International Journal of
Regional Anaesthesia

Case Report

Opioid Sparing Anaesthetic Technique in Downs Syndrome Child with Congenital Heart Disease and Atlanto-Occipital Instability: A Case Report

Himaunshu V. Dongre[1], Sandeep M. Diwan[1], Ganesh P. Bhong[1], Parag K. Sancheti[2]
[1]Department of Anaesthesia, Sancheti Institute for Orthopaedics and Rehabilitation, Pune, Maharashtra, India.
[2]Department of Orthopaedics, Sancheti Institute for Orthopaedics and Rehabilitation, Pune, Maharashtra, India.

Abstract

Downs syndrome, a common chromosomal abnormality is associated with hip and patellar instability and also atlanto-axial instability. Recurrent dislocation of the hip joint leads to potential disability requiring surgical intervention. Femoral varus derotation osteotomy and fixation is one of the procedures performed to stabilise the hip joint [1]. We report a case of Downs syndrome associated with congenital heart disease (CHD) and atlanto-axial instability which successfully underwent femoral varus derotation osteotomy procedure.

Keywords: Downs Syndrome, Atlanto-axial instability, Congenital heart disease

Introduction

Downs syndrome (DS) also known as Trisomy 21, was described in 1866, estimated to be seen 1 in 1000 births. The syndrome is associated with intellectual disabilities in addition to adversely affecting multiple organ systems. Additional chromosome 21 encodes abnormal collagen VI levels leading to hypotonia as well as laxity of ligaments and joints [2]. It contributes to cervical spine instability, hip dislocations, scoliosis and foot deformities. The recurrent dislocation leads to hip joint arthritis, and long term debilitating pain. Surgical intervention for hip containment helps to prevent recurrent dislocations [3].

Case Report

A 7-year-old girl child with DS weighing 17 Kgs, suffering from recurrent hip dislocation was posted for femoral varus derotation osteotomy. On examination, the child was cheerful and had typical features of Down's syndrome, including short neck, low set of ears, flattened nasal bridge and prominent epicanthic folds. There was a pansystolic murmur in the right second intercostal space. Echocardiography revealed Atrial Septal Defect (ASD) and Patent Ductus Arteriosus (PDA). The ASD was 4 mm, and PDA was 1 mm, with a left to right shunt. The X-ray cervical spine revealed increased atlanto axial dense interval (AADI), elongated C2 bone with an increase in AADI on flexion-extension. The opinion of the spine surgeon was sought to evaluate the atlanto-axial instability, which emphasised manual in-line stabilisation of the cervical spine during intubation.

Parents were informed and counselled regarding the potential neurological complications secondary to atlanto-axial instability, and appropriate consent was taken. The iv line was secured in front of the parents in the recovery room. No pre-medication was given in preoperative room, and the patient was wheeled in. Standard monitoring, including ECG, SpO_2, BP ensued, and pre-medication was given using inj. Glycopyrrolate 4 mcg/kg, ketamine 0.25 mg/kg and inj ondansetron 50 mcg/kg. The patient was induced with inj. Etomidate 0.3 mg/kg, and after checking bag-mask ventilation inj. Atracurium was administered 0.5 mg/kg as the neuromuscular blocking agent. Appropriate size LMAs, Igels and video laryngoscope was kept ready. Manual in-line stabilisation was applied during intubation given atlanto-axial instability.

Address of Correspondence
Dr. Himaunshu V. Dongre,
Department of Anaesthesia, Sancheti Institute for Orthopaedics and Rehabilitation, Pune, Maharashtra, India.
E-mail: himaunshu.dongre@gmail.com

Submitted: 11/11/2021; Reviewed: 25/11/2021; Accepted: 19/12/2021; Published: 10/01/2022

DOI: 10.13107/ijra.2022.v03i01.052 | www.ijrajournal.com |

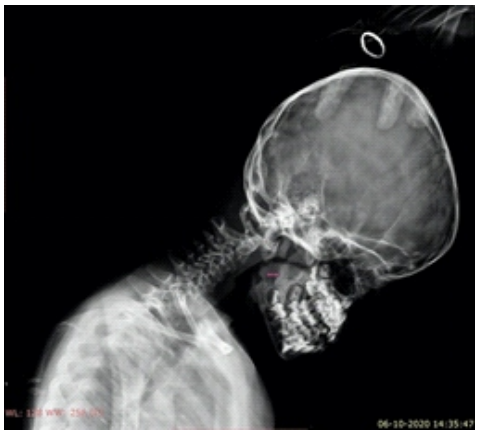

Figure 1: AADI on lateral radiograph 5.41 mm on flexion, with elongated dense of C2

