CARDIAC TUMOURS



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Classification

- Cardiac neoplasms
  - primary or secondary
  - related to the heart muscle or the pericardium
  - direct extension of adjacent primary tumors
  - metastases
- The presence of a cardiac tumor upon clinical examination and electrocardiography was first documented in 1934. Until that time, cardiac tumors were identified postmortem (Majano-Lainez, 1997).
- Angiography first was used to demonstrate a cardiac tumor within a chamber in 1951 (Majano-Lainez, 1997).

- Pseudotumors (Mural thrombi)
- Heterotopias and tumours of ectopic tissue
  - Tumours of the atrioventricular nodal region Teratoma Ectopic thyroid
- Tumours of mesenchymal tissue
  - Hamartoma of endocardial tissue Papillary fibroelastoma Hamartomas of cardiac muscle Rhabdomyoma Histiocytoid cardiomyopathy (Purkinje cell hamartoma)
- Tumours and neoplasms of fat
  - Lipomatous hypertrophy, interarterial septum Lipoma Liposarcoma
- Tumours and neoplasms of fibrous and myofibroblastic tissue
  - Fibroma Inflammatory pseudotumor (inflammatory myofibroblastic tumour)
- Sarcomas
  - malignant fibrous histiocytoma, fibrosarcoma, leiomyosarcoma
- Vascular Tumors and neoplasms
  - Hemangioma Epithelioid hemangioendothelioma Angiosarcoma
- Neoplasm of uncertain histogenesis
  - Myxoma
- Neoplasms of neural tissue
  - Granular cell tumor Schwannoma/neurofibroma Paraganglioma
- Malignant schwannoma/neurofibrosarcoma (rare)
- Malignant lymphomaMalignant mesothelioma
- Metastatic Tumours to the heart

Frequency

- The most common intracardiac mass is thrombus (Schvartzman, 2000).
- Benign primary tumours are more frequent than primary malignant tumours. In autopsy series, the cumulative prevalence of primary cardiac tumours is 0.001-0.3% (Urba, 1986; McAllister, 1979).
- Overall, myxomas account for approximately 30% of all primary cardiac neoplasms. Of atrial myxomas, 90% occur on the left and 90% are solitary (Shapiro, 2001).
- Lipomas are the second most common benign cardiac tumor and account for approximately 10% of all cardiac neoplasms (Hoffmann, 1998; Vanderheyden, 1998).
- Angiosarcomas account for 33% of all malignant primary cardiac neoplasms, and myxomas account for approximately 50% of all benign primary cardiac neoplasms (Shapiro, 2001)

Frequency

- Metastatic tumors to the heart and pericardium are more common than primary neoplasms and occur in as many as 1.5% of patients with malignancies (Schvartzman, 2000).
- The most common sources of metastatic tumours are as follows (in approximate order of frequency):
  - bronchogenic carcinoma of the breast;
  - lymphoma;
  - leukemia;
  - oesophagus;
  - uterus;
  - melanoma;
  - stomach;
  - sarcomas;
  - germ cell tumors; and tumors of the oral/tongue, colon/rectum, kidney, thyroid, larynx, urinary bladder, hepatobiliary system, prostate, pancreas, and ovary (Roberts, 1998).

Paediatric tumours

- Most primary paediatric tumors are diagnosed within the first year of life (Sallee, 1999).
- Among paediatric patients, primary cardiac neoplasms include rhabdomyoma (78%), fibroma (11%), pericardial teratoma (2%), epicardial lipoma (2%), multicystic hamartoma (2%), and unspecified 5%(Beghetti, 1997).
- In children aged 15 years or younger, the most common benign primary cardiac tumors are as follows: rhabdomyoma (the most common benign primary tumor in those <1 y of age and associated with tuberous sclerosis in 30-50%), fibroma, hemangioma, and teratoma (Roberts, 1998).
- In children aged 15 years or younger, the most common malignant primary cardiac tumors are as follows: rhabdomyosarcoma fibrous histiocytoma, angiosarcoma, fibrosarcoma, and myxoid sarcoma (Roberts, 1998).

Adult tumours

- Angiosarcomas tend to be found in patients aged 20-50 years (Kim, 1989; Mader, 1997).
- About 70% of myxomas occur in middle-aged patients (Schvartzman, 2000), usually those aged 50-70 years (Shapiro, 2001).

