The Use of Sugammadex in an Infant with Larsen Syndrome

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Abstract: We describe the case of a 2.5 kg weighing infant with Larsen syndrome that we used sugammadex for reversal of deep neuromuscular blockade. Sugammadex was superior to classical reversal agent neostigmine because of the effect on deep neuromuscular blockade and less side effects. Our patient had cervical instability, small for age and also entubation difficulty with facial anomaly. We discussed the use of sugammadex in Larsen syndrome infant. We wanted to have a safer extubation period and administered sugammadex for reversal of deep neuromuscular blockade. This case has an importance about the first time use of sugammadex such a small infant with Larsen syndrome.

Keywords: Sugammadex, Larsen syndrome, Infant.

INTRODUCTION

Dr. Loren J. Larsen first wrote his description about Larsen syndrome in 1950: multiple congenital dislocations associated with characteristic facial, hand and feet anomalies [1]. It is a complicated syndrome because of the issues relating to respiratory, musculoskeletal, cardiac and central nervous systems [2]. We would like to present a case report of using sugammadex successfully in a difficult entubation Larsen syndrome infant weighing 2.5 kg. The complications during anesthesia with this syndrome can be interesting, especially for pediatric anaesthesiologists.

CASE REPORT

A 3-month-old child weighing 2.5 kg was listed for elective bilateral knee dislocations. Her physical examination showed a small for age 3-month-old child. Her facial appearance was prominent for flat face, widely spaced eyes, depressed nasal bridge and prominent forehead. Airway evaluation indicated Mallampati class 4 airway. She was afebrile, heart rate 160/min, blood pressure 60/30 mmHg, and respiratory rate of 27/min and oxygen saturation 95%. Pulmonary examination revealed a barrel shaped chest with flared ribs. Neurological examination showed generalized hypotonia with poor crying voice. Pediatric cardiology department examined her and echocardiography report was secundum atrial septal defect.

The patient was premedicated with 0.03 mg midazolam intravenously and placed on an operating table with a heater under cover. Anesthesia was induced with sevoflurane by mask because of easy toleration and less myocardial depression when compared with the other inhalational anesthetic agents [3, 4]. After standard monitorization, anesthesia was induced and maintained with nitrous oxide (50%), oxygen (50%), 8% sevoflurane and maintained with %2.5 with the same inhalational agent. At the same time with induction, intravenous access was obtained. We took special precautions for positioning of the big head when compared with the body. Mask ventilation was not convenient in spite of airway device, so we had to administer 0.8 mg/kg rocuronium intravenously. 90 Seconds after the induction of muscle relaxant, we performed the first attempt at direct laringoscopy with a Miller size 0 blade. In the second attempt after the aspiration of blood, we intubated with a 3 mm tracheal tube without cuff. Intubation was really difficult and we could only see the tip of the epiglot. The view was proper with score 3 according to the Cormack Lehane scoring system. Oxygen saturation was 84% simultaneously. If this technique was proved unsuccessful, the backup plan was to perform video laryngoscopy. After intubation we gave our attention to the distended stomach of the child because of the mask ventilation. We administered 2 mg methylprednisolone intravenously because of edema, possibility of the vocal cords with the difficult intubation. We aspirated the air in stomach with a nasogastric tube. Operation lasted nearly 90 minutes. Because of the difficult entubation and ventilation, a discussion took place about the potential role of sugammadex and the patient’s age. We read with interest the report of Buchanan et al. [5] who reported sugammadex used successfully in a 7-month-old infant. We decided to administer 4 mg/kg sugammadex intravenously in the first instance, with

the option to increase the dose if required. We administered 10 milligram of sugammadex intravenously, with rapid return of airway tone and strong respiratory effort. Approximately 1.5 minute after administration of the drug, 3-month-old infant with Larsen syndrome was able to breathe adequately without airway adjuncts.

**DISCUSSION**

Larsen syndrome is a rare congenital defect in collagen formation that occurs in 1 in 100000 births, with equal gender incidence. This syndrome inherited in either autosomal dominant or autosomal recessive patterns and also present as sporadic cases [6, 7]. Generally it is characterized by joint dislocations (hip, knee, elbow), club foot, dwarfism, unusual face with flat faces, frontal bossing, depressed nasal bridge, widely spaced eyes and prominent forehead. Major respiratory malformations are tracheomalacia and bronchomalacia [6]. This syndrome includes spine anomalies like vertebral malformation in the form of failure of segmentation of vertebra and hypoplasia of cervical vertebra. Progressive kyphoscoliosis and cervical spine instability can be seen related to these deformities. Skeletal deformities like dislocations of multiple major joints and hand and feet abnormalities have been described in this syndrome. All these malformations may cause difficult ventilation and entubation situations [8].

Dysmaturity of collagen fibres and striking deficiency of dense mature collagen bundles may be the cause of cardiovascular anomalies like aortic dilatation, atrial septal defect, ventricular septal defect, patent ductus arteriosus, and mitral valve prolapse. These cardiac defects necessitate a thorough clinical cardiac evaluation and an ECG. Prior history of cardiac defects or presence of a murmur requires cardiology consultation [9].

Sensitivity to non-depolarizing muscle relaxants is higher in infants than adults [10]. We preferred 0.8 mg/kg rocuronium for rapid muscle relaxation. We had an advantage that the effect of

**CONCLUSION**

In conclusion sugammadex is a safer choice for reversal of muscle relaxant in Larsen syndrome patients.

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because it permits rapid recovery of respiratory effort and causes less side effects than neostigmine.

**Learning points**
- Larsen syndrome has issues pertinent to anesthesiology relating to the musculoskeletal, respiratory, cardiac and neurological systems.
- Sugammadex can be used effectively and safely in the infant age group.
- Sugammadex is good choice when the speed reverse of muscle relaxant needed.

**REFERENCES**