Cerebral Palsy

Cerebral palsy (CP) is 'an umbrella term covering a group of non-progressive motor impairment syndromes secondary to lesions or anomalies of the brain arising in the early stages of its development. The clinical picture may vary considerably, ranging from mild monoplegia with normal intellect to severe total body spasticity and mental retardation; the common denominator is the presence of a motor disorder arising from a brain lesion or anomaly that is non-progressive and presents early in life.

ANESTHETIC CONSIDERATIONS:

- Risk of aspiration and appropriate aspiration prophylaxis
- Possible difficult airway
- Associated respiratory impairment and increased risk of postoperative pulmonary complications
- Difficult positioning due to spasticity and contractures
- Increased risk of bleeding in spinal instrumentation
- Increased risk of delayed emergence
- Increased frequency of seizure disorders
    - Continue antiepileptic medications in perioperative period
- Medication considerations
    - Succinylcholine does not result in hyperkalemia
    - No association with MH
    - Possible resistance to nondepolarizing NMBAs (likely not clinically significant)
    - Shortened duration of NMBAs secondary to AEDs
    - Continue baclofen – abrupt discontinuation can cause a withdrawal syndrome

ANESTHETIC GOALS:

- Preoperative optimization of respiratory status
    - Treat pneumonias
    - Aspiration prophylaxis
- Anticipate perioperative complications
    - Bleeding with spine instrumentation and consider antifibrinolytics
    - Perioperative aspiration; awake extubation
    - Prolonged emergence
- Appropriate postoperative observation

HISTORY

- Chart review, often multiple procedures
- Evidence of airway abnormalities
- Impaired ability to handle pharyngeal secretions (difficulty swallowing) - tongue thrusting, poor dentition, salivary drooling (hyperactive salivary glands 2° cranial nerve damage)
    - History of glycopyrrolate use
    - Often present for restorations or extractions
- Respiratory involvement – restrictive defect, aspiration pneumonia, recurrent chest infections (cough, dyspnea – difficult to detect if mobilization limited), reactive airway disease d/t injury to parenchyma from chronic insults
- Cardiovascular involvement with severe restrictive lung disease and resultant right-sided heart failure (dyspnea)
- GI – GERD, esophageal dysmotility (poor swallowing, night awakening)
    - Often present for Nissen fundoplication
- MSK – spasticity, dyskinesia, ataxia (muscle pain, spasms)
    - Can present for scoliosis surgery, dorsal rhizotomy and other orthopedic procedures
- CNS – epilepsy (30%), visual & hearing defects (seizure history)
- History of iron-deficiency anemia (fatigue)

PHYSICAL

- **HEENT** – dental malocclusion, dental caries, a/w exam (can be difficult)
- **RESP** – often normal, reduced air entry, bronchial breathing, wheezing
- **CVS** – parasternal heave, loud P2, tachycardia, S3 or S4, distended JVP hepatomegaly
- **GI** – dehydration, pallor, malnutrition
- **MSK** – increased muscle tone, contractures, tremor
- **CNS** – myopia, visual field defects, strabismus
- **HEME** – pallor
- **METABOLIC** – dehydration, malnutrition

INVESTIGATIONS

- **Labs**
    - CBC & differential (Fe-deficiency anemia, PLTs with valproic acid)
    - Lytes, UA, albumin (electrolyte imbalance, malnutrition)
    - ABGs (if restrictive disease)
- **Imaging**
    - EKG
    - ECHO (if right-heart involvement)
    - PFTs (if restrictive lung disease)
    - Endoscopy
- **Special**

- Gait analysis performed before major orthopedic procedures

OPTIMIZATION

- Involve parents in management, as parents have good insight into perioperative care
- Avoid unfamiliar faces if possible
- Optimize respiratory status (bronchodilators, antibiotics, physical therapy)
- Optimize nutrition, fix electrolyte imbalance
- Continue medical treatment (esp. anticonvulsants)
- Aspiration prophylaxis
- Consider antisialagogue
- Sedative premedication (few cases in which the child is predominantly hypotonic, it is prudent to avoid preoperative sedation – loss of a/w tone and increased risk of aspiration is greater in these children), previous history of sedation from parents should be sought
- Topical local anesthetic for venipuncture
- Discuss perioperative analgesia (often regional technique for lower limb surgery)
- Blood products available:
  - Spinal fusion surgery performed in CP, along with other types of neuromuscular scoliosis, is often a much more extensive procedure than spinal fusion performed for idiopathic scoliosis in normal children - the spine is surgically fused from T1 to sacrum and is associated with bleeding that amounts to one or two blood volumes of the patient
  - In general, children who have CP bleed more per segment of spine fused when compared with idiopathic scoliosis patients, reasons are unclear and often attributed to their poor nutritional status or a lifelong non-ambulatory status, there may be a role for factor replacement early (case by case basis)
  - Platelets are often borderline low in these children because of prolonged use of anticonvulsants and platelet function is also affected by anticonvulsants
  - Consider TXA or Aprotinin

ANESTHETIC OPTIONS

- Local, regional, general acceptable depending on procedure
  - Use of regional techniques for postoperative pain management makes postoperative care easier
  - Patients can be monitored for epidural management of pain in much the same way as normal patients having similar surgery
  - Use of clonidine along with bupivicaine and hydromorphone (or fentanyl) is becoming common, and it is believed that clonidine helps reduce the spasticity as well
  - Postoperative hypotension is more likely in some patients when all three agents are used

