

In the name of God

Fetal Cardiac Tumors

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Introduction

- Cardiac masses are abnormal growth of tissues within the heart or pericardium.
- Categorized as **tumors** and **non-neoplastic**.
- Cardiac tumors can comprise muscle(**rhabdomyoma**), fibrous tissue (**fibroma**), vascular tissue(**hemangioma**), mixed tissue (**teratoma**), fatty tissue (**lipoma**) and rarely, metastatic tissue.

Introduction

- Primary or secondary
- Benign or malignant
- Primary cardiac tumors are much more common in children and almost all being benign.(<10% are malignant)
- In adult most of cardiac tumors are secondary and related to metastatic diseases.
- Non neoplastic masses include pericardial cyst, vascular malformation, infectious or inflammatory masses.

Incidence

- Cardiac masses are rare in children, with estimated incidence of 0.03-0.4.
- Over the past decades, the recognition of cardiac masses has increased.
- **Rhabdomyoma** is the most common primary tumor, comprising about 60-80% of cases followed by teratomas (25%) and fibromas (12%).
- Hemangiomas and lipoma are much less common.

Incidence

- Fetal cardiac tumors (FCTs) are rare, and the incidence of these tumors in different series ranges from 0.08% to 0.27%. This low incidence may be related to the difficulties in ultra sonographic screening.
- however, the true incidence of cardiac tumors in prenatal life is difficult to estimate as *these tumors often regress over time*.
- In addition, atrial tumors can be small and may be difficult to recognize, and not all pregnancies are screened with ultrasound.

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- FCTs, after excluding pericardial tumors or cysts, can be divided into two groups: benign tumors including rhabdomyomas, teratomas, fibromas, and myxomas; and malignant tumors including rhabdomyosarcomas and fibrosarcomas.
 - FCTs, especially rhabdomyomas, are often associated with tuberous sclerosis complex (TSC). FCTs have been reported to be associated with TSC at a rate of 30-50%.
 - Although most FCTs are benign, they may cause serious complications, such as intracardiac flow obstruction, heart valve insufficiency, rhythm disturbances, heart failure, hydrops fetalis, and even death.

