

## CASE REPORT

# Anesthetic Management of a Neonate with OEIS Complex for Emergency Surgery: A Complex Challenge for Anesthesiologist!

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### ABSTRACT

Omphalocele, exstrophy of cloaca, imperforate anus, and spinal defect (OEIS) complex is an extremely rare congenital anomaly (1:2,00,000–1:4,00,000) comprising of OEIS. The associated anomalies pose multiple challenges during the anesthetic administration. We are reporting anesthetic management of a baby born with this rare syndrome, posted for an emergency surgery on the 1<sup>st</sup> day of life. Anesthetic management of a neonate with this complex and difficult airway in this relation has never been discussed previously.

**Key words:** OEIS complex; Meningomyelocele, Neonate; Anaesthesia

### INTRODUCTION

Omphalocele, exstrophy of cloaca, imperforate anus, and spinal defect (OEIS) complex is very rare congenital disorder comprising of OEIS [1]. It has an incidence of 1:2,00,000–1:4,00,000 births [2]. Diagnosis is primarily based on the coexistence of this spectrum of anomalies in a patient [2]. This syndrome is commonly associated with other genitourinary, vertebral, limb, renal, and cardiovascular defects [3,4]. The defect occurs due to embryonic failure of convergence of mesodermal migrations and development of urorectal septum [1]. The management of this syndrome is underreported in the field of anesthesiology. We intend to highlight the anesthetic challenges faced in the management of a neonate with this complex posted for an emergency surgery. We have also included a comprehensive overview of this syndrome.

### CASE REPORT

A full-term 1-day-old small for gestation age (weight 2.1 kg and height 42.5 cm) male baby delivered by normal vaginal home delivery, presented with complaints of minimal passage of stools and abdominal distension since birth. He was tachypneic, but was maintaining oxygen saturation between 95 and 97% on oxygen by hood. Signs of dehydration were present (irritability, sunken eyes, parched mucosa, depressed fontanelle,

and tachycardia) at admission and arterial blood gas analysis revealed mild acidosis (pH of 7.31). The child was admitted to the high dependency unit for pre-operative optimization. Intravenous fluid resuscitation and broad-spectrum antibiotics were started.

The baby had a combination of anomalies (omphalocele, exstrophy of bladder, imperforate anus, and spinal defect [sacral meningomyelocele and thoracic kyphoscoliosis]) compatible with the diagnosis of OEIS complex Figure 1. In addition, he had other associated skeletal (bilateral club feet, thoracic kyphoscoliosis, short neck, and rib deformity) and genital (pubic diastasis, and undescended gonads) anomalies. He also had rib and spine deformity, hypotonia, and lower limb areflexia. Cardiovascular and respiratory examination was unremarkable.

Blood investigations including serum electrolytes normal. Chest X-ray showed scoliotic spine with deformed ribs.

After initial stabilization, the baby was posted for emergency laparotomy. Neonatal intensive care unit bed (NICU) was arranged and informed consent for possibility of post-operative mechanical ventilation was obtained. We were anticipating difficult airway because of thoracic kyphoscoliosis, short neck, and meningomyelocele.

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Operation theater was prepared as per the requirements of neonatal surgery including warm intravenous and irrigation fluids, infusion pumps, warm ambience, warming mattress prewarmed (39–40°C), difficult intubation cart (inclusive of Truview PCD™ videolaryngoscope; [TVL]), positioning bolsters, and cotton padding. The baby was shifted to OT in incubator in lateral position. We planned to intubate the child under inhalational induction while preserving spontaneous respiration in lateral position. Gastric decompression was done by suctioning the *in situ* nasogastric tube. After 3 min of pre-oxygenation, anesthesia was induced with sevoflurane (5–7%) and injection fentanyl 1 mcg/kg was administered after securing a 24G intravenous line in upper limb. Laryngoscopy done with Macintosh size “0” blade revealed a modified Cormack-Lehane (CL) grading 4 which did not improve after optimal external laryngeal manipulation (OELM). Repeat laryngoscopy after repositioning the baby to bring sternal notch at the level of tragus using doughnut and generous padding improved the CL grade to 3 with OELM. At this moment, a TVL (size “0” blade with its dedicated stylet) with OELM improved the glottis view to CL grade 2a, and the patient could be successfully intubated with uncuffed 3.0 endotracheal tube. Anesthesia was maintained on intermittent boluses of IV atracurium, oxygen, air, and sevoflurane (to achieve a MAC of 1). Uninvolved parts of body were wrapped in cotton during surgery. Oropharyngeal temperature was monitored and normothermia was maintained. Ventilatory strategy of low tidal volume (12 mL), and high respiratory rate of 36/min with a positive end expiratory pressure of 5 cm H<sub>2</sub>O was initiated. Ventilatory settings were titrated to maintain oxygen saturation of hemoglobin more than 90% and an end-tidal carbon dioxide between 35 and 40 mmHg. Intraoperatively, the neonate was administered Ringer’s lactate (RL) at the rate of 25 mL/h and 10% dextrose at the rate of 8 mL/h using infusion pumps. Intermittent boluses of RL were infused. Total blood loss of 20 mL was well within the allowable blood loss. A divided ileostomy was performed for malrotation of the gut and anal atresia. Intraoperative vitals were stable throughout. Ultrasound-guided bilateral transverse abdominal plane (TAP) block was performed at the end of surgery using 2 mL of 0.125% bupivacaine. The patient showed insufficient respiratory efforts following reversal of neuromuscular blockade and was shifted to NICU intubated for ventilatory support. He was extubated the same day and was discharged to the ward next morning.