- The most common benign primary cardiac tumours are as follows (in approximate order of frequency): myxoma, (>50%), fibroma, hemangioma, granular cell tumor, lipoma, paraganglioma, hamartoma, histiocytoma, and hemangioendothelioma (Roberts, 1998).

- The most common malignant primary cardiac tumours are as follows (in approximate order of frequency): angiosarcoma, fibrous histiocytoma, leiomyosarcoma, osteosarcoma, fibrosarcoma, myxoid sarcoma, rhabdomyosarcoma, and liposarcoma (Roberts, 1998).

Metastatic tumours

- Abdominal tumours can affect the heart; extension of tumor thrombus via the inferior vena cava into the right atrium is a well-recognized complication of advanced renal cell carcinoma
- It also can occur with cervical carcinoma and renal angiomyolipomas (Ogawa, 1999; Vargas-Barron, 1990; Rothenberg, 1986).
- Benign uterine leiomyomatosis also can affect the heart intravenously (Steinmetz, 1996; Okamoto, 1994; Kaszar-Seibert, 1988).
- Germ cell tumours (eg, testicular teratomas) and embryonal cell carcinomas can metastasize to the heart, as can choriocarcinomas (Pickuth, 1992; Paule, 1991; Chandrashekar, 1995).


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Pericardial tumours

- Primary pericardial tumors include malignant spindle tumor, localized fibrous tumor (also called localized fibrous mesothelioma), pericardial cysts, liposarcoma, lipoma , and teratoma (Fukuda, 1989; el-Naggar, 1989).
- Cardiac angiosarcomas tend also to involve the pericardium secondarily and to produce hemorrhagic pericardial effusions (Kim, 1989).
- Pericardial mesotheliomas frequently are associated with asbestos exposure. Most specimens have identifiable asbestos fibers, particularly crocidolite and amosite fibers, as well as chrysotile to a lesser extent (Morinaga, 1989). However, not all mesotheliomas arise in patients with a prior history of asbestos exposure (Watanabe, 1999).
- Lymphomas frequently are found in patients who are immunocompromised and frequently involve the pericardium (Araoz, 1999).
- Liposarcomas are rare tumours, they tend to be extracardiac, and they tend to form large infiltrating masses (Araoz, 1999).

Mortality/Morbidity

- Angiosarcomas have a high mortality rate, and most patients do not survive beyond 6 months of the diagnosis (Shapiro, 2001).
- Mortality rates in patients with other malignant tumours also are high, especially in those with metastases, which tend to involve the heart late in the course of malignant disease.
- Benign tumours can usually be resected and have a good prognosis.

Sex

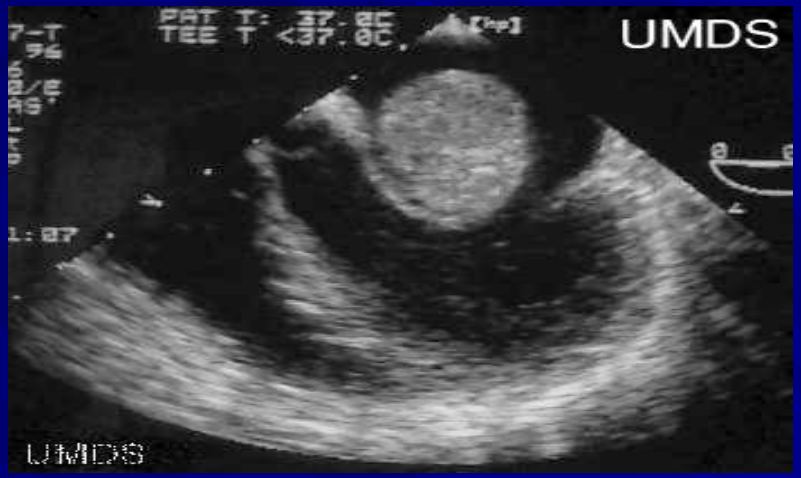
- Cardiac myxomas tend to occur more often in women than in men (Shapiro, 2001).
- Cardiac lipomas have no sex predilection (Schvartzman, 2000).
- Rhabdomyosarcomas and angiosarcomas are more common in males than in females (Shapiro, 2001).

