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ANAESTHETIC MANAGEMENT OF A CHILD WITH SPINAL MUSCULAR ATROPHY TYPE – 2 UNDERGOING PERCUTANEOUS ENDOSCOPIC GASTROSTOMY IN THE NONOPERATING ROOM SCENARIO

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ABSTRACT Spinal muscular atrophies (SMAs) are a group of very rare autosomal – recessive disorders with an incidence of 1: 6000 to10000 causing progressive proximal muscle weakness due to degeneration of spinal motor neurons. Anaesthetic management of children with SMA is highly challenging and requires a muscle relaxant free technique due to erratic response and recovery observed for the same. Also, a judicious use of opioids is mandated to avoid excessive depression of the central respiratory drive. We have discussed the anaesthetic technique used in the clinical scenario encountered by us, where a child with SMA type 2 underwent Percutaneous Endoscopic Gastrostomy. An inhalational induction technique along with blunting of airway reflexes using Local Anaesthetic spray for intubation followed by careful maintenance of anaesthesia, helped with total avoidance of muscle relaxants & opioids. It also aided in ensuring an early recovery while minimizing the perioperative risk in children with SMA.

KEYWORDS :

INTRODUCTION:

Spinal muscular atrophies (SMAs) are a group of autosomal – recessive disorders with progressive degeneration of spinal motor neurons. It has types 1, 2, 3 and 4 in decreasing order of severity with type 1 and 2 being very rare and having high mortality. Anaesthetic management of children with SMA has not been extensively studied with limited data available so far. It is highly challenging due to difficulty in intra venous cannulations & maintanence of airway, muscle weakness, hypersensitivity to non depolarizing muscle relaxants and risk of hyperkalemia. In the non operating room scenario, there is a high risk of laryngospasm especially in case of shared airway procedures. Hereby we are presenting the case report of a child with SMA type 2undergoing percutaneous endoscopic gastrostomy and the anaesthetic approach we undertook.

CASE REPORT :

A 4 year old girl child, weighing 9.3 kilograms diagnosed to have SMA type2 with history of recurrent pneumonia and feeding difficulties on nocturnal Bilevel Positive Airway Pressure – Non Invasive Ventilation (BiPAP-NIV) at home, presented with impending respiratory failure secondary to pneumonia. She was admitted in the Paediatric Intensive Care Unit (PICU) and was given Oxygen supplementation (4 litres via Hudson mask) and was initiated on feeds via Naso Duodenal tube. She recovered well and Percutaneous Endoscopic Gastrostomy(PEG) was planned after 15 days of PICU stay. Her baseline vitals were stable and she was able to maintain a saturation of 98% in room air. Baseline investigations were as follows - Haemoglobin – 12.1 g/dl, Platelet count – 615000, Total count – 12900, serum electrolytes – within normal limits. Chest X ray showed minimal infiltrates on both lungs. Informed consent was obtained from the child's parents. Ensuring an adequate nil per oral status, the procedure was performed under General Anaesthesia in the endoscopy room. After adequate pre oxygenation, inhalational induction was done with sevoflurane 6%. Intravenous(IV) access was established in the left leg with a 22 Gauge cannula following which Lignocaine 20 mg IV & Propofol 50 mg IV in sequential doses were administered to deepen the plane and Lignocaine (10 mg/puff – single puff) was sprayed in the airway prior to intubation. Child was intubated with 5.5 size uncuffed portex endotracheal tube & a mouth prop was inserted to facilitate endoscopy. Anaesthesia was maintained with Sevofluorane 3% and ventilation was assisted with Jackson and Reese circuit. Peri -operatively, child was hydrated adequately with isotonic crystalloids administered at the rate of 10 ml/kg and was given 150 mg (15mg/kg) of Paracetamol IV. Heart Rate, Non Invasive Blood Pressure, Peripheral oxygen saturation, End tidal Carbon-di-oxide Concentration and temperature were monitored throughout. Procedure lasted for 30 minutes and was uneventful. Incision site infiltration was done using local anaesthetic (7 ml of 0.25 % Bupivacaine). Child was extubated on table after she was fully awake and post procedure vitals were stable. She was immediately put on BiPAP support to maintain airway patency and was shifted to PICU for post operative care. Later she was weaned to oxygen supplementation via face mask. Paracetamol (15 mg/kg IV – thrice a day) was prescribed for analgesia post operatively.

DISCUSSION:

SMAs are a very rare group of genetic disorders with an incidence of 1: 6000 to10000 live births. This child fits into a mixed category of type 1 and 2 with predominant features of type 2. Child had bilateral lower limb weakness and mild proximal muscle weakness in the upper limbs suggestive of symmetrical flaccid paralysis with hypotonia. No standardized protocol for management of anaesthesia in SMA children has been established until now.

A retrospective analysis of various anaesthetic management techniques and outcomes for SMA children undergoing spine correction surgeries have been described by Halanski[1] et al pointing to the avoidance of neuromuscular blocking agents due to observance of erratic response and unprecedented delays in muscle recovery.

Stucke & Stuth[2] described the use of Non Depolarizing Muscle Relaxant in a 18 month old child with type 2 SMA to tackle laryngospasm while intubation & noted normal diaphragm recovery but delayed proximal muscle recovery. Graham and Athiraman[3] have conducted a retrospective study of 25 cases of SMA and concluded that peri-operative care can be provided for children with SMA effectively under Total Intra Venous Anaesthesia (TIVA) or inhaled anaesthetic agents along with the judicious use of opioids to improve patient comfort without increased morbidity.

CONCLUSION:

A controlled induction technique by blunting of airway reflexes using local anaesthetic spray and a combined use of inhalational & IV agents for intubation followed by a careful maintenance of anaesthesia, helps with total abolition of usage of neuromuscular blocking agents and ensures early recovery while minimizing the risk of any undue events perioperatively in children with SMA.

The high risk of postoperative respiratory complications can be tackled by strategising the anaesthetic agents and technique to avoid excessive depression of central respiratory drive.

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