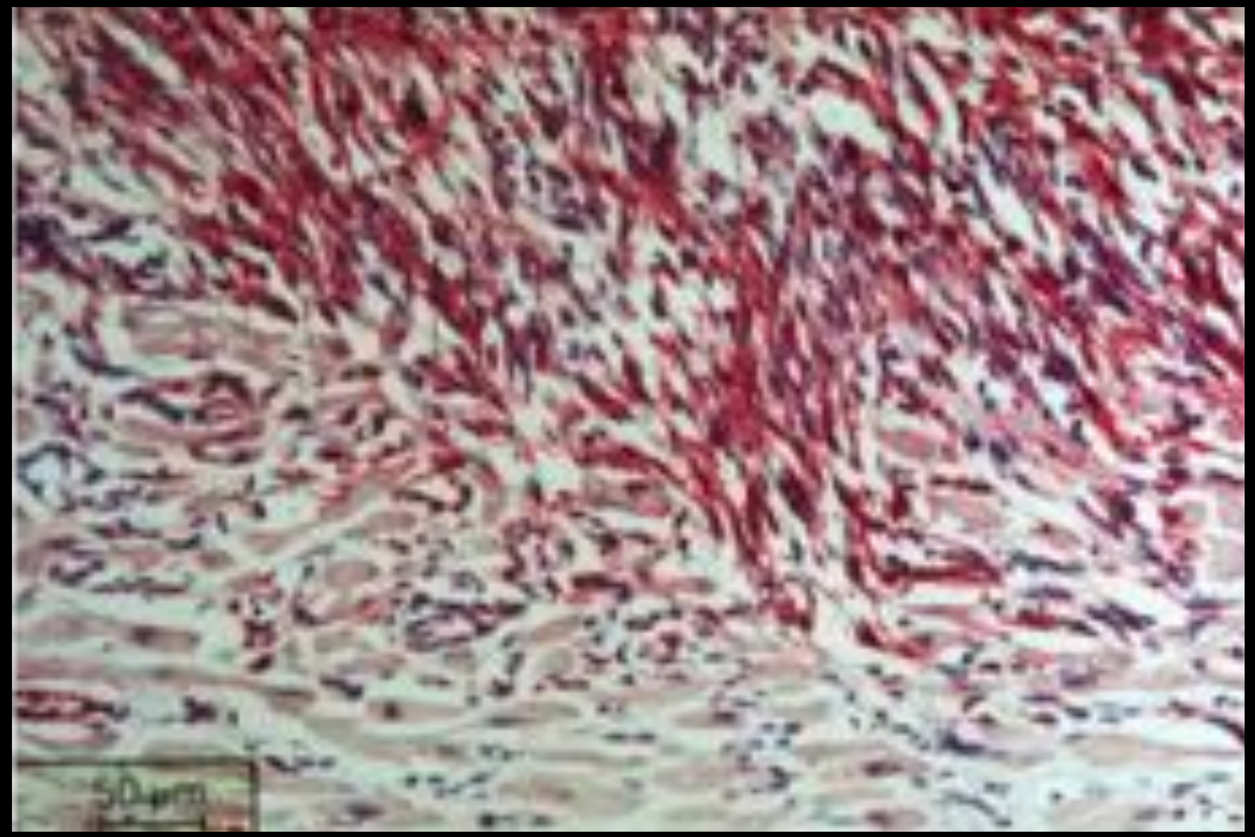
Lymphoma



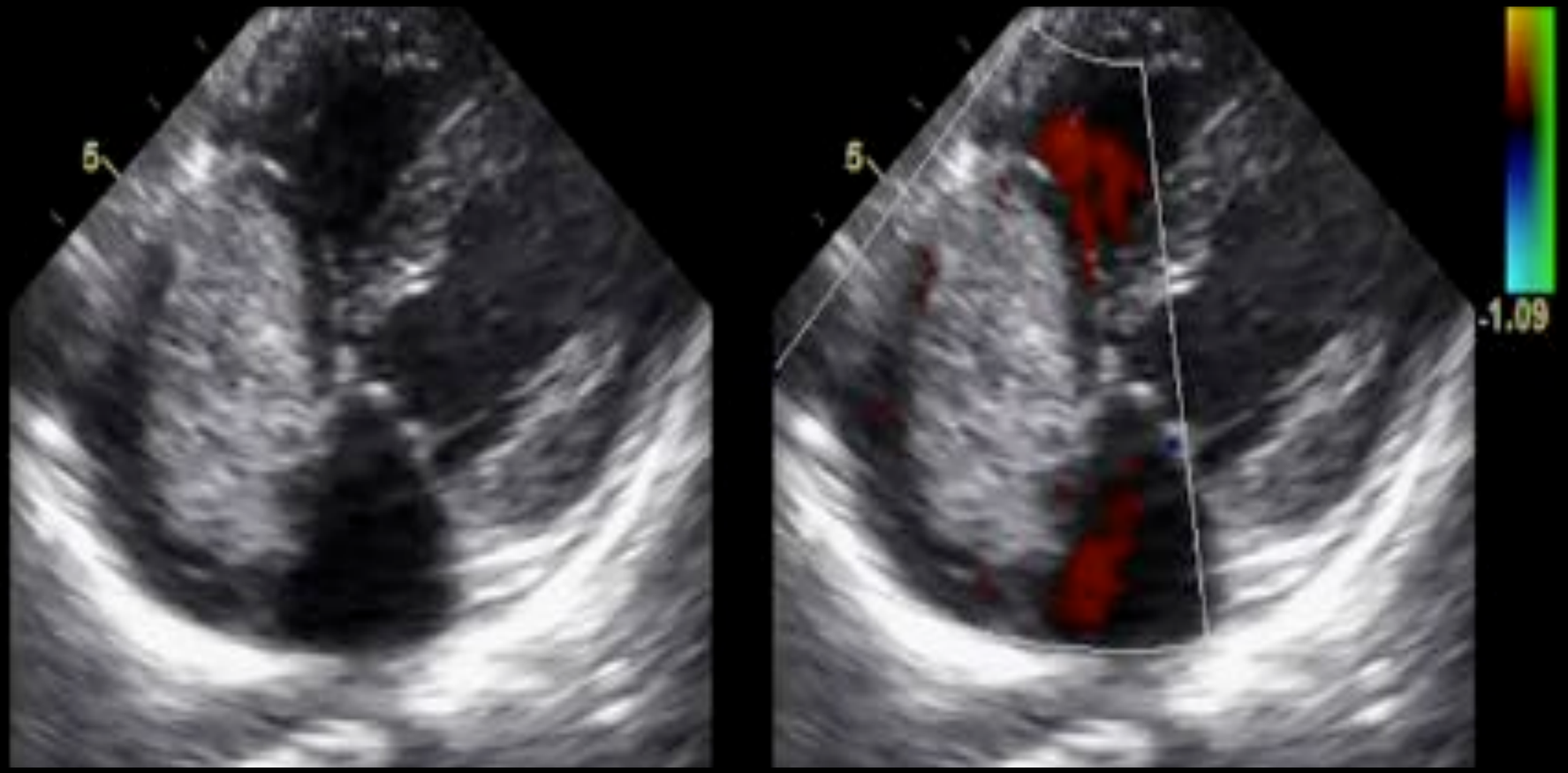


Lymphoma





Sarcoma

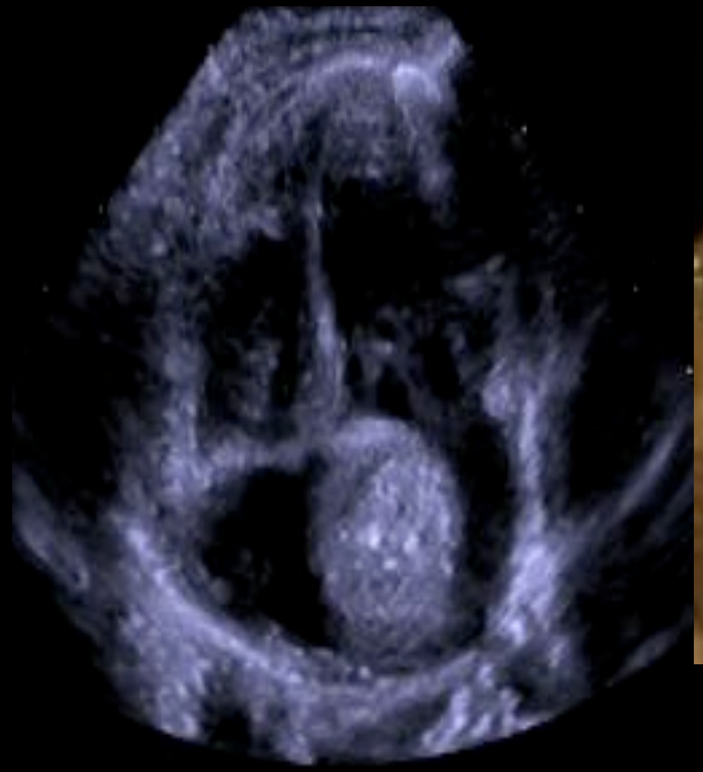


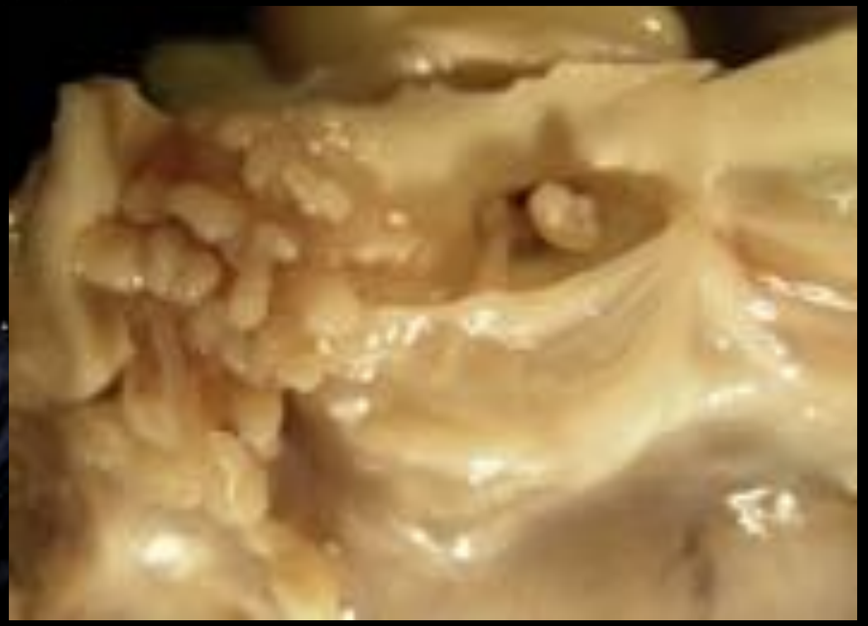
Nephroblastoma - Extension into IVC and right atrium