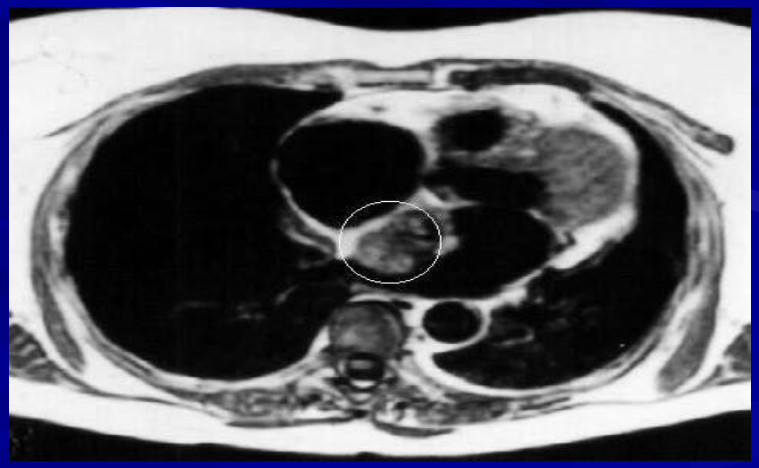
Clinical presentation

- Patients with left heart disease may present with mitral valve obstruction or apparent subacute bacterial endocarditis. Patients with right heart disease may present with tricuspid valve disease, pulmonary embolism, or hypertension (Symbas, 1976).
- Atrial myxomas can cause breathlessness, fever, weight loss, syncope, hemoptysis, peripheral emboli, and sudden death (Shapiro, 2001).
- Angiosarcomas frequently involve the right heart and result in right-sided heart failure. Patients may also present with cardiac tamponade secondary to bloody pericardial effusions or with pleuritic chest pain (Shapiro, 2001; Schvartzman, 2000; Kim, 1989).
- Intrapericardial masses can cause dyspnea as a result of compression of airways (Doshi, 1998; Sievers, 2000).
- Patient with cardiac fibromas may present with congestive heart failure, arrhythmias, sudden death, cyanosis, or chest pain, although one third of patients may be asymptomatic (Burke, 1994; Parmley, 1998).
- Patients with intracardiac phaeochromocytomas may present with chest pain, palpitations, and flushing (Fitzgerald, 1995). Catecholamine release can cause irregular heartbeats, which can be detected by the patient as palpitations. Blood pressure can rise with an increased pulse rate, resulting in more work for the heart.

Imaging

- The degree of confidence is high for detecting tumors on contrast-enhanced CT scanning. Tissue characterization is limited. ECG gating improves image quality and precision.
- Ultrasonographic findings are accurate for identifying tumors inside cardiac chambers provided acoustic access is adequate. Ultrasonography is less helpful if acoustic access is poor or if tumours extend outside the heart or into the great vessels. Although it is more invasive, transoesophageal echo imaging is best for imaging left atrial and left ventricular tumors.



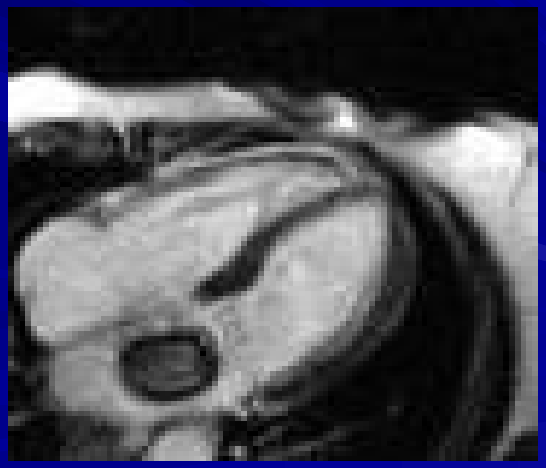


MRI/MRA

Both MRI and MRA are superior to echocardiography in detecting or excluding cardiac tumors, the precise location of cardiac tumors (ie, paracardiac, mural, or intracavitary), the extent of disease, the presence of effusions, and the presence of metastases (Link, 1995). Other effects and complications of tumours are demonstrated well.

