Cerebral palsy and anaesthesia

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Key points

Cerebral palsy describes a spectrum of movement and posture disorders which result from pathological injury to the developing fetal or infant brain.

Commonly associated co-morbidities include dehydration, malnutrition, epilepsy, gastro-oesophageal reflux and impaired lung function.

Significant side-effects and drug interactions related to polypharmacy may have implications for anaesthesia.

Frequently encountered perioperative problems include difficulties with patient positioning and vascular access.

Regional analgesic techniques to reduce postoperative pain, muscle spasms, and respiratory complications are particularly beneficial.

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Cerebral palsy (CP) is a collective term used to describe a diverse group of neurological disorders characterized by varying degrees of motor, sensory, and intellectual impairment.

The condition was first described in 1862 by an orthopaedic surgeon, William Little.[1] He observed that some children, born prematurely or after a complicated labour or delivery, developed stiff spastic legs and postulated that this was caused by cerebral hypoxia at the time of birth. In the late 1890s, Sigmund Freud postulated that many cases of CP were caused, not by complications during the peripartum period, but by abnormalities arising during fetal development. The true significance of Freud's observations was not fully realized until the 1980s when the National Institute of Neurological Disorders and Stroke published a perinatal study which suggested that complicated deliveries could only account for ~10% of all CP cases.

The life expectancy of many patients with CP now extends into adulthood and they frequently present to both specialized and general hospitals for anaesthesia and surgery.

Incidence

The incidence of ~1 in 500 live births has changed little over the past 30 yr, despite improved public health, antenatal care, and an increase in the rate of Caesarean section to prevent neonatal asphyxia. This constancy is attributed to an increase in the survival rate of premature babies, in whom there is a recognized 10- to 50-fold increase in the prevalence of CP.[2]

Aetiology

Eighty per cent of CP cases develop antenatally and 20% post-natally during the first 2 yr of life. The majority of CP cases (~70%) are associated with one or more pathogenic factors outlined in Table 1. In the remaining 30%, no pathogenesis is identified.

Pathophysiology

Advances in neurobiology and magnetic resonance imaging (MRI) have improved understanding of how the human brain develops *in* and *ex utero*. The brain undergoes a series of structural and organizational changes which can be disrupted by different pathogenic insults at different stages of its development. These insults produce unique patterns of injury which are clinically associated with characteristic neurological signs and abnormal appearances on MRI in the post-natal period. For example, bilateral spastic CP is the most common subtype of CP. Periventricular lesions can be identified on MRI in ~90% of those children born prematurely who go on to develop the clinical signs of this subtype of CP (diplegias and tetraplegias) in the post-natal period.[3]

Classification of CP

For many years, CP was classified very simply according to a patient's resting muscle tone, for example, hypertonic (spastic), dyskinetic, ataxic, or mixed. This has been superseded by more comprehensive classification systems that incorporate assessment of a patient's motor function, physical activity, and psycho-social ability. These classifications enable clinicians to plan better day-to-day care for individual patients.[4]

Clinical features of CP

Seventy to 80% of patients present with some degree of muscle spasticity. Where this affects the lower limbs, they will have an abnormal 'scissor' gait and, if still ambulatory, a tendency to walk on 'tip-toes'. Patients develop 'muscle contractures' (stiffening and shortening of muscles) as a result of differential growth rates between long bones and spastic muscle groups. These contractures result in joint deformities and dislocations which go on to cause patients considerable problems with joint pain when walking and sitting.

Although CP is primarily a disorder of posture and movement, the more severe forms impact on patients' neurological, respiratory, gastrointestinal, musculoskeletal, and urological functions.

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Table 1 Pathogenic risk factors for CP

Congenital CP (80%)
Fetal pathogenic factors
Vascular maldevelopments
Congenital genetic/metabolic disorders
Microcephaly
Fetal trauma
Neonatal asphyxia in the peripartum period (6%)
Low birth weight (<2.5 kg)
Prematurity (<32 weeks)
Low Apgar score
Multiple births
Prenatal 'TORCH' infections (toxoplasmosis, rubella, cytomegalovirus, and herpes)
Maternal pathogenic factors
Breech presentation
Pre-eclampsia
Peripartum haemorrhage
Maternal hyperthyroidism
Fetal alcohol syndrome
Acquired CP (20%)—develops during the first 2 yr of life
Intracerebral haemorrhage
Viral encephalitis
Bacterial meningitis
Hyperbilirubinaemia (Kernicterus)
Head injury
Neonatal seizures

Neurological deficiencies, cognition, and communication

CP is the most common cause (60%) of motor impairment in childhood. Two-thirds of patients will have some degree of impaired intellectual and cognitive function, the magnitude of which may be difficult to determine because of problems with communication arising from expressive language disorders and or oro-motor problems affecting speech. Communication difficulties may heighten a patient's perioperative anxiety and consideration should be given to anxiolytic premedication where appropriate.

