VEPTR PLACEMENT OR EXPANSION

Surgical Specialty:	Orthopedic Surgery
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Background:

- General Considerations
  - Vertical expandable prosthetic titanium rib (VEPTR) placement is used to treat early onset scoliosis (EOS) unresponsive to conservative management, thoracic insufficiency, or chest wall defects.[1]
  - It is performed to allow for growth before definitive spinal fusion in children roughly 2-10 years old[2]
  - It involves the placement of one or two telescoping rods that are attached to the ribs at the top and the spine or hips at the bottom of the deformity
  - VEPTR prevents the progression of the scoliotic curve while allowing for normal chest and spine growth.
  - Placement of VEPTR rods is less invasive than spinal fusion surgery with less hemodynamic instability.[1]
  - These devices are expanded intermittently, every 6–9-month to allow for normal spinal growth. VEPTR expansions are performed under general anesthesia involving small incisions[1,3] and are normally done as outpatient procedures.
  - MAGnetic Expansion Control (MAGEC rods) are like VEPTR rods but use a magnet to expand the rods. These rod expansions are done in the clinic and avoid subsequent expansion surgeries[1]
- Patient Considerations
  - Patients with thoracic insufficiency syndromes may have associated cardiac and pulmonary co-morbidities due to the constriction of lung growth, resulting in pulmonary hypertension.[2]
  - Check for underlying renal abnormalities, neurologic abnormalities (i.e., Chiari malformations, tethered cord, syrinx), and bone/muscular diseases due to the patient's underlying syndrome.[4]
- Associated Comorbidities/Syndromes
  - EOS may be idiopathic or may be associated with a genetic syndrome, neuromuscular disease, or congenital malformation
    - There are a large number of uncommon conditions that are associated with thoracic insufficiency syndromes, such as Jeune syndrome, Jarcho-Levin syndrome, Ellis van Creveld syndrome, connective tissue disorders, spinal muscular atrophy (SMA), and spina bifida.[2]

Anesthetic Planning:

- Pre-Anesthetic Evaluation
    - A preoperative neurologic examination should be performed with documentation of any preoperative deficits.[4]
    - The patient may have chronic hypoxia with compensatory metabolic alkalosis and polycythemia related to their restrictive lung disease.
    - Discussions to have with surgeon/family
        - Consent family for general endotracheal anesthesia (GETA) ± an arterial line.
        - Discuss the risks associated with positioning, the possibility of tongue laceration, loss of neuromonitoring signals/wake-up test, postoperative ventilation, and rarely transfusion of blood products.
        - Discuss the need for neuromonitoring with the surgeon and neuromonitoring technician.
- Specific or Unique Room Set-Up Requirements
    - Airway
        - Evaluate for a difficult airway, especially if the patient is syndromic.
    - Drugs/Infusions
        - Consider preoperative anxiolytic: these patients will often have multiple anesthetics for VEPTR expansions.
        - If neuromonitoring is performed, choices for total intravenous anesthesia (TIVA) include
            - Propofol (50-250 mcg/kg/min)
                - with or without dexmedetomidine (0.2-1 mcg/kg/hr)
                - with or without ketamine (0.05 – 0.15 mg/kg/hr)
            - Opioid infusion
                - Remifentanil (0.05-0.5 mcg/kg/min)
                - Sufentanil (0.01 to 0.05 mcg mcg/kg/min)
                - Vasopressor of choice to support the patient's hemodynamics.
    - Monitors
        - Standard American Society of Anesthesiologists monitors, with or without an arterial line.
        - Neuromonitoring of somatosensory evoked potentials (SSEP) and motor evoked potentials (MEP).
    - PICU Bed Availability
        - Expect pulmonary status to worsen in the immediate postoperative period, so a PICU bed may be required depending on co-morbidities.

Intraoperative Considerations:

- General
    - The procedure is performed under a general anesthetic.
    - Although uncommon, significant blood loss can occur. Adequate intravenous (IV) access and a type and screen should be available for initial placement.
    - Hypothermia is common, so warming blankets and a fluid warmer should be used.[3]
- Induction
    - Inhalational induction is typical in this age group unless otherwise contraindicated.
- Positioning
    - Most commonly involves prone positioning.
    - During prone positioning, avoid chest wall compression and pressure on the eyes.[4]
    - Bite blocks should be placed to avoid tongue laceration when performing MEPs.[4]

- Maintenance
    - Maintain general anesthesia with TIVA if neuromonitoring is being performed.
    - Anesthetic plan should allow for a quick "wake-up" test if needed.
- Hemodynamic/Physiologic goals
    - Standard goals are used based on age and co-morbidities; blood pressure should be generally maintained within 20% of preoperative values.[4]
    - If loss of MEPs occurs, blood pressure may need to be increased to improve spinal cord perfusion.
- Surgical Considerations
    - Neuromonitoring is typically employed (MEPs, SSEPs). Confirm modalities planned with the surgeon and neuromonitoring technician.[4]
    - Muscle relaxation must be avoided if MEPs are monitored.[4]
- Emergence/Disposition
    - Patients may be at risk for emergence delirium due to age and repeated procedures.
    - Following initial placement, the patient is typically admitted overnight for monitoring.
- Post-op Pain
    - Multimodal-Consider a PCA/NCA.
    - Muscle spasms are common post-operatively. Consider valium[2]

Case-Specific Complications/Pitfalls

- Baseline neuromonitoring signals may be difficult to obtain secondary to underlying conditions.[4]
- Loss of neuromonitoring signals during the case secondary to spinal cord damage from mechanical distraction or ischemia is a potential complication.
- Patients may not tolerate prone positioning due to difficulty with ventilation from restrictive lung disease, which can further lead to acute right to left shunting in patients with residual intracardiac defects.
- Patients may also not tolerate prone positioning due to increased thoracic pressures reducing stroke volume and/or cardiac index.
- Although rare, an intraoperative pneumothorax may occur.[1]

References

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