Indian Journal of Anaesthesia 2008; 52 (4):397-403 **Special Article**

The Child with Cerebral Palsy and Anaesthesia

A Rudra[1], S Chatterjee[2], S Sengupta[3], A Iqbal[4], S Pal[5], R Wankhede[6]

Summary

Cerebral palsy (CP) is the result of an injury to the developing brain during the antenatal, perinatal or postnatal period. Clinical manifestation relate to the areas affected. Patients with CP often present for elective surgical procedures to correct various deformities. Anaesthetic concerns of anaesthesia are intraoperative hypothermia , and slow emergence. Suxamethonium does not cause hyperkalaemia in these patients, and a rapid sequence induction may be indicated. Temperature should be monitored and an effort made to keep the patient warm. Cerebral abnormalities may lead to slow awakening; the patient should remain intubated until fully awake and airway reflexes have returned. Pulmonary infection can complicate the postoperative course. Postoperative pain management and the prevention of muscle spasms are important and drugs as baclofen and botulinum toxin are discussed. Epidural analgesia is particularly valuable when major orthopaedic procedures are performed.

Key words **Anaesthesia, Analgesia, Cerebral palsy.**

Cerebral palsy (CP) is a common cause of childhood disability. It is defined as "a group of nonprogressive, but often changing motor impairment syndromes secondary to lesions or anomalies of brain arising in early stages of its development"[1].

It is a static encephalopathy and excludes all progressive neurobiological disorders. Other neurological deficits are frequently associated and they add to the disability caused by the motor deficit.

An understanding of the aetiology, clinical problems and their management, including knowledge of the drugs used in this condition will help the anaesthesiologist to manage these patients better during anaesthesia and the perioperative period.

Etiology

Although genetic abnormalities, perinatal anoxia, infection, and trauma have been proposed as etiologic factors in CP, no clear simple cause has been identified. However, a notable feature of CP is that infants generally have very low birth wright[2]; especially birth weight is $\leq$ 1500 gm[3]. Cerebral palsy in the premature infant is commonly due to periventricular leucomalacia as a result of periventricular haemorrhages. Other hypotheses in the term infant include antenatal infection, thyroid disease, and neuronal migration disorders. Postnatal causes include meningitis, viral encephalitis, hydrocephalus, and trauma[4].

Epidemiology

It is the leading cause of childhood motor disability and the prevalence varies from 1.5 to 2.5 per 1000 live births in developed countries. Despite improved antenatal and postnatal care, this incidence has not changed in recent years, partly due to increased survival in premature neonates and partly because neonatal asphyxia is not as common a cause as previously thought. Exact incidence and prevalence figures from our country are not available.

Classifications

Cerebral palsy can be subdivided into several types based on predominant motor pattern into four groups according to symptoms[5] shown in Table 1.

1. Hon. Consultant Anaesthesiologist, Apollo Gleneagles Hospital, Kolkata., 2. Assistant Professor of Anaesthesiology, Medical College & Hospital, Kolkata., 3,6. Consultant Anaesthesiologist, Apollo Gleneagles Hospital, Kolkata., 4,5. P.G. Student, Apollo Gleneagles Hospital, Kolkata. **Correspondence to** : A. Rudra, 1, Shibnarayan Das Lane, Kolkata – 700006,
E mail : sumanc_24@yahoo.co.in *Accepted for publication on:3.6.08*

Table 1 Classification of cerebral palsy according to symptoms

Spastic (70%) : Lesion in cerebrum. Includes quadriplegia, diplegia, hemiplegia. The number of extremities affected (and the degree of spasticity) correlates with level of intelligence.
Dyskinetic (10%) : Lesion in basal ganglia. Includes dystonia (twisting position of torso), athetosis (purposeless movements of extremities), and chorea (quick, jerky proximal movements of extremities).
Ataxic (10%) : Lesion in cerebellum. Includes tremor, loss of balance, and speech.
Mixed (10%) : Lesion in cerebrum and cerebellum. Includes spasticity and athetoid movements.

Associated developmental disabilities

Almost all children with CP will have at least one additional developmental disabilities. Such disabilities include mental retardation, epilepsy, and visual, hearing, speech, cognitive, and behavioural abnormalities, as well as the chronic systemic problems resulting from their disease (gastrointestinal, respiratory and orthopaedic). Generally, the likelihood of such deficits is increased in those with a greater degree of spasticity involving a greater number of limbs.

