Review article

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Peri-operative management of cerebral palsy: Our experience

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ABSTRACT

Cerebral Palsy (CP) is the most common movement disorder in children. Its prevalence ranges from 1–2.5 per 1000 live births. In our hospital, average number of children with CP presenting specially for MRI imaging ranges from 100–150 per year. The clinical picture varies considerably ranging from mild monoplegia with normal intellect to severe spastic quadriplegia with mental retardation. Children with cerebral palsy present with various spastic and dyskinetic movements along with mental retardation, cognitive impairment, sensory loss, seizures, communication and behavioral disturbances, as well as chronic systemic problems and their management requires a multidisciplinary approach. An anesthesiologist plays an active role at various levels ranging from sedation for diagnostic procedures to anesthesia and pain relief for various lifestyle enhancing surgical interventions. Hence, an understanding about the etiopathology, clinical presentation and pharmacological treatment will help the anesthesiologist for hassle-free management during the peri-operative period.

Key words: Anesthesia, Cerebral Palsy, Pain Management, Peri-Operative Management, Regional Anesthesia

INTRODUCTION

Cerebral palsy (CP) is a collective term used for a group of non-progressive, but often changing motor impairment syndromes secondary to lesions or anomalies of brain arising in the early stages of its development[1]. Specific aetiology is yet to be established. Various risk factors associated include: perinatal hypoxia, infection, trauma, prematurity and genetic predisposition. In our hospital, we come across major number of cerebral palsy patients presenting for MRI as part of work up for epilepsy or delayed developmental milestones. Majority of these children are less than 1 year old. Few children present with hydrocepahalus requiring a VP shunt and a few for contracture release procedures. As anesthesiologist, we face various challenges during the peri-operative course depending upon the clinical picture of the disease. As management of patients with CP requires special considerations, we have compiled this article based on our experience along with a review of various textbooks of anesthesia and pediatrics and an online search using pubmed, google search, google scholar and conchrane database.

CLASSIFICATION OF CEREBRAL PALSY

As per the Swedish classification, there are 4 types of CP: Spastic, Dyskinetic, Ataxic and mixed varieties. Spastic CP (70%) develops following injury to cerebral motor cortex and manifests as spastic diplegia, hemiplegia and quadriplegia, usually with good intellect[2]. Dyskinetic CP develops following injury to basal ganglia, and presents as dystonia, chorea and athetosis with impaired speech and drooling of saliva. Ataxic CP follows injury to cerebellum; causing tremors, loss of balance and difficulty in speech. Epilepsy is seen in most cases of spastic quadriplegia and in 25% of those with dyskinetic CP[2].

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Spastic CP forms the major bulk of patients in our hospital. They are known to have best prognosis for functional improvement. Regional analgesic techniques prove especially beneficial for this group of patients as spasticity is the major contributor to musculoskeletal pain, contractures and joint subluxation[3,4]. Presence of severe involuntary movements and tremors make use of general anesthesia mandatory.

ROLE OF ANESTHESIOLOGIST

Management of cerebral palsy patients involves multiple specialities including paediatrics, medicine, orthopaedics, surgery; anaesthesia and physiotherapy. They require multiple surgeries for correction of secondary deformities in order to make their lifestyle more comfortable. With advances in medical technology, many of these patients reach adulthood, also, some cases of parturients with cerebral palsy have been reported. Anesthesia is required for multiple procedures (Table 1).

PRE-ANAESTHETIC CHECK-UP

Patients with cerebral palsy require a thorough pre-anaesthetic check-up for associated systemic anomalies. Multiple hospital exposures make these children irritable and unco-operative. So, history taking and examination requires a lot of patience and should be done in a friendly environment in presence of parents/caretakers.

Decreased cognitive and intellectual level along with hearing and speech defects leads to poor communication causing difficulty in developing a doctor-patient relationship. History is generally provided by the caretaker (parent/gaurdian) and obtained from previous hospital records available.

Poor oral intake may lead to anaemia, reduced immunity, electrolyte imbalance, malnutrition, reduced serum proteins and various other nutrient deficiencies.

