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Letter to Editor

Anaesthetic management of an infant with spinal muscular atrophy [Type 1] for fundoplication and feeding gastrostomy

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Dear Editor,

Spinal muscular atrophies (SMA) include a group of neuromuscular disorders characterized by degeneration of alpha motor neurons in the spinal cord with progressive muscle atrophy, weakness and paralysis.[1] Typically, weakness is symmetric and proximally dominating. From modest proximal limb weakness seen in adulthood to severe widespread weakness with respiratory failure in the neonatal period, the severity of the condition can vary greatly. Lower limbs are more affected than upper limbs, and cases of more severe limb weakness sometimes accompany bulbar and respiratory paralysis.[2] An infantile version of this ailment called SMA type 1 (Werdnig-Hoffmann disease) has an early onset of respiratory failure and progresses quickly.[2]

Here, we describe the case of an 11-month-old female infant with SMA type 1 for fundoplication, feeding gastrostomy, and tracheostomy. She was a term baby born via caesarean section with birth weight 3.35 kg. She required numerous hospital stays due to recurrent cases of aspiration pneumonia, and at the age of two months, SMA-Type 1 was identified. She was hospitalised at the age of 11 months for respiratory failure and aspiration pneumonia.

In the pre-anaesthetic check-up, she was alert, hypotonic, on BiPAP (IPAP-12, EPAP-6), with respiratory rate of 30/min at Oxygen- 1 L/min, SpO2 was 100%, Heart rate was between 136-138/min and was in regular sinus rhythm. On auscultation air entry was equal bilaterally with crepitations in both lower lobes. Blood investigations were normal. Chest X-ray showed bilateral infiltrates (Figure 1). Echocardiography revealed normal contractility. Her height and weight were 70 cm and 6.2 kg respectively. 120 ml each of packed red cell concentrate and plasma were arranged. Inhalational induction was done with Oxygen and Sevoflurane at 8%. Fentanyl 16 mcg and Propofol 8 mg were given intravenously (IV).

Patient was bag-mask ventilated for 3 minutes and endotracheal intubation was performed using an uncuffed endotracheal tube of size-4 without any muscle relaxants. Analgesia was achieved with Ketamine 5 mg IV, additional Fentanyl bolus of 2 mcg IV, Propofol as IV infusion and Paracetamol 40 mg IV infusion. Caudal block was avoided. Mechanical ventilation with peak airway pressure <20 cm water and respiratory rate 30/min was set. SpO2 was 100% throughout surgery, End tidal CO2 was maintained between 35-38mm Hg and heart rate was in the range 100-120/min. The total duration of the procedure was 4 hours 30 minutes with stable vitals. 500 ml crystalloids were administered intra-operatively. Tracheostomy was performed post-procedure with a cuffed tube (size-4) - bilateral breath sounds were equal and the infant was mechanically ventilated.

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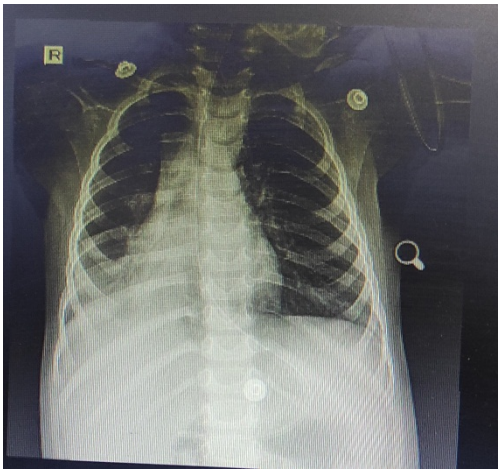

Fig. 1: Pre operative X-ray showing bilateral infiltrates in the lower zone

Post-operative Arterial Blood Gas analysis results were as follows: pH 7.31 ; PaO_2- 86.4 ; $PaCO_2$-39.7 ; SO_2 - 96.4%. She was shifted to Paediatric ICU in control mode. Chest X ray was taken next day (Figure 2) and was switched to BiPAP. BiPAP settings were titrated according to the respiratory status and chest physiotherapy was initiated. Feeds were started via gastrostomy and slowly escalated. She was discharged after 15 days on BiPAP via tracheostomy insitu (5/10 cm H_2O). Mic-Key button gastrostomy tube was placed after a month.

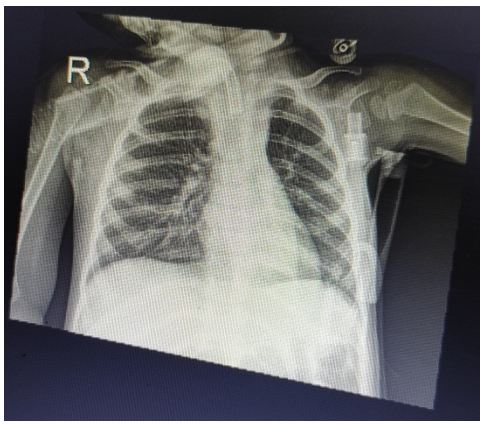

Fig. 2: Post operative X-ray day 1

SMA patients who have a low density of acetylcholine receptors at the neuromuscular junction may experience a delayed recovery as a result, which can lead to atelectasis, aspiration pneumonia, and the requirement for extended mechanical ventilation.[3,4] Rhabdomyolysis and hyperkalemia in denervated muscles are brought on by succinylcholine. Muscle relaxants, both depolarizing and non- depolarizing were therefore avoided.[5]

The anaesthetist continues to struggle with these individuals; therefore each patient's care needs to be tailored.

Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

References

1. Mercuri E, Bertini E, Iannaccone ST. Childhood spinal muscular atrophy: controversies and challenges. *Lancet Neurol*. 2012;11(5):443–52.
2. Arnold WD, Kassar D, Kissel JT. Spinal muscular atrophy: diagnosis and management in a new therapeutic era. *Muscle Nerve*. 2014;51(2):157–67.
3. Garvie JM, Woolf AL. Kugelberg-Welander Syndrome (Hereditary Proximal Spinal Muscular Atrophy). *Br Med J*. 1966;1(5501):1458–61.
4. Beach TP, Stone WA, Hamelberg W. Circulatory collapse following succinylcholine: report of a patient with diffuse lower motor neuron disease. *Anesth Analg*. 1971;50(3):431–7.
5. Azar I. The response of patients with neuromuscular disorders to muscle relaxants: a review. *Anesthesiology* . 1984;61(2):173–87.

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