

Sevoflurane in child with carpus callosum agenesis syndrome, a series of four cases

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Key points

The clinical features most commonly seen in patients with agenesis of the carpus callosum syndrome are microcephaly, macrocephaly, seizures, developmental delay or retardation, hypotonia, spasticity and multiple somatic anomalies including arthrogryposis, eye anomalies, syringomyelia in the spinal cord and facial dysmorphisms. Main issues of anaesthetic concerns are difficult airway, altered respiratory mechanics, myopathy, gastric reflux, interaction of polypharma therapy for seizures and anaesthetic agents, associated cardiovascular disorders. The response to general anaesthesia with sevoflurane and muscle relaxants and its postoperative outcomes were observed in these unreported cases.

Abstract

Agenesis of the corpus callosum (ACC) occurs when the band of tissue connecting the two hemispheres of the brain does not develop typically in utero. It has been correlated with inherited genetic factors, prenatal infection or injuries, prenatal toxic exposures, structural blockage by cysts or other brain abnormalities, metabolic disorders fetal alcohol syndrome, craniofacial abnormalities, and chromosome errors such as unequal chromosome x inactivation or deletion of the short arm of Chromosome 1¹. It is diagnosed sometimes accidentally presenting with a broad range of clinical manifestations and no unique prognosis.² The clinical features most commonly seen in patients with agenesis of the corpus callosum or hypoplasia of the corpus callosum are microcephaly, macrocephaly, seizures, developmental delay or retardation, hypotonia, spasticity and multiple somatic anomalies including arthrogryposis, eye anomalies, syringomyelia in the spinal cord and facial dysmorphisms.³ Some syndromes that frequently include ACC are Aicardi syndrome, Andermann syndrome,

Shapiro syndrome, acrocallosal syndrome, septo-optic dysplasia (optic nerve hypoplasia), Mowat-Wilson syndrome and Menkes syndrome⁴. We report four cases of carpus callosum agenesis syndrome undergoing seven procedures under general anaesthesia with sevoflurane. Common issues of anaesthetic concerns are difficult airways, altered respiratory mechanics, myopathy, gastric reflux, interaction of polypharma therapy for seizures and anaesthetic agents, associated cardiovascular disorders, response to general anaesthesia with sevoflurane and muscle relaxants and its postoperative outcomes were observed.

Keywords: Sevoflurane, carpus callosum agenesis syndrome, difficult intubation, pediatric.

Case reports

Case 1. One and half month old female baby weighing 3kg was posted for trabeculectomy and trabaculotomy under general anaesthesia. Patient was born at full term with normal delivery, cried immediately after birth weighing 2.5 kg. Patient had facial dysmorphism, microcephaly, craniosynostosis, congenital bupthalmos of left

eye, right sided facial palsy, micrognathia, low set ears, sacral hairs and global developmental delay. Complete hemogram, liver and renal profile was normal. ECHO showed normal cardiac function. The sonography of abdomen showed mild pelvicalyceal fullness across left kidney. MRI of brain showed complete corpus callosum agenesis and colpocephaly. The procedure took about 130 minutes.

Case 2. Ten months old female baby weighing 9kg was posted for bilateral cataract under general anaesthesia at a week interval. Patient was born at full term with normal delivery, cried immediately after birth weighing 3 kg. Patient had bilateral cataract, central hypotonia, global developmental delay and lactose intolerance. There was a recent history of staphylococcal skin scalded syndrome three months back before surgery. Complete hemogram, liver and renal profile was normal. Ig G antibodies for cytomegalovirus and rubella were strongly positive. The karyotype was normal. ECHO showed normal cardiac function. MRI of brain showed hypoplastic corpus callosum with mild hydrocephalus. MRI of spinal cord showed syringomyelia in cervical and dorsal spinal cord sparing a small portion of upper dorsal cord. Procedure took around 100 minutes.

