# Hypertrophic Cardiomyopathy Involving Right Ventricle-Clinical, Hemodynamic And Angiographic Findings

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**Summary:**

**A case of fifteen months old child presented with clinical picture suggestive of RVOT obstruction was diagnosed to have HCM involving Right Ventricle. The child also had associated unusual congenital anomalies like cleft palate and congenital bilateral talipes equinovarus.**

**Introduction:**

Hypertrophic Cardiomyopathy (HCM) involves ventricular septum and frequently left & right ventricular free walls. Fifteen per cent patients with typical clinical and hemodynamic features of HCM show systolic gradients across right ventricular outflow tract. It is rare to find right ventricular involvement in absence of pressure gradient across left ventricular outflow tract. The case reported here has unusual features as it is presented at very early age of 8 months; associated with unusual anomalies like cleft palate, bilateral talipes equinovarus and has documented isolated RVOT gradient in absence of LVOT gradient.

**Case Report:**

The patient S.S., 15 months old female child presented with complaints of failure to thrive and dyspnea on excessive crying and feeding since age of 7 months. There was no significant history if form of repeated lower respiratory tract infections, cyanosis, cyanotic spells, convulsions. The child was full term caesarian section delivery of non-diabetic mother. There was no pre/intra/post natal complications. Her physical mile stones were delayed, however mental mile stones were normal. Her elder brother (4 years of age) was normal on history and clinical examination.

On physical examination, the child was thin (80th percentile for the age). Pulse rate was 110/min, regular and normal in character. BP was 90/70mm of Hg in right upper extremity. There was no cyanosis or clubbing. Mean JVP was normal and there was prominent a wave (7 cms from sternal angle) on examination of jugular venous pulse. There were other congenital anomalies in form of cleft palate and bilateral talipes equinovarus. Apex impulse was normally located. S1 was normal. S2 was normally split and was of normal intensity. There were no additional sounds. There was grade 3/6 ejection systolic murmur in 2nd left intercostal space radiated to 3rd intercostal space and left sternal border. Other systemic examination was normal.

The ECG showed right axis deviation (+170), abnormal Q waves in II, III, aVF; rS pattern in chest leads without any evidence of ventricular enlargement.

2 D Echocardiography and M mode examination revealed asymmetrical septal hypertrophy (ASH) (IVS thickness 9mm); LVPW thickness of 3 mm with

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IVS:LVPW of 3:1; moderate systolic anterior motion of anterior mitral leaflet; hypertrophied RV. LV free wall was normal and calculated EF was 88% (Fig. 1 showing SAM of AML).

Doppler examination showed peak systolic gradient of 64 mm of Hg across RVOT. There was no L to R shunt across patent foramen ovale.

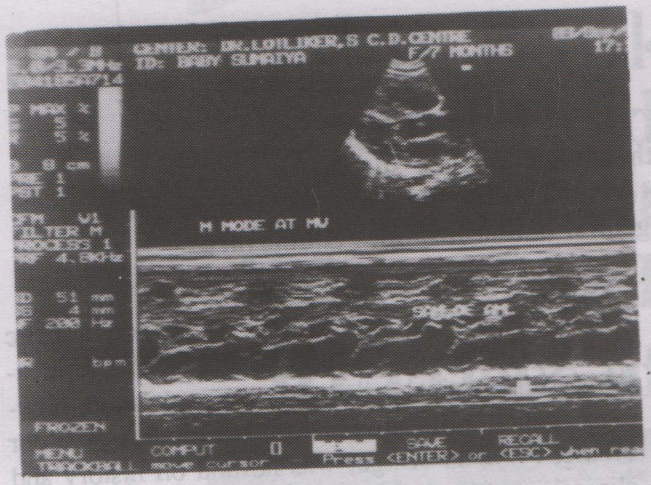


Fig. 1
Showing Systolic anterior motion of AML on M mode echocardiography.

Cardiac catheterization was done using femoral as access to both right & left heart. There was no intracardiac shunt. There was peak systolic gradient of 34 mm of Hg at RVOT and RV body junction (systolic pressure of 27 and 61 mm of Hg respectively). MPA pressure was normal. RA pressure showed prominent a wave of 12 mm of Hg. LV pressure was 90 mm of Hg with cuff systolic, brachial artery pressure of 90 mm of Hg. There was no LVOT gradient in basal state. No manoeuvres were done to provoke LV gradient.

RV angiogram in lateral view revealed obstruction at junction of RVOT and RV body. LV angiogram revealed hypertrophied papillary muscle and near obliteration of LV cavity during systole (Fig. 2).

**Discussion:**

RVOT obstruction in case of HCM can be due to protrusion of hypertrophied septum (Brei et al 1960; Soulie, Joly, Carlotti 1962); hypertrophy of free wall (Liversay, Wagner and Ambrust 1960; Daoud, Gallaner and Kaplan 1961); hypertrophy of septal and parietal parts of cristae supraventricularis (Braunwald 1964). The predominant site of obstruction is at junction of RVOT and RV body. This patient demonstrated mild RV gradient at same level. This was not associated with any gradient across LVOT in basal state. The provocative interventions were not done in view of very young age. The dynamic nature of obstruction is demonstrated by cine angiographic studies (Cohen et al 1964, Steiner 1964, Oakley 1964); by necropsy specimen and by surgical findings (Taylor et al 1964). Double chambered Right ventricle is an important differential diagnosis. However, DCRV shows constant filling defect at site of obstruction on angiography. The asymmetrical septal hypertrophy, elongation of RV cavity and low infundibular stenosis suggest HCM. DCRV is commonly associated with other anomalies like VSD/ pulmonary stenosis. HCM in young may be associated with other like multiple lintigines, secundum ASD, coarctation of aorta, aortic valve stenosis, tuberous sclerosis.

The patient we have described is believed to have unusual manifestation of HCM. RV gradient in absence of basal LV gradient, age of presentation (8 months), Right axis deviation on ECG in absence of ventricular enlargement, associated cleft palate and congenital bilateral talipes equinovarus (CTEV) are unusual features.

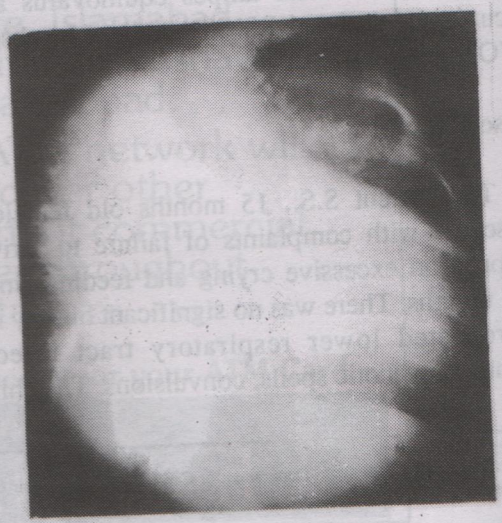

Fig. 2
Showing left ventricle in ROA 30° view.

**Summary:**

Fifteen months old child when presented with clinical picture suggestive of RVOT obstruction was found to have HCM involving RV on echocardiography and cardiac catheterization. Follow up of child in view of progression of RV gradient, development of LV gradient, response to provocative interventions is required to judge the clinical progression of the disease.

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