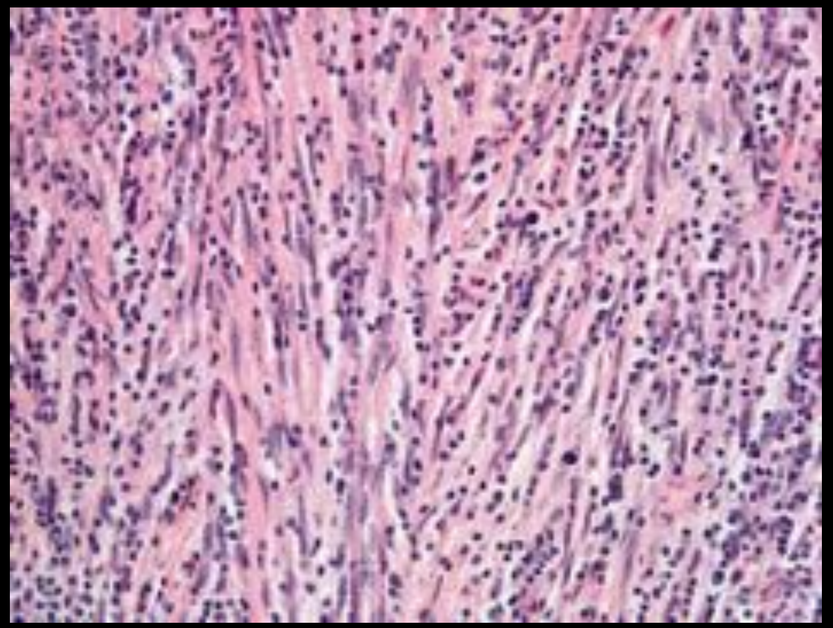
Other rare anomalies

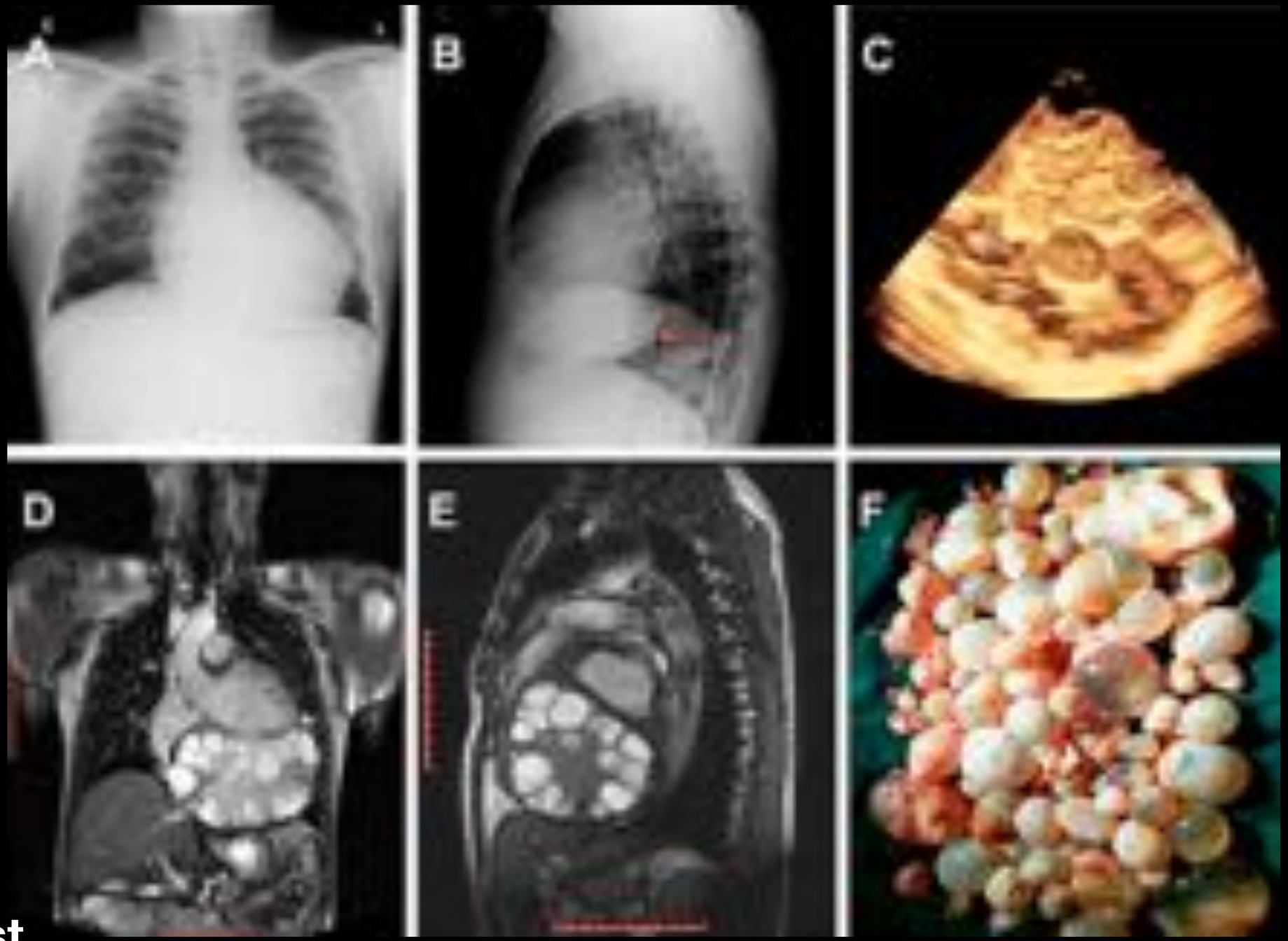
Inflammatory myofibroblastic tumor

- IMFTs are proliferations of uncertain histogenesis, which vary in appearance from inflammatory, reactive-appearing proliferations to low-grade sarcomas.
- In the heart, they invariably arise from the endocardium, including valve leaflets, are variably cellular, and usually have abundant myxoid matrix and surface fibrin.









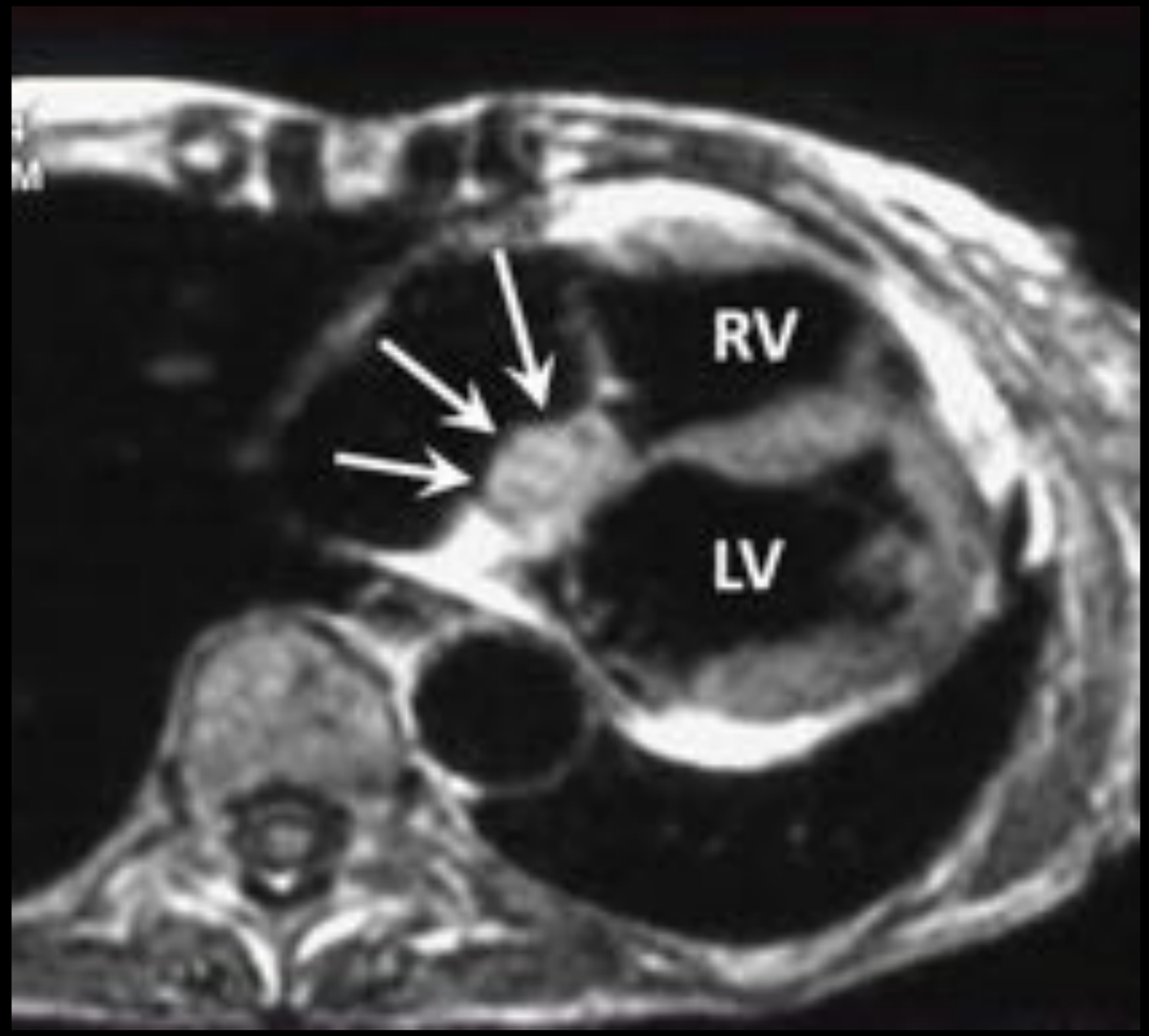
Hydatid cyst

Moorthy N et al. JACC 2013;62

Cystic tumor of the atrioventricular node
&
Histiocytoid cardiomyopathy
Purkinje tumors

