Congenital Abnormalities of CNS

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Abstract: Abnormalities in the development of the neural tube result congenital anomalies of the central nervous system, which can range from mild to extensive anomalies and involve any part of nervous system. In this chapter, common neurosurgical congenital anomalies are discussed. In the early part of this chapter, cranial anomalies are summarized, including congenital hydrocephalus, cerebral aqueductal stenosis, craniosynostosis, and encephalocele. Common spinal defects like meningocele, myelomeningocele, lipomyelocele, and tethered cord syndrome are discussed in the middle of this chapter. Craniospinal anomalies, especially Chiari malformation and syringomyelia, are very important in clinical practice. Finally, arachnoid cysts are discussed briefly.

Abbreviations

AqS	aqueductal stenosis	BPC	Blakes pouch cyst
CNS	central nervous system	CSF	cerebrospinal fluid
CSO	Craniosynostosis	CT	computed tomography
CVJ	cranio-vertebral junction	DWC	Dandy–Walker cyst
DWV	Dandy–Walker variant	DWM	Dandy–Walker malformation
ETV	endoscopic third ventriculostomy	HA	Headache
HCP	hydrocephalus.	ICP	intracranial pressure
MM	myelo-meningocele	MRI	magnetic resonance imaging
OFC	occipito-frontal circumference	PICA	posterior inferior cerebellar artery
SCM	split-cord malformation	TC	tethered cord syndrome
VP shunt	ventriculo-peritoneal shunt		

1. Introduction

Disorders of the anatomy of the central nervous system (CNS) that develop in the womb and are present at birth are known as congenital anomalies of the CNS. The symptoms and prognoses for congenital malformations vary depending on their type and severity. Some are diagnosed at birth, while others may remain undetected until adolescence or adulthood.

Causes of congenital anomalies:

- Genetic factors (most common);
- Multifactorial etiology: alcohol, tobacco, smoking, drugs/medications, vitamins, environmental toxins, toxic chemicals, and several viruses that can infect the fetus while in the uterus;
- Idiopathic mechanisms.

Common congenital CNS abnormalities:

A. Primary Cranial Anomalies
   (i) Cerebral aqueductal stenosis;
   (ii) Congenital hydrocephalus;
   (iii) Craniofacial anomalies (a. craniosynostosis; b. encephalocele);
   (iv) Dandy–Walker malformation;
   (v) Arachnoid cysts;
   (vi) Others.

B. Primary Spinal Anomalies
   (i) Spinal dysraphism;
   (ii) Tethered cord syndrome;
   (iii) Klippel–Feil syndrome;
   (iv) Others.

C. Primary Craniospinal Anomalies
   (i) Chiari malformation;
   (ii) Neural tube defects;
   (iii) Others.

2. Cranial Congenital Anomaly

2.1. Cerebral Aqueductal Stenosis with Hydrocephalus

Aqueductal stenosis (AqS) is the narrowing of the Sylvian aqueduct such that the CSF flow in the ventricular pathway is obstructed (Figures 1–3).

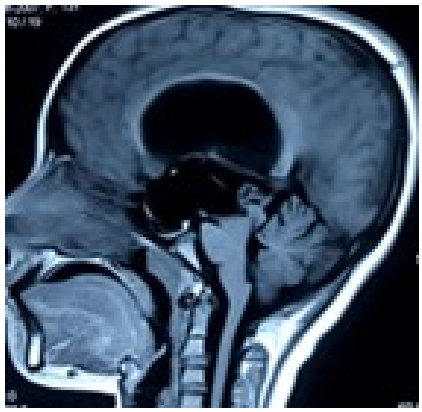

Figure 1. MRI of the brain (sagittal view) showing triventricular HCP due to cerebral aqueductal stenosis (CAS). Source: Figure by authors.

2.1.1. Etiology

1. Congenital: The anomaly may occur alongside neurofibromatosis (NF) or Chiari malformation.
2. Acquired: The anomaly may be due to the causes cited below:
   (a) Inflammation as a sequalae hemorrhage and infections, e.g., tuberculosis (TB), intra-uterine infections, etc.;
   (b) Tumors, particularly brainstem astrocytoma (including tectal plate gliomas) and lipoma;
   (c) Quadrigeminal plate arachnoid cysts.

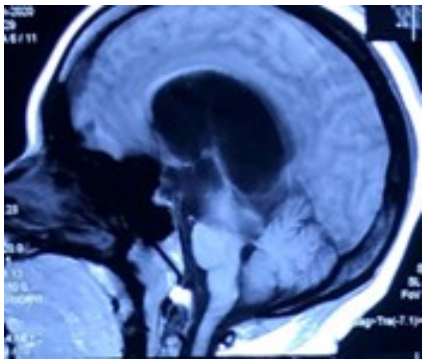

Figure 2. MRI of the brain (sagittal section) showing CAS. Source: Figufre by authors.

