



## REVIEW

# Epidemiology and outcome of primary cardiac tumours prenatally, in neonates and children: A single center experience from tunis

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## 1. Introduction

During the past decade the number of cardiac masses detected in paediatric population has increased significantly because of the widespread use of non invasive imaging techniques. Echocardiographic diagnosis of tumours can be made in antenatal period.

The Aim of the study was to determine the incidence of primary cardiac tumours in our tunisian pediatric population (from fetus to childhood), emphasizing on echocardiographic findings and outcome.

## 2. Methods

It was a retrospective, descriptive study; that included 27 patients diagnosed with primary cardiac tumours hospitalized in the cardiopediatric department of the Rabta hospital between January 2000 and January 2017.

Only Patients under 15 years of age were included.

The pseudo tumor including hydatid cysts and thrombi were excluded.

Clinical features of postnatal examinations were documented from records and echocardiographic images reviewed from the computer database. Data included age at diagnosis, clinical presentation, physical examination findings (cyanosis, heart murmur, arrhythmia, heart failure), electrocardiogram (ECG) results, initial and last echocardiography findings (number of rhabdomyomas, location, myocardial dysfunction), indication for surgery, and outcome.

Babies were subjected to cardiac follow-up with clinical evolution, color Doppler echocardiography and Holter-ECG.

The diagnosis was based on echocardiographic findings, magnetic imaging resonance (MRI) data only in case of inconclusive echocardiography and on histological evidence in operated patients.

## 3. Statistical analysis

Quantitative variables are expressed as means  $\pm$  standard deviations. Qualitative variables are expressed as percentages.

**Résultats** :the main results of our study were reported in Table 1.

Twenty seven cases of primary cardiac tumours were diagnosed: Rhabdomyomas (n = 17), Fibromas (n = 3), Teratomas (n = 3), Myxoma (n = 2), calcified amorphous tumor (n = 1) and

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**Table 1**  
The main characteristics of our population.

	Patients (n)%
<i>Type of tumors</i>	
Rhabdomyomas	17
Fibromas	3
Teratomas	3
Myxoma, calcified amorphous tumor unknown origin	2
	1
	1
<i>Age at diagnosis</i>	
Antenatal	11
Neonatal < 1 years	10
1–11 years	6
<i>Clinical Presentation</i>	
Incidentally	8
Cardiac murmur	5
Cyanosis	3
Heart failure	7
Arrhythmias	2
Syncope	2
<i>Tumor side</i>	
Pericardium	3
Left atrium	2
Left ventricle (LV)	15
LV + right ventricle	7
<i>Outcome of tumors</i>	
Death	6
Surgery	9
Partial regression	3
Total regression	5
Stable size	4

tumor of unknown origin (n = 1). Eleven primary cardiac tumors were diagnosed prenatally between 22 and 36 weeks of gestation with a mean of  $28 \pm 3.5$ . Ten cases were detected in the neonatal period, within the first year of life and 6 cases between the age of 1 and 11 years old. There were 15 females (55%) and 12 (44%) males.

Fetal tumors were monitored until birth. Echocardiographic follow up showed ventricular rhabdomyomas (n = 3 cases) that grew in size (Fig. 1) then regressed after the 32nd week of gestation, an intrapericardial tumors (n = 3cases) with median size

of 41x34mm evoking teratomas (Fig. 2) and a stable sized tumors in 5 fetuses. All fetuses were born in full term and then transferred to the cardiology department, where the initial diagnosis was confirmed.

Postnatal circumstances of discovery were diverse. Tumors were discovered incidentally (n = 5) because of signs of heart failure (n = 7), due to cardiac murmur (n = 8), due to cyanosis (n = 3), due to arrhythmia (n = 2) and after a syncope (n = 2).

Rhabdomyomas account for 63% of cardiac tumors (n = 17), they were predominant in fetuses and neonates but rare in child with respectively (n = 9, n = 7 and n = 1). Tuberous sclerosis (TS) was associated with in 9 patients (53%). These tumors were diagnosed based on echocardiographic findings (multiples mass) and the correlation with TS. Four rhabdomyomas cases in neonates died before surgery because of acute heart failure due to obstruction (Fig. 3).