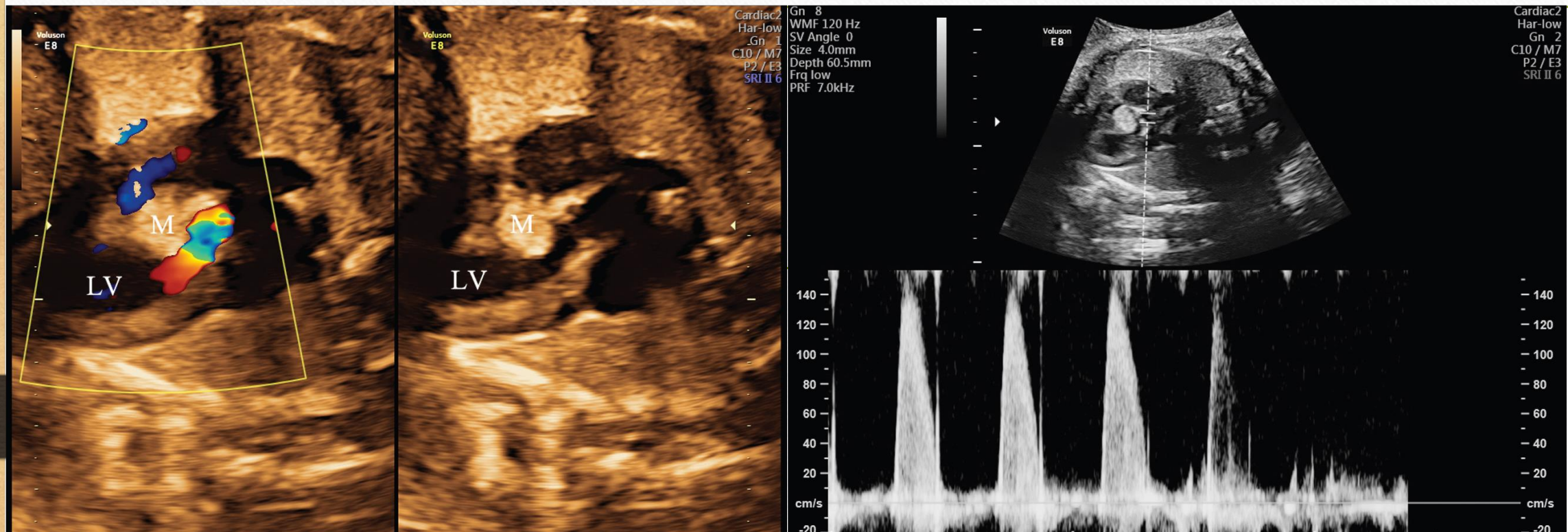
Rhabdomyoma

- Rhabdomyomas are the commonest subtypes and present as homogenous hyper echogenic masses that are generally multiple, with variable sizes.
- They may be intramural or intracavitary, and generally occupy the interventricular septum or right ventricle, close to the moderator band, but they can be located in any cardiac chamber.
- Rhabdomyomas are **hormone-dependent tumor**, which explains their well-known capacity for spontaneous regression or reduction, along with their close relationship with tuberous sclerosis.

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- Most rhabdomyomas are located in the fetal ventricular septum , but they have been present in all cardiac chambers.
 - The tumors can be clinically silent or cause hemodynamically significant obstructions, heart failure, cerebral embolization, arrhythmias, and sudden cardiac death.
 - Symptoms can be due to a variety of anatomic prerequisites including displacement, mobility, space occupation, coronary infiltration, and flow obstruction.



Fetal echocardiography (four-chamber view) at 26 weeks, showing multiple tumors (arrows) in left ventricle (LV); minimum diameter was in tumor at lateral wall of LV (approximately 3.8 mm; short arrow). Pathological examination revealed multiple rhabdomyoma



(a) Fetal echocardiography (four-chamber view) at 24 weeks, showing tumor (M) in basal segment of interventricular septum and obvious left ventricular (LV) outflow tract obstruction on color Doppler flow imaging. Narrowest diameter of basal segment of LV outflow tract, 2.8 mm. Pathological results revealed rhabdomyoma.

(b) Maximum velocity of LV outflow tract in systole was approximately 140 cm/s (normal value at 24 weeks of gestational age: 54~80 cm/s) measured by pulsed-wave Doppler.

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- Poor prognostic indicators include **development of intracardiac flow obstruction, alteration of the atrioventricular valve function with consequent regurgitation, arrhythmia, cardiac dysfunction and hydrops.**
 - Early detection of congenital tumor is critical to improving outcome.
 - The affected fetus most commonly presents on detection of the mass or masses themselves, but some will present with fetal arrhythmia or fetal hydrops.

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- The tumor tends to appear between 20 and 30 weeks' gestation, although the earliest diagnosis reported was at 17 weeks.
 - rhabdomyomas appear as nodular masses in the atrial or ventricular myocardium and usually occur at multiple loci. Most rhabdomyomas are homogeneously hyperechoic.
 - Fetal cardiac tumors can also produce arrhythmias including tachy and Brady arrhythmias as well as pre-excitation during pregnancy or following birth.

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- Rhabdomyomas can often impair the conduction system of the fetal heart and lead to heart rate disorders such as extrasystoles, supraventricular tachycardia or, less commonly, prolongation of the PR space, nonspecific alterations of ST follow-up, Wolf-Parkinson-White syndrome or aberrant atrioventricular conduction.
 - Wacker-Gussmann et al evaluated the heart rate of 10 fetuses with rhabdomyoma, by means of electrophysiology, and observed that even the asymptomatic fetuses presented conduction disorders.

Management

- Management of fetuses with diagnoses of cardiac tumors requires serial echocardiograms, and the approach will vary according to the symptoms.
- An **expectant approach** is used in cases of **asymptomatic tumors**.
- The echocardiographic evaluation in cases of **obstructions** should be very detailed, so as to be alert with regard to the presence of reverse flow in the ascending aorta and pulmonary trunk, caused by obstruction of the right and left outlets.

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- Another important sign is **increased flow in the circumflex artery**, which may nourish the tumor mass.
 - In cases of obstruction of the blood flow, the **management should be conservative** and **preterm delivery should be avoided**, except in cases in which the fetus is in the third trimester and presents severe hemodynamic disorders.
 - In these cases, delivery needs to be induced and should be done in a tertiary-level center with a multidisciplinary team, given that immediate surgical resection to relieve the flow sometimes becomes necessary.