ANESTHETIC SETUP

- **Drugs**
  - **Baclofen** should not be stopped abruptly but may cause postoperative bradycardia & hTN
  - Resistance to **non-depolarizing NMB** (probably not clinically significant)
  - **Ketamine** and methohexital may be avoided in epileptic patients
  - N_2O and opiates may worsen nausea
  - **SCh** use in CP controversial (**classic answer is that SCh does not trigger hyperK in CP**)
    - Question is of abnormal potassium release
    - Dierdorf and coworker's study concluded that there was no significant difference in potassium release after SCh in children with CP when compared with normal children
      - Unfortunately, events such as hyperkalemia following SCh administration are rare so the study design used will not be expected to capture it
    - A study by Theroux and colleagues examined the potency of SCh, demonstrated increased sensitivity toward succinylcholine in children with CP
      - Showed that the effective dose (ED) of SCh to depress 50% of baseline twitch (ED50) is less in children who have cerebral palsy when compared with healthy children
      - The same relationship is true for the effective dose of SCh to depress 95% (ED95) of baseline twitch
    - In a more recent study, the same investigators examined the abnormal distribution of acetylcholine receptors in muscle biopsies obtained from children who have CP
      - Used a double-staining method in which AChE stained the limits of NMJ, which was followed by a second stain in which alpha-bungarotoxin stained the AChRs
      - Allowed the examination of the spread of AChRs in relationship to the boundaries of the NMJ
      - Up to 30% of the children had AChRs over and beyond the boundary of the NMJ
      - This direct evidence compounded by indirect evidence from clinical studies renders the routine use of SCh in children who have CP questionable at best
- **Equipment**
  - Standard CAS and 5-lead ECG
  - Temperature monitors and warming devices
    - The more affected the child, the greater the likelihood that they are unable to regulate their temperature
  - SSEPs & MEPs if scoliosis surgery
    - Often thought to be somewhat meaningless as the baseline evoked response is diminished or even absent in children who have CP
    - However, the monitoring capability is evolving, and children who have CP are being monitored with increasing frequency

MANAGEMENT OF ANESTHESIA

- **Induction**
  - ETT is better sized to age, not weight
  - Salivary secretions may make ventilation difficult
  - Overbite may make intubation difficult
  - Cannulation often difficult

- RSI may be required (GER + secretions) but often impractical
- Risk of using **SCh** in CP (best to avoid, see above), may have to give larger dose of **non-depolarizing NMB**
- **Propofol** for intravenous induction is a good choice, as many children with CP have reactive airway disease, and, propofol, unlike thiopental, decreases the airway tone
- Inhalation sometimes favored (in semi-sitting position if concerns of reflux)

- **Maintenance**
  - Careful positioning
  - Consider antiemetics
  - IV fluids
  - MAC may be lower in CP (esp. if on anticonvulsants)
  - Use warming devices
  - Consider regional (epidural) techniques for lower limb surgery
- **Emergence**
  - Awake if prone to reflux

DISPOSITION & MONITORING

- Maintain normothermia
- Susceptible to nausea and vomiting
- Avoidance / treatment of muscle spasm (intravenous diazepam, epidural clonidine)

COMPLICATIONS

- Latex allergy
- Hypothermia
- Prolonged recovery time
- Postoperative N/V (worse with opiates)
- Postoperative muscle spasm
- Retention of secretions and postoperative chest infections

PATHOPHYSIOLOGY

- Cerebral palsy (CP) is a collection of motor system disorders including choreoathetosis, hypotonia, ataxia, dystonia, spasms, or mixed forms
- Originates from a non-progressive neurological insult sustained perinatally or before 2 years of age
- Incidence rising in the US and currently estimated to be 2.4 per 1000 live births
- Etiology of CP varies and includes any event causing cerebral injury that is non-progressive
  - Predominantly affects the motor system, and the resultant spasticity and hypertonicity are progressive
  - Notable feature is that infants generally have very low birth weight
    - Long-term studies of the outcome of very premature infants have documented significant motor, cognitive, and behavioral deficits in most of these patients, especially when birth weight is > 1500 g
    - MRIs show that such infants have decreased cerebral gray and white matter volume, as well as an increased volume of cerebrospinal fluid
    - These infants make up more than 25% of the children diagnosed with CP
- Clinically, CP can be classified into four groups according to symptoms, shown in Box 1

Box 1. Classification of cerebral palsy according to symptoms

Spastic: Lesion in cerebrum. Includes quadriplegia, diplegia, hemiplegia. The number of extremeties affected (and the degree of spasticity) correlates with level of intelligence.

Dyskinetic: Lesion in basal ganglia. Includes dystonia (twisting position of torso), athetosis (purposeless movements of extremities), and chorea (quick, jerky proximal movements of extremities).

Ataxic: Lesion in cerebellum includes tremor, loss of balance, and speech.

Mixed: Lesion in cerebrum and cerebellum. Includes spasticity and athetoid movements.

Table 1
Ashworth Scale scoring system

Ashworth Score	*Degree of muscle tone*
1	No increase in tone
2	Slight increase in tone, giving a 'catch' when the affected part(s) is moved in flexion or extension
3	More marked increase in tone but affected part(s) easily flexed or extended
4	Considerable increase in tone; passive movements difficult
5	Affected part(s) rigid in flexion or extension

- Treatment
  - Combined surgical, medical and physiotherapy approaches
  - Assessments regularly monitor motor function, independence, self-care and general care
  - Scoring systems such as the Ashworth Scale (monitoring muscle tone, Table 1) are useful tools to monitor function
  - Regular gait assessment in ambulatory patients will detect deterioration in function early enough to allow noninvasive therapy
  - Currently, a more aggressive approach to nutritional care helps prevent malnutrition, infection and renal complications

REFERENCES

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