Table 1: Common indications of anesthesia in patients of cerebral palsy

Imaging techniques	MRI of brain, spine, hips etc.
Orthopaedic surgeries	Hip reconstruction, tendon lengthening, contracture release, scoliosis correction
CNS surgeries	VP shunt, dorsal horn rhizhotomy
Dental surgeries	Extraction, restoration
GIT	GI endoscopy, feeding gastrotomy, Nissen fundoplication
ENT	Cochlear implants
Ophthalmic	Squint correction
Others	Pain management Botulinum toxin injection Intrathecal baclofen pump insertion

Table 2: Anaesthetic implication of commonly used drugs in cerebral palsy

Drugs	Anaesthetic implications
Carbamazepine[5]	• Oliguria • Hypertension, arrhythmias • Aplastic anemia, thrombocytopenia, chronic leucopenia • Sedation, dizziness • Nausea, vomiting • Hepatic enzyme induction, jaundice • High dose causes inadequate ADH secretion causing hyponatremia
Sodium valproate[5]	• Impaired platelet function • Hepatotoxicity • Bone marrow suppression
Phenytoin[5]	• Gastric irritation • Hepatic enzyme induction • Hyperglycemia • Gum hyperplasia • Peripheral neuropathy

Baclofen [6–8]	• Bradycardia and hypotension • Delayed arousal after GA • Exacerbation of effect of morphine and fentanyl • Abrupt cessation may cause seizures, hallucinations, disorientation, dyskinesias and pruritus • May be delivered via intrathecal pump which interferes with spinal anaesthesia and may predispose to infection
Benzodiazepines[5]	• Sedation • Respiratory depression • Dependence and withdrawal symptoms
Dantrolene[1,5]	• Muscle weakness, dose reduction of muscle relaxants is required • Altered LFT • Prolonged therapy may cause pleural effusion
Botulinum toxin[1,9]	• May potentiate action of muscle relaxants • Can unmask subclinical Eaton - Lambert myasthenic syndrome
Cisapride[1]	• Prolonged QT interval • Increased risk of Torsades de Pointes

Presence of irregular/loose teeth and temporomandibular joint dysfunction can lead to difficult laryngoscopy and intubation. Poor oral hygiene predisposes them to oropharyngeal infection which may be transmitted to lower airways with airway intrumentation and also aggravate post-operative sore throat following endotracheal intubation. Many patients are likely to have history of pneumonia and chronic obstructive/restrictive lung disease making intra-operative respiratory management challenging.

Bony deformities of chest wall and scoliosis along with muscle spasticity, contractures and involuntary movements make neuraxial anaesthesia techniques difficult and unpredictable.

These patients are on various medications like anticonvulsants, anti-spasticity (Baclofan, Botulinum toxin), anticholinergic, anti-reflux, antacids, laxatives, anti-depressants and anti-anxiety drugs. Many of these interfere with different anaesthetic agents and hence detailed history and compliance needs to be recorded (Table 2).

History of previous anesthesia exposure provides information regarding difficult airway, difficult neuraxial block, latex and other allergies and post-operative ICU stay with/without ventilatory support.

Pre-operative investigations include a complete blood count, renal function tests, liver function tests, serum electrolytes and serum proteins. Coagulation profile, Chest X-ray, ECG and 2-D echo is required in selective cases.

PERIOPERATIVE MANAGEMENT

Pre-Operative

Patients with CP require pre-operative optimization of associated medical condition using chest physiotherapy, antacids, bronchodilators, antibiotics, nasogastric/parentral feeding, IV hydration, correction of haemoglobin, serum protein and electrolyte levels. IV cannulation may be difficult due to soft tissue contractures, multiple punctures and uncooperative behaviour.

Written and informed consent from the parent/caretaker needs to be documented clearly after explaining the associated risks.

Children with CP have poor ability to handle oropharyngeal secretions, possibly due to hyperactive salivary glands, tongue thrusting, poor head control or impaired cranial nerve function due to pseudobulbar palsy[10]. Gastroesophageal reflux is common due to oesophageal dysmotility and spasm, poor lower oesophageal sphincter tone and spinal deformity[11]. Pre-medication with an anticholinergic like atropine/glycopyrrolate and antacid can help counter these problems. However, anticholinergics are likely to thicken respiratory secretions and increase chances of lung infection.

