

(CASE REPORT)



## Anesthetic management of paediatric cataract surgery with Marinesco-Sjogren Syndrome

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### Abstract

Marinesco-Sjogren syndrome (MSS) is a rare autosomal recessive disorder characterized by cataracts, muscle weakness, mental retardation, and cerebellar ataxia. Patients with MSS may be posted for surgery early in the age for cataract aspiration, which requires unique anesthetic considerations involving myopathy, hypotonicity, growth and mental retardation.

**Keywords:** Marinesco-Sjogren syndrome (MSS); Cataract; Paediatric; Myopathy; Respiratory Depression; Ataxia.

### 1. Case report

A 8-year-old child weighing 16kg (<10th percentile for his age) was posted for lens aspiration and IOL placement of right eye. He was born to 3rd degree consanguineous marriage, and had similarly affected younger sibling who was also advised for lens aspiration and IOL placement. His birth history was uneventful, But child had delayed mile-stones and global-developmental-delay with decreased visual-acquity (RE>LE). The child had difficulty in holding objects and showed high stepping scissoring gait, for which he was evaluated by pediatrician and was diagnosed as Marinesco Sjogren syndrome (MSS).



**Figure 1** Hypotonia noted in both upper and lower limbs.

### 2. Examination

PR: 112 BPM, CRT: < 3 sec, BP: 96/66 mmHg, Spo2: 97%. MRI Brain showed mild diffuse cerebral atrophy. There was hypotonia of both upper and lower limbs along with scoliosis. Roving type nystagmus seen. All routine investigations

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were normal. Airway examination was normal. On auscultation b\l chest was clear and S1S2 heard with no added sounds or murmur.

### **2.1. Anaesthetic management**

Child parents were explained about the need for surgery and after obtaining written informed consent, Child was shifted to Operation theatre, by providing an audio-visual exposure using a smartphone, so that he could get accustomed to OT environment and also the separation anxiety will be reduced. Monitors like ECG, SPO<sub>2</sub>, and NIBP were connected. The child was preoxygenated and premedicated with glycopyrolate, fentanyl, and midazolam. And then induced with propofol, which was deep enough to tolerate peribulbar block. The block was given by using 0.5% Bupivacaine + 2% lignocaine. Here the initial sedation was enough to maintain the child immobile intraoperatively thereby providing adequate surgical period. At the end of the surgery, The child was observed in recovery room for an hour and then shifted to ward. Child got discharged to home the next day. The Recovery period was uneventful.

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### **3. Discussion**

MSS is characterized by myopathy, mental retardation, cerebellar ataxia and cataract. Literature regarding the anesthetic management of patients with MSS described prolonged recovery following administration of vecuronium for intubation and requirement of post-op ventilation. Therefore to avoid these complications in our case, who was posted for cataract aspiration and IOL placement of right eye, Peribulbar block under propofol induction with monitored anaesthetic care was planned as the surgery was limited to ocular area. Child was premedicated with glycopyrolate & sedation maintained with midazolam and fentanyl, which was adequate enough to avoid voluntary limb movements with maintaining spontaneous ventilation thereby avoiding the need for intubation. General-Anaesthesia, was only kept reserved as a backup and to be used only if necessary. Paracetamol suppository was used as analgesic thereby avoiding other opioids, which could delay the recovery. Problems associated with scoliosis such as difficulty in positioning the patient. GA and intubation problems, such as risk of malignant hyperthermia and ventilation perfusion mismatch were also avoided just by adopting a slight change in our anaesthesia plan.

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### **4. Conclusion**

Patients with MSS show delayed recovery from NMB's along with increased risk of malignant hyperthermia during GA. Respiratory depression and the need for ventilatory support are common in them due to muscle weakness. Therefore regional block with meticulous monitored anaesthetic care and cautious anaesthetic drug usage were helpful in avoiding these complications and also providing satisfactory intraoperative and recovery period.

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### **Compliance with ethical standards**

#### *Disclosure of conflict of interest*

No Conflict of interest to be disclosed.

#### *Statement of informed consent*

Informed consent was obtained from all individual participants included in the study.

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### **References**

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