

Anaesthetic Management of Emergency Cesarean Section in a Patient with Holt Oram Syndrome

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Holt-Oram syndrome (HOS) is an autosomal dominant condition with complete penetrance. manifested in 1:100000 live births and characterized by forelimb deformities¹, congenital heart disease and/or cardiac conduction abnormalities. It is linked to a single-gene TBX5 "protein-producing" mutation² with gene map locus 12q24 and is the most commonly occurring heart-hand syndrome. This condition with a high rate (30-85%) of new nonfamilial cases was first described by Holt and Oram in 1960 in a 4-generation family with atrial septal defects and thumb abnormalities.¹ The most common cardiac disorder¹ is an ostium secundum ASD, followed VSD and ostium primum ASD.³ ECG abnormalities such as various degrees of AV block have also been reported.⁴

The anaesthetic implications in such patients include difficulty in catheterizing vessels, difficulty in tracheal intubation, lung ventilation and problems with blood pressure monitoring. There is a potential susceptibility to arrhythmias, hemodynamic instability and even cardiac arrest in the perioperative period. These patients could have a wide range of co morbidities like restrictive lung disease, end stage renal failure, stroke, cardiomyopathy and cardiac dysfunction.⁵

CASE REPORT

A 26 yrs old (weight-60Kgs, height-155cms) booked G₄P₂L₁A₁ with bad obstetric history presented at 42 weeks of gestation with vertex presentation for emergency caesarean section. Our patient had undergone closure of ASD by open heart technique 3 yrs back presently not on any medication.

Pre anaesthetic evaluation did not reveal any history of chest pain, dyspnoea, palpitation, cough, swelling of feet or fatigue disproportionate to normal pregnancy. She was found to have Both Upper limbs cubitus varus deformity, relatively shorter forearms and absence of thenar eminence.³ Heart rate was 57/min and Pulse was 30/min irregularly irregular with pulse deficit of 27. BP-140/ 90mmhg. Lungs were clear and heart sounds were normal, but had multiple missed beats and ejection systolic murmur in pulmonary area.

Preoperative haemogram, RBS, electrolytes were normal, ECG revealed Atrial flutter with AV dissociation, CX-Ray showed cardiomegaly with straightening of left border, 2D echo revealed R and L atrial enlargement with normal LV function, LVEF 60%, no clots or wall motion abnormality.

Patient received aspiration, Infective endocarditis prophylaxis and IM Lornoxicam 8mg preoperatively. Monitoring was established with 5 lead ECG, Non invasive blood pressure, Pulse oximeter, Temperature probe, Neuromuscular monitor, Central venous pressure monometer, and Urine output. Biphasic defibrillator, transcutaneous pacemaker and Inj. Isoprenaline were kept ready.

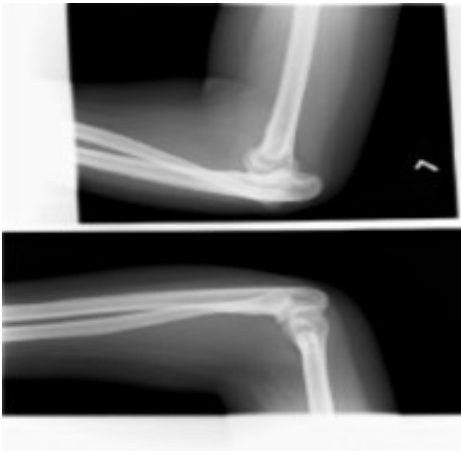
Right subclavian vein was cannulated with 16G triple lumen CVP catheter for Perioperative fluid management. Inj Amiodarone 300mg given to control the arrhythmia.

Keeping in mind the cardiac anomalies and after discussion with cardiologist, general anaesthesia with controlled ventilation was planned. She was premedicated with IV Glycopyrrolate 0.2mg, preoxygenated for 5mins, surgeons were asked to paint and drape the surgical field, then rapid sequence intubation carried with 6.5 cuffed OETT, under inj Propofol 100mg and inj Suxamethonium 100mg. Anaesthesia was maintained with oxygen, isoflurane and inj. Vecuronium bromide. The conduct of surgery was uneventful, a live female baby weighing 2 kgs extracted with the APGAR of 6 and 8 at 1 and 5mins. 15 units syntocinon was infused. At the conclusion of surgery Transverse Abdominis Peritonius block done bilaterally with 0.25% of inj. Ropivacaine 15cc on each side. Extubation carried out under deeper planes of anaesthesia to avoid sympathetic stimulation. Neuromuscular block reversed with inj. Neostigmine 2.5 mg and inj. Atropine 1.2mg. Post extubation patient was maintained with 100% O₂ through Bain's circuit till patient was fully awake. Later patient could maintain SpO₂ between 94-98% on room air. Perioperatively fluid was managed under CVP guidance and urine output. Postoperative course was uneventful.