## DISCUSSION

OEIS complex represents a spectrum of congenital malformations due to defective embryonic development of ventral abdominal wall and urorectal septum [1,4]. In our patient, omphalocele was very small but imperforate anus, pubic diastasis, and spinal defects were present. Major anesthetic challenges in our case were



Figure 1: The neonate with OEIS: MMC (a); positioning during surgery over doughnut (b); club foot (c); and kyphoscoliosis (d)

the early neonatal period, difficulty in the management of airway, and positioning for surgery. Anesthetic concerns in a case of OEIS and the management in the present case have been presented in Table 1.

To prevent compression of meningocele, initial attempt at intubation was undertaken in lateral position. After failed laryngoscopy, the baby was positioned supine. However, supine positioning was difficult in this baby due to his kyphoscoliotic spine and large meningocele. Placement of doughnut below meningocele caused hyperflexion at neck affecting ventilation, so an extra padding at back and below the shoulder was placed to aid in positioning. After second failed attempt at direct laryngoscopy by a senior operator, VL helped improve glottis view and allowed intubation. Intubation in such neonates has been deemed to be difficult even by neonatologists who routinely intubate this group of patients [5]. It has been mentioned that they may require multiple attempts at intubation which can worsen their airway outcome, especially in patients with congenital anomalies [5]. Videolaryngoscopy has been suggested as a measure to increase effectiveness at intubation in children [6]. TVL with its 46° anterior deflection provides a non-line of sight vision, avoids the need to align the optical axis for laryngoscopy, and improves the laryngoscopic view as compared to direct laryngoscopy in infants [7]. MMC child has been successfully intubated in the past using lateral position/doughnut/silicone supports [8]. Kyphoscoliosis, omphalocele, and abdominal distention due to imperforate anus can lead to low lung compliance and high airway pressure in these patients. We used low volume ventilatory strategy and high frequency to maintain airway pressures in acceptable limits. Application of PEEP avoided atelectasis due to diaphragmatic splinting. During the surgical procedure in supine position, the pressure on the MMC sac was minimized using doughnut and generous padding to prevent damage and prevent its compression.

Table 1: Anesthetic concerns, their implication, and management in a patient with OEIS complex

Anesthetic concerns	Implication	Management in the present case
Neonatal age group	Prevention of hypothermia, hypoglycemia, optimization of intravascular volume status, and risk of apnea after anesthesia	Heating mattress, warm IV fluids, 10% dextrose as maintenance fluid, post-operative ventilator support
Anticipated difficult airway	Difficulty in intubation due to large meningomyelocele, kyphoscoliosis and short neck	Use of padding and doughnut to align the sternal notch with tragus, TVL with its non-line of sight vision improved glottis view
Colostomy closure requiring the supine position	Placing of doughnut below meningomyelocele can flex the neck and affect ventilation	extra-padding at back and below the shoulder placed to aid in positioning
Kyphoscoliosis, omphalocele, and imperforate anus	Low lung compliance and high airway pressure	Ventilatory strategy of low volume, high frequency
Associated undiagnosed congenital syndrome	Emergent nature of surgery may not allow adequate time for evaluation	Careful clinical examination of all systems
Meningomyelocele	Positioning, increased intracranial pressure	Care to prevent sac compression

Anesthetic management of OEIS complex has never been discussed in literature. Advances in medical and surgical care of these patients have helped improve the survival of these patients who may therefore come for multiple surgical procedures [9]. Most cases present in early infancy for emergency surgery due to its association with imperforate anus manifesting into intestinal obstruction. Adequate post-operative pain management is crucial in these cases for minimizing stress response with its associated adverse systemic sequelae as well as for optimization of respiration. TAP block provides effective post-operative analgesia after abdominal surgeries [10].

In conclusion, we have described the anesthetic management of a rare syndrome presenting complex anesthetic challenges. Most cases present in early neonatal period and later undergo multiple staged procedures increasing the associated risks and complications. Knowledge of associated anomalies and their anesthetic implications is therefore essential for optimal outcomes in these patients. The use of videolaryngoscopes may be considered during initial attempts at intubation in children with congenital anomalies.

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