Beneficial effects of use of antianxiety/sedative medications to allay patient anxiety needs to be weighed against the risk of over sedation, drug interactions and respiratory depression. Benzodiazepines are useful for sedation in children with neurological deficit and seizure disorder because of their pleitropic effects: anxiolysis, muscle relaxation, sedation, and anterograde amnesia. They are effective anticonvulsants with neuroprotective effects[12].

Patient positioning is difficult due to fixed contractures. Improper positioning along with poor muscle mass, absent fat deposit and dry, thin skin can lead to pressure ulcers and nerve damage. These children are at high risk of pathological fractures due to various risk factors like poor nutrition, use of anticonvulsants, decreased weight bearing, rickets at prematurity and immobilisation. Bone mineral density in these

children is lower than the normal population[13] hence utmost care during positioning with adequate padding is essential.

Intra-Operative

Mode of anesthesia depends upon age of the patient, site and type of surgery, co-operation level, cognitive function and previous experiences. Regional anesthesia's the preferred technique either alone or in combination with general anesthesia.

Along with mandatory routine monitoring of pulse rate, ECG, NIBP, SpO_2, $ETCO_2$ and urine output; neuromuscular monitoring, monitoring of temperature, BIS values and MAC of inhalational anesthetic agents are necessary. Absolute BIS values obtained in patients with cerebral palsy are lower than their normal counterparts while awake and during anesthesia. But, they exhibit a similar pattern of change as obtained in normal children[14].

Doses of all the drugs need to be titrated according to body weight, ongoing medications, liver and renal function. Barbiturates like thiopental are known to provide neuroprotection by causing a decrease in cerebral blood flow, cerebral metabolic rate, maintain cerebral perfusion pressure, cause reductions in ischemia induced glutamate release and inhibition of intracellular calcium release. Due to its anticonvulsant properties thiopental can be the preferred agent in children with history of epilepsy[12]. Apart from routine IV route, rectal thiopental (15–25 mg/kg) has also been used for sedation of pediatric patients during CT scan[15]. Propofol is the induction agent of choice in presence of hyperreactive upper airways[10]. ED50 of Propofol was found to be reduced by approximately 25% in children with cerebral palsy as compared to normal healthy children undergoing MRI under sedation with Propofol, so dose titration according to clinical response is required[16]. It is preferable to avoid epileptogenic drugs such as enflurane, etomidate, methohexitone, ketamine, tramadol and pethidine. Etomidate increases cerebral vascular resistance and has the potential to expand the ischemic core penumbra in injured brain tissue[12]. Risk of myoclonus also precludes the use of Etomidate in these patients.

Pharmacokinetics of muscle relaxants is also altered in patients with cerebral palsy. In these cases there is constant and repeated spasm of skeletal muscles. Also, muscular denervation leads to extra junctional proliferation of acetylcholine receptors which produces an increased response to succinylcholine and a resistance to effects of non-depolarising muscle relaxants[17]. Long term phenytoin therapy causes significant change in Ach receptor number and function. Various studies have shown a decreased ED_{50} of Succinylcholine (increased sensitivity) requiring a lower dose and some resistance to Vecuronium requiring a higher dose in patients with CP[18]. On the contrary, severe myopathies can lead to prolonged action of depolarizing muscle relaxants such as Succinylcholine and a theorotical possibility of rise in serum potassium levels but this effect is not seen practically[10]. Hence, neuromuscular monitoring is essential to guide the dose of muscle relaxants and the time for reversal. However, the response to NMJ monitoring may be unreliable due to muscle hypotonia and recovery of respiratory muscles may not be as rapid as that of limb muscles.

MAC value of inhalational anesthetics is also lower in children with CP and is further reduced in those taking anticonvulsants. MAC value of Halothane was decreased by 20% in children with cerebral palsy with severe mental retardation and was further reduced by 10% in those taking long-term anticonvulsant medication[19]. Similar effect was seen with Desflurane[20]. Central nervous system impairment with upper motor neuron dysfunction of various degrees along with pain insensitivity/ indifference in patients with severe mental retardation is probably the cause for increased sensitivity to inhalational anesthetic agents. Sevoflurane, more than isoflurane, exhibits epileptogenic activity in epilepsy patients[21,22].