Contrast-enhanced MRI may define the extent of the tumor, but it is limited in discerning benign from malignant disease. MRI has advantages over CT, because intravenous contrast material is not required for imaging.





Myxomas

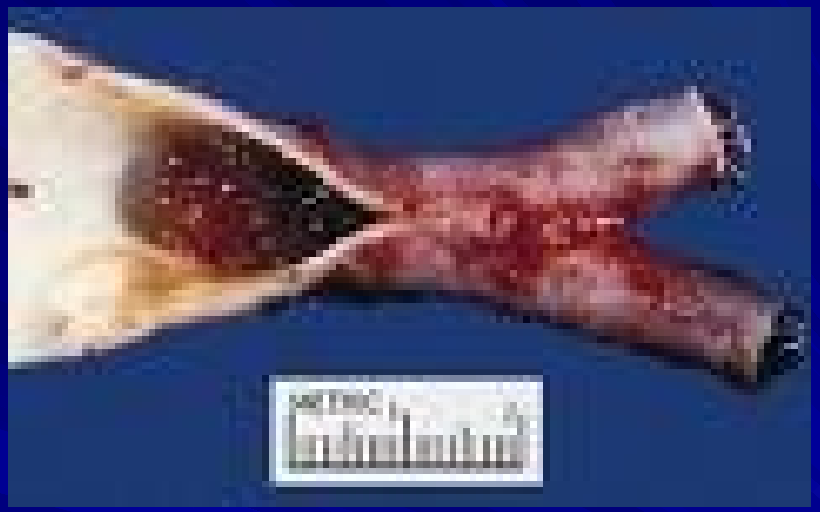
- Myxomas usually are found in the left heart, but they do occur, less often, on the right (Symbas, 1976).
- Myxomas tend to be solitary and are characteristically attached to the interatrial septum adjacent to the edge of the fossa ovalis in approximately 85% of patients (Schvartzman, 2000).
- Can invade the interatrial septum and the opposite atrium (Peachell, 1998). Tumours may be pedunculated or polypoid (Schvartzman, 2000).
- Firm, oval masses are associated with dyspnoea and show small vessels, tortuous vascularity, haemorrhage, and fibrosis. Soft papillary tumors tend be associated with neurologic symptoms and brain infarcts (Shimono, 1995).
- Almost one half may have surface thrombus, and almost one half may have fibrosis (Burke, 1993).
- Patients with left-heart myxomas may present with mitral valve obstruction or features suggesting subacute bacterial endocarditis. Patients with right-heart myxomas may present with tricuspid valve disease, pulmonary embolism, or pulmonary hypertension (Symbas, 1976; Seemann, 1996).

SURGICAL MANAGEMENT

- Surgical resection is the only effective therapeutic option for patients with cardiac myxoma and should not be delayed because death from obstruction to flow within the heart or embolization may occur in as many as 8% of patients awaiting operation.
- Median sternotomy approach with ascending aortic and bicaval cannulation is usually employed.
- Manipulation of the heart before initiation of cardiopulmonary bypass is minimized in deference to the known friability and embolic tendency of myxomas.
- For left atrial myxomas the vena cava are cannulated through the right atrial wall with the inferior cannula placed close and laterally to the inferior vena cava right atrial junction .
- Caval snares are always used to allow opening of the right atrium, if necessary. If extensive exposure of the left atrium is needed, or a malignant left atrial tumor is suspected, we mobilize and directly cannulate the superior vena cava, which allows it to be transected if necessary for additional exposure .
- Body temperature is allowed to drift down but there is no attempt to induce systemic hypothermia unless the need for reduced perfusion flow is anticipated.
- Modern cardioplegic techniques yield a quiet operative field and protect the myocardium from ischemic injury during aortic cross-clamping. Cardiopulmonary bypass is started and the aorta is clamped prior to manipulation of the heart.