Examination of the baby, revealed similar upper limb defect as her mother, but the baby was found to be cyanotic

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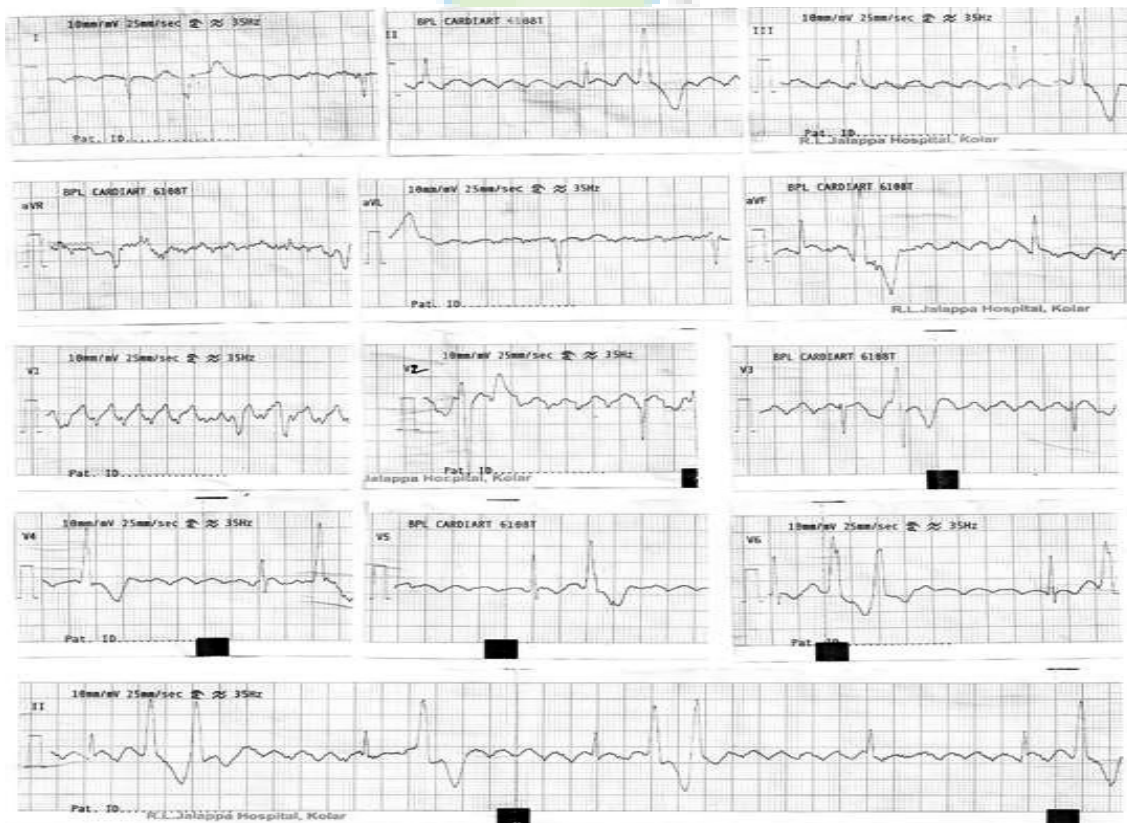


within couple of hours of delivery, suspecting congenital cyanotic heart she was referred to higher cardiac centre for evaluation. 2D ECHO and coronary angiography done revealed congenital heart disease associated with situs solitus, Ostium secundum ASD, VSD, Severe Pulmonary Stenosis and Coarctation of Aorta. Baby died on day seven of birth.

DISCUSSION

Preoperative examination for non-cardiac surgery in adults with HOS sometimes discloses cardiac anomalies, so it is important to precisely evaluate the cardiovascular system before any surgery in patients with hypoplastic thumbs.

A scoring system to assess severity has been recommended by Gall et al and modified by Gladstone and Sybert, as follows.⁶



Scoring system to assess skeletal abnormalities in HOS

- 0 - No abnormality on physical or radiological examination
- 1 - Minor abnormalities, including reduced thenar eminence, clinodactyly, or hypoplasia of the thumb
- 2 - Present arms and forearms, with one or more bones missing
- 3 - Phocomelia

Scoring system to assess cardiac abnormalities in HOS

- 0 - Asymptomatic, with no abnormal physical findings
- 1 - Conduction defect
- 2 - Structural heart abnormality that does not require surgery
- 3 - Structural heart abnormality that requires surgery but is not life threatening
- 4 - Potentially lethal malformation

Out of the three reported case of anaesthetic management in HOS, two case reports pertaining to a parturient were conducted under combined Spinal Epidural block.

In our patient, diagnosis of Holt- Oram syndrome was based on previous history of corrected ASD, upper limb anomalies, and atrial flutter with AV dissociation.

Both the above reported cases were conducted in specialized centers as elective cases, but in peripheral institution like ours with limited resources we preferred the below mentioned technique which suited our setup. We opted General anaesthesia with controlled ventilation for better control of haemodynamics. Our patient received aspiration and endocarditis prophylaxis as per our hospital protocol even though, the AHA recommendations state that delivery by caesarean section and vaginal delivery in the absence of infection does not require endocarditis prophylaxis except, in patients at high risk. Inj. Lornoxicam 8mg i.m. preemptively was given as it would avoid the use of opioid for premedication and also obtund the sympathetic response to laryngoscopy and intubation. Cardiac arrhythmia was converted to sinus rhythm by inj. Amiodarone 300mg slow i.v infusion, but in cases like ours careful monitoring of the heart rate and ECG is important as the heart rate was already on the lower side. We used slow titration of propofol

for induction of anaesthesia & maintained haemodynamic stability throughout the procedure. As our patient was well compensated preoperatively she withstood general anaesthesia and surgery uneventfully.

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