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## Anesthetic management in a patient with muscle-eye-brain syndrome

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**Introduction:** Muscle-eye-brain syndrome is one of the rare congenital autosomal recessive muscular dystrophies and was first described in 1974 in Finland. Overall the world, only 30 cases have been reported, most of whom were in Finland. 12 cases have been reported from Turkey so far. In this report, anesthetic management in this rare syndrome is presented.

**Case:** Pediatric surgery planned gastrostomy for a 7 year old boy with muscle-eye-brain syndrome. The patient was agitated and it was observed that muscular tone increased, head was in hyperextension and contracture developed in elbow and wrists. Patient was on lioresal for spasticity and the cause of gastrostomy opening was the difficulty in swallowing owing to hyperextension of the head. ECG, pulse oximeter and non-invasive blood pressure was used in standard monitoring in addition to placement of heat probe. Patient was sedated with sevoflurane induction and venous access was made with 22G cannula. 2.5 mg/kg propofol induction was made and subsequently 2.5% LMA was placed. In anesthesia maintenance, sevoflurane 2%, N<sub>2</sub>O 60% mixture was used. No increase was seen in body temperature and etCO<sub>2</sub> (end tidal carbon dioxide). No intraoperative problems were experienced and patient was awakened without any problems and transferred to clinic. Discussion: (POMGnT1) genes are implicated for muscle-brain-eye (MBE) disease. In MEB disease, congenital muscular dystrophy, eye anomalies (myopia, glaucoma, retinal hypoplasia), brain malformation (type 2 lysencephalia, hypoplasia in brain stem, and cerebellum, and high creatine kinase values occur. In patients, who are hypotonic at birth, spasticity develops in time. In diagnosis, clinical and MRI findings are used. In the literature, four cases have been reported in whom CK levels increase excessively after the administration of succinylcholine under general anesthesia, but the relation between malignant hyperthermia and MEB syndrome is still far from clear.

**Conclusion:** In the literature, there is not sufficient information on anesthesia practice in patients with MEB syndromes, as it occurs rarely.

### Biography

Ozkan Onal has completed his medical education at Gazi University Medical Faculty and he was specialized in anesthesiology in Hacettepe University Medical Faculty. He has more than 15 publications in reputed journals in the field of anesthesia.

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