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Original Research Paper

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ANESTHETIC MANAGEMENT OF A CHILD WITH HOLT-ORAM SYNDROME POSTED FOR DEFORMITY CORRECTION OF RADIAL CLUB HAND: A CHALLENGE

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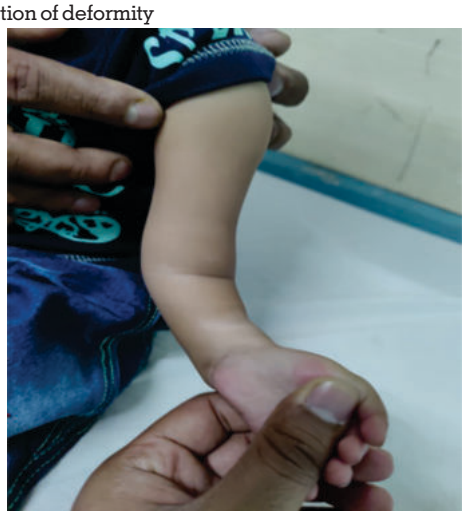
ABSTRACT Holt- Oram Syndrome is an Autosomal Dominant disorder characterized by abnormalities in upper limb and congenital heart disease. We present to you an interesting case of a 7 month old child, a diagnosed case of Holt-Oram Syndrome with Acyanotic Heart disease (patent foramen ovale) and left radial club hand deformity. The child was posted for deformity correction under General Anesthesia. Pre-operatively, thorough history taking and investigations were done. After meticulous OT preparation, patient was induced and maintained intra operatively. Goals were directed towards maintenance of hemodynamic stability, maintenance of deeper plane of anesthesia and smooth recovery by providing analgesia in the form of Ultrasound guided nerve block. Our challenges included susceptibility to arrhythmia, hemodynamic instability, tendency for blood loss and chances of cardiac arrest in peri-operative period. Duration of the surgery was roughly 3 hours with significant blood loss. Transfusion with 70 ml PRBC was done intra-operatively. The child maintained stable vital parameters throughout the surgery and in the post operative period.

KEYWORDS : Holt-Oram syndrome, child, acyanotic heart disease, general anesthesia, hemodynamic stability, anesthetic challenge

INTRODUCTION

Holt-Oram syndrome is an Autosomal Dominant disorder characterized by a heterozygous mutation in the TBX5 gene.[1] It comprises of abnormalities in the upper limb (radial, thenar, carpal bones) and congenital heart disease (Atrial Septal Defect, Ventricular Septal Defect, Conduction defects). It is also called the "Heart-Hand syndrome".[2] The incidence is 1 in 1,00,000 individuals with no predilection for male or female gender. Both physical characteristics and family history of the disorder can aid in the diagnosis of Holt-Oram Syndrome. Further investigations including Electrocardiogram, 2D echocardiography, X-rays and genetic testing can confirm diagnosis.

We present a case of a 7-month-old Male child, a diagnosed case of Holt-Oram Syndrome at 4 months of age with Acyanotic Congenital Heart disease (patent Foramen Ovale with Left to Right Shunt and trivial Tricuspid Regurgitation) and radial club hand defect. The child was posted for surgical correction of deformity



Picture 1

Anaesthetic Goals

- Maintenance of Hemodynamic stability with respect to avoidance of tachycardia, fluctuations in blood pressure and prompt recognition and management of any arrhythmia or fluctuations of any Hemodynamic parameter
- Maintenance of Deeper plane of anesthesia by provision of adequate analgesia intra operatively and post operatively

Case Report

Pre-operative Evaluation

7 months old Male child weighing 7 kg was brought with complaints of deformity of left upper limb since birth (radial club hand)

The child was evaluated at the age of 4 months and diagnosed as a case of Holt-Oram Syndrome with Acyanotic Congenital Heart disease and Radial club hand defect.

On history taking, birth was uneventful with complete immunization status and no significant medical or surgical history. It was also revealed that the child's father had a similar defect in upper limb since birth which was not adequately evaluated.

Clinical examination was suggestive of a faint systolic murmur on auscultation of heart sounds with vital parameters and other systemic examination being within normal limits.

Investigations such as Complete Hemogram, Electro-cardiography (ECG) and 2D echocardiography were done

Complete Hemogram revealed hemoglobin of 11 gm/dl, Total Leukocyte Count of 8500 and Platelets of 2.3 lakhs/cu.mm ECG was suggestive of Normal sinus rhythm 2D echo-cardiography suggestive of Acyanotic Congenital Heart Disease; Patent Foramen Ovale measuring 2.4 mm with Left to Right Shunt; no Pulmonary arterial Hypertension; Trivial tricuspid regurgitation and normal biventricular function.

The child was posted for surgical correction of Radial club hand defect under general anesthesia with Ultrasound guided axillary block for intra operative and post operative analgesia.

Intra Operative Management

Thorough OT preparation was done with all emergency resuscitation equipment including defibrillator with pediatric

pads, difficult intubation trolley , Central venous catheter of appropriate size Ambient OT temperature was ensured and warm fluids were kept ready

After confirming the informed written consent of the parent and Nil per oral status, the child was taken inside the OT. Standard ASA monitors including precordial stethoscope were attached, IV cannulation was done on right upper limb with a 24 G IV cannula under spontaneous ventilation with Oxygen and Sevoflurane Premedication given with Inj Glycopyrrolate IV 0.028 mg and Inj. Midazolam IV 0.35 mg, Inj Fentanyl IV 10 mcg

Induction was done with Inj Propofol IV 7 mg and Inj Ketamine IV 5 mg Gentle laryngoscopy was done and intubation with uncuffed Endotracheal Tube size 4.0 done

After tube position was confirmed, Ultrasound guided Axillary Block was given on the operative limb with 0.2% Ropivacaine 3.5 ml

Anesthesia was maintained on oxygen, air, sevoflurane and atracurium bolus Care was taken to avoid any bubbles in the IV lines

The surgery lasted for 3 hours with intra operative blood loss being roughly 100 ml

The child was transfused with 70 ml PRBC (10 ml/kg) Vital parameters were stable throughout the surgery and the child was extubated uneventfully

The child was later shifted to Pediatric Intensive Care unit post operatively for observation.

Picture 2

Post Operative Management

Since Holt- Oram syndrome predisposes the patient to the development of hemodynamic instability in the post operative period too, adequate monitoring and management of the patient post operatively is crucial

The child maintained stable vital parameters post operatively. Analgesia with the axillary block provided post operative pain relief and the child was shifted to the ward after 24 hours of observation.

DISCUSSION

Anesthetic management of a case of Holt-Oram Syndrome poses great challenges in terms of difficult IV cannulation, difficult tracheal intubation, arrhythmias, hemodynamic instability and cardiac dysfunction

Hence, we opted for gentle induction with necessary emergency equipment kept ready.

We avoided extreme fluctuations in heart rate and blood pressure by maintaining a deep plane of anesthesia a with maintenance of adequate pain relief.

Excessive blood loss and subsequent hemodynamic instability were managed with timely blood transfusion and tourniquet application.

Thorough pre operative evaluation [3] with cardiac work up is critical in successful management of such cases.

CONCLUSION

We hereby conclude a successful anesthetic management of a case of Holt-Oram syndrome with goals directed towards maintenance of hemodynamic stability, deeper planes of anesthesia with adequate intra operative and post operative pain relief.

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