Liberal use of opioids is to be avoided due to increased risk of respiratory depression, nausea, vomiting and constipation. It is beneficial to use multimodal analgesia in the form of regional analgesic techniques and NSAIDS combined with opiods. Use of epidural local anesthetic combined with an opiod shows markedly reduced pain scores over first 24 hours with decreased need of post-operative ICU stay[23].

Airway management is difficult in patients with CP due to poor dentition. Temporomandibular joint dislocation secondary to muscle spasticity may be present. Patients often present with pseudobulbar palsy and/or oromotor dysfunction. Rapid sequence induction is preferred, however in an unco-operative veinless patient, gaseous induction with the patient inclined at 20–30 π head-up tilt is the only available option[10]. Endotracheal tube is preferred over supraglottic airways due to high risk of aspiration. As the children are small for age, a smaller size of endotracheal tube may be needed.

Children with cerebral palsy are more prone to hypothermia due to poor temperature regulation, lean body mass and muscle atrophy. Warming blankets and use of warm IV fluids can help prevent major change in body temperature[24].

Reversal of anaesthesia and tracheal extubation is again a challenging step due to emergence agitation, poor pharyngeal muscle tone and high risk of post-operative respiratory depression and aspiration. Excessive secretion requires frequent suctioning. One can opt to step down from endotracheal tube to laryngeal mask airway or directly plan for an awake extubation

depending on the clinical scenario. Presence of parent/caretaker may help comfort the agitated patient. Good analgesia aids smooth extubation. Various agents like midazolam, clonidine and dexmedetomidine have been used to reduce emergence agitation in children. Use of Dexmedetomidine 0.5 μgm/kg IV has been shown to reduce intra-operative Sevoflurane requirement, extubation time, emergence time and ICU stay[25].

Post-Operative

Patients with cerebral palsy require observation in a high dependency unit during the post-operative period. Most common post-operative complication in these patients includes hypothermia and hypotension. Others are delayed emergence, aspiration pneumonitis, airway obstruction, bronchospasm, laryngospasm, epilepsy and neuropathy[26]. Post-operative chest physiotherapy will help those with poor cough and recurrent chest infections. Decreased gut motility makes them prone to constipation, often necessitating use of laxatives. Severe cases may also need post-operative assisted ventilation and even tracheostomy during the weaning process. Vigilant nursing care, IV hydration, feeding through gastric tube or parentral nutrition, antibiotics, anticonvulsants and effective analgesia aid post-operative recovery.

Post-Operative Pain Management

Post-operative pain assessment and management is difficult in children and is further complicated by mental retardation in patients with CP. Children with cognitive impairment are unable to adequately communicate their level of pain, and undertreatment of pain leads to increase in agitation and irritability among them. Validity of VAS scoring and FACES scale is controversial and depends highly on the intellectual level of the patient. Revised FLACC (Table 3)[27] and PAICP[28] scores can be used for pain assessment wherever possible. PAICP score uses various images of painful and painless day-to-day activities and patients are asked to tell the level of their discomfort during these activities. It is particularly useful for patients undergoing hip reconstruction surgery. Correct assessment of pain are important as fear of respiratory depression generally leads to restriction of opioid dose by the care providers[29] thereby providing inadequate analgesia.

Table 3: FLACC-Revised (revised descriptors for children with disabilities shown in [brackets])

Categories	0	1	2
Face	No particular expression or smile	occassional grimace or frown, withdrawn, disinterested [appears sad or worried]	Constant grimace or frown. Frequent to constant quivering chin, clenched jaw. [distressed looking face: expression of fright or panic]
Legs	Normal position or relaxed	Uneasy, restless, tensed [occassional tremors]	Kicking or legs drawn up [marked increase in spasticity, constant tremors or jerking]
Activity	Lying quitely, normal position, moves easily	Squirming, shifting back and forth tense. [mildly agitated (eg. head back and forth, aggression); shallow, splinting respiration; intermittent sighs]	Arched, rigid or jerking [severe agitation, head banging, shivering (not rigors), breath holding, gasping or sharp intake of breath, severe splinting]
Cry	No cry (awake or asleep)	Moans or whimpers; occassional complaint [occassional verbal outbursts or grunts]	Crying steadily, screams or sobs, frequent complains [repeated outbursts, constant grunting]
Consolability	Content, relaxed	Reassured by occassional touching, hugging or being talked to, distractable	Difficulty to console or comfort [pushing away caregiver, resisting care or comfort measure]