A Total tumor resection was performed in 3 rhabdomyomas (Table 2). The absence of significant obstruction or resistant arrhythmias.

Over time with a mean follow-up of 92.2 months, a complete or partial cardiac rhabdomyoma regression was observed in respectively 5 and 3 cases, 2 rhabdomyomas remained stable in size.

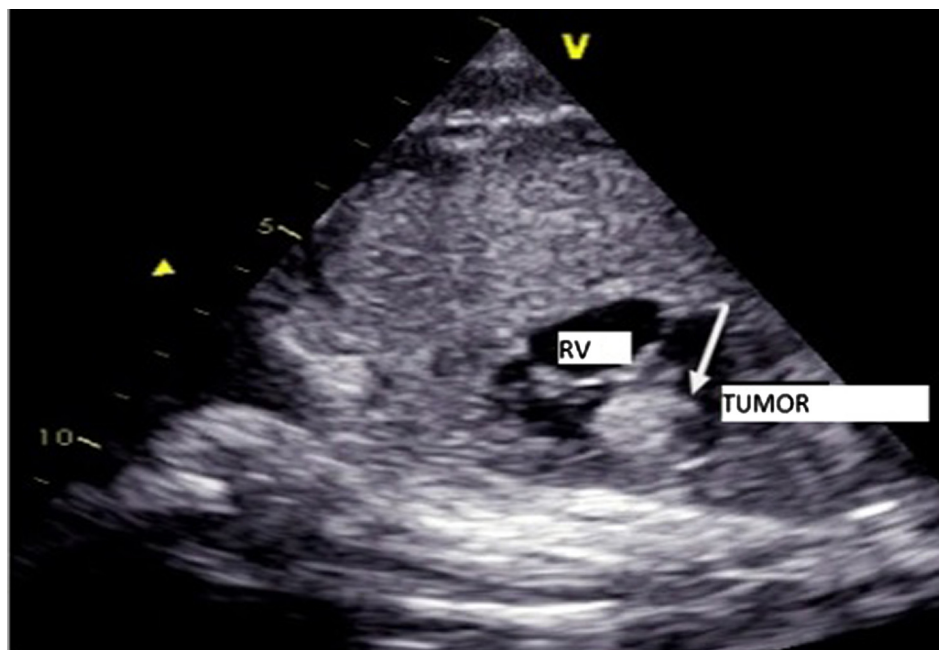
Patients with TS had mental deficiency of variable degrees. Patients with seizures received anticonvulsive treatment and were followed-up with by pediatric neurologists.

Two teratomas were confirmed by echocardiography and MRI (pericardium localisation), these tumors caused compression leading to death before surgery intervention. One teratoma was operated in the presence of compression signs (Table 2).

Two neonates with respectively a fibroma tumor and tumor of unknown origin did not require surgery intervention because of absence of obstruction signs. Over the time there was an increasing rate of surgical intervention. All characteristics of operated tumors were presented in the Table 2.

#### 4. Discussion

Cardiac tumors rare in paediatric practice with a prevalence of 0.0017 to 0.28 in autopsy series (1), whereas their incidence during foetal life has been reported to be approximately 0.14%.<sup>1</sup>