Up to 50% of patients have either focal or generalized forms of epilepsy (particularly those with a history of neonatal encephalopathy) which may be poorly controlled. Other problems include visual and auditory impairment and abnormal touch and pain perception.

Respiratory system

A significant proportion of children born prematurely will have underlying chronic lung disease secondary to neonatal respiratory distress syndrome. There is an increased risk of aspiration pneumonitis and consequently chronic lung scarring because of swallowing difficulties, oesophageal dysmotility, abnormal lower oesophageal sphincter tone, and spinal deformity which lead to gastro-oesophageal reflux (GOR). Decreased immunity, secondary to poor nutrition, respiratory muscle hypotonia, and a weak cough in conjunction with GOR make patients more susceptible to recurrent chest infections which exacerbate the underlying chronic lung disease. In the long-term truncal muscle, spasticity can lead to scoliosis, restrictive lung defects, pulmonary hypertension, and ultimately cor pulmonale and respiratory failure.

Gastrointestinal system

Patients are generally small for their age. An inability to chew food, swallow food, or both secondary to pseudo-bulbar palsies, oro-motor dysfunction, or both coupled with hyperactive salivary glands leads to drooling and poor feeding, which in turn leads to malnutrition, dehydration, anaemia, and electrolyte imbalances. Many patients receive supplemental nutrition overnight via a naso-gastric or percutaneous gastrostomy feeding tube. Poor dental hygiene, awkward, and loose teeth are commonplace and coupled with temporomandibular joint dislocation secondary to muscle spasticity can make airway management during anaesthesia challenging. A review of previous anaesthetic records may highlight previous difficulties with airway maintenance or intubation.

Where the medical treatment of GOR with proton pump blockers has proved unsuccessful, patients may have undergone surgical fundoplication or where this has failed, oesophago-gastric dissociation.

Musculoskeletal and skin

Fixed flexion deformities of the limbs and trunk as a result of muscle contractures, joint dislocations, scoliosis, and decubitus skin ulcers can make patient positioning, vascular access, and invasive monitoring for anaesthesia challenging (Fig. 1).

Non-weight-bearing long bones become osteopenic and vulnerable to fracture. CP muscle contracts poorly when surgically incised. This can result in significant blood loss during major surgery which may necessitate the use of red cell salvage devices, antifibrinolytic drugs such as tranexamic acid, and homologous blood transfusion. Heat loss is a major consideration in CP patients because they have thin skin, very little subcutaneous fat, and atrophic musculature. Morphologically, they can be likened to 'giant neonates' because they have large surface area to body weight ratios and cannot conserve heat effectively. Often these patients come to theatre with borderline hypothermia (<35°C) and they should be actively warmed throughout the perioperative period.

Immobility limits a patient's ability to exercise and the anaesthetist's ability to assess their cardiorespiratory reserve. It is worth noting that the incidence of ischaemic heart disease in the adult CP patient population is higher than in the general adult population.

Urological disorders

A significant number of patients are incontinent due to a number of factors, including neuropathic bladder, general immobility, and, communication, learning, or both difficulties. Patients who have undergone intermittent or indwelling urinary catheterization or repeated surgery should be considered to be at increased risk of latex allergy as a result of multiple previous exposures.

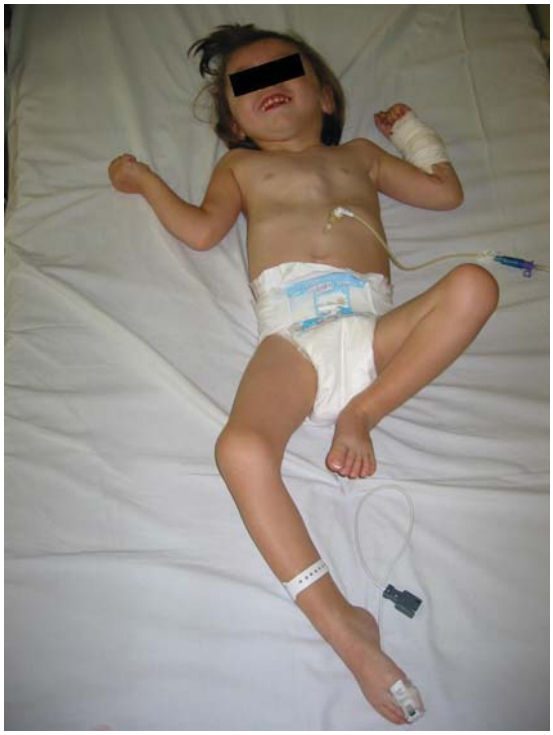

Fig 1 Typical cerebral palsy posturing.