Left atrium

- For left atrial tumours the superior vena cava is extensively mobilized as is the inferior vena caval–right atrial junction, allowing increased mobility and exposing the left atrial cavity.
- Left atrial myxomas can be approached by an incision through the anterior wall of the left atrium anterior to the right pulmonary veins.
- This incision can be extended behind both cava for greater exposure

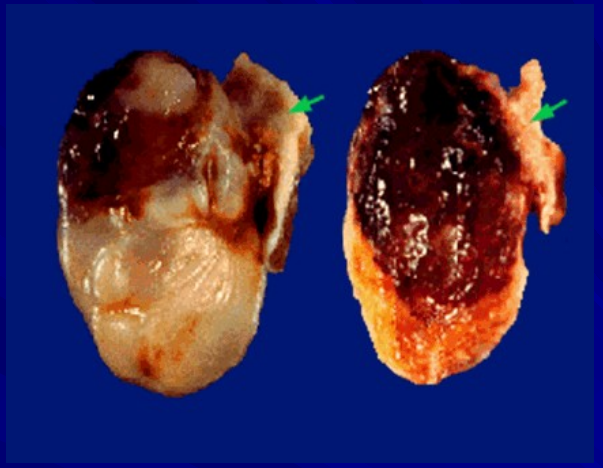


Right atrium

- Right atrial myxomas pose special venous cannulation problems and intraoperative echocardiography may be of benefit. Both vena cava may be cannulated directly. When low- or high-lying tumour pedicles preclude safe transatrial cannulation, cannulation of the jugular or femoral vein can provide venous drainage.
- Resection of large or critically placed right atrial myxomas often requires careful preoperative planning, intraoperative transesophageal echocardiography, and special extracorporeal perfusion techniques to ensure complete removal of the tumour, protection of right atrial structures, and reconstruction of the atrium.
- Because myxomas rarely extend deep in the endocardium, it is not necessary to resect deeply around the conduction tissue. The tricuspid valve and the right atrium as well as the left atrium ventricle should be inspected carefully for multicentric tumours in patients with right atrial myxoma, with or without familial myxoma.

Tumour resection

- Regardless of the surgical approach, the ideal resection encompasses the tumour and a portion of the cardiac wall or interatrial septum to which it is attached.
- Whether excision of full thickness wall is necessary or excision of only an endocardial attachment is sufficient to prevent recurrence is controversial.





Ventricular tumours

- Ventricular myxomas usually are approached through the AV valve or by detaching the anterior portion of the AV valve for exposure and resection and reattachment after resection.
- Occasional small tumours in either outflow tract can be removed through the outflow valve.
- If necessary, the tumour is excised through a direct incision into the ventricle, but this is unusual.
- It is not necessary to remove full thickness of the ventricular wall since no recurrences have been reported with partial thickness excisions.
- As with right atrial myxoma, the presence of ventricular myxoma prompts inspection for other tumours because of the high incidence of multiple tumours.

Mortality/Recurrence

- Removal of atrial myxomas carries an operative mortality rate of 5% or less.A review of 202 resections indicates that operative mortality is related to advanced age or disability and comorbid conditions.
- Excision of ventricular myxomas carries a higher risk (approximately 10%) but the experience is small.
- Recurrence of nonfamilial sporadic myxoma is approximately 1% to 4%

Follow up

- Uncertainty concerning the true malignant potential of myxomas has increased as reports that myxomas generally classified as "malignant" are found on subsequent review to be sarcomas with myxoid degeneration. However, this issue also remains unsettled because of reports of metastatic growth of embolic myxoma fragments in the brain, arteries, soft tissue, and bones. (Castells E, 1993)
- Symptomatic lesions of possible metastatic myxoma should be excised if possible. (Seo S, 1980)
- The extent to which patients should be subjected to long-term echocardiographic surveillance after myxoma resection is not standardized. It would seem prudent to closely follow patients who are treated initially for multicentric tumors, those whose tumours are removed from unusual locations in the heart, all tumours believed to have been incompletely resected, and tumours found to have an abnormal DNA genotype.
- Patients undergoing resection of tumours thought to be myxoma but with malignant characteristics at pathologic examination should have long-term, careful follow-up.

Lipoma

- Lipomas are well-encapsulated tumours consisting of mature fat cells that may occur anywhere in the heart but also are found in the pericardium, subendocardium, subepicardium, or intra-atrial septum.
- Occur at any age and have no sex predilection.
- Lipomas are slow growing and may attain considerable size before producing obstructive or arrhythmic symptoms. Many are asymptomatic and are discovered incidentally on routine chest x-ray, echocardiogram, or at surgery or autopsy.
- Large tumours that produce severe symptoms should be resected. Smaller, asymptomatic tumours encountered unexpectedly during cardiac operation should be removed if excision can be performed without adding risk to the primary procedure.
- These tumours are not known to recur.