**Fig. 1.** Echocardiography showed and antenatal rhabdomyoma.

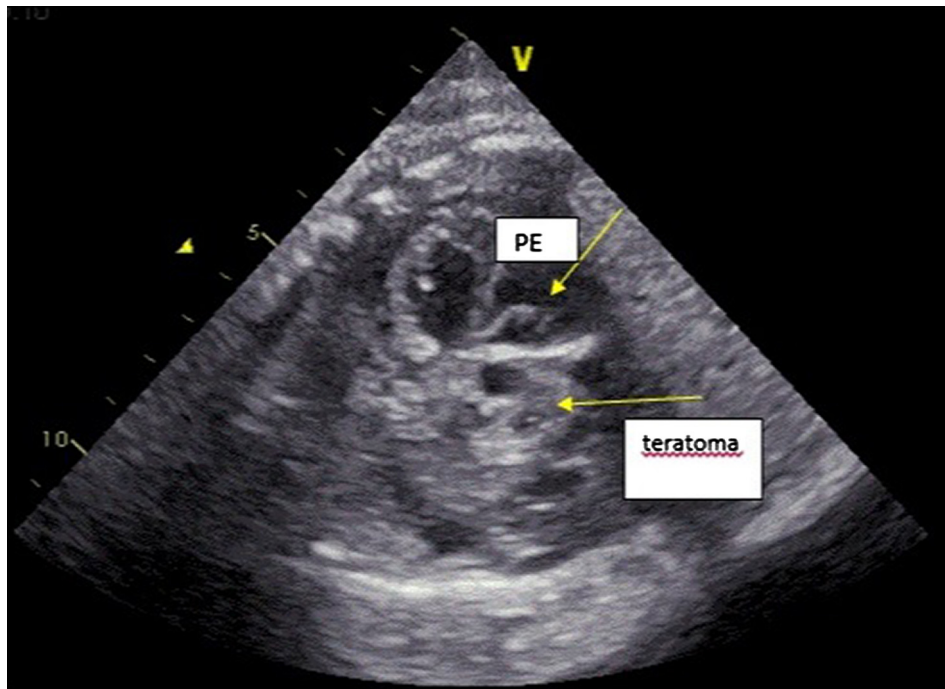


Fig. 2. Antenatal teratoma PE:pericardium effusion.

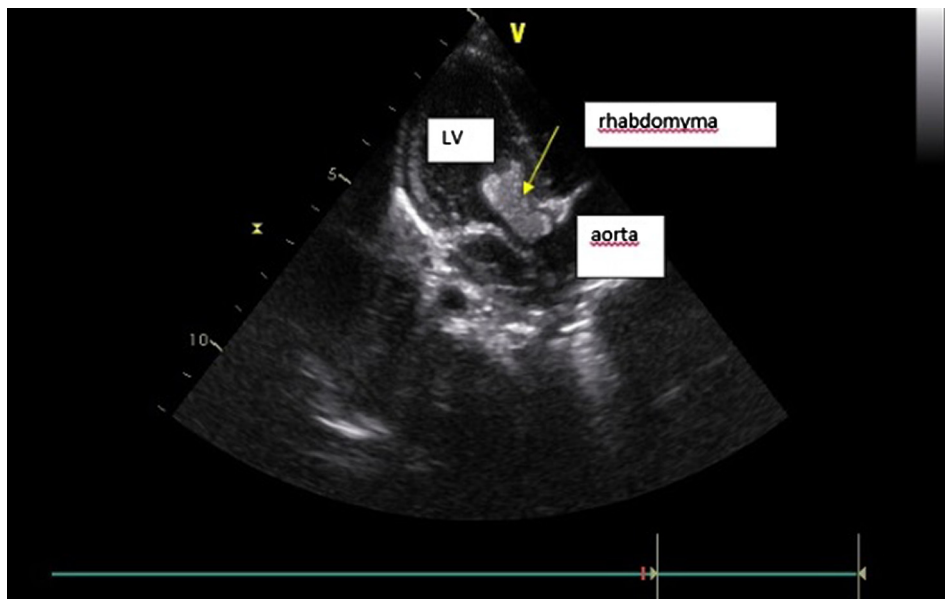


Fig. 3. Rhabdomyoma with aortic obstruction.

The frequency and type of cardiac tumours in children are different from those in adults.

The majority of primary cardiac tumours in children are benign.<sup>2,3</sup>

Clinical manifestations of cardiac tumors vary widely from asymptomatic presentations to life-threatening cardiac events. Similar results were reported in our study.

