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Case Study

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PERI AND POST OPERATIVE MANAGEMENT OF TOF WITH HYPERTROPHIC CARDIOMYOPATHY

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ABSTRACT

Tetralogy of Fallot(TOF) is among the commonest cause of cyanotic congenital heart disease and the association of hypertrophic cardiomyopathy(HCM)is extremely rare. The completer surgical repair has been offered in very few cases with high mortality rate 55%. We present a case of this rare association who underwent surgical repair and had challenges in Intensive Care Unit(ICU).

KEYWORDS: Hypertrophic Cardiomyopathy(HCM), Tetrology of Fallot, intensive care unit.

INTRODUCTION

Tetralogy of Fallot(TOF) is among the commonest cause of cyanotic congenital heart disease and found in 7–10% of all congenital cardiac anomalies.[1,2] TOF may be associated with other congenital heart diseases but HCM is a rare entity4, 5.789 TOF with HCM possess challenges regarding myocardial protection which should be carried out vigilantly and meticulously to prevent any myocardial injury. This can lead to problem in weaning off cardiopulmonary bypass, or other related problems in post-operative period. We present a case of TOF with HCM which fulfilled the criteria as AHA guidelines,[3] underwent complete repair. These patients usually have ventricular diastolic dysfunction that could result in high left ventricle end diastolic pressure as in our case that resulted in prolonged ventilation and ICU stay. we faced a number of other challenges in managing our patient but the early recognition of complications and prompt management was key to success in our patient.

CASE REPORT

A 03 year-old child presented with history of cyanosis and tet spells, diagnosed as Tetralogy of Fallot with hypertrophic cardiomyopathy. The parameters were as interventricular septum(IVS) systolic diameter was 16mm which is Z+14.82 and IVS diastolic diameter was 20mm (Z+8.6), LV posterior wall thickness was 10mm(Z+7.1), LV end diastolic volume 7.6ml/14.6ml/m² and LV end systolic volume 2.3ml/4.42ml/m²using Z scores Boston model[picture 1,2]. The LVEDV and LVESV were calculated post operatively. The LV function was good with adequate sized pulmonary arteries. Cardiac catheterization revealed no MAPCA, normal origin of coronaries, no additional VSD and left pulmonary artery was 9mm and right pulmonary artery was 12mm in diameter. On examination he was cyanosed with height 85cm, weight 12 and HR 110/min, O2 sat 60% on air. He was prepared for surgery according to institutional protocol and informed written consent was taken. Complete surgical repair was done, trans annular mono-cusp 15mm was placed and ASD 5 mm was left open. Total cardio-pulmonary by-pass time was 153 mins and cross clamp time was 106 mins. Came off CPB with high inotropes (inotropic score of 23) and pacing DDD @ 120/min. Epicardial echo showed left ventricular diastolic dysfunction, markedly hypo kinetic inter ventricular septum., VSD patch was stable and right ventricular outlet tract gradient was 25mmHg.

Postoperative ICU progress. After surgery the child was shifted to ICU, soon after shifting his blood pressure(BP) decreased vasopressors increased and vasopressor ionotropic score reached 33.04. Adrenaline infusion was not increased to avoid LV cavity being squashed. We were able to maintain BP on CVP of 17 to 18 mmH2O which was beneficial both for TOF's restrictive RV physiology and LV diastolic dysfunction secondary to HCM. Abdominal ascites also developed and was drained by inserting central venous catheter 7.5 Fr triple lumen instead of standard PD catheter. Hepatomegaly was also noted. Lasix infusion and Digoxin was commenced. 2600 ml peritoneal and 515ml pleural fluid was drained in the next 17 hrs, half of which was replaced with 5% albumin and additional dose of 20% albumin @ 2ml/kg once or twice daily was transfused to maintain albumin levels of 30mg/dl.

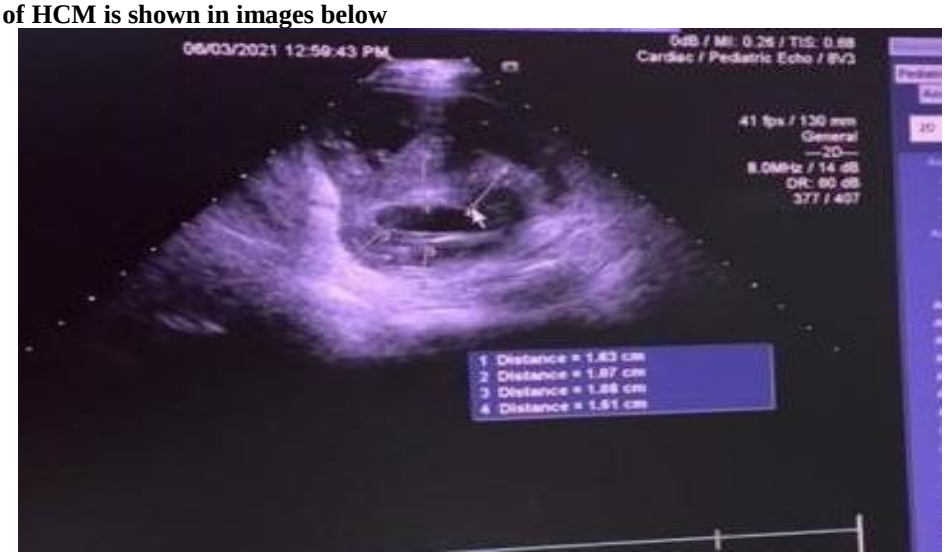
On post op day 1 the child was weaned off ventilator but sudden sharp decrease in blood pressure, tachycardia and