Lipomatous hypertrophy of the interatrial septum

- Nonencapsulated hypertrophy of the fat within the atrial septum is known as lipomatous hypertrophy.
- This abnormality is more common than cardiac lipoma and is usually encountered in elderly, obese, or female patients as an incidental finding during a variety of cardiac imaging procedures.
- Various arrhythmias and conduction disturbances have been attributed to its presence. (McAllister HA, 1979).The main problem is differentiation from a cardiac neoplasm when the lesion is discovered on echocardiography.
- Arrhythmias or heart block are considered by some as an indication for resection, but data is lacking as to the long-term benefits from resection. (Vander Salm 2000)



Papillary fibroelastoma of the heart valves

- Arise characteristically from the cardiac valves or adjacent endocardium.
- The AV and semilunar valves are affected with equal frequency.
- They are capable of producing obstruction of flow, particularly coronary ostial flow, and may embolize to the brain and produce stroke. (Israel DH,1991)
- They are usually asymptomatic until a critical event occurs but now are found more often because of the more frequent use of echocardiography.
- Papillary fibroelastomas of the cardiac valve should be resected whenever diagnosed because of their known tendency to produce life-threatening complications.
- Valve repair rather than replacement should follow the resection of these benign tumours whenever technically feasible, using conservative margins of resection.
- Cytomegalovirus has been recovered in these tumours suggesting the possibility of viral induction of the tumour and chronic viral endocarditis.



Rhabdomyoma

- Rhabdomyoma is the most frequently occurring cardiac tumour in children. It usually presents during the first few days after birth.
- Associated strongly with tuberous sclerosis, a hereditary disorder characterized by hamartomas in various organs, epilepsy, mental deficiency, and sebaceous adenomas.
- Over 90% of rhabdomyomas are multiple and occur with approximate equal frequency in both ventricles.
- The most common presentation is heart failure caused by tumour obstruction of cardiac chambers or valvular orifice flow. Clinical findings may mimic valvular or subvalvular stenosis.
- Early operation is recommended in patients who do not have tuberous sclerosis before 1 year of age.
- Unfortunately, symptomatic tumours often are both multiple and extensive, particularly in patients with tuberous sclerosis who unfortunately have a dismal long-term outlook. In such circumstances, surgery offers little benefit.



Fibroma

- Fibromas are the second most common benign cardiac tumour, with over 83% occurring in children.
- These tumours are solitary, occur exclusively within the ventricle and the ventricular septum, and affect the sexes equally. Fewer than 100 tumours have been reported and most are diagnosed by age 2 years. These tumours are not associated with other disease, nor are they inherited..
- The majority of fibromas produce symptoms through chamber obstruction, interference with contraction, or arrhythmias.
- Surgical excision is successful in some patients, particularly if the tumour is localized, does not involve vital structures, and can be enucleated.However, it is not always possible to completely remove the tumour and partial removal is only palliative although some patients have survived many years. (Reece IJ, 1984)

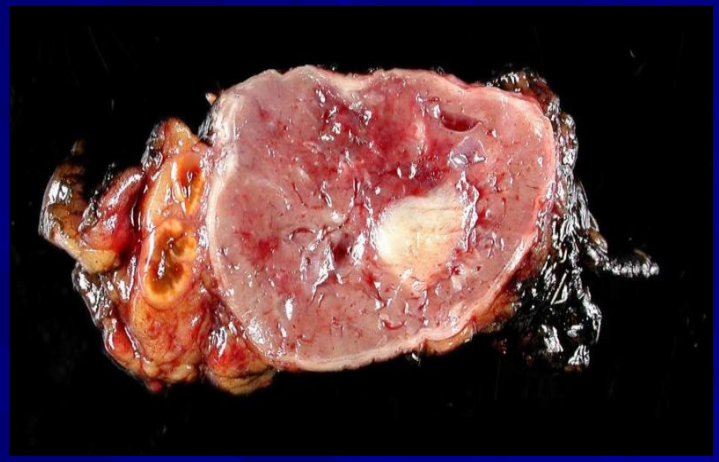


Mesothelioma of the AV node

- Mesothelioma of the atrial ventricular node, also termed polycystic tumour, Purkinje tumour, or conduction tumour.
- It is a relatively small, multicystic tumour that arises in proximity to the atrial ventricular node and may extend upward into the interventricular septum and downward along the bundle of His.
- Mesothelioma is associated with heart block, ventricular fibrillation, and sudden death. Cardiac pacing alone does not prevent subsequent ventricular fibrillation.
- Surgical excision has been reported. (Balasundaram S, 1992)