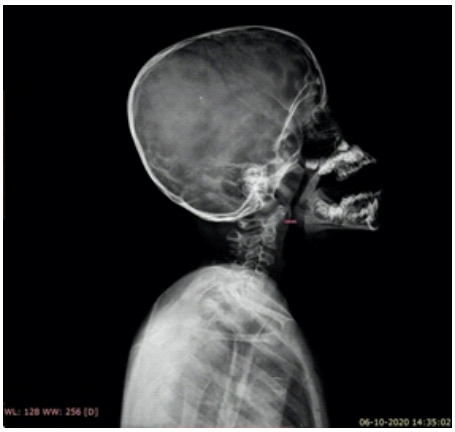

Figure 2: AADI on lateral radiograph 1.85 mm on extension

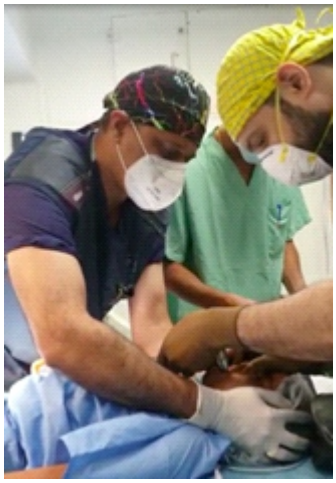

Figure 3: Manual inline stabilization for intubation.

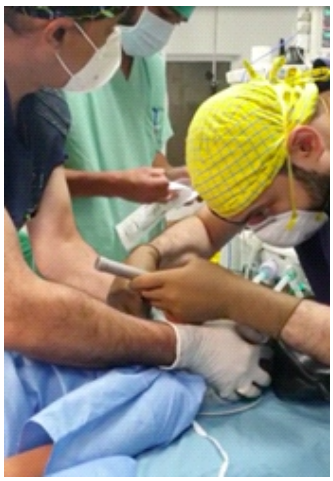

Figure 4: Manual in line stabilization during direct laryngoscopy

After adequate ventilation laryngoscopy was done using flexitip blade number 2, maintaining manual in-line stabilisation. Laryngoscopy revealed Cormac Leyhan Grade 3 and intubation was achieved in the first attempt using micro cuff tube size 4.5 mm after lifting the epiglottis using the flexitip. A soft cervical collar was applied after fixation of the tube. We witnessed one episode of bradycardia just before intubation which returned to pre-induction level immediately after intubation. A left lateral decubitus position was given maintaining cervical spine stability for caudal analgesia. Caudal epidural analgesia was administered with 22G hypodermic needle using 1 ml/kg of 0.25% bupivacaine and 1 mc/kg of clonidine. The patient was turned into supine position taking care of the cervical spine, and right femoral derotation osteotomy was carried out in the supine position. Anaesthesia was maintained with 50% oxygen with air and sevoflurane. There was no change from the post-induction baseline heart rate and blood pressure readings following incision or surgical manipulation of right femur. The patient was extubated on the table after reversal of neuromuscular blocking agent with the cervical collar in situ. The patient was shifted to the recovery room, and the postoperative course was uneventful.

Discussion

Downs syndrome (DS) patients pose numerous challenges to anaesthesiologist as the syndrome is associated with abnormal cardiovascular, respiratory, gastrointestinal, immune, musculoskeletal, endocrine, nervous system features. The additional chromosome 21 leads to excessive expression of collagen VI, which leads to cardiac endocusion defects, musculoskeletal laxity and airway abnormalities [2]. Atlanto-axial instability is well described in downs syndrome and seen in 6-27% of patients [4]. The American Academy of Paediatrics has released a positional statement in 1984 and stated that lateral radiographs are indicated for diagnosing atlanto-axial instability [5]. The atlanto-axial dental (AAD) distance in a neutral position on a lateral radiograph is crucial, and if >3 mm, is considered as moderate atlanto axial instability (AAI). In our case, the AAD distance was 4 mm on lateral radiograph with an elongated C2 dome. There was an increase in the atlanto-axial dense interval (AADI) on flexion and extension. These patients are asymptomatic for months to years or can present with an abnormal gait, easy fatigability, hyperreflexia, and may progress to hemiplegia, quadriplegia. It is prudent to hold the cervical spine in a neutral position during intubation to prevent the anterior migration of atlas,

which may result in cord compression. Manual in-line stabilisation is useful in keeping the cervical spine in a neutral position. (Ref) After induction of anaesthesia soft collar can be applied to prevent extreme flexion and extension [5].