2.1.2. Aqueductal Stenosis in Infants

AqS (Chowdhury et al. 2017; Greenberg 2010) is a common etiology of congenital hydrocephalus (HCP) (accounting for up to 70% of cases), and, rarely, it is the outcome of hydrocephalus.

Types of congenital AqS: The four varieties of congenital AqS described by Russell (Nag and Falconer 1966):

1. Forking: As a result of the partial fusion of the median fissure, the aqueduct is split into many independent channels. These channels may re-connect to form a single aqueduct, or they may come to an abrupt halt and form a dead-end. This variety usually occurs in conjunction with other developmental anomalies (spina bifida and lipomyelocele).

2. Periaqueductal gliosis: Here, the aqueduct starts out partially obstructed. To overcome this partial block, the pressure in the third ventricle is increased. This produces more stress on the aqueduct, resulting in greater injury to the epithelial lining of the third ventricle, causing gliosis and glial cell proliferation, which ultimately obstructs the aqueduct.

3. True stenosis: This variety consists of narrowing present since birth. This narrowing results from an unusual folding of the neuro-epithelial plate.

4. Septum formation: Here, a membrane composed of glial cells forms across the aqueduct (Figure 3). The commonest site is at the lower and distal portion of the aqueduct. It obstructs the canal totally.

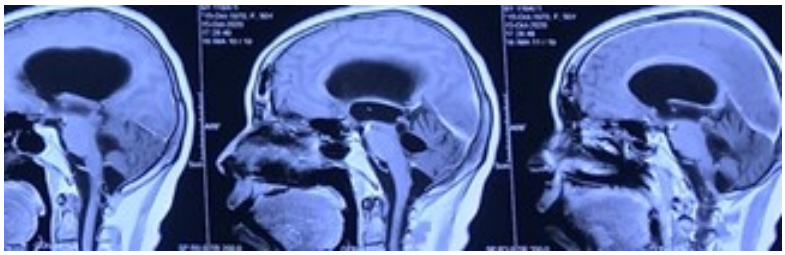

Figure 3. MRI of the brain (sagittal sections) showing CAS with HCP due to septations. Source: Figure by authors.

2.1.3. Aqueductal Stenosis in Adulthood

Most patients present with this anomaly between the ages of 1 and 5, but it may emerge at as late as 47 years of age. The cause of this late presentation is unknown. AqS may be an overlooked cause of "normal pressure hydrocephus" in an adult (Vanneste and Hyman 1986). Although most cases remain benign, there may be elevated ICP and sudden death.

2.1.4. Clinical Features

Most cases occur in childhood, but some may present in adulthood.

Symptoms:

- Headache: This is the most common symptom, and it is caused by raised ICP.
- Visual disturbance: This consists of blurring of vision, loss of acuity, and upward gaze palsy.
- Cognitive difficulty and developmental delay.
- Gait disturbance and frequent falling.
- Endocrine abnormalities, including menstrual irregularity, hypothyroidism, and hirsutism.

Signs:

- Papilledema (the commonest sign);
- Reduced peripheral vision and increased blind spots, while visual fields may be normal;
- Intellectual impairment;
- Others: ataxia, pyramidal tract signs, hemiparesis/paraparesis, spasticity, Babinski sign, anosmia.

2.1.5. Evaluation

MRI is the primary choice for investigation. It will show the disappearance of the usual CSF flow void in the aqueduct. Contrast should be injected to exclude tumors. CT scans can also be used for diagnosis and evaluation. USG can also be used as screening test for younger children.

2.1.6. Treatment

- Endoscopic third ventriculostomy: Possibility of failure of ETV is higher in younger children (age < 06 months);
- Ventriculo-peritoneal shunt.

2.2. Congenital Hydrocephalus

Hydrocephalus (HCP) is an unusual collection of CSF within the ventricular system of the brain. Congenital hydrocephalus is when a child is born with an excessive accumulation of CSF within the brain (Figures 1–6).

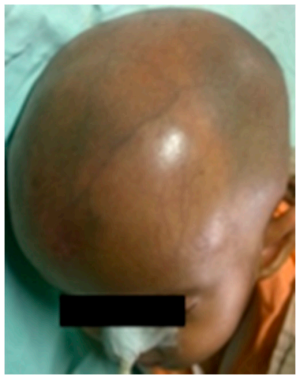

Figure 4. Head of a patient with HCP showing enlarged head, tense fontanels, and a shiny scalp with a sunset eye sign. Source: Photo by authors.

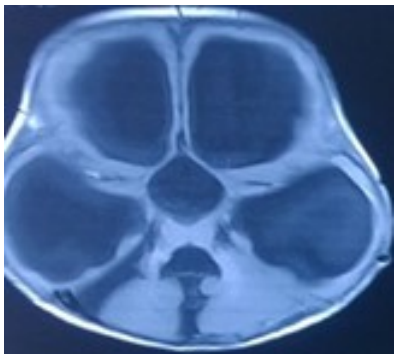

Figure 5. MRI of the brain (axial section) showing HCP with Dandy–Walker Variant (DWV). Source: Figure by authors.

