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*RESEARCH ARTICLE*

# RESPIRATORY DISTRESS REVEALING CARDIAC TUMOR IN INFANT.

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## Abstract

Primary tumors of the heart are uncommon in neonate and during childhood. Most tumors are benign, and these rhabdomyoma is the most common, followed by teratoma, fibroma, oncocytic cardiomyopathy, vascular tumors, and myxoma. Malignant and metastatic tumors are described but are rare.
They may cause complications, which are sometimes life threatening, including arrhythmias, ventricular outflow/inflow obstruction, cardiac failure, and even sudden death. (1)
Echocardiography, Computing Tomography (CT) and Magnetic Resonance Imaging (MRI) of the heart are the main non-invasive diagnostic tools , cardiac catheterisation is seldom necessary. Tumour biopsy with histological assessment remains the gold standard for confirmation of the diagnosis. Surgical resection of primary cardiac tumours should be considered to relieve symptoms and mechanical obstruction to blood flow. The outcome of surgical resection in symptomatic, non-myxomatous benign cardiac tumours is favourable. Patients with primary cardiac malignancies may benefit from palliative surgery but this approach should not be recommended for patients with metastatic cardiac tumours. Surgery, chemotherapy and radiotherapy may prolong survival. The prognosis for malignant primary cardiac tumours is generally extremely poor



## Introduction:-

Primary cardiac tumors are rare in all age groups with an incidence of 0.0017-0.027% at autopsy.(2). In infantile population, the incidence of primary cardiac tumors was reported to be 0.25%. (3)

When faced with a child suspected of cardiac neogrowth, non invasive imaging should be undertaken as the first step of assessment. We report two cases of primary cardiac tumor, its presentation, image evaluation, and final outcome, along with a review of the literature, focusing on the tools of a diagnosis and the therapy .

**Case report n°1**

A newborn infant whose apgar score at 1 min was 10 becoming 5 in the 5th min with progressive installation of respiratory distress was admitted in the neonatal intensive care unit, the clinical exam found a heart beat at 160 ,

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respiratory rate about 60 breaths per minute, 65% of oxygen saturation, signs of respiratory struggle, diffuse crackling, homogeneous hepatoglia and heart murmur .

The analysis showed: Mixed acidosis, negative infectious exams, correct ionogram, Chest X-ray provided evidence of a bilateral interstitial peribronchial thickening. Further Transthoracic echocardiogram showed the presence of several hyperechogenic tumor formations in the right and left ventricles, interesting the inter-ventricular septum as well, the largest of which causes mechanical obstruction of the left ventricle inflow , with preserved ejection fraction, dysfunctional right ventricle with significant pulmonary hypertension. The newborn was intubated , ventilated, diuretics were administated with good initial evolution, then brutal degradation occurs on day 4 causing death by cardiogenic shock.

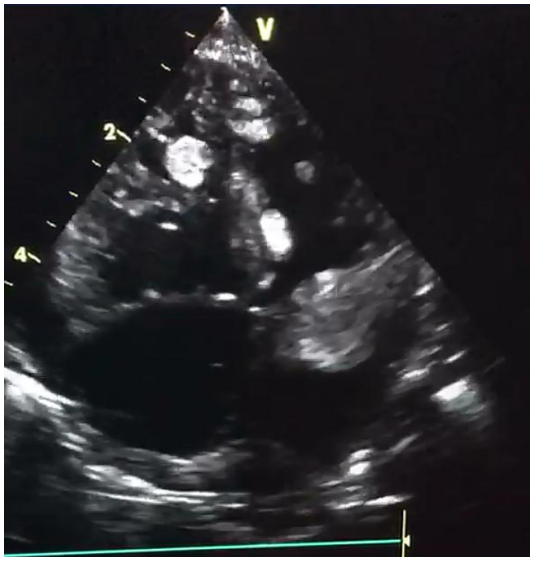


**Figure 1**:-Transthoracique Echocardiography of a newborn showing multiple rhabdomyomas found on both ventricles and in the inter-ventricular septum with obstruction of the left ventricle inflow . Rhabdomyomas appears brighter than the surrounding myocardium.( apical 4-chamber view )

**Case report n° 2**

A five-year-old , previously healthy male infant seen in the emergency department with an acute respiratory distress .The clinical examination showed an afebrile child, signs of respiratory struggle, signs of right overload and presence of a diffuse systolic murmur , the EKG showed a right bundle brunch block and a first degree auriculo-ventricular block, Transthoracic echocardiogram showed a huge mass depending on the right cavities with important pericardial effusion, in order to better characterize the mass, a CT scann was performed which confirmed the presence of a septal mass with both right intraventricular and atrial development suggesting a fibroma, primary cardiac lymphoma or myxoma. The young patient died few days after the diagnosis.