Subglottic stenosis, tracheal stenosis has been reported in Downs syndrome. It is suggested that Downs syndrome patients should be intubated with an endotracheal tube 0.5-1.0 mm diameter smaller than the age-appropriate endotracheal tube size [6]. Kenji et al. have suggested the use of preoperative airway assessment using ultrasonography to know the exact size of the endotracheal tube [7]. They have also advocated the use of appropriate size micro cuff tubes in such patients. In our case, we used a 4.5 mm size micro-cuff tube which was 0.5 mm less than the age-appropriate tube.

Cardiac Endocusion defects are commonly seen is Downs syndrome and can present as Atrial Septal Defect (ASD), Ventricular Septal Defect (VSD), Tetralogy of Fallot (TOF), which are seen in 40-63.5% of patients [8]. All the defects except TOF result in increased pulmonary vascular resistance. Etomidate, a non-barbiturate hypnotic agent with minimal effect on the haemodynamics, is frequently used in paediatric patients with CHD [9]. Medical air was used instead of Nitrous oxide in anticipation to prevent the increase in pulmonary vascular resistance.

Caudal analgesia is safe and effective regional anaesthesia technique (RAT) in paediatric patients for sub umbilical procedures with an overall complication rate of 0.12% [10]. Common drugs used for caudal are bupivacaine 0.125% - 0.25% and ropivacaine 0.1-0.375%. The current recommendations for bupivacaine and ropivacaine are, not to exceed 2.5 mg/ml and 2 mg/ml, respectively, in paediatric patients [10]. The volume used depends upon the dermatomal level required for surgery. The current guidelines recommend 0.5 ml/kg for sacral, 1 ml/kg for lumbar and 1.5 ml/kg for thoracic dermatomes. Clonidine, an alpha 2 agonist and preservative-free morphine are the two approved drugs for epidural use [10]. Clonidine can be used in the doses of 1-2 mic/kg and morphine 10-30 mic/kg. In our case, we used 0.25% bupivacaine 17 ml and clonidine 15 micrograms as per recommendations. Caudal analgesia provided a pain-free postoperative period. It also reduced the intra-operative requirement of inhalational anaesthetic (sevoflurane) and opioids.

Inhalational anaesthetic agents cause myocardial depression in all patients. The patients with DS have low levels of circulating catecholamines. The combined effect may lead to profound bradycardia and hypotension in DS [6]. The reason for bradycardia and hypotension witnessed in our case after induction can be attributed to low catecholamine levels and mask ventilation with sevoflurane.

In our case, we witnessed one episode of bradycardia just before intubation which returned to pre-induction level immediately after intubation.

Children with neurodevelopmental disability have 1.8 times higher chance of opioid-induced respiratory depression [11]. Given this, we used an analgesic dose of ketamine as pre-medication, etomidate as an induction agent and caudal epidural analgesia to provide opioid-free anaesthesia.

Conclusion

Downs syndrome is a multiorgan disease. Regional anaesthesia technique combined with a sub-anaesthetic dose of ketamine and etomidate as an induction agent provides an opioid-free strategy for managing DS patients with CHD for non-cardiac surgeries. In-line stabilisation during intubation and smaller size micro cuff tube is useful in avoiding complications arising secondary to atlanto-axial instability and subglottic stenosis.

Abbreviations used: DS- Downs Syndrome; CHD – Congenital Heart Disease; ASD- Atrial Septal Defect; PDA – Patent Ductus Arteriosus; VSD – Ventricular Septal Defect; TOF – Tetralogy of Fallot; AADI – Atlanto Axial Dense Interval; AAD – Atlanto Axial Dense; RAT – Regional Anaesthesia Technique; AAI – Atlanto Axial Instability

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the Journal. The patient understands that his name and initials will not be published, and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil **Source of support:** None

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How to cite this article: Dongre HV, Diwan SM, Bhong GP, Sancheti PK | Opioid Sparing Anaesthetic Technique in Downs Syndrome Child with Congenital Heart Disease and Atlanto-Occipital Instability: A Case Report | International Journal of Regional Anaesthesia | January-June 2022; 3(1): 31-34.