Antenatal diagnosis has improved during the past decade, Fetal cardiac tumours develop between 20 and 30 weeks of gestation, therefore a second trimester fetal anomaly does not completely rule out a cardiac tumour. We also reported 11 cases of antenatal tumors diagnosed at a medium age of  $28 \pm 3.5$  weeks of gestation.<sup>4,5</sup>

Rhabdomyoma is the most common cardiac tumor diagnosed in childhood and represents more than 60% of all cardiac tumors.<sup>1</sup> The same frequency was reported in our study (63%) and 50% among our patients suffered from TS].

Irrespective of size Rhabdomyomas spontaneously diminish or disappear completely.<sup>6,7</sup> That is why surgery is necessary only for children with life-threatening hemodynamic abnormalities or arrhythmias. Similarly in our study, rhabdomyomas were managed conservatively. In fact only 3 cases underwent surgery in the presence of complications. Late course was marked by parietal and complete tumor regression of the majority of rhabdomyomas cases.

**Table 2**  
Characteristics of operated patients.

	Age (years)	Clinical presentation	Tumors characteristics	Surgery and outcome
Patient 1	21	Pericardium Compression signs	<b>Teratoma</b> (intrapericardial compresses right atrium and ventricle)	Total excision Doing well, no recurrence
Patient 2	3	Heart failure	<b>Fibroma sitting in</b>  the ventricular septum obstructed the left and right outflow tract with maximum pressure gradient of 100 mmhg	Excision of tumor and subaortic obstruction, residual gradient of right outflow. No recurrence
Patient 3	11	Syncope	<b>Myxoma of left atrium</b>	Total excision, Doing well, no recurrence
Patient 4	8	syncope	<b>Myxoma</b> of left atrium obstructed mitral valve	Total excision, recurrence, reoperated
Patient 5	9	Right Heart failure	<b>Fibroma</b> localised on right ventricle	Inextirpable tumor, then ventricular tachycardia reduced and then lost view
Patient 6	5	Right heart failure	<b>Calcified Amorphous tumor</b> Multiple tumors extended to pulmonary infundibulum	Excision, tumour then recurrence with acute pulmonary embolism, death
Patient 7	5 days	Heart failure	<b>Rhabdomyomas</b> ventricular that obstruct aortic outflow	Total excision, doing well No Recurrence
Patient 8	10 days	Heart failure	<b>Rhabdomyoma</b> ventricular that obstruct aortic outflow	Total excision, doing well no recurrence
Patient 9	1 months	Résistant ventricular arrhythmias	<b>Multiples Rhabdomyomas</b>	Total excision, no recurrence, remains in sinus rhythm

Others histopathologically confirmed tumours in our series were fibroma, teratoma and myxoma. Fibroma is the second most common cardiac neoplasm in paediatric population.<sup>8</sup> It is located in the right heart and leads often to right-sided failure symptoms. Spontaneous regression of fibroma has not been reported. Therefore, it is usually advisable (if feasible) to excise the tumour as early as possible, because it tended to grow fast and cause severe hemodynamic complications.<sup>9,10</sup> This constation was reported in our fibromas cases so that surgery was indicated.

The third most frequent type of primary tumours in children is teratoma. It is usually single and may reach a considerable size, sometimes even larger than an infant's heart. It is usually located in the pericardium<sup>11,12</sup> and causes pericardial effusion. Our three teratomas cases corroborated with the data found in the literature.

In our study among histopathologically confirmed cases we reported two cases of myxomas, which developed in children patients. According to literature<sup>13,14</sup> this tumor represent more than 10–15% of cardiac tumours in children and were rare in fetuses and neonates. Surgery was recommended in order to prevents the development of the dangerous complications (obstruction, embolism).

## 5. Limitations

The main limitations of our study are its retrospective nature and its small population size, both of which are due to the scarcity of infant cardiac tumor cases. Therefore, a multi-centric study is needed to corroborate our conclusions.

## 6. Conclusion

Our study highlighted that early fetal detection of cardiac tumours is possible at early age of pregnancy, rhabdomyoma was the most commonly encountered tumour in our paediatric population and reported a favorable course of the majority of cases with a low morbidity. That's why a monitoring becomes mandatory since

hemodynamic obstruction and severe arrhythmias could occur and require a surgical treatment.

## Conflict of interest

No conflict of interest.

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