2.2.1. Common Causes of Congenital Hydrocephalus

(i) Aqueductal stenosis;
(ii) Neural tube defects;
(iii) Dandy–Walker syndrome;
(iv) Type 1 and type 2 Chiari malformations;
(v) Genetic mutation.

2.2.2. Symptoms and Signs

(a) In young children:
- Unusually large head;
- Cranial growth > facial growth;
- Thin, transparent scalp;
- Scalp veins show engorgement along with flow reversal from intracranial sinuses due to ↑ICP;
- Fontanelles are full and bulging;
- Irritability, poor head control, nausea, and vomiting;
- Downward gaze;
- Hyperactive reflexes.

(b) In older pediatric patients (with unyielding cranial vault) and adults
- Features of raised ICP: headache, nausea, vomiting, papilledema;
- Gait changes;
- Downward gaze;
- Abducens nerve palsy;
- Initially, slowly increasing ventricles may be asymptomatic.

2.2.3. Treatment

- ETV;
- VP shunt.

2.2.4. Fourth-Ventricular Outflow Obstruction (FVOO)

This is a very rare congenital anomaly where atresia of the foramen Magendie and Luschka cause hydrocephalus with pan-ventriculomegaly (Figure 6). The treatment options are ETV or a VP shunt.

2.2.5. Foramen of Monro Stenosis/Atresia

It is an extremely rare anomaly causing univentricular hydrocephalus.

2.3. Craniosynostosis (Craniostenosis)

Craniosynostosis (CSO) is a condition where one or more sutures close too early, causing impairment of normal brain development and skull growth, increased pressure inside the head, and a change in the skull or facial structures from a normal symmetric appearance. CSO is rarely associated with hydrocephalus (HCP) (Golabi et al. 1987).

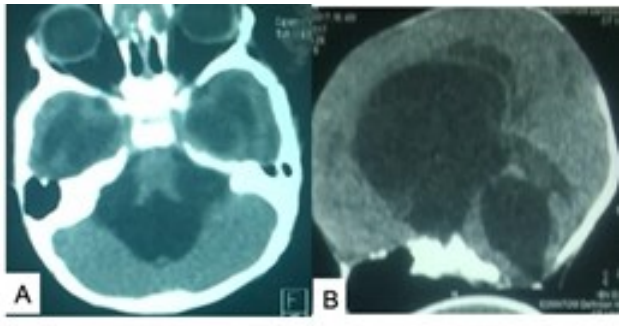

Figure 6. CT scan of the head (**A**) axial and (**B**) sagittal views) showing fourth-ventricular obstruction (FVOO) due to foramen Magendie and Luschka atresia. Source: Figure by authors.

2.3.1. Types

(a) Sagittal synostosis
- This is the commonest form of CSO involving a single suture, and 80% of cases are male.
- It results in dolichocephaly or scaphocephaly (a boat-shaped skull) associated with frontal bossing, a marked occiput, and a palpable, keel-like sagittal ridge.
- In this condition, OFC remains near to normal, but the biparietal diameter is significantly reduced.
- Affected individuals may present with features of elevated ICP.
- Treatment—Surgery: Within the first 36 months of life, a linear strip craniectomy should be performed, removing the sagittal suture from the bregma to the lambda. The strip's width should be at least 3 cm.

(b) Coronal synostosis
- About 18% of CSO cases are coronal synostosis, and they are more common in females.
- In Crouzons syndrome, this condition is associated with abnormalities of sphenoid, orbital, and facial bones.
- In Apert's syndrome, features of Crouzons syndrome plus syndactyly are present (Renier et al. 1996).

  Unilateral coronal CSO is called plagiocephaly, featuring a flattened or concave forehead on the afflicted side above the eye (the normal side appears to protrude unnaturally) and a supra-orbital edge that is higher than that one the normal side (presenting a harlequin eye sign in a skull X-ray). Amblyopia can occur when the orbit spins out on the abnormal side.
- If left untreated, the cheeks flatten, and the nose moves to the usual side (the root of the nose develops a tendency to deviate towards deformity). Bilateral coronal CSO (e.g., Aperts) brachycephaly is characterized by a broad, flattened forehead.
- The corrective surgery in this case is a simple strip craniectomy of the affected suture or a frontal craniotomy (unilateral or bilateral), advancing the lateral canthus by taking off the orbital bar.

(c) Metopic synostosis
- Here, the frontal bone is divided into two similar parts at birth by the frontal/metopic suture, which results in a pointed forehead with a midline ridge (trigonocephaly).

- Many of affected individuals are cognitively handicapped and have a 19p chromosomal issue.