Case 3. Three year old female weighing 20kg was posted for electroretinography under general anaesthesia. Patient was born at full term with normal delivery, cried immediately after birth weighing 3.3 kg. Patient had seizures, myoclonic jerks, macrocephaly, quadriplegia, global developmental delay and symptomatic west syndrome. Complete hemogram, renal and liver profile was normal. MRI of brain showed complete corpus callosum agenesis, colpocephaly, fusion of bilateral thalami. Urine amino acid negative and ketones negative. EEG showed lateralized seizures from left hemisphere. Visual evoked potential showed mild conduction block in left side. Gastroesophageal reflux disease was diagnosed owing to repeated lower respiratory tract infections. Ileostomy was done 2 month back. The last episode of lower respiratory tract infection was one week ago and

she was on antibiotics. Patient had uncontrolled seizures and was on long term multiple antiepileptic (sodium valproate, liveracetam and vigabatrin) drugs with maximum dose. Sodium valproate level was normal. Patient was posted for electroretinography to rule out vigabatrin toxicity under general anaesthesia. The procedure took about 60 minutes.

Case 4. 10yr old female weighing 15 kg was posted for bilateral cataract extraction under general anaesthesia at a week interval. Patient was born at full term with normal delivery, cried immediately after birth. Patient had severe intentional tremors, global developmental delay, spastic paraparesis, ataxia without seizures. Complete hemogram, renal and liver profile was normal. Cardiac function was normal. MRI of brain shows 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 potential was normal. The procedure took about 90-100 minutes for each eye.

All patients were shifted to operation theatre with standard fasting protocol. All antiepileptic were continued on morning of the surgery. All patients were accompanied with their parents into the operation theatre to reduce the anxiety. Parameters such as electrocardiography, non invasive blood pressure, saturation and temperature probes were applied. Inhalation induction was done with sevoflurane in graded incremental manner upto 8 vol % via facemask with 100% oxygen, while monitoring the hemodynamic parameters. Intravenous line was secured in all the patients except in three year old child requiring multiple attempts. Intravenous glycopyrrolate 0.08 mg/kg, fentanyl 1.5 mcg/kg was given. Intubation was visualized and intubated with appropriate endotracheal tube without muscle relaxant in all patients except in one and half month baby where the malampatti score was 3. Succinylcholine 1 mg/kg was given in that patient and intubated with the help of stylet. Ryles tubes of appropriate size was inserted and fixed through nasal

route in all patients, suctioned and connected to the collector bag. All Vital parameters were monitored and maintained throughout the procedure. Patients were carefully positioned in supine position. All the pressure points were padded with cotton. Euthermia was maintained. Paracetamol suppository 15 mg/kg was inserted to decrease the postoperative pain and pyrexia. Patients were maintained with oxygen, nitrous oxid (1:1) and sevoflurane in closed circuit. Extubation was done in fully awake condition. Propofol one mg/kg was given 5 minutes before extubation in 3 year and 10 year child to avoid emergence delirium. All patients maintained saturation in the postoperative period. All were shifted to pediatric intensive care unit for 24 hours. Ryles tube was removed after 4 hours of extubation as oral fluids was allowed. No adverse events were noted in the postoperative period. All patients were discharged on third postoperative day.

Discussion

Agenesis of the corpus callosum occurs when the band of tissue connecting the two hemispheres of the brain does not develop typically in utero. It has been correlated with inherited genetic factors, prenatal infection or injuries, prenatal toxic exposures, structural blockage by cysts or other brain abnormalities, metabolic disorders and chromosome errors such as unequal chromosome x inactivation or deletion of the short arm of Chromosome 1¹. It is diagnosed sometimes accidentally presenting with a broad range of clinical manifestations and no unique prognosis.² The clinical manifestations of anomalies involving the corpus callosum are more dependent on the degree of disturbance of development of the brain in general and midline structures in particular than the absence of the corpus callosum itself³. Focal cortical dysplasias such as zones of disorganized lamination occur and may be epileptogenic³. Furthermore, when the callosal defect is only part of a more pervasive disorder, the clinical picture is usually dominated by the primary condition³. The clinical features most commonly seen in patients with agenesis of the corpus callo-

