# CARDIAC RHABDOMYOMA WITH SEVERE RIGHT VENTRICULAR OUTFLOW TRACT OBSTRUCTION PALLIATED WITH DUCTAL STENTING – A CASE REPORT

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## ABSTRACT

We present this case of cardiac rhabdomyoma in a term baby presenting with peripheral and central cyanosis. On echocardiography multiple masses were noted occupying the right ventricle and the adjacent myocardium. There was evidence of right ventricle outflow tract obstruction causing functional pulmonary atresia. A small patent ductus arteriosus was seen supplying the lungs. After instituting supportive treatment and consulting the cardiac surgical team palliation was decided. Balloon atrial septostomy and ductal stenting was done on 7th day of life.

**Keywords**: Balloon atrial septostomy, Patent ductus arteriosus, stenting, Rhabdomyoma.



## INTRODUCTION

In paediatric practice primary cardiac tumours are rare having a prevalence of 0.0017-0.28 at autopsy, 0.02-0.08% in live-born infants, and 0.12% in prenatal studies[1,2]. Rhabdomyoma being the most common primary cardiac tumor in children, accounts for up to 60% of all primary cardiac tumors[3]. Although the nature and histology of the mass is benign, owing to the location and involvement of the cardiac chambers, impingement on valves and restriction to blood flow in diastole can cause serious and life threatening hemodynamic compromise. In addition, serious arrhythmias, as a result of involvement of the conduction system can lead to sudden death[4].

We are reporting a case of a term baby girl who was diagnosed with cardiac rhabdomyoma on the 5th day of life on echocardiography and was subsequently palliated with patent ductus arteriosus (PDA) stenting and balloon atrial septostomy (BAS).

## CASE REPORT

A term baby girl born to consanguineous parents at 39 weeks of gestation with a birth weight of 2.9kg presented to the local pediatrician with complaints of central and peripheral cyanosis. After being referred to the cardiology outpatient department, she was found to have central and peripheral cyanosis, nasal flaring, tachypnea with a respiratory rate of 60/min, heart rate 110/min and oxygen saturation of 83% in air. Chest radiograph showed mild cardiomegaly. Baseline electrocardiography (ECG) was normal. Echocardiography revealed multiple masses occupying the right ventricular intracavitary space and myocardium, the largest measuring 18mm x 16mm. The tricuspid valve could not be visualized and there was minimal flow through the valve. The right ventricle outflow tract (RVOT) was also involved causing functional pulmonary atresia. There was a small patent ductus arteriosus (PDA) measuring 3 mm supplying the lungs (fig-1).

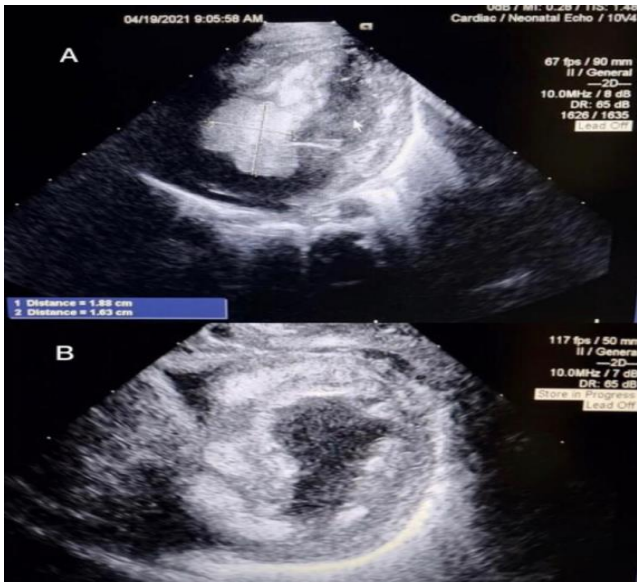


**Figure-1A: Echocardiography images of cardiac rhabdomyoma. Four chamber view showing a portion of the mass measuring 18mm x16mm, occupying almost all of the right ventricle (RV). Figure-1B: Parasternal short axis view at the mid left ventricle (LV) level showing the myocardium studded with multiple masses.**

The baby was admitted and nasal prong oxygen, antibiotics along with oral prostaglandin started. Cardiac surgical team was consulted and after reaching a consensus, palliation was decided because of the extensive involvement of the myocardium. The baby was palliated on day 7 of life with balloon atrial

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septostomy and PDA stenting to improve blood supply to the lungs (fig-2).

