

## PRENATAL DIAGNOSIS OF THE RIGHT VENTRICLE TUMOR - CASE REPORT



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**Abstract**

The study case is a foetus with a diagnosed heart tumor in week 23 of pregnancy. The tumor was found in the right ventricle with a concomitant local change of echogenicity of the right atrial wall; it did not cause any haemodynamic disturbances, impair systolic function or disturb inflow into the right ventricle. Foetal check-up in week 30 has shown a complete regression of the lesion in the right ventricular lumen. Because of no foetal movements felt, in week 36 the pregnant patient reported to the hospital where foetal heart asystole was diagnosed and the pregnancy was terminated by labour induction. Foetal autopsy has shown no nodose lesions in the heart or any other lesions which could have been the cause of an abrupt foetal death in utero.

**Key words:** fetal cardiac tumor, fetal echocardiography

**INTRODUCTION**

Heart tumours are rare pathological lesions observed in prenatal examinations<sup>1</sup>. Their incidence is from 0.001 to 0.28%, in the foetal period it is usually estimated at approximately 0.14%<sup>2</sup>.

90% are benign lesions. In the foetal and neonatal period, the most common histopathological type of lesions are rhabdomyomas, while teratomas, fibromas or haemangiomas are much more rare<sup>2</sup>. Disorders observed in foetuses and neonates with heart tumours largely depend on the location and size of the lesion, and include arrhythmias, heart failures due to inflow or outflow disorders, valve regurgitation, cardiac muscle contractility disorders, hydrops foetalis<sup>3</sup>. They can also lead to premature birth as well as cause intrauterine foetal death<sup>2,4,5,6</sup>.

Heart tumour diagnostics, both in the foetal and in the neonatal period, is based on ultrasound examinations. If the tumour is discovered early (around week 18 of pregnancy) and decision is made to continue the pregnancy, haemodynamic condition of the foetus and tumour(s) growth rate must be monitored<sup>7</sup>. Usually, such lesions are diagnosed in foetal hearts in the third

trimester of pregnancy<sup>2</sup>. Benign lesions (in particular rhabdomyoma) frequently tend to spontaneously regress in the foetal and neonatal-infant period, showing good natural course, in particular in asymptomatic cases which do not cause haemodynamic disorders<sup>2,8,9</sup>. Surgical treatment of primary heart tumours is considered in symptomatic neonates if it is possible to remove the lesions causing significant blood flow disorders or impairing cardiac valve function<sup>2,4</sup>. Continuous monitoring of neonates is necessary in each case due to high incidence of combination of tuberous sclerosis with benign heart tumours<sup>2,4</sup>.

Case study presentation

21-year-old primigest woman, with no complications, came for a routine foetal examination in week 23 of pregnancy. The 2<sup>nd</sup> genetic assay (the 1<sup>st</sup> was not performed for this patient) found a single female foetus with correct build, and placenta on the posterior uterine wall. Foetal organ examination revealed correct heart size (HA/CA – 0.33), four cardiac cavities, two symmetrical atria, patent oval opening, two symmetrical, morphologically diversified ventricles with continuous intraventricular septum (Figure 1).

With adverse position of the foetus, basal projection of the heart found a nodose lesion of 8mm in diameter,

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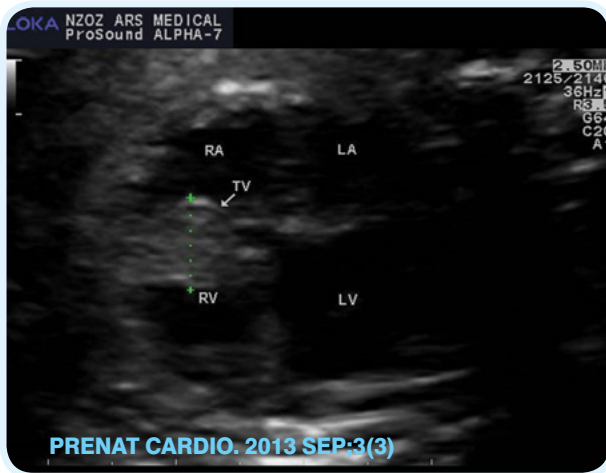


Figure 1. Echocardiographic image of four cardiac cavities in basal projection (2D) with atria and ventricles. Visible tumor projecting onto the ring of the tricuspid valve; imaging performed in pregnancy week 23 (Photo by ARS Medical)

projecting to the outline of the tricuspid valve (Figures 1-3). The lesion was not accompanied by haemodynamic disorders – the right and left ventricle without inflow disorders, with normal systolic function, without blood outflow restriction, with normal setting and flow through large arteries. The patient was referred to the Department of Diagnosis and Prophylaxis of Congenital Malformations of the Polish Mother's Memorial Hospital – Research Institute (ZDiPWW ICZMP) for verification of the diagnosis and further diagnostic tests. The first examination at the ZDiPWW ICZMP (in apical projection) confirmed presence of a tumour sized 10 x 9mm, located in the right ventricle right under the tricuspid valve (Figures 2, 3). The tumour caused no haemodynamic disorders, was accompanied by disturbed echogenicity of the right atrial wall and a nodosity in the tricuspid valve ring region. Follow-up examination performed after three weeks (week 30 of pregnancy), foetal biometry was found to be normal for 30-31 HBD, with no abnormalities in general build of the foetus, normal heart build, total regression of the tumour