(d) Lambdoid synostosis
- Males are more likely to have this condition than females (with a male/female ratio = 4:1), and the right-sided suture is affected in 70% of instances.
- It commonly appears between the ages of 3 and 18 months; however, it can appear as early as 1–2 months.
- Positional flattening (lazy lambdoid) causes confusion, which might be caused by limited movement in patients who always rest in the supine position with their head on the same side.
- Abnormal postures: congenital torticollis, congenital cervical spine abnormalities

2.3.2. Clinical Findings

- The occiput is flattened. It could be either unilateral or bilateral.
- Unilateral lambdoid synostosis results in a rhomboid skull, with the ipsilateral ear located anterior and inferior to the contralateral ear when severe; bilateral lambdoid synostosis results in brachycephaly with anterio-inferiorly displaced auricles in severe cases (Muakkassa et al. 1984).

2.3.3. Treatment

- Nonsurgical management (McComb 1991).
  Despite the fact that most patients recuperate, permanent deformity is a common occurrence.
- In 85% of situations, repositioning is effective. In this process, patients are placed on the side that is not affected or on their abdomen. Torticollis-related occipital flattening in infants should be treated aggressively with physical therapy, and remission should be seen within 3–6 months.
- Molding helmets (Clarren 1981) may be used to treat more severe involvement. Surgery is required only 20% of the time. The best time to conduct surgery is when an afflicted child is between the ages of 6 and 18.
- Surgical alternatives include a simple unilateral suture removal and repair by a craniofacial team.

2.4. Encephalocele

The term encephalocele (also known as cephalocele or meningoencephalocele) refers to intracranial material protruding through a calvarial or skull-base defect. Until proven differently, a nasal polyp like a mass in a baby should be regarded as an encephalocele.

2.4.1. Some Related Terminologies

Cranium bifidum: congenital fusion failure of cranial bones in the midline.
Encephalocele: when meninges and cerebral tissue protrude through the defect.
Meningocele: when meninges and CSF come through the defect.

2.4.2. Classification

The following classification scheme is based on the work by Suwanwella and Suwanwella (Suwanwela and Suwanwela 1972):

1. Occipital, often affecting vascular structures
2. Cranial vault, constituting about 80% of encephaloceles in the developed world
   (a) Interfrontal;
   (b) Anterior fontanelle;
   (c) Interparietal, often including a vascular component;
   (d) Temporal;
   (e) Posterior fontanelle.
3. Fronto-ethmoidal: (synonym—sincipital) (constituting 15% of encephaloceles), wherein there is an exterior opening on the face in one of the following three areas:
   (a) Nasofrontal: an outer defect in the nasion;
   (b) Naso-ethmoidal: a defect between the nasal bone and nasal cartilage;
   (c) Naso-orbital: a defect in the antero-inferior part of medial orbital wall.
4. Basal: This form constitutes 1.5% of encephaloceles;

(a) Transethmoidal: projection into the nasal cavity through a defect in the cribriform plate;
(b) Spheno-ethmoidal: the defect projects into posterior nasal cavity;
(c) Transsphenoidal: through the patent craniopharyngeal canal (foramen cecum), it projects into the nasopharynx or sphenoid sinus;
(d) Fronto-sphenoidal or spheno-orbital: the pathway is the superior orbital fissure, protruding into the orbit;

5. Posterior fossa: commonly contains cerebellar tissue and a ventricular part.

Basal Encephalocele

This consists of the developmental failure of skull-base ossification. This is the sole group of encephaloceles that does not generate soft tissue swelling that can be seen. CSF leaks or recurrent meningitis are possible symptoms. The majority of babies are stillborn; however, some make it to the age of 17. Basal encephaloceles consist of the following subtypes:

(a) Midline basal encephalocele
  (i) Transsphenoidal: A defect in the foramen cecum that protrudes into the sphenoid sinus or nasopharynx;
  (ii) Transethmoidal: A defect in the Cribriform plate;
  (iii) Sphenoethmoidal: A defect in the Sphenoid and ethmoid bones.
(b) Lateral basal encephalocele
  (i) Sphenomaxillary: A defect in the maxillary sinus and orbital fissure that protrudes into the pterygopalatine fossa;
  (ii) Sphenoorbital or fronto sphenoidal: A defect in the superior orbital fissure that protrudes into the orbit.

Cranial Vault Encephalocele

These encephaloceles may be of the following subtypes:

- Anterior fontanelle;
- Posterior fontanelle;
- Interfrontal;
- Interparietal;
- Temporal.

Frontoethmoidal (Sincipital) Encephalocele

In this type, there is an external opening in the face in one of three areas:

(a) Nasofrontal: an outer gap in the nasion;
(b) Naso-ethmoidal: a defect that stays between nasal bone and cartilage;
(c) Naso-orbital: a gap in the antero-inferior zone of the medial orbital wall.

Occipital Encephalocele

In this type of encephalocele, there is a failure in primary neural tube closure. The defect may be in the occipital bone or foramen magnum. It often involves vascular structures. The surgical removal of the sac and its contents is followed by a watertight dural closure. Hydrocephalus is common in this condition and may require different treatment. If there is a substantial quantity of cerebral tissue in the encephalocele, the ventricles expand into the mass, or there is HCP, the prognosis is usually poor. An occipital encephalocele may be of two subtypes:

Low occipital: A defect that continues into the foramen magnum.