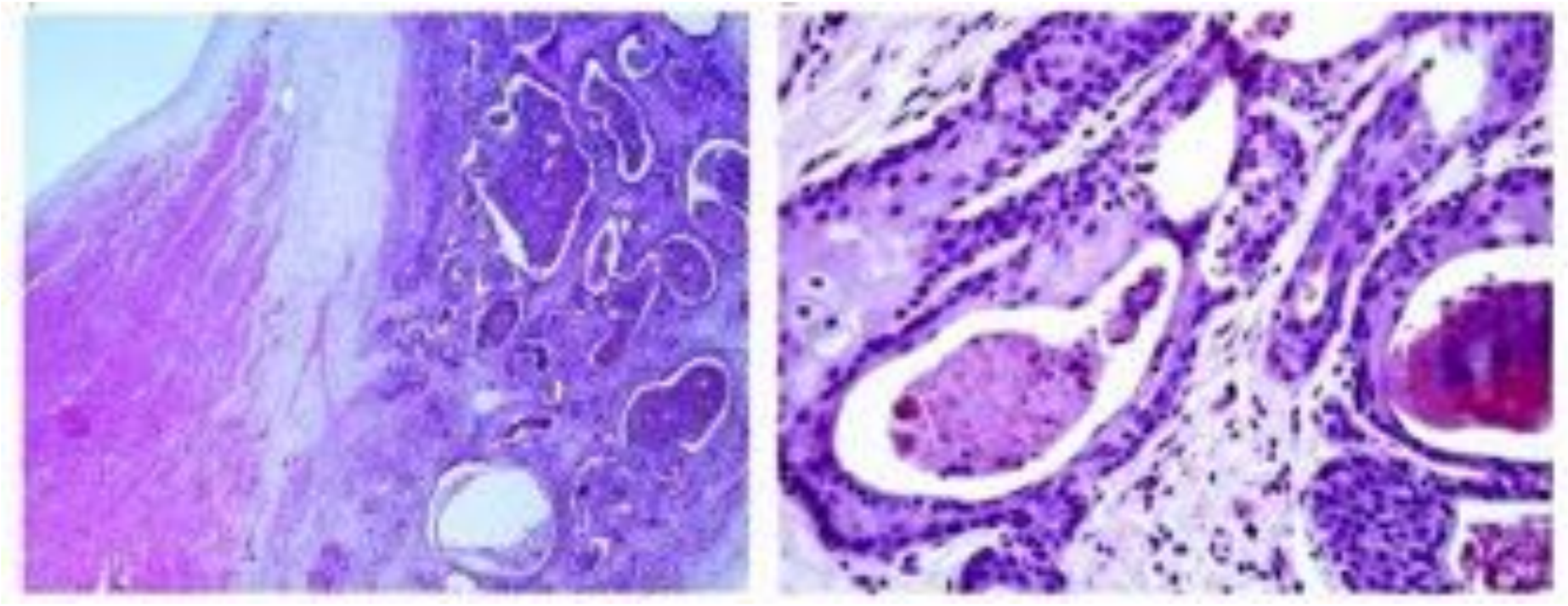
Cystic tumor of the atrio-ventricular node

- Rare, benign
- Revealed as AV block or sudden death
- Mainly in female



Hoey ETD et al. QIMS 2014

Cystic tumor of the atrio-ventricular node



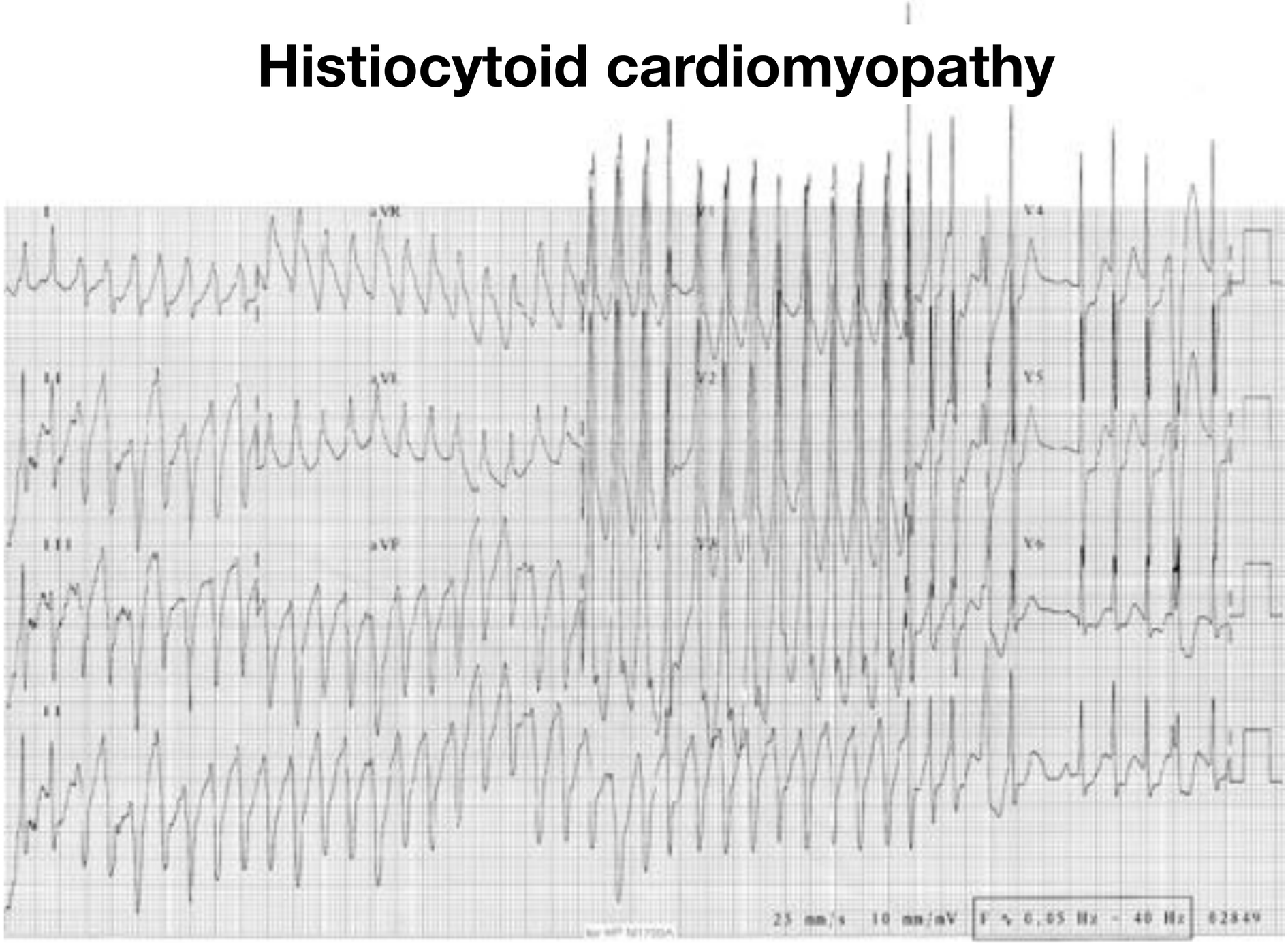
Sharma G et al. Cardiovascular Pathology 19 (2010) e75–e78