Diagnosis

The diagnosis of CP is essentially clinical. In severe and long-standing cases, the diagnosis of CP is not difficult. Diagnosis of mild cases requires experience. Some observations and signs are useful in prediction and early identification of CP(Table 2).

Table 2 Signs useful for early diagnosis of CP

• Decreased spontaneous motility
• Stereotyped abnormal movements
• Delayed social smile
• Excessive extensor tone, dystonia
• Delayed appearance of postural reflexes and developmental milestones
• Persistent asymmetry in posture, movement, and reflexes.
• Associated signs like roving eyes, no visual following, nystagmus, persistent squint, and lack of auditory response.

Anaesthesia

Patients with CP require special consideration because of their various disabilities. It must not be forgotten that they have feelings and emotions which are sensitive to the way they are handled and their understanding may be greater than seems apparent on first meeting. These children often require multiple surgical procedures. Orthopaedic operations to improve function of the extremities are common, and some patients require surgical correction of progressive spinal deformities. Common procedures include surgical soft tissue procedures that reduce muscle spasm around the hip girdle, including an adductor tenotomy or psoas transfer and release.

Preoperative assessment

Thorough preoperative preparation is important in ensuring an uneventful perioperative course. These children are ideal for assessment by a multidisciplinary team which include anaesthesiologists. A complete history and physical examination is essential before anaesthetizing these children, including a discussion of postoperative pain management with the parents. Cognitive and communication problems may make preoperative assessment more difficult. Parents and caretakers are best to be involved during this period not only to gain information but also allay the fears of the child. The following systems may be involved and should be considered preoperatively:

Gastrointestinal

Gastroesophageal reflux is common and may cause chronic respiratory problems. Esophagitis is not uncommon and night wakening may indicate reflux or spasm. Patients may present for Nissen fundoplication, gastrostomy, or esophageal dilatation for strictures. Children who have CP have an impaired ability to handle pharyngeal secretions, and pooling of secretions in the oropharyngeal area is common. The etiology of increased salivation is caused by hyperactive salivary glands and impaired cranial nerve function[6]. Some pa-

tients may have already been placed on antisialogogue; the most common agent used is the anticholinergic, glycopyrrolate. It is necessary to suction the oropharynx immediately following induction of anaesthesia (or even before). Anticholinergics given intravenously, once this access is secured, will aid in minimizing this problem.

Respiratory

Respiratory disorders are usually a common cause of death. Recurrent pneumonias, pulmonary aspiration and chronic lung disease are aggravated by the inability to cough and poor nutritional state which decreases immune response. Respiratory pathology of some significance may go unnoticed in case where communication is poor and exercise tolerance test cannot be properly assessed because of neurological impairment.

Scoliosis may compound the problem with a restrictive lung pattern. Noisy breathing may be indicative of upper airway obstruction which may worsen on induction or postoperatively.

The airway should be assessed for potential difficult laryngoscopy because of dental caries, loose teeth, and an increased incidence of temporomandibular joint dysfunction[7].

Following intubation of the trachea, suctioning via an endotracheal tube is a good practice in children with increased secretions, especially if rhonchi or expiratory wheeze is present. Often breath sounds will clear using this simple maneuver.

Epilepsy

Epilepsy occurs in 30% of patients with CP. It is most common in spastic hemiplegia and least common in ataxic and choreoathetoid forms[8]. Anticonvulsant therapy should be continued including day of surgery, and also postoperatively and should be administered per schedule.

Visual and hearing deficits

Visual and hearing defects are present in 40% of CP patients including strabismus, myopia, retinopathy of prematurity and cortical blindness.

Intellectual disability

Intellectual disability occurs in two-third of patients with CP and is severe in 50% of those affected. Sixty percent of children with hemiplegia will have normal intelligence but it is present in less than 30% with spastic quadriplegia. Depression and emotional liability are common problems in adolescence.

Contracture

Contractures may make positioning and intravenous access difficult.

Medical therapy

Baclofen, is a GABA receptor agonist with action in the dorsal horn of the spinal cord. It is commonly given orally to reduce muscle spasms and the associated pain and may delay the development of contractures. It can also be administered intrathecally via an implantable pump, allowing a lower dose and fewer side effects[9]. Baclofen is rarely implicated in delayed awakening, bradycardia or hypotension during general anaesthesia. Abrupt withdrawal can result in seizures, hallucinations, itching, and disorientation lasting up to 72 hours[9]. Side effects such as urinary retention and hypotension may respond to reducing the dose. Pump malfunction must be considered if overdose is suspected and intravenous flumazenil and physostigmine along with respiratory support and removal of cerebrospinal fluid via lumbar puncture may be needed.