sum or hypoplasia of the corpus callosum are microcephaly, macrocephaly, seizures, developmental delay or retardation, hypotonia, spasticity and multiple somatic anomalies including arthrogryposis, eye anomalies, and facial dysmorphisms.³ Although only one quarter of these children will have midline facial anomalies, this is the single most frequent category of somatic malformation³. Colpocephaly is a cephalic disorder that refers to the disproportionate enlargement of the occipital horns of the lateral ventricles and is usually diagnosed early after birth due to seizures. It is a non-specific finding and is associated with multiple neurological syndromes, including agenesis of the corpus callosum, Chiari malformation, lissencephaly, and microcephaly⁵. The neonate with agenesis of the corpus callosum is most often recognized by identification of the accompanying somatic or facial anomalies³. The slightly older infant may also be identified by accompanying anomalies or during the evaluation of seizures³. Both neonates and infants may have feeding difficulties and swallowing apraxia. In children the usual clinical presentation is developmental delay, difficulty in school, or seizures, and the agenesis of the corpus callosum is identified incidentally as a result of the neuroimaging studies performed as part of the evaluation³. Agenesis of the corpus callosum is also seen as a feature of many chromosomal and genetic mental retardation syndromes as well as several hereditary metabolic diseases where the clinical presentation is dominated by the primary disease³. Some syndromes that frequently include ACC are Aicardi syndrome, Andermann syndrome, Shapiro syndrome, acrocallosal syndrome, septo-optic dysplasia (optic nerve hypoplasia), Mowat-Wilson syndrome and Menkes syndrome. The main issues of anaesthetic concern were difficult airways, altered respiratory mechanics, myopathy, gastric reflux, interaction among polypharma therapy for seizures and anaesthetic agents, associated cardiovascular disorders, response to general anaesthesia with sevoflurane and muscle relaxants and its postoperative out-

come. These patients have hard of hearing, vision impairment and impaired face processing for pain. So, the attender was accompanied with the child to reduce anxiety during induction and to avoid injury and assess pain in the post operative period. Difficult intravenous cannulation was anticipated and measures for central cannulation was considered as these patients will be on multiple antiepileptic drugs. Fibre optic intubation and difficult intubation cart was always be made ready since these patients usually have micrognathia, short neck, and spasticity. Use of succinylcholine was avoided in view of myopathy. Use of non depolarizing muscle relaxant was also avoided initially in view of difficult intubation. So it was better that intubate without muscle relaxation. Use of fibre optic bronchoscopy was considered if found unsuccessful. Inserting ryle tube after the intubation and keeping it throughout the procedure was very helpful to prevent aspiration as these patients have swallowing difficulties and gastric reflux. Carefull positioning was done as these cases have cervical instability or torticollis, contractures, weakness/hypotonia, movement disorders and seizures. Care was taken to avoid hyperventilation/hypocarbica that leads to reduction of CBF and worsening metab/EEG. Sevoflurane was used as both induction and maintenance as it is known for rapid induction, hemodynamically stable, less incidence of malignant hyperthermia, short duration of surgery and rapid recovery. Emergence delirium was prevented by giving propofol 1 mg/kg 10-15 minutes before extubation. Care in interpretation of cause or nature of twitching, tonic, shivering, confusion, and anaesthetics with effects on seizures should be considered. Interaction among polypharma therapy for seizures, anaesthetic agents and global developmental delay response in the child was kept in the mind. Possibility of status epilepticus and need for post operative ventilation was also be kept in mind. Cautious monitoring of vit parameters,

temperature, seizures was done in postoperative period in the intensive care unit

Conclusions

Sevoflurane was safely used as an induction and maintenance agent in patients with corpus callosum agenesis syndrome. Trachea was intubated without muscle relaxant with sevoflurane and fentanyl combination in majority of cases (6/7 cases). Carefull positioning, temperature monitoring, adequate ventilation, preventing aspiration by inserting ryles tube, assessing pain with hemodynamic parameters and parental presence and continuing all antiepileptic drugs in pre and post period was very helpful in successful management of cases with corpus callosum agenesis syndrome.

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