Treatment of CP

Treatment is supportive rather than curative and targeted towards:

- improving posture and mobility by reducing spasticity, muscle spasms, and contractures;
- the symptomatic relief of the associated medical problems, for example, epilepsy, GOR, chest infections.

Combination therapies have been shown to be more successful than single-treatment protocols. They include physiotherapy, psychological counselling, occupational, speech, and behavioural therapy. These are offered in conjunction with:

- antispastic medications, for example, baclofen;
- neuromuscular denervation techniques, for example, i.m. botulinum toxin injections, radiofrequency ablation of dorsal horn ganglia and dorsal rhizotomy;
- surgery, for example, tenotomies, arthrodeses, osteotomies tendon transfer/lengthening, and multi-segmental spinal fusion procedures.

Drug therapy

The majority of patients will be taking a combination of some or all of the following medications: anticonvulsants; antispasticity medications; antacids; analgesics; antisialogogues; antidepressants; laxatives; and prophylactic antibiotics to combat recurrent respiratory and urinary tract infections. Many of these medications influence the conduct of anaesthesia, for example, some anticonvulsants are known to induce liver enzymes and over judicious use of laxatives can lead to electrolyte imbalance and dehydration.

Anticonvulsant and antispasticity medications

These are often used in combination to centrally suppress seizure activity and muscle spasm.

Anticonvulsants

CP patients are often prescribed combinations of anticonvulsants which may potentiate or inhibit one another's pharmacodynamic actions and accentuate side-effects, depending on their ability to induce or inhibit hepatic enzymatic function. Side-effects are wide ranging and include gastrointestinal and neurological disturbances, blood dyscrasias, and skin disorders.

Many of the drugs have long half-lives and a tendency to accumulation. They can reduce MAC values by up to 30%[5] and cause over-sedation, a slower recovery from anaesthesia, and airway compromise.

Antispasticity medications

Many patients rely on a degree of spasticity to maintain their posture and limb function.

Inadvertent overdose or overzealous use of antispasticity medications can lead to sedation and hypotonia, which may potentiate problems with mobility, swallowing, airway patency, and the ability to cough in the perioperative period. For this reason, these drugs tend to be reserved for use in the treatment of mild to moderate spasticity only.

Baclofen, a $GABA_B$ receptor agonist, decreases spasticity by inhibiting the release of the excitatory neurotransmitters aspartate and glutamate in the dorsal horn of the spinal cord (Rex lamina II and III). It can be given orally or intrathecally via a subcutaneously implanted continuous infusion device attached to a subarachnoid catheter inserted at T_{12}–L_1. Intrathecally, it is effective in smaller doses and is associated with fewer of the systemic side-effects which include muscle weakness, respiratory depression, incontinence, and drowsiness. The devices can last for 6–7 yr and need refilling every 3–6 months depending on their size.

These devices can be easily overlooked, and their presence should be actively sought by the attending anaesthetist. Patients who cannot tolerate oral baclofen in the immediate postoperative period may develop acute withdrawal symptoms which include anxiety, disorientation, painful acute muscle spasms, status dystonicus, seizures, bradycardia, and hypotension. These complications can be attenuated by the administration of rectal diazepam or an i.v. midazolam infusion titrated to effect. Patients should, in these circumstances, be monitored in a high dependency environment for signs of sedation, hypotonia, and respiratory depression. Botulinum toxin type A (Botox™, Dysport™) is widely used in the treatment of muscle spasticity. The toxin is injected into the bodies of the

affected muscle groups, where it binds irreversibly to the acetylcholine (ACh) receptors on the motor endplates. The onset of action is within 1–3 days and its effect lasts for 3–4 months depending on the rate of regeneration of new motor endplates.

Side-effects are rare because systemic absorption is minimal. Overdose or inadvertent intravascular injection could, in theory, lead to respiratory muscle weakness.

Other less commonly used antispasticity drugs include tizanidine, an α_2-agonist, which inhibits the release of aspartate centrally; vigabatrin which has both anticonvulsant and antispasticity actions mediated through inhibitory actions on GABA transaminase and dantrolene which inhibits the uptake of calcium in the sarcoplasmic reticulum of the affected muscle groups. Although dantrolene causes less sedation than many of the other antispasticity drugs, its use is limited by its effect on liver function.

Perioperative management of patients with CP

CP patients are anaesthetized for a wide range of diagnostic and therapeutic procedures (Table 2).

Preoperative management

One of the greatest challenges that the anaesthetist faces is to establish the patient's level of communication and cognition. This is one of the benchmarks by which the quality of the anaesthetic care will be measured by the patient and parent or guardian. It is important to recognize the invaluable contribution that the parent or guardian can make, not only in terms of their expert knowledge of the patient's past medical history, but also their ability to facilitate communication and ease anxiety levels. It is worth remembering that CP patients are regular hospital attendees and there preconceptions of anaesthesia may have been marred by previous bad experiences, for example, painful postoperative muscle spasms.