Botulinum neurotoxin (Botox), is indicated when spasticity is interfering with function. It produce reversible muscle denervation and temporary reduction in muscle tone. Generalized weakness from systemic toxicity is rare. Lower extremity injections have been given

in hamstrings or gastrocnemius muscle[10]. The onset of effect is 12 hours to 7 days with effect for 2 to 6 months. Botox injections are performed under general anaesthesia or in the older child with EMLA. It can also decrease pain scores and muscle spasm postoperatively when given prior to adductor tendon release surgery of the hip. Potentiation of muscle relaxants has not been substantiated clinically[11].

Preoperative sedation and preparation

Sedation may be valuable to decrease anxiety but children with poor pharyngeal coordination may need special precautions. Loss of airway tone and increased risk of aspiration is greater in hypotonic children. Before preoperative sedation is ordered, parents should be consulted to learn how well the child previously tolerated. However, most patient will handle premedication without incident, and reducing the dose may be more appropriate than omitting premedication altogether. If obtaining intravenous access is necessary before induction, then use of EMLA cream (lidocaine 2.5% and prilocaine 2.5%) can be helpful. Often, a combination of EMLA cream, nitrous oxide (50%-70%) with oxygen, and preoperative sedation will help in facilitating intravenous access. If premedication is chosen, oral midazolam 0.25–1 $mg.kg^{-1}$ or ketamine 3–6 $mg.kg^{-1}$ with glycopyrrolate 0.02 $mg.kg^{-1}$ may be considered. Antacids, antiemetics and drugs to reduce secretions may also be indicated. The extremities of these children are often cold and vasoconstricted. Use of a warming device, such as warm towels, will aid in vasodilatation and make it easier and quicker to access a peripheral vein. Latex allergy is more frequent in these children[12] especially if they have had multiple operations. A history of such sensitivity should be sought in all patients with CP.

Intraoperative management

Careful positioning is paramount in the child with spastic CP to avoid nerve and muscle damage. Fixed contracture may add to difficulty in positioning.

Propofol for intravenous induction is a good choice since many children with CP have reactive airway disease and propofol unlike thiopentone decreases the airway tone[13].

Tracheal intubation should be performed if this is a concern or if there is a history of gastroesophageal reflux. Children with CP are often small for their age. Tracheal tube size selection should be based on their age as this usually provides the most appropriate fit[4]. If a rapid sequence induction is considered necessary, poor cooperation may allow only a 'modified' technique is best. The risk between securing an airway rapidly and a slower inhalational induction in an uncooperative child must be balanced.

Drug responses may vary from the normal response in the child with CP. There may be resistance to non depolarizing muscle relaxants[14], although this is of doubtful clinical significance. Sensitivity to suxamethonium probably does not occur[15]. Train-of-four responses in a severely affected limb may not represent the actual state of neuromuscular blockade[16] and the degree of block can be underestimated.

Mean alveolar concentration (MAC) of halothane are lower in children with CP when compared with normal children[17]. The MAC of halothane is 0.9 in healthy children, in children who have CP, it is 0.71. It is interesting to note that children who have CP and who were receiving anticonvulsants had an even lower MAC; 0.63. Similarly, anaesthetic agents lead to a greater hypnotic state in these children as evidenced by lower bispectral index score (BIS) when exposed to similar concentrations of sevoflurane and compared with patients who are otherwise normal[18].

Hypothermia is a problem due to hypothalamic dysfunction and the malnourished child. Often, these children will come to the operating room with borderline hypothermia ($<35.0^0C$). Measures to conserve temperature should include warming of intravenous and irrigating fluids plus warming blankets or forced air warmers. Otherwise, it is likely that $1–2^0C$ could be lost be-

fore the patient is prepared and draped.

Combined general and regional anaesthetic techniques are highly recommended especially in lower limb surgery or dorsal rhizotomy where patients are susceptible to severe postoperative muscle spasms. In patients undergoing infraumbilical orthopaedic procedures or Nissen fundoplication, a continuous epidural infusion of 0.2-0.5 $ml.kg^{-1}.hr^{-1}$ of 0.1% bupivacaine with fentanyl 2 $mcg.ml^{-1}$ for 72 hours provided satisfactory pain control[19]. However, due to high incidence of nausea and vomiting with opioid, use of bupivacaine 0.125% with clonidine 2.5 $mcg.ml^{-1}$ at 0.2-0.3 $ml.kg^{-1}.hr^{-1}$ provide better analgesia, less muscle spasm and vomiting[4]. Clonidine may also decrease spasticity. In clinical practice, narcotics appear to have greater potency in children who have CP. Doses need to be reduced, and greater vigilance is necessary to ensure maintenance of a patent airway post extubation.