Histiocytoid cardiomyopathy

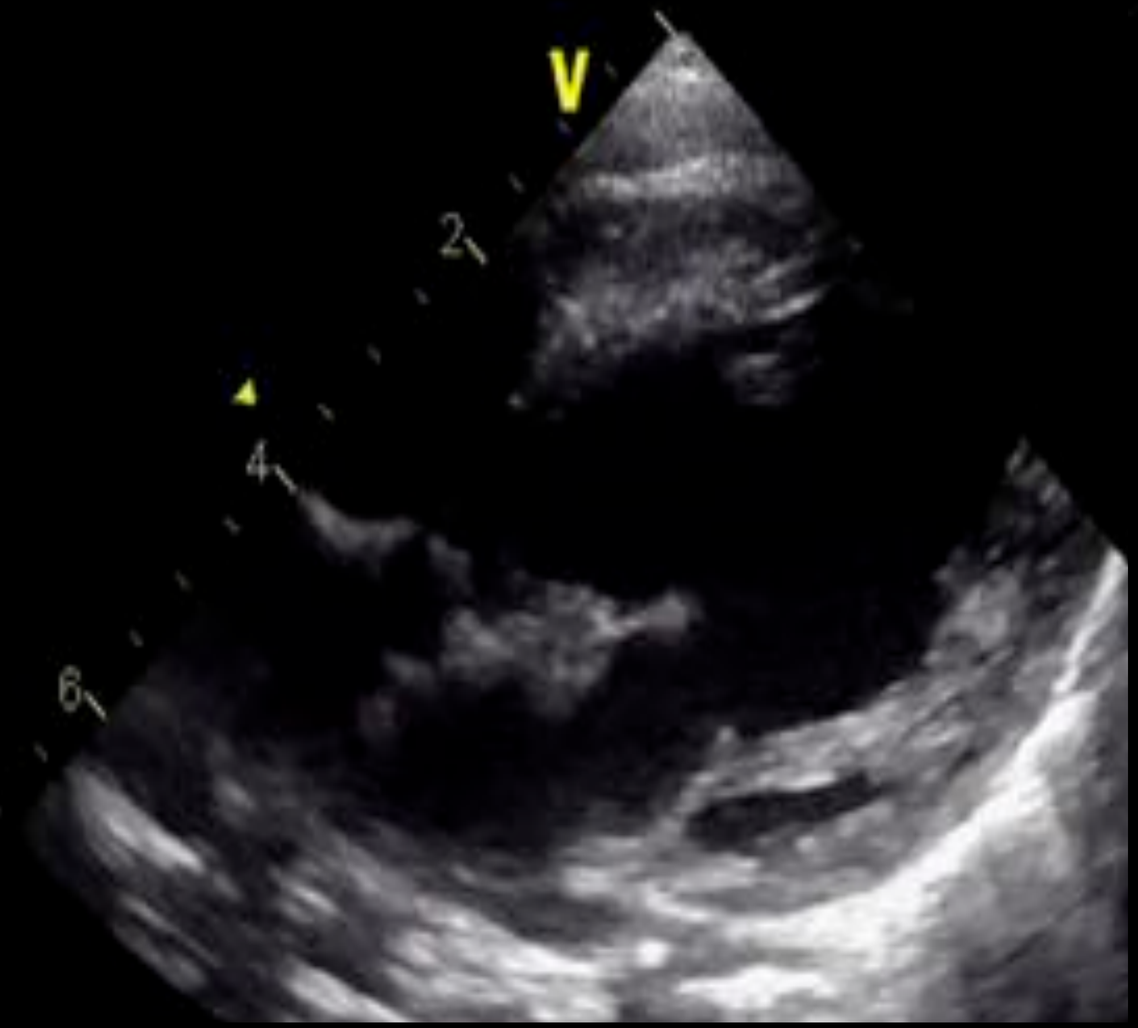
- Rare, arrhythmogenic disorder caused by multifocal hamartomatous proliferation of cardiac cells with oncocytic features.
- Synonyms include Purkinje cell hamartoma and cardiac hamartoma.
- The female:male ratio is 3:1.
- Approximately 5% of reported cases have occurred in families.
- Arrhythmias associated with histiocytoid cardiomyopathy include paroxysmal atrial tachycardia, atrial fibrillation, ventricular fibrillation, ventricular tachycardia, premature atrial contractions, premature ventricular contractions, Wolff– Parkinson– White syndrome, and right or left bundle branch block.

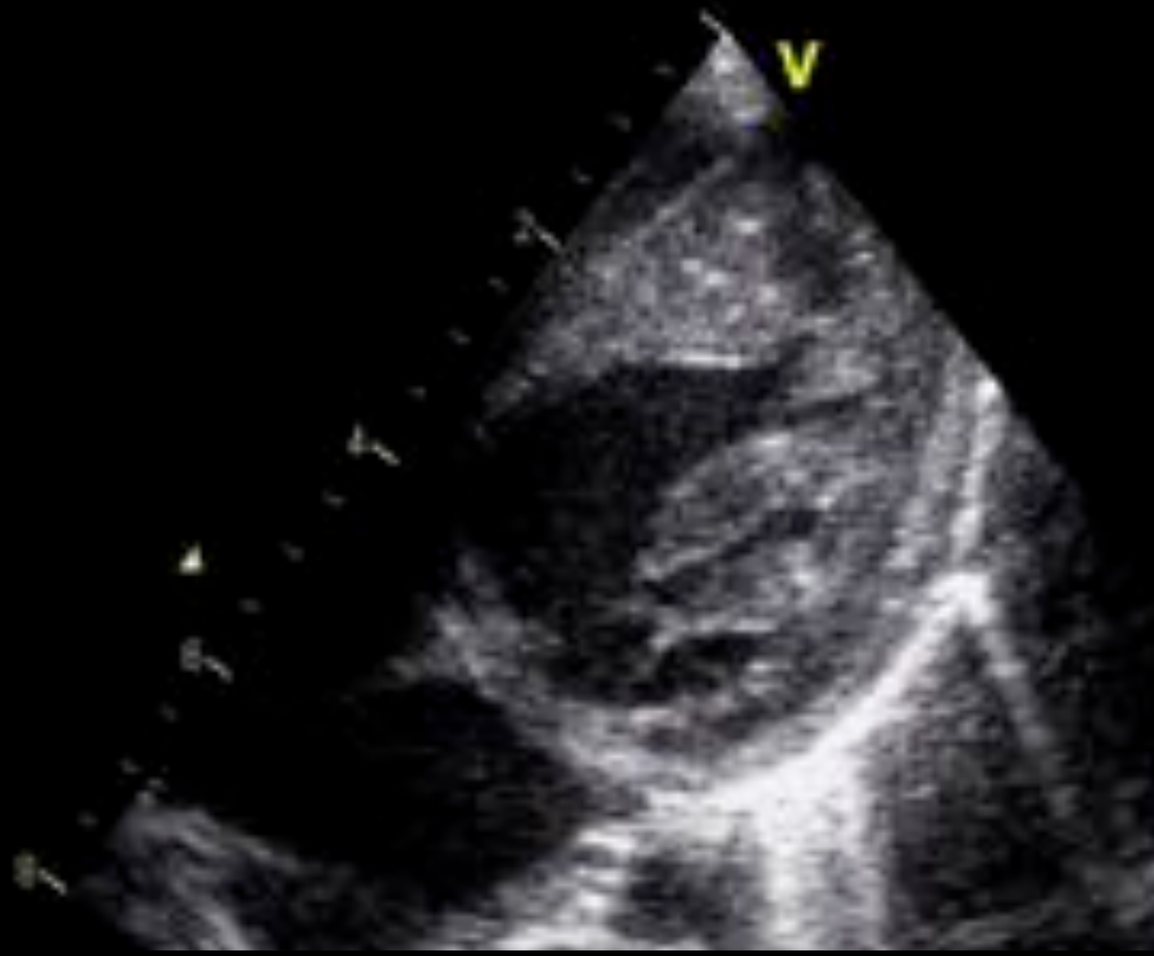
- In infants with intractable arrhythmias, electrophysiological mapping is indicated if antiarrhythmics are ineffective in ablating arrhythmias and allowing regression of the lesions. Treatment includes surgical excision or direct-vision cryoablation of the multiple small nodular tumors.
- Mortality is high (20%).