High occipital: A defect that stops at the foramen magnum.

Posterior Fossa Encephalocele

Generally, this type contains cerebellar tissue and a ventricular part.

2.5. Other Cranial Congenital Abnormalities

2.5.1. Dandy–Walker Malformation (DWM)

This malformation is a deformed and wrapped-in-a-neuroglial-vascular-membrane larger posterior cranial fossa with partial (hypoplasia) or total aplasia of the vermis and cystic enlargement of the fourth ventricle. Dandy and Blackfan first characterized this condition in 1914, and Benda called it Dandy–Walker deformity 40 years later (Incesu and Khosla 2008). Hydrocephalus is a common occurrence in this condition.

Differential Diagnoses of DWM

Developmental disorders associated with posterior fossa CSF (or CSF-like) accumulations include (Calabro et al. 2000) the following:

1. Dandy–Walker malformation (DWM);
2. Dandy–Walker variant (DWV), which is defined as a case wherein one or more of the Dandy–Walker requirements are missing. Vermian hypoplasia and cystic distension of the fourth ventricle without expansion of the posterior fossa are examples of this (Figure 5).
3. Persistent Blakes pouch cyst (BPC): This consists of panventriculomegaly with a connected fourth ventricle and a posterior fossa cyst, with or without hypoplasia of both the cerebellar vermis and the medial portions of the cerebellar hemispheres.
4. Retrocerebellar arachnoid cyst: A cyst that pushes the fourth ventricle and the cerebellum anteriorly, a process that has the potential to have a mass effect. Here, the vermis is in good condition (Figure 7).
5. Joubert's syndrome: In this case, the cerebellar vermis is absent or underdeveloped.
6. Enlarged (mega) cisterna magna: Subsequent to an enlarged cisterna magna, an enlarged posterior fossa develops. The vermis and fourth ventricle are normal, and the cerebellum does not have a large impact.

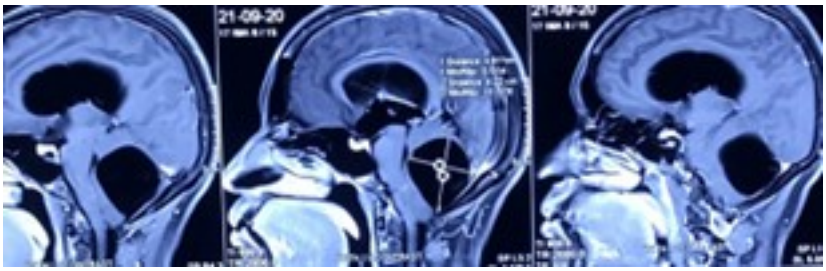

Figure 7. Contrast MRI of the brain showing a retrocerebellar arachnoid cyst causing HCP. Source: Figure by authors.

Differentiating Features

DWC: DWM and DWVs may refer to the Dandy–Walker Complex, a group of developmental disorders (DW) (Forzano et al. 2007).

BPC nor retrocerebellar arachnoid cysts: Although neither BPC nor retrocerebellar arachnoid cysts (Figure 7) undergo vermian agenesis, they may appear to be DWMs. The fourth ventricular choroid plexus is normal in arachnoid cysts, missing in Dandy–Walker malformations, and pushed into the superior cyst wall in BPC.

Arachnoid cyst and mega cisterna magna: An intrathecal enhanced CT scan (wherein iodinated contrast is injected into the CSF through a ventricular catheter, and then CT is performed) will reveal a large cisterna magna that interacts with the ventricular system, but DWM as well as most arachnoid cysts are non-interactive.

Pathophysiology and Risk Factors

The cause of DWM is not known, but it is probably caused by dysembryogenesis of the roof of the fetal rhombencephalon, and not as a result of the failure of the genesis of the fourth ventricular outlets, as previously thought (Forzano et al. 2007). This produces cerebellar vermian aplasia with a large posterior fossa cyst connected to an expanded fourth ventricle (Incesu and Khosla 2008).

Associated abnormalities: Stringed CNS anomalies are aplasia of the corpus callosum in 17% of cases, and 7% have an occipital encephalocele. Extra physical findings include heterotopias, spinal dysraphism, spinal cord syrinx, a small head (microcephaly), dermoid tumors, porencephaly, and Klippel–Feil syndrome. An expanded

posterior fossa with superiorly displaced torcular herophili is also common. Atresia of the Magendie as well as Luschka foramen may be present (Raimondi et al. 1969).

Systemic abnormalities may be associated with each other and include (Hirsch et al. 1984) facial anomalies (e.g., hemangiomas, a palatal cleft, macroglossia, and a dysmorphic face), ocular anomalies (e.g., coloboma, dysgenesis of the retina, and small orbits and eyes (microphthalmia)). Cardiovascular abnormalities may include atrial/ventricular septal defects, patent ductus arteriosus, coarctation of the aorta, and dextrocardia. When considering surgery for these patients, the possibility of a cardiac abnormality must be taken into consideration.