Post-operative muscle spasms are most common cause of pain following contracture release procedures[30]. Surgically exposed nerve endings and muscle stretch receptors activate local spinal reflexes resulting in muscle spasm and thereby causing pain[31]. Epidural anesthesia is the most effective method in immediate post-operative period[32]. Local anaesthetic along with opiods and α-agonists (Clonidine, Dexmedetomidine) helps to relive spasm and hence cause pain relief.

Various other drugs like intrathecal Baclofen, botulinum toxin injection into the affected muscle, tizanidine, dantrolene, vigabatrin and benzodiazepines are used with variable benefits. Those not responding to medical therapy require a selective dorsal rhizotomy[31].

Patient controlled analgesia is not an option due to poor communication and continous infusion rates of analgesic agents need to be preset. Insertion of pain pump catheter at the surgical

site and delivery of local anesthetic solution through it has also been used successfully for post-operative pain relief[33]. Benefits of systemic opioid usage need to be weighed against the risk of respiratory depression, nausea, vomiting and constipation.

REGIONAL ANAESTHESIA IN CEREBRAL PALSY

It is difficult to conduct a case under sole regional anesthesia in children with cerebral palsy due to unco-operative behaviour. Bony deformities of chest wall, scoliosis, fixed muscle contractures and inability of the patient to stay still for long time due to athetoid movements pose difficulty in providing regional anesthesia. With the advent of ultrasonographic techniques, we can overcome these problems. Altered curvatures of spine lead to variable and unreliable effects of local anesthetic agents.

Spinal anesthesia alone or in combination with light sevoflurane anesthesia is a reliable technique in selected children with cerebral palsy undergoing orthopaedic procedures[34]. Single shot caudal epidural, continuous epidural, single shot sciatic nerve block, continuous sciatic block, infraclavicular brachial plexus block and popliteal nerve block have been used with IV Propofol sedation and resulted in an awake and pain free child at the end of surgery who can breathe deep and cough effectively[31].

Use of combined general and regional anesthesia can provide excellent pain relief and decrease the dose of sedative anesthetic agents thereby reducing post-operative complications. Epidural analgesia is looked upon as the first choice for peri-operative pain management in children with cerebral palsy[30,32]. Also, as per studies, caudal block reduces the requirement of Sevoflurane by 36% in children with cerebral palsy undergoing lower limb surgery[35].

PARTURIENT WITH CEREBRAL PALSY

Few case reports are available reporting uneventful cesarean delivery in patients with cerebral palsy. Both spinal and general anesthesia has been used successfully in these cases. Regional anesthesia in such patients can remove inhibition of athetoid movements and trigger a harmful athetoid crisis, which in turn can precipitate fetal hypoxia[36]. However, Halemani K R et al.,[37], report successful use of spinal anesthesia for cesarean delivery. Aiudi C et al.,[38], report two cases undergone Selective Dorsal Rhizotomy (SDR) during childhood, who underwent cesarean section under spinal anesthesia with smooth peri-operative course. Ali Sakr Esa W et al., have reported a case of epidural labour analgesia for a woman with CP with an intrathecal Baclofen pump in situ[39].

SUMMARY

Anesthetic management of a patient with cerebral plasy is a challenging job as each patient presents with a different profile and requires individualised management. In a nutshell, the various problems encountered are pediatrics age, poor cognition, epilepsy, poor nutrition and hydration state, difficult airway, altered pharmacokinetics of anesthetic and analgesic agents, drug interactions, soft tissue contractures and bony deformities, latex allergy, hypothermia and multiple exposures to medical and surgical treatments. A detailed review of history and investigation is excellent guide for management. These patients require a calm, patient and skilled clinician for management of various problems.

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