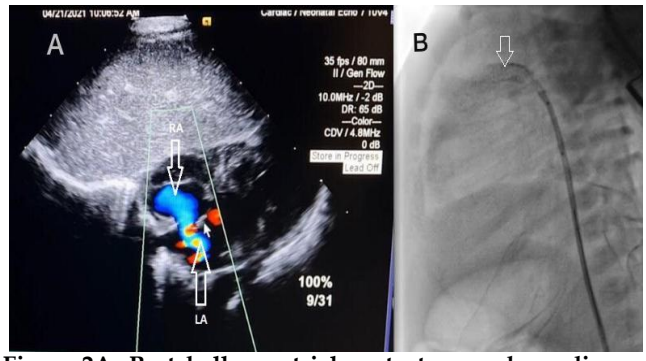


**Figure-2A: Post balloon atrial septostomy echocardiography, subcostal coronal view showing adequate septostomy with good flow, shunting right to left (arrows indicating RA and LA) with cursor pointing at the inter atrial septum. Figure-2B: Aortogram in left anterior oblique (LAO) 90º showing the PDA stent (arrow) in place.**

After the procedure the oxygen saturation improved and baby was successfully shifted back to post catheterization care and subsequently discharged the following day with a plan to closely follow the case and look for mass regression on future visits.

## DISCUSSION

Being the most frequently diagnosed cardiac tumour in utero and in early childhood, cardiac rhabdomyoma (CR) accounts for 45–80% of all primary cardiac tumors[5]. It is a benign hamartomas with a strong association with tuberous sclerosis where as many as about 50% of patients with rhabdomyoma have tuberous sclerosis. The usual location is ventricles though atrial rhabdomyomas may also occur. The atrioventricular junction rhabdomyoma may lead to ECG abnormalities.

Association between multiple CR and tuberous sclerosis has been described with the incidence of tuberous sclerosis in patients with CR between 60-80%[6]. Conversely 43-72% of patients with tuberous sclerosis also have CR[7]. Therefore a careful search of the heart chambers should be made if a solitary tumour is diagnosed so that smaller lesions are not missed.

Many CR are diagnosed on routine antenatal sonography where multiple intracardiac masses can be seen at anomaly scan carried out around 20 weeks[8]. The in-utero presentation also includes non-immune foetal hydrops and foetal death. Postnatal presentation depends on the size, location and the level of obstruction caused by the tumour and includes murmur, weak peripheral pulses, cyanosis, arrhythmias and heart failure[9].

Echocardiography shows rhabdomyomas to be a solid echogenic mass associated with the ventricular myocardium septum which may protrude into and deform the cardiac chambers. Diffuse myocardial thickening can also be seen in case of multiple small lesions. Magnetic resonance imaging shows the same features and is particularly helpful when surgical resection is under consideration.

Because the majority of cardiac rhabdomyomas regress spontaneously, surgery is not routinely required[10]. An 11.8 times quicker regression without any serious side effects has been reported by Aw *et al,* in CR treated with low dose of everolimus, (4.5 mg/m$^2$/ week in daily divided doses), compared to natural regression[11]. Similar effect was observed by Chang *et al*, in CR associated with tuberous sclerosis[12]. Cases of CR with obstruction of either right or left outflow tracts or refractory arrhythmias need surgical intervention where surgical excision may be considered[13]. In cases where complete excision is not possible due to the location and size of the tumour, like the one we encountered, palliative measures like insertion of PDA stent and balloon atrial septostomy are considered[14]. The maternal use of oral sirolimus for noninvasive treatment of the antenatally diagnosed foetal cardiac rhabdomyoma has also been described[15].

## CONCLUSION

In cases where the CR is inoperable, the patient can be palliated accordingly to buy time and wait for tumor regression.

## CONFLICT OF INTEREST

This study has no conflict of interest to be declared by any author.

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