Treatment

- If HCP is absent, conduct a follow up.
- If HCP is present, the shunting of the cyst of the posterior fossa is required.
  Solely shunting the lateral ventricles is not recommended as there is a risk of potential trans-tentorial herniation (Mohanty et al. 2006).
  The patency of the aqueduct of Sylvius must be investigated; otherwise, a VP shunt should be inserted simultaneously. When the aqueduct is patent, an endoscopic third ventriculostomy can be attempted.
- Prognosis: Because the severity of the deformity varies, the prognosis varies as well. According to certain pediatric neurosurgery studies, fatality rates range from 12% to 50%, albeit this figure is improving with newer shunting procedures. Only half of affected patients have a normal IQ. Common symptoms include ataxia, spasticity, and impaired fine motor coordination. Seizures affect 15% of afflicted people.

3. Spinal Congenital Anomalies

3.1. Spinal Dysraphism (Spina Bifida)

The word "spinal dysraphism" refers to a group of disorders that affect the spine, spinal cord, or nerve roots and are present at birth. It occurs as a result of the neural tube's incomplete construction or the abnormal development of structures connected to the neural tube and surrounding somites.

3.1.1. Types

A. Spina bifida occulta;
B. Spina bifida aperta, which consists of the following entities:
   a. Meningoceles;
   b. Myelomeningoceles;
   c. Lipomeningoceles;

3.1.2. Spina Bifida Occulta

A lack of a spinal process and a varied amount of lamina are both congenital. There are no apparent meninges or neural tissue. There may be overlaying cutaneous manifestations, and a deficiency may be visible.

3.1.3. Meningocele

- Only meninges herniate through this defect; no neural elements are present. The spinal cord is usually normal. The mass is usually seen in the midline of the lower back. Most meningoceles are well covered with skin (Figure 8).
- Surgery may be delayed or avoided in asymptomatic infants with normal neurologic results and full-thickness skin covering.
- To prevent meningitis, patients with leaking cerebrospinal fluid or a thin skin covering should undergo prompt surgical therapy, which includes sac excision and repair with a watertight dural closure.

3.1.4. Myelomeningocele (MM)

A myelomeningocele is a congenital defect in vertebral arches featuring cystic swelling over the area of the defect encompassing meninges, the spinal nerve root, and the spinal cord itself and that has left the vertebral canal (Figures 9–11).

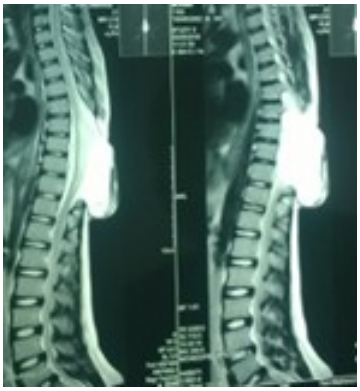

Figure 8. MRI of dorsal spine (sagittal views) showing dorsal meningocele. Source: Figure by authors.

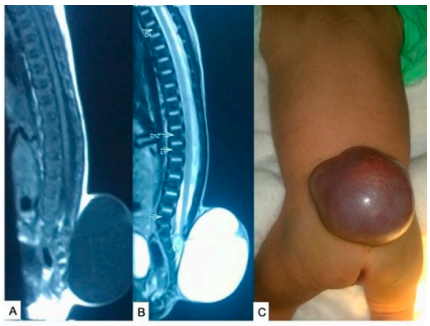

Figure 9. (**A**,**B**) MRI of dorsal spine (sagittal views) showing a lumbo-sacral myelomeningocele (MM). (**C**) Myelomeningocele after positioning a patient for surgical excision. Source: Figure by authors.

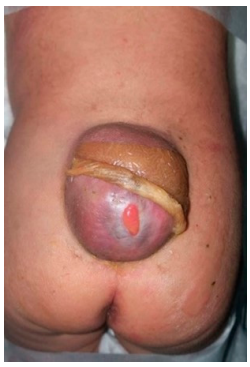

Figure 10. Lumbo-sacral MM seen after positioning of patient for surgical excision. Source: Photo by authors.

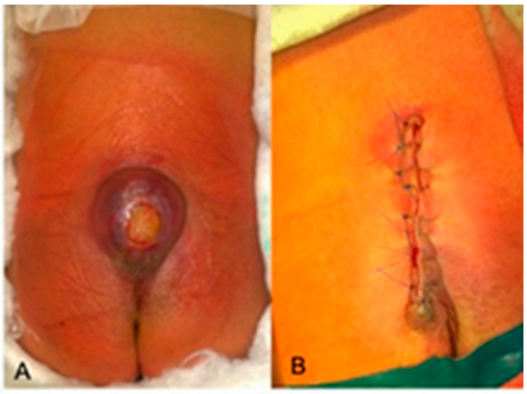

Figure 11. (**A**) Lumbo-sacral MM seen after positioning the patient for surgical excision; (**B**) perioperative view after excision. Source: Photos by authors.