Histiocytoid cardiomyopathy



Histiocytoid cardiomyopathy

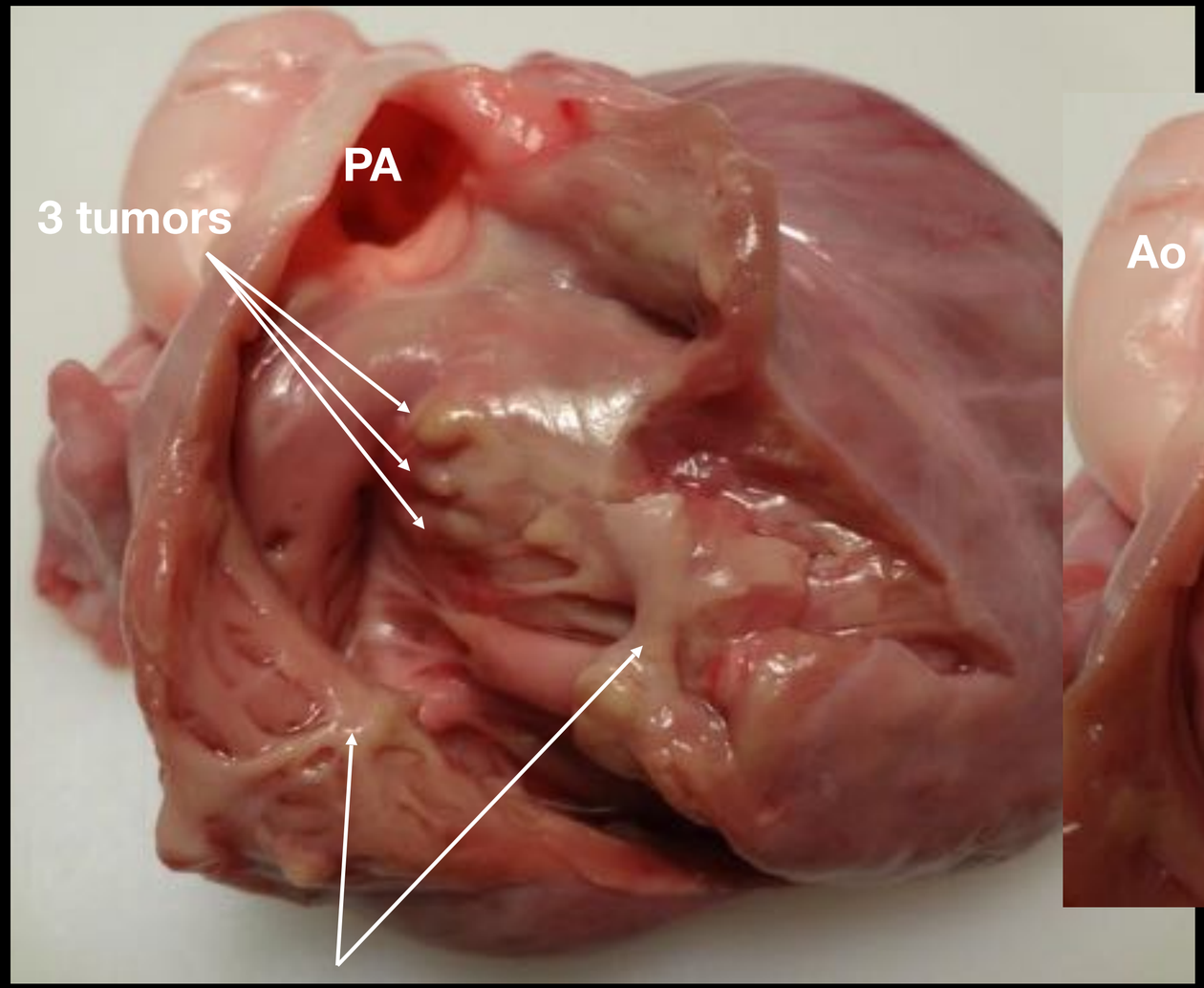


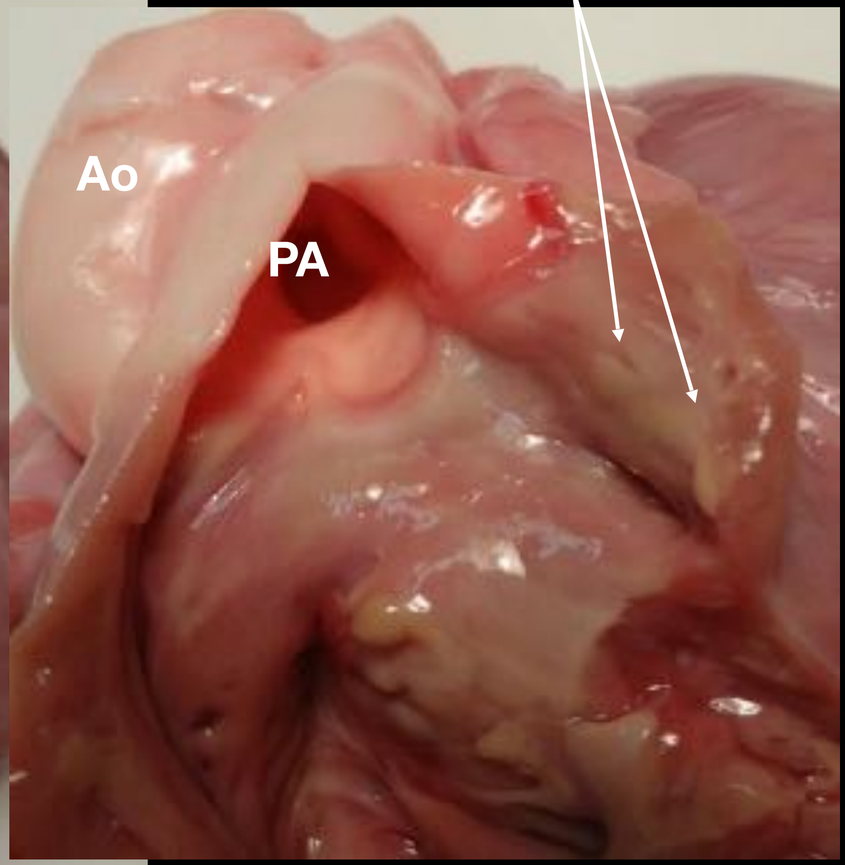


Histiocytoid cardiomyopathy

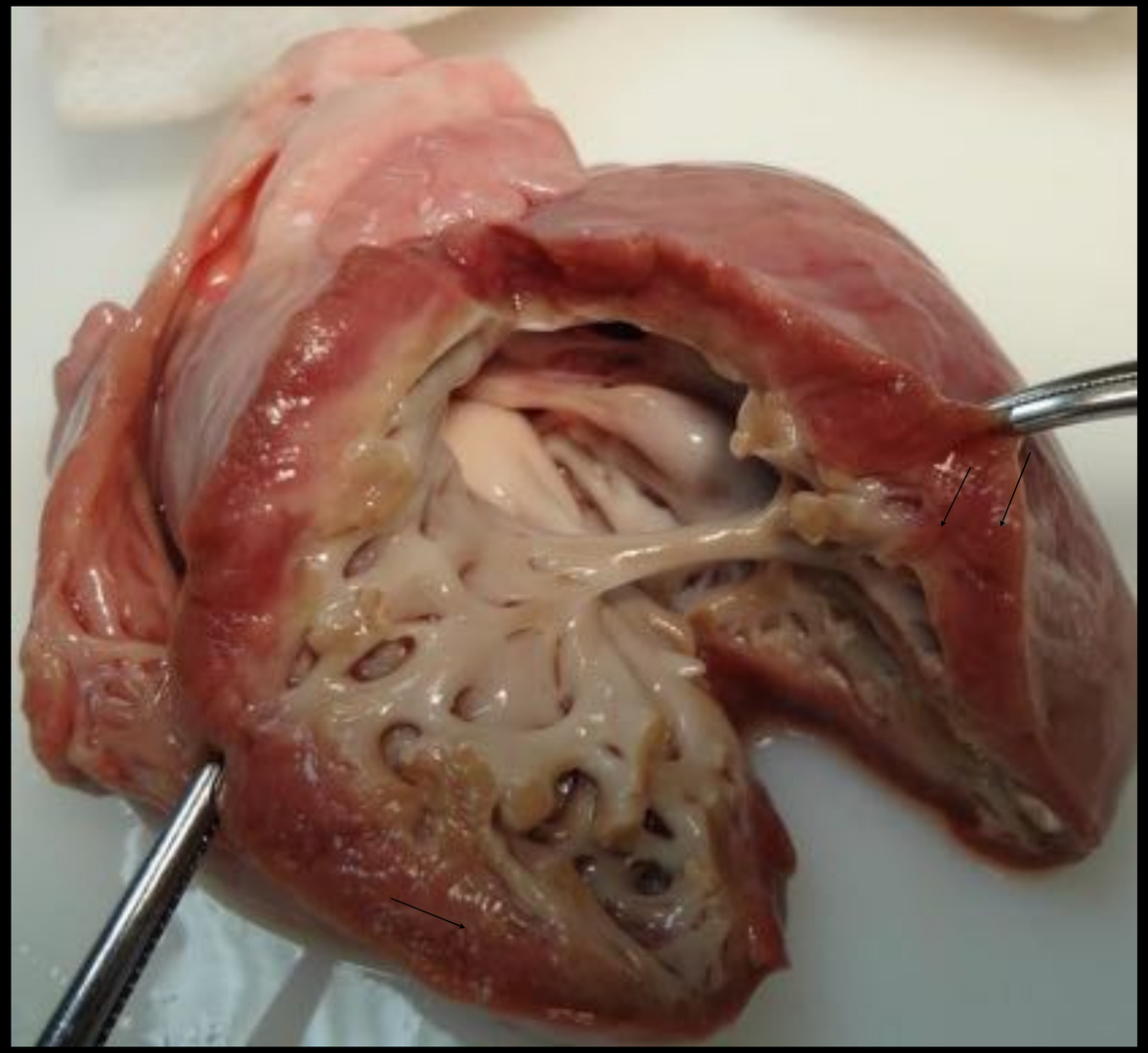
- Pathologically, there are typically subendocardial yellow–tan nodules or plaques.
- They can also be seen in the inner myocardium and subepicardial areas.
- The lesions may be grossly difficult to identify, but there is generally a subtle color difference separating the lesion from a normal myocardium.
- The histologic findings are pathognomonic, with nests of foamy-appearing myocytes resembling macrophages.

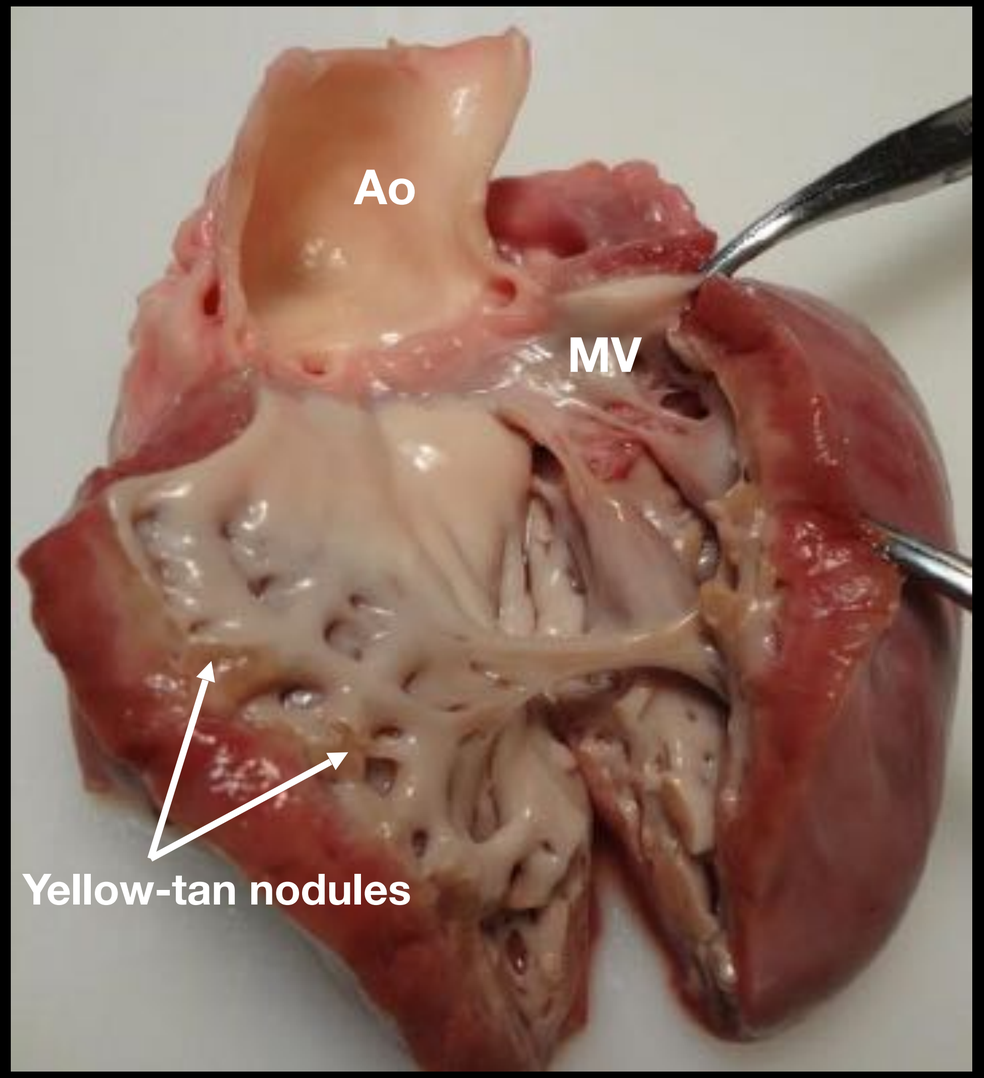
Histiocytoid cardiomyopathy
Right ventricle



