Abstract:
Hocm is one of the causes of sudden deaths, it is comon in the 3rd and 4th decade, it is not comon in adolescents, but if present, surgical resection done early is the main mode of treatment.

Keyword:
hypertrophic obstructive cardiomyopathy, left ventricular outflow obstruction, myectomy.

CASE SUMMARY:
13 YEAR OLD GIRL HAD COMPLAINTS OF OCCASSIONAL CHEST PAIN AND WAS DIAGNOSED TO HAVE HYPERTROPHIC OBSTRUCTIVE CARDIOMYOPATHY CAUSING LEFT VENTRICULAR OUTFLOW OBSTRUCTION. SHE WAS CONSERVATIVELY MANAGED WITH BETA-BLOCKERS AND REFERRED TO CVTS DEPT. SHE WAS EVALUATED THOROUGHLY, HER FAMILY HISTORY SHOWED NO SIMILAR COMPLAINTS, HER ROUTINE BLOOD TEST WERE NORMAL, ECG DONE SHOWED LEFT VENTRICULAR HYPERTROPHY, ECHO DONE SHOWED BASAL SEPTAL HYPERTROPHY CAUSING LEFT VENTRICULAR OUTFLOW OBSTRUCTION. PATIENT HAD SMALL A-V MALFORMATION IN HER RIGHT ELBOW 3X3CMS VASCULAR OPINION OBTAINED SHOWED IT TO BE BENIGN LESION FOR WHICH NO SURGICAL MANAGEMENT WAS REQUIRED. PRE OP ECG

PREOP ECHO:
HYPERTROPHIED BASAL INTERVENTRICULAR SEPTUM SYSTOLIC ANTERIOR
MOTION OF MITRAL LEAFLET+ MITRAL REGURGITATION-MILD TRICUSPID REGURGITATION-TRIVIAL PEAK GRADIENT ACROSS LVOT-72 LV DIMENSIONS 4.3/3.6/63%

MANAGEMENT-MYECTOMY

GENERAL ANAESTHESIA

PROCEDURE MEDIAN STEANOTOMY, THYMUS DISSECTED, PERICARDIUM OPENED, PERICARDIAL CRADLE FORMED, AORTA CANNULATED, SUPERIOR AND INFERIOR VENA CAVA CANNULATED, CARDIOPULMONARY BYPASS ESTABLISHED, CORE COOLED TO 28 DEGREES, AORTA WAS CROSS CLAMPED, ROOT COLD CADIOPLEGIA WAS GIVEN, HEART ARRESTED IN DIASTOLE, RIGHT SUPERIOR PULMONARY VEIN VENTED, AORTOTOMY WAS DONE 15mm ABOVE RIGHT CORONARY ARTERY, RIGHT CORONARY CUSP OF THE AORTIC VALVE WAS RETRACTED AND HYPERTROPHIED SEPTUM IDENTIFIED, HYPERTROPHIED BASL SEPTUM WAS INCISED WITH 11 BLADE CUTTING ONLY THE MUSCLE ABOUT 3mm IN THICKNESS, TAKING CARE NOT TO INJURE THE UNDERLYING BUNDLE, LEFT VENTRICULAR OUTFLOW TRACT WAS ADEQUETLY RELEASED.

THERE WAS NO INTERVENING OBSTRUCTION, MITRAL VALVE WAS FOUND TO BE NORMAL, AORTOTOMY WAS CLOSED. CORE REWARMED TO 37 DEGREES, CROSS CLAMP WAS RELEASED, HEART PICKED UP IN SINUS RHYTHM AND WAS GRADUALLY WEANED OFF CARDIOPULMONARY BYPASS, DECANNULATION DONE, PROTAMINE GIVEN, RIGHT VENTRICULAR PACEMAKER WIRE FIXED, RIGHT PLEURO PERICARDIAL DRAIN KEPT, STERNUM CLOSED WITH WIRES. PEAK GRADIENT-20 MEAN GRADIENT-9 NO AORIC REGURGITATION MITRAL REGURGITAION-MILD

ECG-SINUS RHYTM POST OP ECG

DISCUSSION:

Hocm is recognised as a heterogeneous sarcomere disease and mutations have been described in the beta-myosin chain gene (chromosome 14q11-q12), in cardiac troponin-t (chromosome 1), in chromosome 15q2 and in two other conditions. Microscopic findings in the ventricular septum are distinctive and hypertrophy is maximal in the cephalad portion of the septum. The free edge of aml beyond the point of cooptation hinges on the remainder.
of the leaflet in a cephalad direction towards the aortic annulus, this brings the free edge of aml in contact with the ventricular septum. This systolic anterior motion of aml is a constant feature of classic hcm. Coronary arteries are larger in diameter than normal in hcm. Muscular bridging of the left anterior descending artery is more common in hcm and has been reported to cause transmural mi. Symptoms include angina, dyspnoea, syncopal attacks, arrhythmias. Three cardinal signs of hcm include 1. late onset systolic ejection murmur between the left sterna edge and apex, 2. bifid arterial pulse, 3. palpable left atrial contractions.

Although the majority of patients are asymptomatic throughout life, some present with severe limiting symptoms of dyspnea, angina, and syncope; some may even die suddenly from cardiac causes. The mechanisms of hypertrophic cardiomyopathy are complex and include dynamic left ventricular outflow tract obstruction, mitral regurgitation, diastolic dysfunction, myocardial ischemia, and cardiac arrhythmias.

Treatment strategies are directed at symptom relief and the prevention of sudden death.

Surgical resection (myectomy) offers relief of all symptoms and prevents sudden death. Myectomy poses a great risk to the underlying bundle of his, if accidently interrupted can lead to heart block.

**CONCLUSION:**
This case report is to highlight the importance of early intervention in cases of left ventricular outflow obstruction (hcm) to decrease mortality and morbidity.

**REFERENCES:**


15. Ommen SR, Nishimura RA, Squires