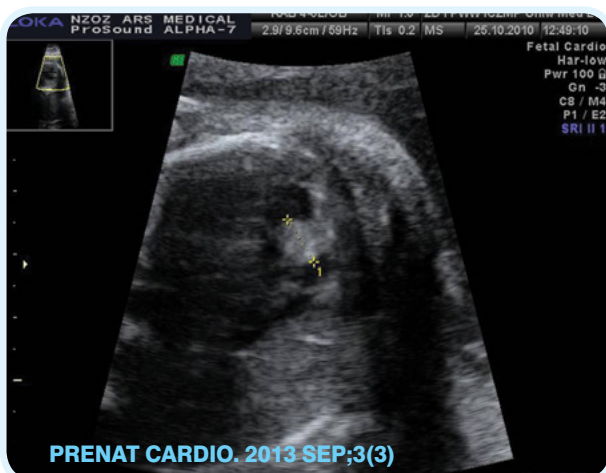


Figure 2. Echocardiographic image of the heart (2D) in pregnancy week 27; visible mass of the tumor in the lumen of right ventricle under the tricuspid valve (Photo by Department for Diagnosing and Prevention of Congenital Malformations at the Polish Mother's Memorial Research Institute)

in the right ventricle, with atypically nodose tricuspid valve ring and increased AFI (amniotic fluid index) – 25.

In week 36 of pregnancy, the patient called at the hospital because she felt no foetal movements. Foetal heart asystolia was diagnosed and the pregnancy was terminated by induced labour. Foetal autopsy failed to reveal any lesions in the right atrium or right ventricle, or to indicate any other cause of intrauterine foetal death.

## DISCUSSION

In the presented case, the tendency for lesion regression as well as the presence of potential additional foci in the right atrial wall suggest a benign histopathological type of the primary lesion. As no cause of intrauterine foetal death, maternal causes or lesions in foetal autopsy and afterbirth examination were found, an incidental coincidence of heart tumour diagnosis and subsequent intrauterine foetal death is possible. References, however, report the possibility of intrauterine foetal death in spite of benignity of nodose lesions in histopathological examination, lack of haemodynamic disorders and close monitoring of the pregnant patient<sup>9</sup>. Symptomatic factors increasing the risk of intrauterine foetal death include early onset of the lesion(s) in the heart, its histopathological type, large size and hydrops foetalis<sup>10</sup>. The above-mentioned comorbid symptoms apply to cases in which the diagnosed heart tumour was observed as a permanent lesion. This, however, does not rule out the fact that factors unrelated with the histopathological type of the lesion, tumour mass and location as genetic and metabolic disorders, combined with origination of primary foetal heart neoplasms may also adversely affect the course of pregnancy regardless of the initial regression of the heart lesion.

Qualification of foetuses with prenatally diagnosed heart tumours for natural birth or Caesarean operation depends on the foetus's competence, foetal cardiac

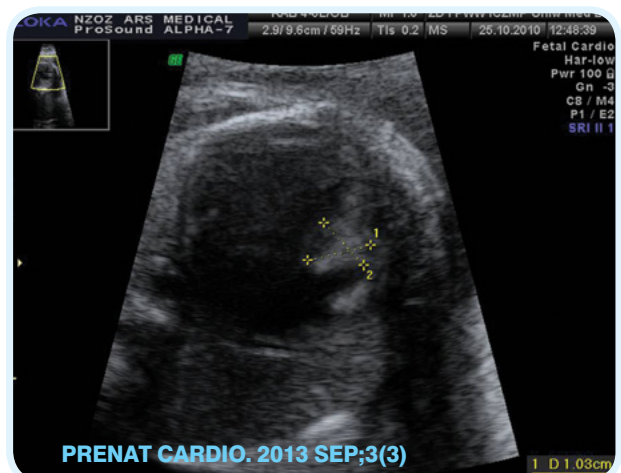


Figure 3. Echocardiographic image of the heart (2D) in pregnancy week 27; visible mass of the tumor in the lumen of right ventricle and abnormal echoes in the area of right atrial wall and the tricuspid valve (Photo by Department for Diagnosing and Prevention of Congenital Malformations at the Polish Mother's Memorial Research Institute)

rhythm, possibility of postnatal cardiosurgical treatment and attitude of the baby's parents<sup>7</sup>.

In the following pregnancy of the patient (if the previous one ended in giving birth to a baby with a heart tumour), it is recommended to perform the first ECG of the foetal heart in weeks 13 and 18, and every 4 weeks afterwards.

Each pregnant patient with diagnosed foetal heart tumours requires regular monitoring at a referral centre<sup>7</sup>.

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