# PEDIATRIC NEUROSURGERY

# Congenital anomalies of cranium and spinal region


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## SPINA BIFIDA OCCULTA

**Definition**
o neural defect
o usually CNS (cauda and PNS not involved)
o radiological diagnosis (not associated with neurologic defect, only bony deficit)

**Epidemiology**
o 20-30% of the general population

**Etiology**
o failure of fusion of the posterior arch

**Clinical Features**
o no obvious external markings
o no obvious clinical signs
o presence of skin dimple or hair tuft should increase suspicion of an underlying anomaly (occult spinal disraphysm)

**Investigations**
o plain film: absence of the spinous process along with minor amounts of the neural arch
o most common at L5 or S1

**Treatment and Results**
o requires no treatment

## MENINGOCELE

**Definition**
o a defect consisting of a herniation of meningeal tissue and CSF
through a defect in the spine

**Etiology**
o 2 theories
• primary failure of neural tube closure
• rupture of a previously closed neural tube due to overdistension (Gardner; unpopular theory)

**Clinical Features**
• usually no disability
• low incidence of associated anomalies and hydrocephalus

o plain films, CT, MRI, ultrasound, cardiac echo, GU investigations

**Treatment and Results**

o surgical excision (excellent results)

# MYELOMENINGOCELE

**Definition**

o a defect consisting of a herniation of meningeal tissue and CNS tissue through a defect in the spine

**Etiology - same as meningocele**

**Clinical Features**

o sensory and motor changes distal to anatomic level producing varying degrees of weakness, anesthesia, urine and fecal incontinence

**Investigations**

o plain films, CT, MRI, ultrasound, cardiac echo, GU investigations

**Surgical Indications**

o preserve intellectual, sensory and motor functions

o prevent CNS infections

**Results**

o 40-85% ambulatory

o associated with hydrocephalus (80%)

o complications: ventriculitis, ICH

# HYDROCEPHALUS IN PEDIATRICS

**Etiology**

o congenital

• aqueductal anomalies

• primary aqueductal stenosis in infancy

• secondary gliosis due to intrauterine viral infections

(mumps, varicella, TORCH) or germinal plate hemorrhage

• Dandy Walker (2-4%)

• Chiari malformation, especially Type II

• myelomeningocele

o acquired

• post meningitis

• post hemorrhage (SAH, IVH)

• masses (vascular malformation, neoplastic)

**Clinical Features**

o symptoms and signs of hydrocephalus are age related in pediatrics

o increased head circumference

o irritability, lethargy, poor feeding and vomiting

o bulging anterior fontanelle

o widened cranial sutures

o ”cracked pot” sound on cranial percussion

o scalp vein dilation (increased collateral venous drainage)

o sunset sign - forced downward deviation of eyes

o episodic bradycardia and apnea

**Management**

o similar to adults (see Hydrocephalus Section)

# CHIARI MALFORMATIONS

**Definition**

o malformations at the medullary-spinal junction

**Clinical Features**

o Type I (cerebellar ectopia): cerebellar tonsils lie below the level of the foramen magnum

• average age at presentation 41 years

• brain compression: suboccipital headache, nystagmus, ataxia, spastic quadraparesis

• foramen magnum compression syndrome (22%)

• central cord syndrome (65%)

• cerebellar syndrome (11%)

• hydrocephalus

• syringomyelia

o Type II: part of cerebellar vermis, medulla and 4th ventricle extend through the foramen magnum often to midcervical region

• present in infancy

• findings due to brain stem and lower cranial nerve dysfunction: swallowing difficulties, apneic spells, stridor, aspiration, arm weakness

• syringomyelia, hydrocephalus in > 80%

o Type III: displacement of posterior fossa structures with cerebellum herniated through foramen magnum into cervical canal (rare, usually incompatible with life)

o Type IV: cerebellar hypoplasia without cerebellar herniation

**Investigations**

o MRI or CT myelography

**Treatment**

o surgical decompression - indications

• Type I: symptomatic patients (early surgery recommended)

• Type II: neurogenic dysphagia, stridor, apneic spells

# CRANIOSYNOSTOSIS

**Definition**

o premature closure of the cranial suture(s)

**Classification**

o saggital - most common

o coronal

o lambdoid - least common

o metopic (forehead)

o multiple suture synostosis or pansynostosis

**Epidemiology**

o most cases are sporadic

o familial incidence is 2% of saggital and 8% of coronal synostosis

**Clinical Features**

o skull deformity

o raised ICP

o ophthalmologic problems  due to increased ICP or bony abnormalities of the orbit

o hydrocephalus may accompany multiple craniosynostoses

**Investigations**

o plain radiographs, CT scan (3D)

**Management**

o surgery for cosmetic purposes, except in cases of elevated ICP