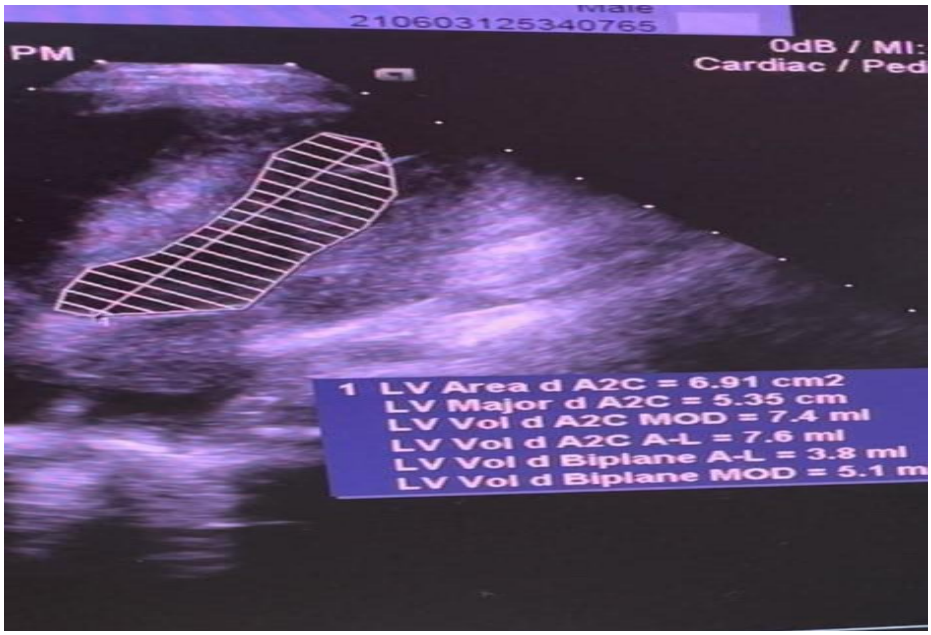
tachypnea was noted that led to re-ventilation and further increase in inotropes. Nasal Endo tracheal tube of 5.5mm internal diameter inserted in order to decreased sedation requirement which would facilitate in weaning from mechanical ventilation later on. The child also developed oliguria followed by anuria for which peritoneal dialysis was started. Serum creatinine levels increased gradually, antibiotics were changed and adjusted accordingly. Peritoneal fluid increased and was being drained @ 4000 to 5500 ml per day and pleural fluid in the range of 200 to 300ml per day.

On 2nd post op day liver functions including coagulation were impaired and required albumin and FFPs transfusion. Blood Cultures were sent in view of rising TLC and multi organ dysfunction which later on confirmed presence of Klebsiella pneumonae in blood and antibiotics were changed. Trophic nasogastric tube feeding was also started which was gradually increased and high daily calories(80-90kal/kg/day) were aimed and achieved on post op day 10.

From post op day 4 inotropes were gradually decreased and ventilatory support and FiO2 decreased. Urine out put started and later on increased by post op day 5, peritoneal dialysis stopped. Lasix infusion restarted and central venous pressures of 15cmH20 were aimed. Gradually ionotropic support decreased and tapered off supports on 12th post op day and milrinone was started at 0.1 mic/kg/hr and increased to 0.3mic/kg/hr. Later on milrinone was overlapped with 0.2 to 0.3mg/kg captopril to off load heart. On vent spontaneous trials started on post op day 9 and intermittent T-piece from 10th post op day. The child was successfully extubated on 13th post op day. On 18th post op day child was shifted from ICU. There have been three follow up visits since discharge from the hospital and the child is doing fine.

Echo findings of HCM is shown in images below





DISCUSSION

Kang-Hong Hsu at el. described 11 cases of TOF with HCM from 1978 to 2008.This rare association appears more often in males. This study further described that four of them received total correction They presented 2 cases out of which one received complete correction and BT shunt was performed in other both of them could not survive.[9]

In only few case palliative procedures like cavo pulmonary shunt and systemic-to-pulmonary artery anastomosis was done.[7,10] One of the patient was 13 year old adolescent and other were 2 infants. One of the later 2 died.[11] In study by Carvalho (8 and colleagues a 15 month old baby underwent cavo pulmonary shunt.

In French Literature two studies described 3 patient with this association 2 of them were managed medically while in one of the patient 15 years old, corrective surgery was done but unfortunately died due to cardiogenic shock in early post-operative period.[11,12]

Management of TOF includes treatment and prevention of cyanotic crises and prophylaxis with propranolol in all patients who are waiting for surgery or are not considered good candidates for surgical treatment. For suitable anatomy patients total surgical correction is the treatment of choice, and a palliative procedure with a systemic-to-pulmonary artery anastomosis is performed to increase pulmonary blood flow in patients with unfavorable anatomy,[13] Primary repair in association with HCM could be challenging due to impaired diastolic ventricular function resulting in increased ventricular end diastolic pressure leading to increase pulmonary artery pressure and ultimately right ventricular failure.

Immediate post-operative care can be quite challenging due to haemodynamic changes resulting from bi ventricular dysfunction. Vigilant hemodynamic monitoring and timely management is important. These patients require prolong ICU addressing every aspect of patients care including ventilatory support, hemodynamic changes, infection control, and caloric intake.

Currently no consensus are available for surgical management of this rare association. Physiology of both TOF and HCM should be taken into account while planning surgery and post-operative intensive care. Increased pulmonary venous flow after either cavo or aortopulmonary shunt or corrective surgery of TOF in a patient with HCM may result in pulmonary edema due to small volume and increased LV end-diastolic pressure (LVEDP) in HCM,[14] However, HCM must be considered while managing these patients. Long term follow up with regular echocardiographic monitoring of progression of HCM is important as these patients might require myomectomy and preventive use of implantable cardioverter defibrillator to prevent sudden cardiac death.

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