Phaeochromocytoma

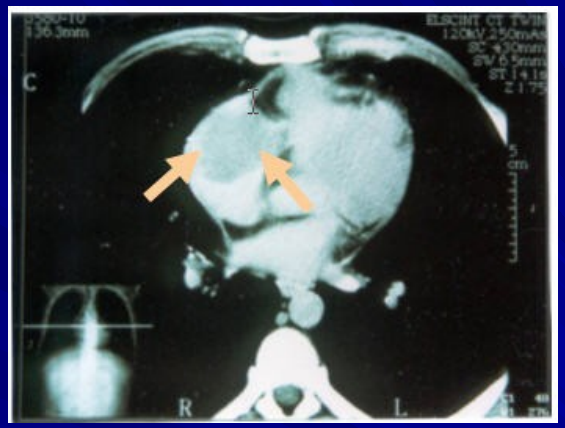
- Cardiac phaeochromocytomas arise from chromaffin cells of the sympathetic nervous system and produce excess amounts of catecholamines, particularly norepinephrine.
- Approximately 90% of phaeochromocytomas are in the adrenal glands. Fewer than 2% arise in the chest.
- Approximately 60% occur in the roof of the left atrium.
- The patients usually present with symptoms of uncontrolled hypertension or are found to have elevated urinary catecholamines. The tumour is usually located by scintigraphy. Because these tumours are vascular and may be near major coronary arteries, coronary arteriograms are advisable.
- After the tumour is located, it should be removed, using cardiopulmonary bypass with cardioplegic arrest.

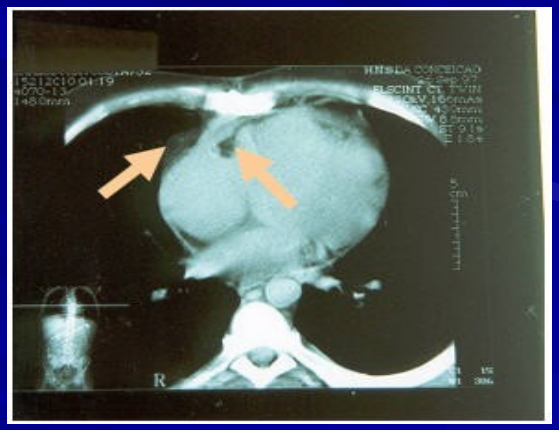


- Patients require preanesthetic alpha and beta blockade, and careful intraoperative and immediate postoperative monitoring.

Angiosarcoma

- Angiosarcomas are two to three times more common in men than women and have a predilection for the right heart.
- 80% arise in the right atrium.
- These tumours tend to be bulky and aggressively invade adjacent structures, including the great veins, tricuspid valve, right ventricular free wall, interventricular septum, and right coronary artery. Obstruction and right heart failure are not uncommon.
- Most of these tumours have spread by the time of presentation, usually to the lung, liver, and brain.
- Without resection 90% of the patients are dead within 9 to 12 months after diagnosis despite radiation or chemotherapy.
- Carefully selected patients without evidence of spread on metastatic evaluation who have undergone complete surgical resection with subsequent chemotherapy have been reported.





Rhabdomyosarcoma

- Rhabdomyosarcomas do not evolve from rhabdomyoma and occur equally in the sexes.
- The tumours are multicentric in 60% of patients and arise from either ventricle. These tumours frequently invade cardiac valves or interfere with valve function because of their intracavitary bulk.
- The tumours are aggressive and may invade pericardium.
- Surgical excision of small tumours may be rational but local and distant metastasis and poor response to radiation or chemotherapy limit survival to less than 12 months in the majority of these patients. (Burke AP, 1994)