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- In cases of **malignant arrhythmias** such as supraventricular tachycardia and atrial flutter, drug treatment with anti-arrhythmia agents should be started **immediately**, remembering that the arrhythmia may often be refractory to treatment.
 - In cases of **fetal hydrops**, with significant pericardial effusion, intrauterine pericardiocentesis may be necessary.

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- One report describes effective management of hydropic fetus with trans placental sirolimus.
 - The **mTOR inhibitor** (such as sirolimus and everolimus) successfully used to treat rhabdomyoma associated arrhythmias.

Teratoma

- The majority of teratomas arise from the pericardium and are of mixed echogenicity with cystic or calcified structures.
- Almost all are associated with pericardial effusion.
- However, it should be kept in mind that teratomas can also originate from intracardiac cavities.

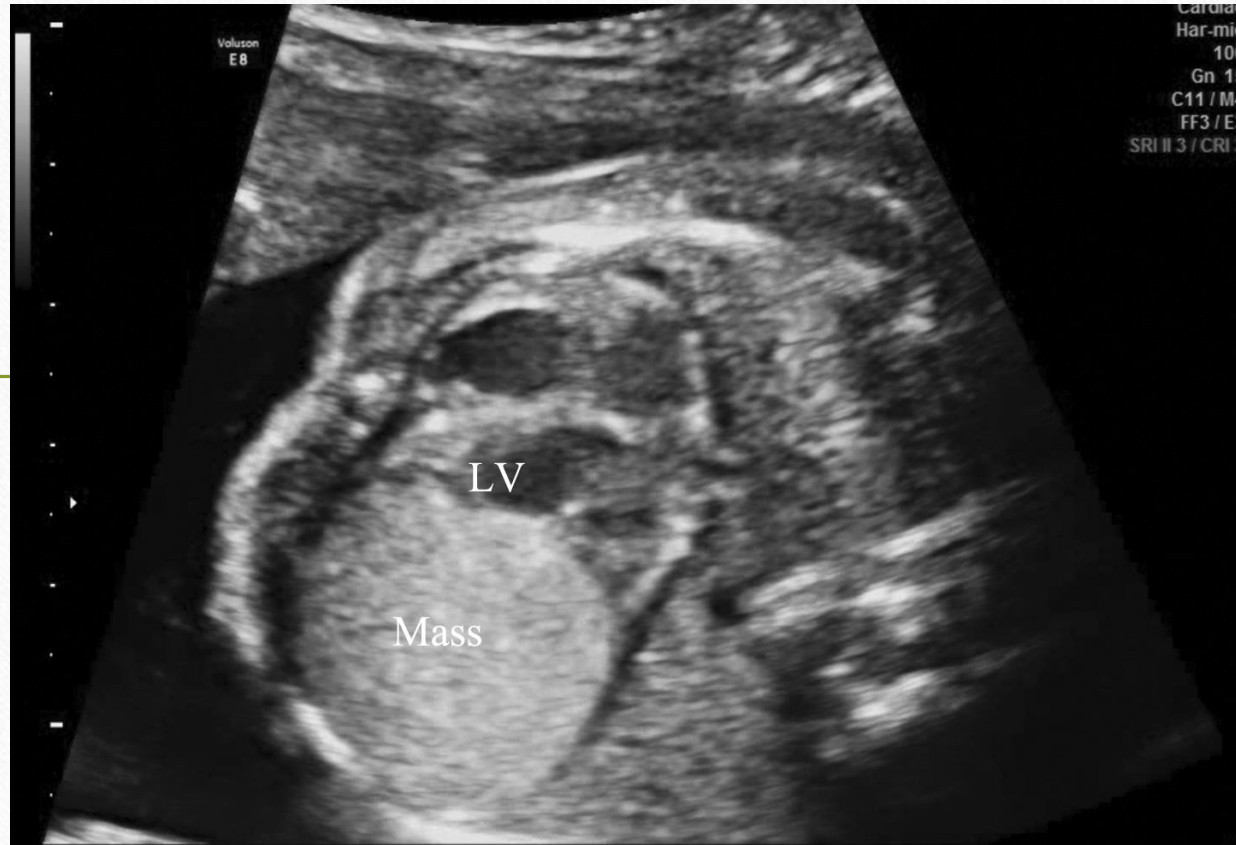
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- Intrapericardial teratomas are attached to the root of the pulmonary artery or aorta, with both solid and multicystic areas.
 - The presence of a pericardial effusion in association with a pericardial mass is almost diagnostic of a teratoma.
 - Echocardiographic assessment of cardiac tamponade is critical in the patient with a large pericardial effusion.
 - This should include 2D assessment of right atrial and right ventricular diastolic collapse, and also Doppler interrogation of respiratory variable cardiac filling and cardiac output.