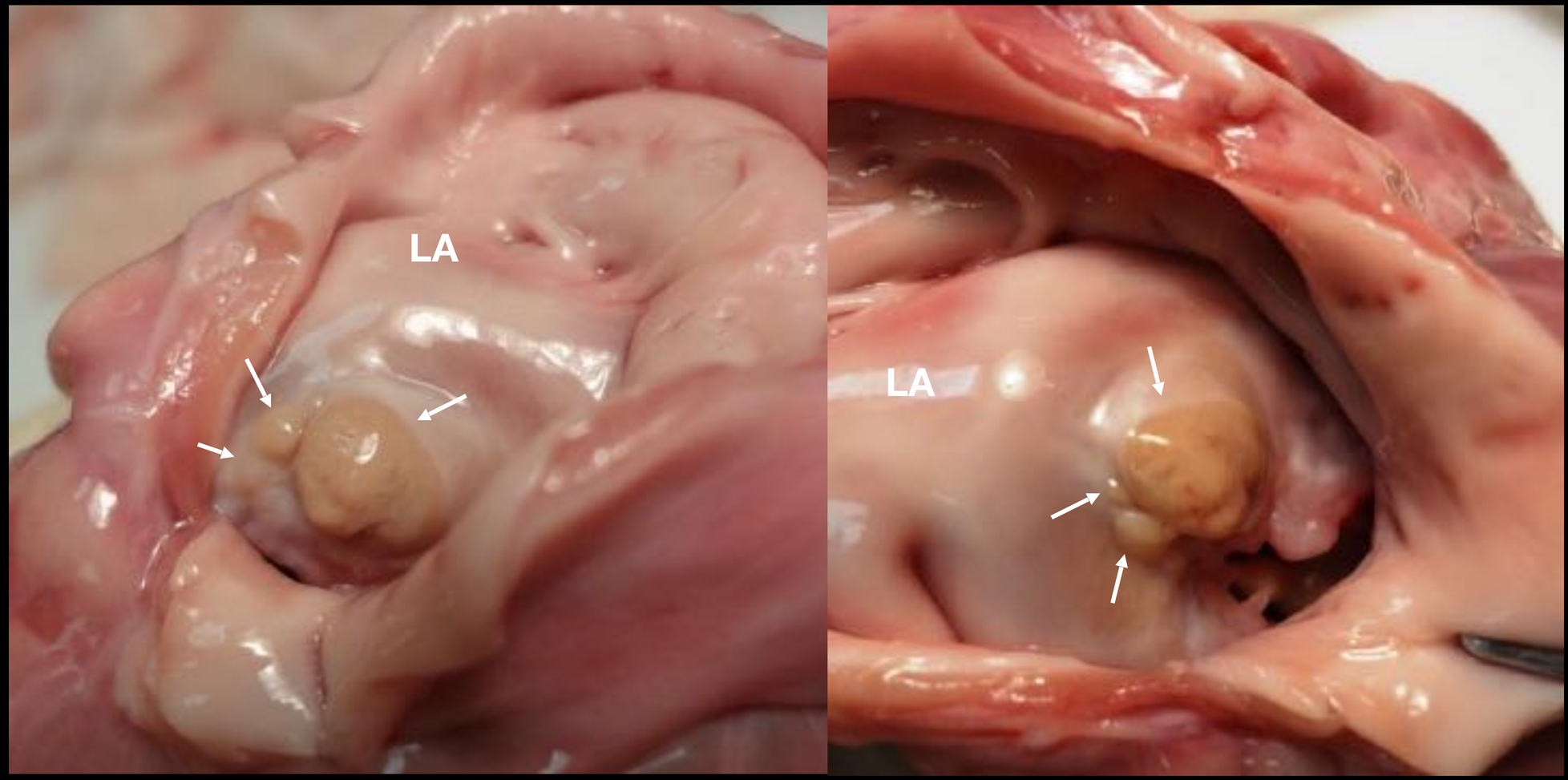


Subendocardial, epicardial, or valvular yellow-tan nodules



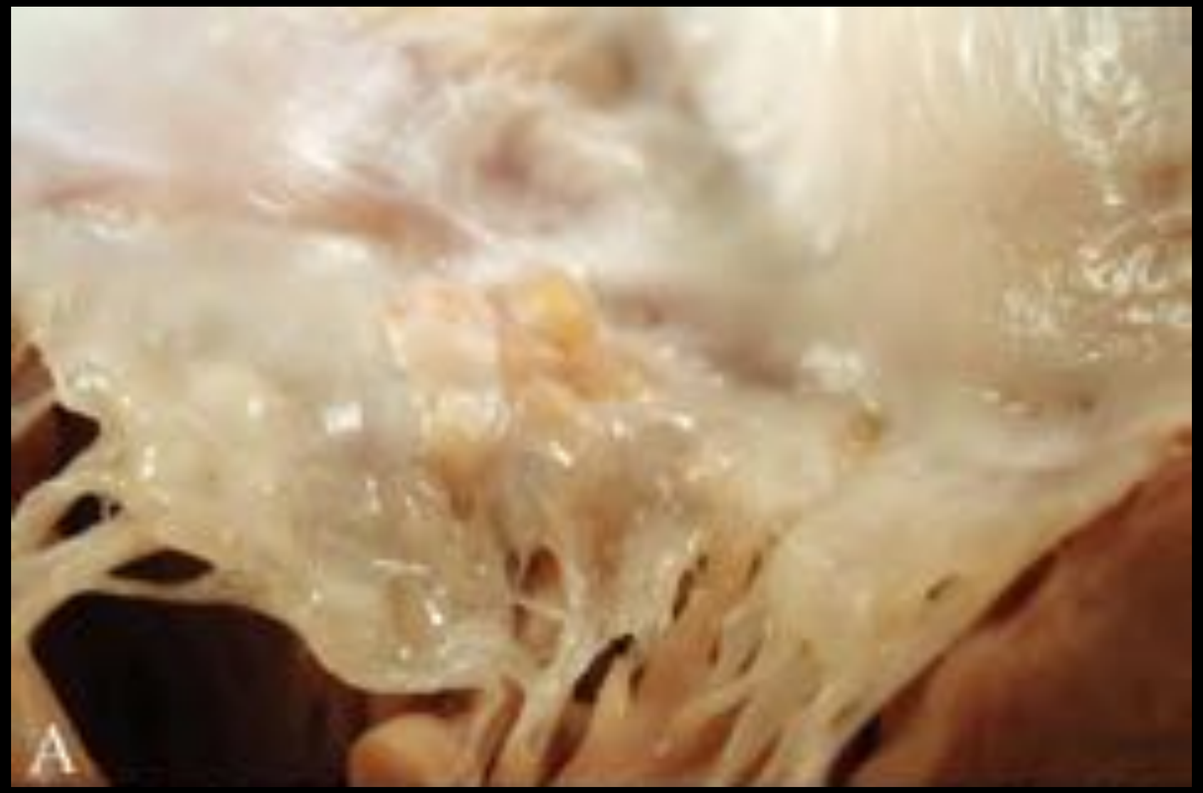


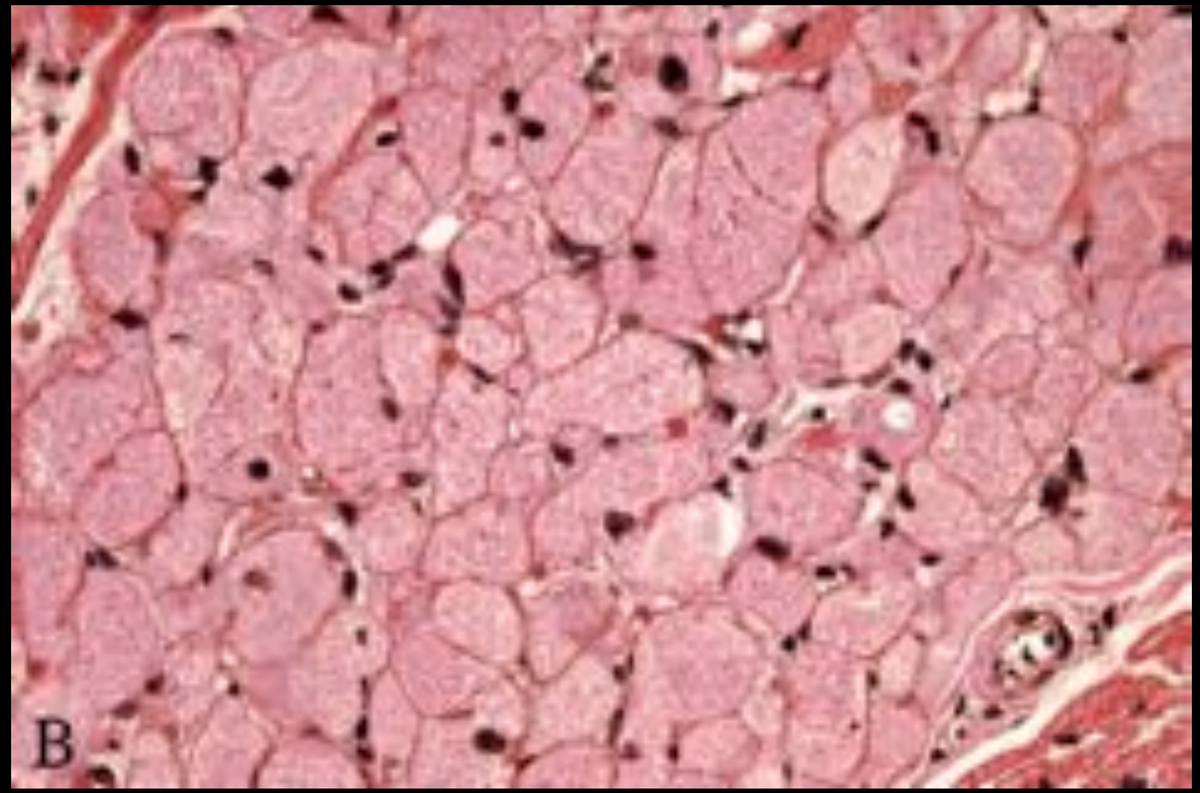
Histiocytoïd cardiomyopathy



Multiple tumors in the left atrium

Histiocytoïd cardiomyopathy





Vacuolated oncocytic cells

Pericardial tumors

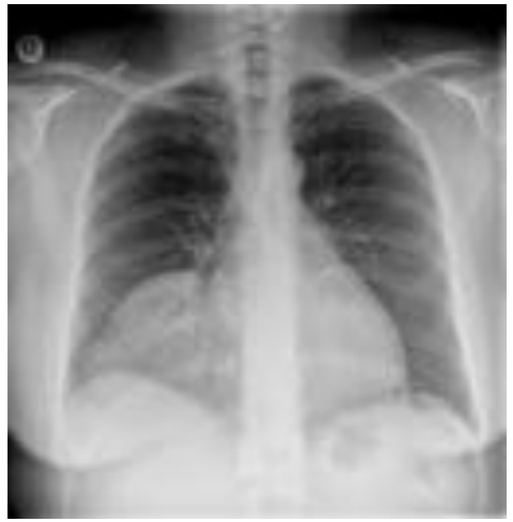
see the course Pericardial anomalies

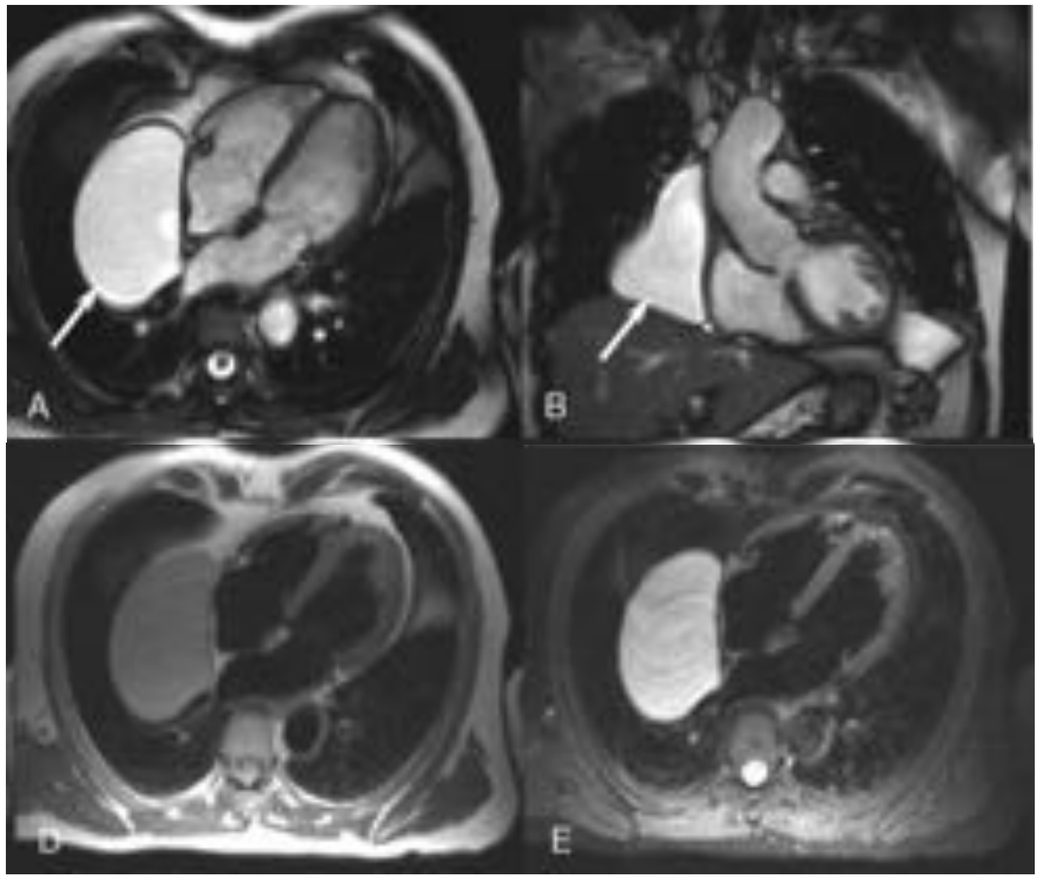
Pericardial tumors

- Pericardial tumors can be benign or malignant (1-3 per 10,000 patients in autopsy series)
- Of the benign pericardial tumors, **pericardial cysts are the most common**.
- Other benign pericardial tumors include **angiomas**, **lymphangiomas**, **fibromas**, **teratomas**, and **lipomas**.
- **Mesothelial cardiac excrescences and mesothelial papillomas** are among the benign tumors of the pericardium. They are small collections of mesothelial cells mixed with fat cells, macrophages with no intervening stroma. Histologically, mesothelial papillomas appear as a cuboidal epithelioid cell arising from the pericardial surface. It is also called an adenomatoid tumor. It is usually an incidental finding at autopsy.
- Surgical excision of a benign pericardial tumor is usually curative.
- **Metastatic pericardial tumors occur 20–40 times more commonly** than the primary pericardial tumors.