Many patients present with significant co-morbidity and an ASA classification of III or more. Emphasis should be placed on the assessment of hydration state and renal function, drug history, and cardio-respiratory function. A full history and examination should guide the anaesthetist as to what preoperative investigations and treatments are required. A full blood count, renal indices and electrolytes (within the past 6 months), and a group and save or blood cross-match would, in the authors' opinion, be a basic requirement for patients undergoing major surgery.

A chest X-ray and ECG may highlight cardiopulmonary disease in adults, but these investigations are not routinely requested in adolescents and children unless they are suspected of or known to have congenital heart disease or acute respiratory problems Patients with respiratory compromise may benefit from perioperative chest physiotherapy, antibiotics, and elective postoperative admission to a high dependency or intensive care unit. All medications, particularly those of the anticonvulsants and antispasticity variety, should be continued in the perioperative to avoid problems with acute withdrawal and worsening of seizure control. Sedative premedication is best avoided in hypotonic patients because it may accentuate problems with upper airway muscle tone and increase the risk of aspiration perioperatively.

Table 2 Types of surgery in CP patients

Diagnostic procedures: radiological imaging of central nervous and musculoskeletal systems, e.g. MRI of brain, spine and hips
Pain interventional procedures: botulinum injections for muscle spasticity, radiofrequency ablation of dorsal horn ganglion, surgical dorsal rhizotomy
Orthopaedic: management of muscle contracture and bony deformities; hip spica, neurectomy, tenotomy, arthrodesis, pelvic/femoral osteotomies, tendon transfer/tendon lengthening, spinal fusion for scoliosis correction
Dental: extractions/restorative treatment
Neurological: ventriculo-peritoneal shunt
General surgical: manual evacuation, upper and lower GI endoscopy, percutaneous endoscopic or open gastrostomy, fundoplication, oesophagogastric dissociation, hernia repairs
Ophthalmic and ENT: squint correction, grommet insertion, cochlear implants

Intraoperative management

Propofol is the preferred anaesthetic induction agent in patients with reactive upper airways, because unlike thiopental, it decreases tone in the smooth muscle of the airway. Succinylcholine is not contraindicated in patients with CP. Although some studies have demonstrated the presence of extra-junctional ACh receptors in up to 30% of CP patients, other studies have demonstrated no significant difference in potassium release after succinylcholine administration to children with CP when compared with non-affected children.[6]

Contrary to what one might expect, non-depolarizing neuromuscular blocking agents are less potent and have a shorter duration of action in patients with CP owing to the up-regulation of ACh receptors. Clinically, this is offset by the fact that these highly water-soluble drugs are redistributed through a smaller volume of total body water because these patients are often relatively dehydrated compared with other groups of patients.

In those patients with proven GOR, it would seem prudent to secure the airway with a tracheal tube. Many anaesthetists choose to perform a rapid sequence induction in order to achieve this, but there is no evidence to suggest that this is any safer than a gas induction with the patient inclined at 20–30° head-up tilt. A gas induction is often the only option in the 'veinless' uncooperative patient.

Postoperative management

After operation, CP patients are best managed on a high dependency or intensive care unit because they are prone to hypoxia, hypovolaemia, and hypothermia.

They often have a poor cough reflex and reduced respiratory drive, making them prone to secretion retention, basal atelectasis, and pulmonary collapse. Further attention is often required with

regard to blood loss, hydration, and circulatory status and temperature control.

Communication difficulties can make the assessment of postoperative pain difficult and postoperative analgesia should be based on 'continuous' rather than 'on demand' regimens. Acetaminophen and non-steroidal anti-inflammatory drugs should be given regularly and supplemented with i.v. morphine infusions or local anaesthetic-based epidural infusions or other regional blocks as appropriate. Systemically and extradurally administered opioids should be used with caution in CP patients because they can accumulate resulting in over-sedation, suppress of the cough reflex, and depression of respiration in this already vulnerable patient group.

Hypothermia, postoperative pain, and anxiety can all trigger acute muscle spasms which are often more painful and distressing to the patient than the operation itself. Caudal or epidural analgesic techniques combining extradural clonidine with a local anaesthetic agent are proving to be beneficial in combating pain associated with both the operative procedure and muscle spasms. In the authors' experience, extradural clonidine is more effective in the management of postoperative muscle spasms than a combination of either extradural or systemic opioids administered in conjunction with enteral or parentral benzodiazepines, and it produces less sedation.

Other postoperative considerations include poor nutritional status, which can lead to delayed wound healing and the presence of musculoskeletal contractures which can increase the likelihood of pressure sores developing in the immobile patient.

Conflict of interest

None declared.

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