Hydrocephalus in MM

A total of 65–85% of MM patients develop hydrocephalus (HCP), and 5–10% of MM cases may have overt hydrocephalus at delivery (Stein and Schut 1979). HCP affects almost 80% of MM patients before they reach the age of six months. The majority of MM patients have a Chiari type-2 malformation. Closing the MM defect can turn latent hydrocephalus into active hydrocephalus via blocking a CSF outflow route. The lower the abnormality in the neuro-axis (sacrum), the less likely it is that hydrocephalus will occur. However, regardless of the spinal level, the danger of hydrocephalus development should always be recognized.

Clinical Features

- Approximately 75% of MMs are present in the lumbosacral region. MMs induce bowel and bladder incontinence, as well as anesthesia in the perineal area, when it is present in the lower sacral region, but it does not impair motor function.
- MMs in the mid-thoracic region typically feature a growing neurologic deficit and may even present with complete paralysis of all muscles of the lower limbs.
- MMs in the upper thoracic or cervical region are normally accompanied by a minimal neurodeficit and are not associated with hydrocephalus in the majority of instances, but they may occur alongside neurogenic bladder and intestine dysfunction in certain situations.

Surgical Management

Timing of MM Surgical Closure: Although there is no evidence that early closure of an MM defect leads to improved neurologic function, there is evidence that it leads to a lower infection rate. If the membrane is intact, the MM should be closed within 24 h (after 36 h, the lesion will have been colonized, and the chance of postoperative infection is high).

MM Defect Closure and VP Shunting at the Same Time: Most neurosurgeons forestall shunting for at least three days after MM repair in cases without HCP. MM repair and VP shunt insertion can be performed simultaneously in MM cases with obvious HCP at birth without an increased risk of infection and with a shorter hospital stay (Epstein et al. 1985; Hubballah and Hoffman 1987).

These procedures could also lower the likelihood of MM repair failure previously existing at the time preceding shunting. In this combined procedure, the patient is positioned so that they are lying down with their head rotated to the right (to reveal the occiput on the right side) and their right knee and thigh flexed to reveal their right flank (one can consider utilizing the left flank to avoid confusion with an appendicectomy incision scar later in life).

Surgical Technique of MM Repair

General principles (McLone 1980): Keep the exposed neural tissue wet to avoid desiccation. Make use of a latex-free setting (this decreases the chance of stimulating a latex allergy and hypersensitivity due to maternal antibodies that may have transferred across the placenta). Scrubbing chemical or antimicrobial solutions should not come into contact with the neural placode. Monopolar cautery should not be used. Avoid putting tension on the neuronal placode at any time during the closure.

(i) The placode should be sharply separated from the surrounding arachnoid membrane and ectodermal elements. Retained fragments of cutaneous epithelium, which can cause a dermoid cyst, should be removed from the placode.
(ii) Reconstitute the neural tube by gently folding the placode towards the midline and securing the pia with small, nonabsorbable monofilament sutures.
(iii) The thickened filum terminal, if it can be located, should be sectioned.
(iv) Identify the dural–dermis boundary. Circumferentially incise the dura at the boundary, separate it from subcutaneous tissues, and move it towards the midline, where it should be closed.
(v) Ensure water-tight dural closure.
(vi) Move the skin by separating it from the underlying normal fascia.
(vii) Do not apply tension to the neural placode at any point during closure (Figure 11).

Postoperative Care

(i) The patient is kept in prone position to avoid the placement of pressure on the incision.

(ii) A barrier dressing below the incision is used to avoid contamination from urine or stool.
(iii) Daily measurements of OFC and weekly head ultrasounds are obtained to check for progressive ventriculomegaly (especially in children who do not have shunts).
(iv) Routine bladder catheterization is ensured.
(v) Orthopedic and urological consultations for the correction of limb, hip, and bladder function are conducted.

3.2. Other Spinal Anomalies

3.2.1. Klippel–Feil Syndrome

The fusion of two or more cervical vertebrae is a congenital condition. The fusing of merely the bodies of the vertebrae (congenital block vertebrae) to the complete vertebrae is possible (including the neural arch). Flattened vertebral bodies and hypoplastic or missing disc gaps are common in involved vertebral bodies. Hemivertebrae can also be found. The neural foramina are smaller and more oval-shaped than typical foramina. Cervical stenosis is a rather uncommon condition. Iniencephaly is an uncommon condition characterized by a complete lack of the posterior components, a large foramen magnum, and a permanent hyperextension posture.