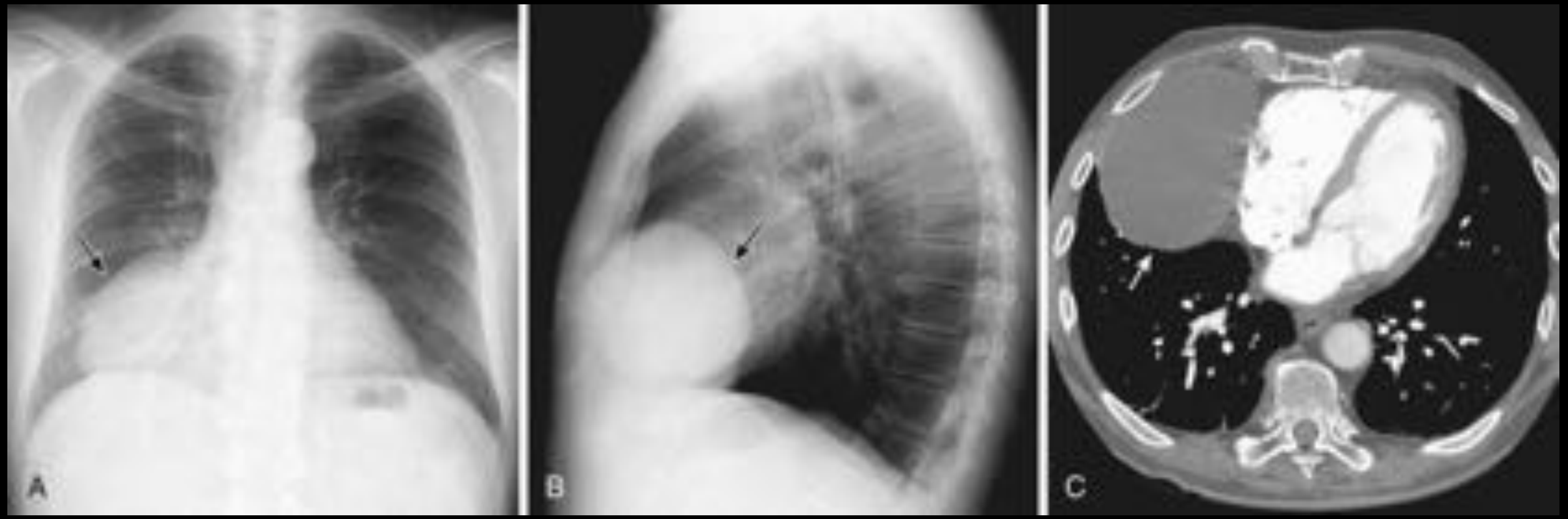
Pericardial cysts

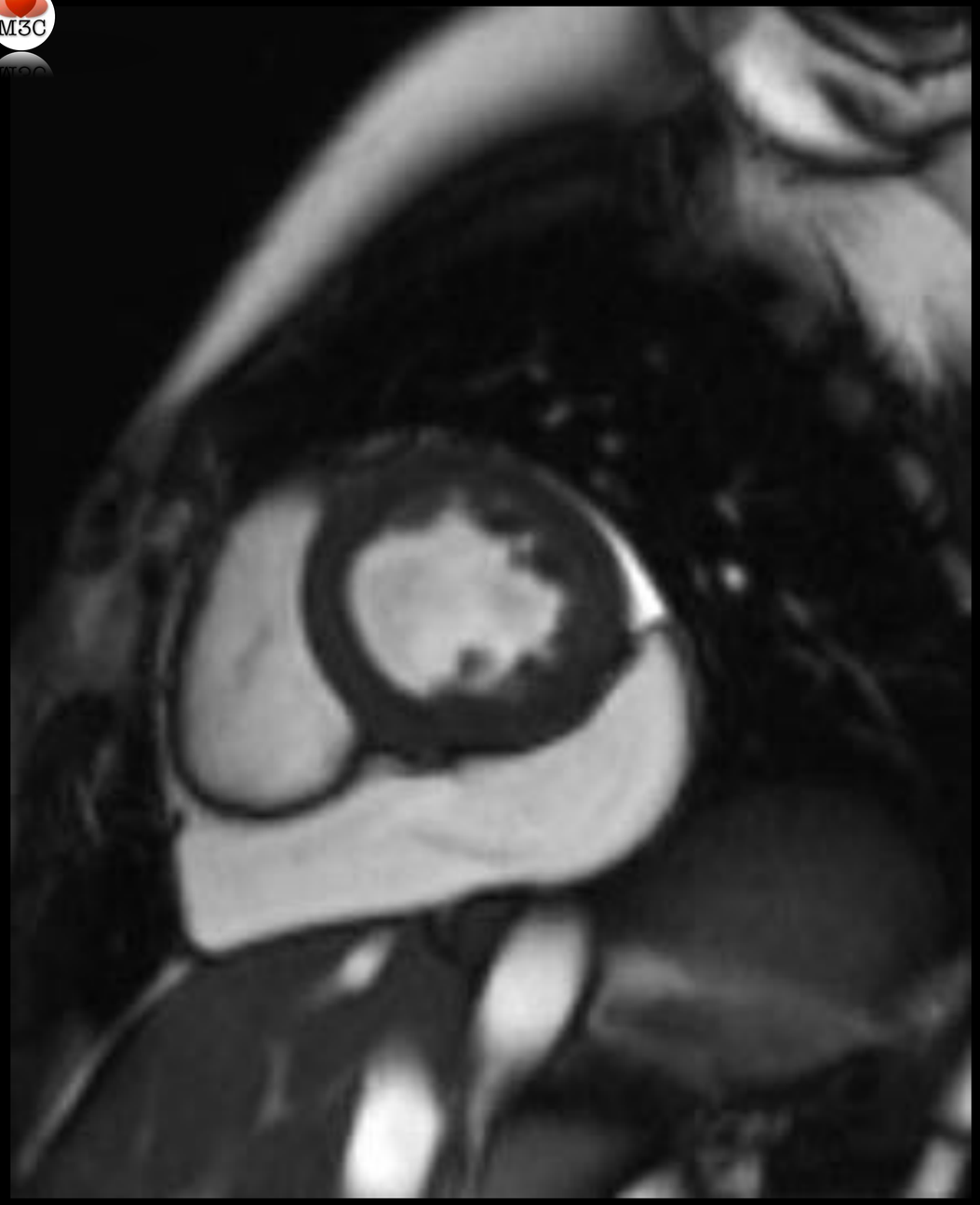
- Rare lesions and commonly located in the right pericardiophrenic angle.
- Usually filled with clear fluid.
- Patients are generally asymptomatic, and the lesion is often discovered on a routine chest film. The appearance is typically stable over a long period. In most cases, no cardiac surgery is necessary.
- On CMR, pericardial cysts appear as paracardiac masses with long T1 and T2 values and flow void, indicating fluid-filled structures. They have low signal intensity on T1-weighted images and increased signal intensity on T2-weighted images. After the injection of gadolinium, intracystic septae may be seen. In addition, a line of low signal intensity, representing the pericardial layer, can often be visualized. The significant advantage of CMR is its ability to differentiate these lesions from other mediastinal masses and avoid explorative surgery to determine the diagnosis.



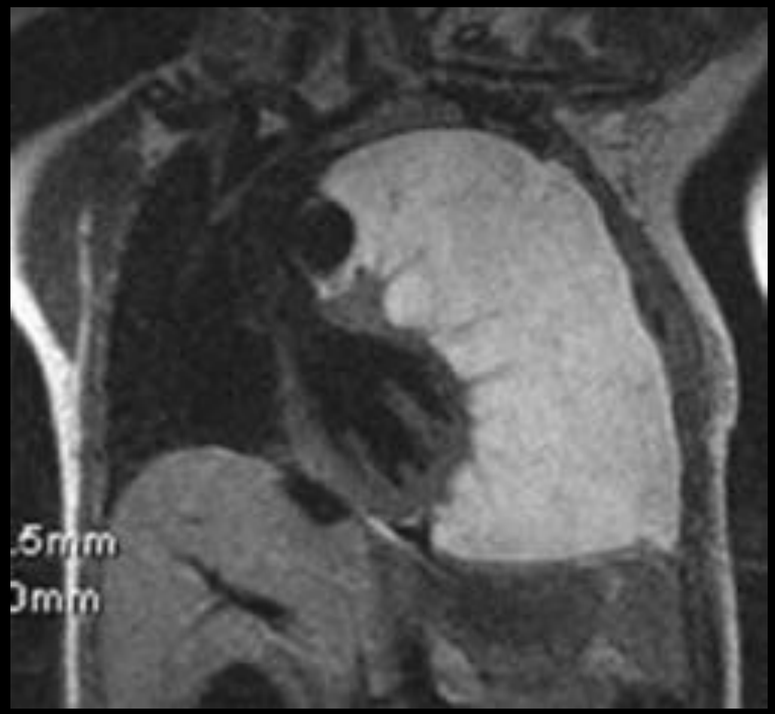


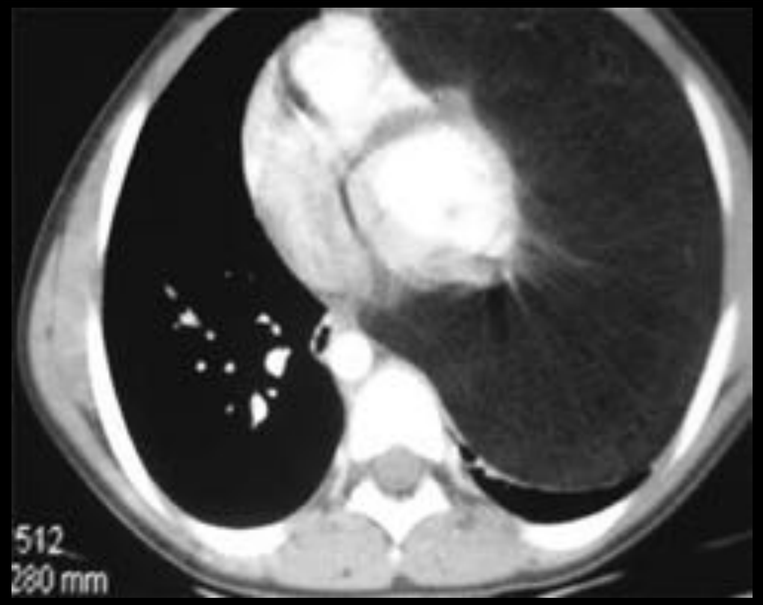
Pericardial cysts





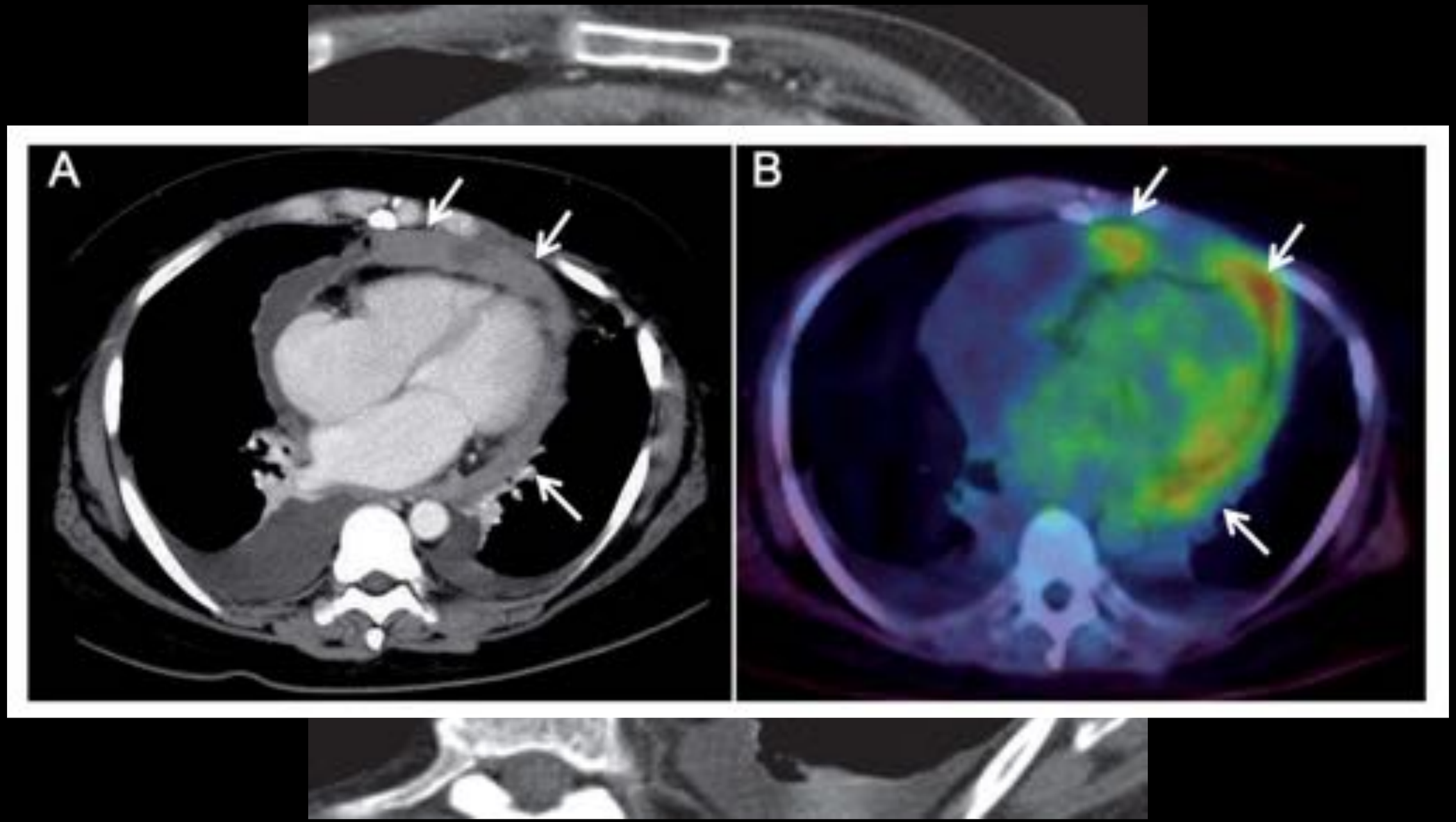
Pericardial lipoma - MRI T2





Pericardial lipoblastoma - MRI T1

Restrepo SC et al.
Radiographics 2013



Mesothelioma

Restrepo SC et al. Radiographics 2013

Collective ignorance is the motivation
Curiosity is the strength
Research is the path

Individual experience is the brake
Indifference is the weakness
Argument from authority is the threat