Patients undergoing scoliosis surgery may have a higher volume of blood loss than idiopathic scoliotic patients, especially if anterior and posterior procedures are performed[20]. More segments are usually involved with the intramuscular scoliotic fusion procedures[21]. A recent report shows that cerebral palsy patients may have abnormalities of coagulation factor levels. Platelets may be borderline low because of prolonged use of anticonvulsants and their function may also be affected by anticonvulsants[22,23].

Postoperative care

The immediate postoperative period can present specific problems in the recovery room for children suffering from CP.

- Emergence from anaesthesia may be delayed due to hypothermia. Delayed emergence may also be due to residual volatile anaesthetic agents. MAC is reduced relative to normal controls and it is conceivable that MAC awake is also reduced.

- Drooling, if present preoperatively, require frequent suctioning for maintenance of airway. Moreover, careful attention is needed to protecting the airway in children with pseudobulbar palsy from regurgitation of gastric fluids and vomiting.

- Irritability on emergence from anaesthesia is not uncommon. This may be due to pain, urinary retention or being in an unfamiliar environment. Parental presence in the recovery room can help with the latter.

- Children, who suffer from epilepsy may be unable to have orally administered anticonvulsants if they are nil by mouth or experiencing postoperative nausea and vomiting. Most anticonvulsants, however, have long elimination half-lives (24-36 h) and, if their levels are within the therapeutic range, a 24 hour period can elapse without significantly increasing the risk of seizures.

- Those with poor cough, recurrent respiratory infections, and impaired clearance of secretions will benefit most from postoperative chest physiotherapy.

- Contractures can cause difficulties with positioning. If epidural analgesia is used for postoperative pain management, prolonged pressure over bony prominences may lead to troublesome skin breakdown, especially in malnourished children.

- Children with CP are prone to constipation. Oral, intravenous, epidural opioids may compound this problem. Therefore, attention to normal bowel habits for that child and the prescription of laxatives, supplemented by enemas, may be necessary.

Postoperative pain management

The surgical procedures, particularly those for relieving spasticity, are extremely painful. Therefore, adequate pain relief is important in the postoperative care of cerebral palsy patients. Moreover, assessment of pain in these children is very difficult because of conceptual problems associated with intellectual disability or poor verbal communication skills. Behavioural indicators such as facial grimacing, groaning, moaning or altered sleep patterns may be present in the non-pain state and can be inconsistent and difficult to interpret.

Moreover, postsurgical pain must be differentiated from hunger, anxiety, discomfort due to positioning and other benign causes, e.g., headache toothache, otitis media, and menstrual pain in females.

Postoperative pain following extensive lower limb orthopedic surgery is treated in these children with continuous epidural (caudal or lumber) infusions with[16]:

- Lidocaine (1.5 to 2 $mg.kg^{-1}.hr^{-1}$) plus fentanyl (0.5 $mcg.kg^{-1}.hr^{-1}$).
- Bupivacaine (0.625 to 1 $mg.ml^{-1}.hr^{-1}$) with or without fentanyl (0.5 $mcg.kg^{-1}.hr^{-1}$).
- Hydromorphone (2 to 4 $mcg.kg^{-1}.hr^{-1}$)

Muscle spasms are virtually universal and are treated prophylactically with intravenous diazepam 0.1–0.2 $mg.kg^{-1}$ or addition of clonidine to the postoperative epidural infusion at a dose of 0.08 to 0.12 $mcg.kg^{-1}.hr^{-1}$.[16]

Patients with CP who undergo extensive surgical procedures that necessitate analgesic infusions should be monitored as far any child receiving analgesia by the same route.

Conclusion

Cerebral palsy may be due to antenatal, perinatal or postnatal causes. Knowledge of pathology associated with different types of CP, related conditions and their drug treatment will enable the anaesthesiologist to predict likely problems which might impinge on anaesthesia and the perioperative period. The choice of anaesthetic drugs should be tailored to the individual need and surgical regiment. Severely affected children may have gastroesophageal reflux and problems with pharyngeal coordination. This may make them vulnerable to aspiration during induction and extubation, and should mandate a cautious approach to these potentially anaemic, malnourished children. Suxamethonium does not cause hyperkalaemia in the children with CP. The response to nondepolarizing muscle relaxants appears to be normal. Careful postoperative care should be provided to adequately maintain the vitals.

