

Sevoflurane in a child with cerebellar ataxia and central hypomyelination syndrome with hypoplastic corpus callosum

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We report a rare case of cerebellar ataxia and central hypomyelination syndrome with hypoplastic corpus callosum that has never been reported in the literature and studied the response to general anesthesia and its post-operative outcome. A 10-year-old female child weighing 15 kg was posted for cataract extraction and intraocular lens implantation of both eyes at a week interval under general anesthesia. She was born to parents with second degree consanguineous marriage. Clinically, patient had microcephaly, underweight for the age group; head turned to a side suggesting spasticity in the cervical muscles and decreased thyromental distance. The patient had severe intentional tremors that occurred even in initiating talk.

Patient had dysarthria because of spastic tongue muscles, difficulty in feeding and could not stand or walk. Patient had history of global developmental delay, spastic paraparesis, ataxia and mental retardation. There was no history of seizures. Blood investigations including complete hemogram, liver and renal profile was normal. Echocardiogram of heart was normal. Magnetic resonance imaging of brain showed diffuse hypomyelination of white matter of both cerebrum and cerebellum, anterior and posterior limbs of bilateral internal capsule and hypoplastic corpus callosum. Visual evoked potential, somatosensory evoked potential and brainstem evoked response potential was normal.

Patient was shifted to operation theater after standard fasting protocol. Parents accompanied the child to

the operation theatre to reduce the anxiety of the child. Difficult intubation cart was kept ready. All monitors; such as electrocardiography, non-invasive blood pressure, pulse oximeter and temperature probe was applied. General anaesthesia was induced with sevoflurane in an incremental manner up to 8 vol. %. Intravenous line was secured. glycopyrrolate 0.12 mg, fentanyl 20 µg and propofol 20 mg was given. Tracheal intubation was performed using cuffed endotracheal tube no.5 in the second attempt, without muscle relaxant successfully. Anesthesia was maintained with oxygen, nitrous oxide and sevoflurane 2.5 vol. % in low flow anesthesia circuit (1;1). All parameters such as electrocardiography, non-invasive blood pressure, end tidal carbon dioxide; saturation and temperature were monitored continuously. The procedure took 90 min. Trachea was extubated when patient was fully awake. A total of 15 mg propofol was given 5 min before extubation. Saturation was maintained in the post-operative period.

The child was kept under observation in the paediatric intensive care unit for 24 hours. There were no adverse events in the post-operative period. She was discharged on day 3. After a week, patient was posted for cataract extraction of other eye under general anesthesia. All standard precautions were followed as above. The procedure went uneventful and the child was discharged on day 3.

Childhood ataxia with central nervous system hypomyelination/vanishing white matter disease (CACH/VWM) phenotypes range from a congenital or early infantile form to a sub-acute infantile form (onset age < 1 year), an early childhood onset form (onset age 1-5 years), a late childhood/juvenile onset form (onset age 5-15 years) and an adult onset form. Both the childhood and juvenile forms have been observed in sibs.^[1]

The neurologic signs include ataxia, spasticity and variable optic atrophy. Chronic progressive decline can be exacerbated by rapid deterioration during febrile illness or following minor head trauma or fright.^[2] The antenatal onset form presents in the third trimester of pregnancy with oligohydramnios and decreased fetal movement.^[3] Clinical features that may be noted soon after birth include feeding difficulties, vomiting,

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hypotonia, mild contractures and cataract (sometimes oil droplet cataract) and microcephaly. Apathy, intractable seizures and finally apneic spells and coma follow. Other organ involvement can include hepatosplenomegaly, renal hypoplasia, pancreatitis and ovarian dysgenesis.^[3]

The clinical course is rapidly and relentlessly downhill; the adverse effect of stress factors is less clear. So far, all infants with neonatal presentation have died within the 1st year of life.^[3] A rapidly fatal severe form of CACH/VWM is characterized by onset in the 1st year of life and death a few months later^[4] or become comatose spontaneously or acutely following mild head trauma or febrile illness.^[5] Considering the known adverse effect of fever, it is important to prevent infections and fever as much as possible (e.g., through the use of vaccinations, including anti-flu vaccination); low-dose maintenance antibiotics during winter time, antibiotics for minor infections and antipyretics for fever are appropriate.

The main anesthetic concerns in this case were difficult intubation, malignant hyperthermia (MH) and recovery. Due to microcephaly, decreased thyromental distance and spastic neck leading to decreased cervical movement, difficult intubation cart was made ready and available. Drugs causing MH was avoided. Succinylcholine was avoided in view of myopathy and MH. Use of non-depolarizing muscle relaxant was also of concern in view of difficult intubation. Atracurium was the drug of choice if intubation was not successful without relaxant after making sure of adequate bag and mask ventilation. In general, sevoflurane and desflurane have been reported to be less potent triggers of MH.^[6] Good recovery was also of prime concern. Propofol could have also been used in maintenance of anesthesia, but we used sevoflurane as both induction and maintenance as it is known for stable hemodynamic parameters, least incidence of MH, short duration, self-resolving of hyperthermia after discontinuation and rapid recovery.

Emergence delirium of sevoflurane was avoided by giving propofol 5 min before extubation. Continuous

monitoring of core temperature and end tidal carbon dioxide was done throughout the procedure. Propofol can be used as an alternative to sevoflurane if in case of suspected MH. Since, these patients are very sensitive to fever, continuous monitoring of temperature was done in post-operative period also. Neuromuscular monitoring is done if relaxant is used and it should be given in titrated doses.

In conclusion, patient with CACH syndrome and hypoplastic corpus callosum tolerated general anesthesia with sevoflurane well with no adverse events in the post-operative period.

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