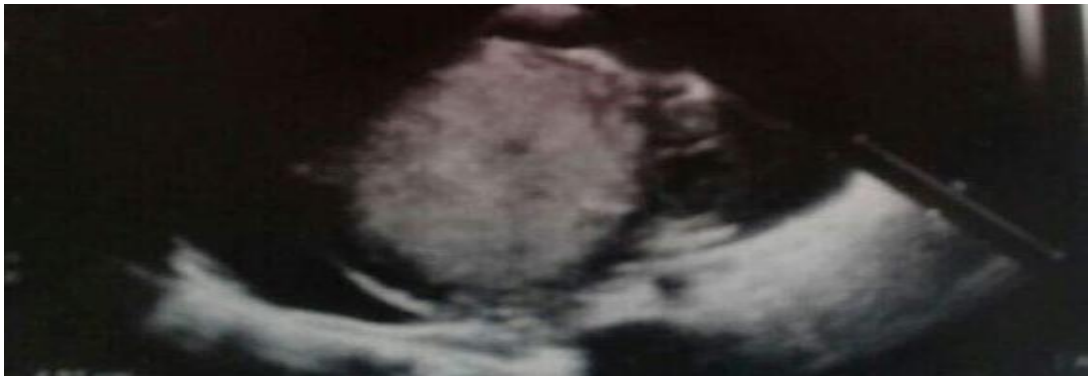

**Figure 2**:-Transthoracic echocardiogram showing a huge mass depending on the right cavities (apical 4-chamber view)

## Discussion:-

Cardiac tumors are known to originate in any part of the heart, including the myocardium, epicardium, endocardium, and pericardium.

Among these, benign tumors make up the majority, whereas 10% are malignant. The most commonly found benign pediatric cardiac tumor is rhabdomyoma, which comprises >60% of the total number of benign primary tumors. Cardiac Fibroma and cardiac teratoma are the second and third most frequently occurring tumors, respectively.(1- 4- 5-6) a third of these patients are diagnosed at <1 year of age.(6) The mean age of presentation is 13 years. (7)

**Table 1**:-Frequency Distribution of Cardiac Tumours in Children (source)

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

Source : Isaacs [8 , 9, 10 ] and Burke and Virmani [11].

**Clinical features**

The clinical manifestations of cardiac rhabdomyomas in neonates are notably variable. Symptoms are attributed to the presence of intracardiac obstruction, myocardial involvement, and rhythm disturbances . A cardiac murmur, often systolic along the left or right sternal border, may be the initial manifestation . Signs of cardiac failure may be present at birth. Since rhabdomyomas may be large or multiple, there is a potential for complex and/or multiple cardiac arrhytmias . Recurrent arryhthmias or sudden unexpected death may be the result of rhabdomyomas that interfere with the conduction system or obstruct left or right ventricular blood flow . Cardiac arrhythmia is one of the most frequent antenatal signs and is a definite indication for sonography .( 1 ,8,9,10)

**Diagnosis methods**

Noninvasive diagnostic imaging techniques have recently developed to such an extent that cardiac tumors have become amenable to effective and immediate treatment (12). However, computer tomographic scanning and magnetic resonance imaging techniques are expensive and time-consuming, which can slow down diagnosis and delay treatment.

On the other hand, TTE often allows a rapid and precise diagnosis of the intracardiac mass, giving the surgeon all the information he needs to perform a safe and complete resection of the neoplasm [13]. In addition, the peculiar echographic features of intracardiac masses can permit a reasonably reliable diagnosis of its nature.

Open surgical or endomyocardial biopsy can be used to reveal the histology of the lesion before surgical resection . A thorough metastatic check should be carried out with CT or MRI imaging before removal of malignant cardiac tumours. A bone marrow biopsy or bone scan may be necessary.

## Treatment

### Medical treatment

Cardiac rhabdomyoma the most common cardiac tumor diagnosed in neonates and infants is known to have a spontaneous regression in most cases, a case report( 14 ) of a critically ill neonate, resuscitated after cardiac arrest secondary to massive locally invasive cardiac rhabdomyoma, who was successfully treated with everolimus (mammalian target of rapamycin [mTOR] inhibitor). Rapid tumor regression was observed on echocardiography, whereas this evolution may be confounded by the natural disease course of regression, this case suggests that mTOR inhibitors may play a significant role in the treatment of large cardiac rhabdomyomas in critically ill neonates.

### Resection

Neonates and infants with cardiac tumors were considered for surgical intervention based on the severity of symptoms. In general, indications for resection were hemodynamic/respiratory compromise, severe arrhythmia, and a significant embolization risk. Approach to and strategy for tumor resection varied according to location and hemodynamic status ,without damage to adjacent structures.

Surgical resection remains the treatment of choice despite its high mortality rate and relatively poor late outcome in patients with large masses. However, some benefit arising from subtotal resection, staged surgery, or palliative shunting has been reported. (15)

Primary cardiac malignancies may benefit from palliative surgery but this approach cannot be recommended for patients with metastatic cardiac tumours. Surgery, chemotherapy and radiotherapy may prolong survival. The prognosis for malignant primary cardiac tumours is generally extremely poor. Transplantation If the resection of the tumour is not achievable in severely symptomatic patients, orthotopic cardiac transplantation may be considered. This treatment modality is only an option for patients with unresectable cardiac tumours with no evidence of metastatic involvement of the heart.

## Conclusion:-

cardiac tumor in infant are rare and most cases are benign, but they may cause a series of cardiovascular events, such as intracardiac flow obstruction, heart valve insufficiency, arrhythmia, heart failure, and hydrops fetalis, or even fetal death.

Today, TTE can provide accurate anatomic information about the tumor location, extent and characteristics, helping to provide a clear and timely surgical plan. Everolimus treatment (mammalian target of rapamycin [mTOR] inhibitor) should be considered in fetal cardiac rhabdomyomas .

Surgical resection remains the treatment of choice despite its high mortality rate and relatively poor late outcome .

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