Postoperative pain management and the prevention of muscle spasms are important with drugs such as baclofen and botulinum toxins. Epidural analgesia is particularly valuable when major orthopaedic procedures are performed.

References

1. Salem MR, Klowden AJ. Anesthesia for orthopedic surgery. In : Pediatric Anesthesia. Gregory GA (editor) 4th edition, Churchill Liviingstone 2002 ; p. 65A.
2. Kuban KC, Leviton A. Cerebral palsy. N Engl J Med 1994 ; 330 : 188-95.
3. Inder TE, Warfield SK, Wang H, et al. Abnormal cerebral structure is present at term in premature infants. Pediatrics 2005; 115 : 286-94.
4. Nolan J, Chalkiodis GA, Low J, Olesch CA, Brown TCK. Anaesthesia and pain management in cerebral palsy. Anaesthesia 2000; 55 : 32-41.
5. Mutch L, Alberman E, Hagberg B, Kodama K, Perat M. Cerebral palsy epidemiology where are we now and where are we going ? Developmental Medicine and child Neurology 1992 ; 34 : 547-51.
6. Reddihough D, Johnson H, Staples H, Hudson I, Exarchos H. Use of benzhexol hydrochloride to control drooling of children with cerebral palsy. Developmental Medicine and child Neurology 1990 ; 32 : 985-9.
7. Pelegano JP, Nowysz S, Geopferd S. Temperomandibular joint contracture, in spastic quadriplegia : effect on oral – motor skills. Developmental Medicine and child Neurology 1994; 36 : 487-94.
8. Eicher PS. Batshaw ML. Cerebral palsy. Pediatr Clin N Am 1993 ; 40 : 537-51.
9. Albright AL. Intrathecal baclofen in cerebral palsy movement disorders. J Child Neurol 1996 ; 11 : S 29-35.
10. Forssberg H, Tendorff KB, Botulinum toxin treatment in cerebral palsy : intervention with poor evaluation ? Developmental Medicine Child Neurology 1997; 39 : 635-40.
11. Fiacchino F, Grandi L, Soliveri P, Carella F, Bricchi M. Sensitivity to vecuronium after botulinum toxin administration. J. Neurosurg Anesthesiol 1997 ; 9 : 149-53.
12. Delfico AJ, Dormans JP, Craythorne CB, Templeton JJ. Intraoperative anaphylaxis due to allergy to latex in chil-

dren who have cerebral palsy : a report of six cases. Developmental Medicine and Child Neurology 1996; 39 : 194-7.

13. Shapiro JM, Management of respiratory failure in status asthmaticus. Am J Resp Med 2002 ; 1 : 409-16.
14. Moorthy SS, Krishna G, Dierdorf SF. Resistance to vecuronium in patients with cerebral palsy. Anesth Analg 1991 ; 73 : 275-7.
15. Dierdorf SF, McNiece WL, Rao CC, et al. Effect of succinylcholine on plasma potassium in children with cerebral palsy. Anesthesiology 1985; 62 : 88-90.
16. Zuckerberg AL, Yaster M. Anesthesia for orthopaedic surgery. In : Motoyama EK, Davis PJ,(editors). Smith's Anesthesia for Infants and Children. St. Louis, MO : CV Mosby, 2006 ; pp. 756-8.
17. Frei FJ, Haemmerle MH, Brunner R, Kern C. Minimum alveolar concentration for halothane in children with cerebral palsy and severe mental retardation. Anaesthesia 1997 ; 52 : 1056-60.
18. Choudhury DK, Brenn BR. Bispectral index monitoring : a comparison between normal children with quadriplegic cerebral palsy. Anesth Analg 2002; 95 :1582-5.
19. Bren BR, Rose JB. Epidural analgesia in children with cerebral palsy. Can J Anaesth 1998; 45 : 1156-61.
20. Bobechko WP. Blood loss in scoliosis surgery. Spine 1991 ; 5 : 75-82.
21. Brenn BR, Theroux MC, Dabney KW, et al. Clotting parmeters and thromboelastography in children with neuromuscular and idiopathic scoliosis undergoing posterior spinal fusion. Spine 2004 ; 29 : 310–4.
22. Sherard Jr ES, Couri D. Treatment of childhood epilepsy with valproic acid results of the first 100 patients in a six-month trial. Neurology 1980; 30 : 31-5.
23. Kreuz W, Funk M, Meyer-Schrod R, et al. Valproate therapy induces von Willebrand disease type 1. Epilepsia 1992; 33 : 178-84.