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- In a literature review published by Bader et al., 31 cases of prenatal intrapericardial teratoma were identified. The diagnosis was typically suspected because of the presence of a pericardial effusion in the second or third trimester. Most tumors were located at the base of the heart. More than 75% of the fetuses developed hydrops. Fetal pericardial drainage was performed in 11 of the 31 reported cases with technical success, although reaccumulation of fluid was common
 - When intrapericardial teratoma is detected in fetal life, mortality is high.
 - Tumor size can be large and associated with pericardial effusion, the combination of which leads to progressive constraint of filling, cardiac tamponade, fetal hydrops and death.

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- Treatment of fetal intrapericardial teratoma before birth is a challenge.
 - To date, management most often includes observation with temporizing measures of pericardial fluid drainage or delivery, once the fetus reaches a viable gestational age for postnatal surgical resection.
 - Prenatal resection, if possible, would be an ideal means of treatment.
 - Open fetal surgery for resection of lung lesions is feasible with good results, when performed for impending hydrops prior to onset of severe hemodynamic compromise.

Fibroma

- The cardiac fibroma is a benign connective tissue tumor derived from fibroblasts.
- Cardiac fibromas are generally large tumors, averaging 5 cm in diameter, with the larger lesions able to obstruct outflow tracts and compress cardiac chambers.
- Their growth is typically within the myocardial mass itself, and they occur much more frequently within the anterior free wall of the left ventricle or the interventricular septum than in the posterior left ventricular wall or right ventricle.



Fetal echocardiography (four-chamber view) at 24 weeks, showing large tumor, with maximum diameter of approximately 33 mm, at apex of left ventricle (LV). Pathological examination confirmed tumor to be a single fibroma.

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- Although less common than the rhabdomyoma, fibromas are more often associated with symptoms, and more often require surgery. In contrast to the behavior of rhabdomyomas, spontaneous regression of fibromas is unusual.
 - Cardiac fibromas are solitary lesions that arise from the ventricular septum or the free wall of the left or right ventricle.
 - They often appear circumscribed, and are typically hyperechoic compared with normal myocardium. Calcification is frequent and an important diagnostic imaging feature.

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- Fibromas rarely present during fetal life.
 - They present as large single masses and, differently from rhabdomyomas, do not regress after birth.
 - For this reason, they may cause obstructions.
 - Fibromas require clinical follow-up at birth because they may lead to sudden death.
 - Because they are giant masses, resection may be difficult and **therefore heart transplantation is indicated.**

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- In the symptomatic patient, severe LVOT obstruction may be profound and can mimic hypertrophic obstructive cardiomyopathy, with limited ventricular filling and dynamic outflow obstruction.
 - the neonate who presents with severe LVOT obstruction may have ductal-dependent systemic blood flow.
 - Doppler interrogation of the transverse arch should be performed to look for retrograde ductal supply of the transverse arch.

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- Spontaneous regression of the cardiac fibroma is rare. Therefore, in the patient with symptoms, surgical resection or palliation is often considered.
 - Reports suggest that many ventricular fibromas can be completely resected with excellent early and mid-term results.
 - Cardiac transplantation is usually not required, even in those patients with massive obstructive tumors.
 - Surgical debulking and resection has been shown to cure arrhythmias in nearly all patients.

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- In summary, attention is drawn to cardiac tumors because of their diversity of forms of clinical manifestations.
 - Early diagnosis by means of fetal echocardiograms is essential, in order to evaluate the prognosis and schedule the best prenatal and delivery management, with the multidisciplinary team.