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Split Cord Malformation, Type I (Bony Spur)

Introduction

Split cord malformation (SCM), though rare, is widely documented in the literature. It usually occurs in isolation but is termed **complex spina bifida** when combined with other congenital spinal anomalies.

The terms **"diastematomyelia"** and **"diplomyelia"** have been replaced by "split cord malformation," as proposed by Pang et al. SCM is further classified into:

- **Type I**: Midline bony spur with two separate dural sacs.
- **Type II**: Hemicords within a single dural sac separated by a rigid fibrous septum.

Embryological Basis

As proposed by Pang et al., the primary embryological error occurs during the closure of the primitive neurenteric canal. This leads to the formation of an **accessory neurenteric canal** through the midline embryonic disc, maintaining communication between the yolk sac and amnion. This anomaly permits mesenchymal infiltration, causing the neural tube to:

- Split into two components separated by a fibrocartilaginous or bony septum (diastematomyelia).
- Remain as a single dural tube separated by fibrous tissue (diplomyelia).

Signs and Symptoms of Occult Spinal Dysraphism (OSD)

1. Cutaneous Markers

- Hypertrichosis (hairy patch).

2. Orthopedic Deformities

- Foot and leg deformities with asymmetry.
- Scoliosis.

3. Urologic Problems

- Neurogenic bladder.

- Recurrent urinary tract infections (UTIs).
- Incontinence.

4. Neurological Symptoms (Age-based)

- **Infants**:
  - Decreased spontaneous leg movements.
  - Absent reflexes.
  - Leg atrophy hidden by baby fat.
- **Toddlers**:
  - Foot asymmetry.
  - Delayed walking milestones.
- **Older Children**:
  - Abnormal gait.
  - Asymmetric motor and sensory dysfunction.
  - Painless foot burns.
  - Upper motor neuron (UMN) signs, e.g., hyperreflexia.

Diagnostic Studies

1. **MRI**:
   - Investigation of choice.
   - Allows visualization and differentiation of neural tissue, making it ideal for congenital spinal disorders.
2. **CT Scan**:
   - Useful for 3D orientation of the bony spur.

Surgical Intervention

Aim of Surgery

The primary goal is to prevent neurological deterioration and avoid further spinal cord injury.

Surgical Challenges

- Dysraphic soft tissues (muscle, ligamentous structures, and bony abnormalities) overlying the SCM.
- Attachment of dural sleeves to the bony spur.
- Three-dimensional orientation of the bony spur.

Surgical Procedure

1. **Laminectomy**:
   - Performed from lateral to medial, leaving the middle part attached to the spur.
2. **Drilling**:
   - A high-speed, small-diameter diamond microdrill is used to thin the bony spur while avoiding dura or spinal cord injury.

- The forward-cutting rongeur No. 1 is suitable for neonates.

3. **Excision of the Spur**:
    - Initially thinned with a pneumatic drill, followed by removal with a forward-cutting punch.
    - Dysraphic elements (muscle, ligamentous tissue, and bony spur) are removed cranially and caudally, except for a small central portion.
    - Excision is performed extradurally.
4. **Bleeding Management**:
    - Venous bleeding from plexuses is controlled immediately using heavy bone wax application.
5. **Dural Repair**:
    - The dura is opened in an elliptical manner above and below the spur.
    - Ventral dural margins are sutured in the midline in front of the hemicords for a watertight closure.

Conclusion

Surgical intervention is recommended for any child with progressive neurological deterioration, motor or sensory deficits, or urinary dysfunction. Other indications include pain, spasticity, gait abnormalities, and persistent or progressive scoliosis. If a neurological deficit is already present, surgery should be performed promptly upon diagnosis.