Presentation

Classical clinical triad (all three are present in <50%):

1. A low posterior hairline;
2. A shortened neck (brevicollis);
3. Limitation of neck motion (may not be evident if <3 vertebrae are fused, if fusion is limited only to the lower cervical levels (Gray et al. 1964), or if hypermobility of non-fused segments compensates for this lack of mobility). Limitation of movement is more common in terms of rotation than flexion–extension or lateral bending.
   Symptoms are not due to the fused vertebrae but result from non-fused segments that possibly become hypermobile, leading to instability or degenerative arthritic changes.
4. Visceral abnormalities:
   (a) Kidneys: ↑albumin excretion in urine, ↑BUN, and ↑ creatinine levels.
   (b) Respiratory: wheezing.
   (c) CVS: irregular heartbeat, complete heart block.

Investigations

(i) Urine: ↑albumin; ↓urinary volume.
(ii) Blood: ↑BUN and ↑creatinine.
(iii) Radiology: vertebral fusion between C2-5, scoliosis, spina bifida, atlanto-axial dislocation.

Treatment

A. Pain killers, muscle relaxant, and physiotherapy. If the condition does not improve, surgery is required.
B. The surgical options are as follows:
   (i) Correction of cranio-vertebral instability and decompression of the spinal cord;
   (ii) Cervical discectomy and fusion;
   (iii) Scoliosis surgery.

3.2.2. Tethered Cord Syndrome (TCS)

Tissue attachments that impede the mobility of the spinal cord within the spinal canal cause this neurological condition. The conus medullaris is unusually low here. TCS is generally accompanied by a short and thickened filum terminale or an intradural lipoma. (Figure 12).

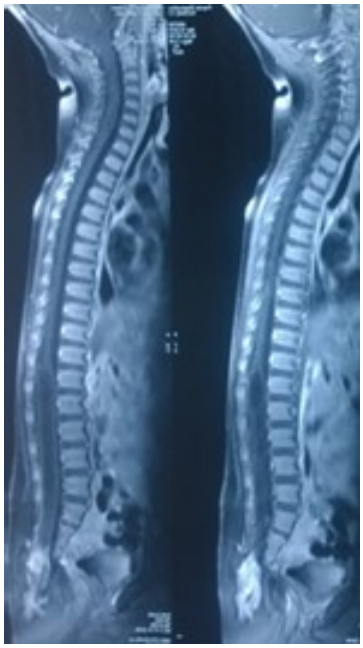

Figure 12. MRI of the spine (sagittal views) showing tethered cord and syringomyelia with sacral lipoma. The patient also had Chiari malformation. Source: Figure by authors.

TCS is most commonly seen in cases of myelomeningoceles (MMs). If an MM patient has deteriorating scoliosis, progressively increasing spasticity, increasing gait disturbances, and worsening urodynamics and the condition is painful, the individual should be considered to have tethered cord syndrome until proof suggests otherwise (Park et al. 1985).

Progressive scoliosis can be observed at the same time as a tethered cord is seen. Early release of the spinal cord may induce an elevation in scoliosis status.

Presentations

TCS is mostly noted in children. Children and adults have different clinical presentations. The symptoms and signs may develop during or after the lengthening of the spine. Comparison of childhood versus adult tethered cord syndrome is shown in Table 1.

Table 1. Comparison of childhood versus adult tethered cord syndrome.

Clinical Presentation	Adult TCS	Pediatric TCS
Pain	Common	Rare
Swelling	Less common	Frequent
Leg weakness	Frequent	Less frequent than adult
Sensory impairment	In 15%	In 18%
Sphincteric dysfunction	In 65% adult TCS	In 18% pediatric TCS
Orthopedic deformity	In 30%	In 16%
Cutaneous stigmata (tufts of hair, dimples, and capillary angioma/naevus flammeus)	In 80–100% cases	In <50% cases
Lipomeningocele	In 40%	In 13%
Meningocele & meningomyelocele	Very rare	Relatively frequent
Dermal sinus	In 10% patient	In 1% patient
Aggravating factor	Trauma	Growth spurts

Source: Authors' compilation based on data from Pang and Wilberger (1982); Shukla et al. (2018).

Evaluation

Radiography: A conus medullaris with a thickened filum terminale (diameter > 2 mm) and a low-lying conus medullaris (below L2) are observed. It can be difficult to tell the difference between a tethered cord and a conus that is naturally low-lying (here, the filum diameter is normal).

Pre-operative cystometrogram: This is especially necessary if the patient is continent

Surgical Treatment

(a) If only a thickened and shortened filum is observed, untethering by sectioning the filum should be performed.
(b) If lipoma is also present, it may be excised along with filum (if it can be easily separated from nervous tissues) or debulking.
(c) Perioperative differentiation of filum terminale from nerve roots:
    (i) Filum has typical squiggly vessels on its surface.
    (ii) Microscopically, filum has a typical whiter appearance than the nerve root.
    (iii) Ligament-like strands can be observed running within it.

Outcome

- It is normally impossible to permanently untether a cord in the case of an MM; but, after 2–4 untetherings, a growing MM-afflicted child's growth may be complete, and tethering may be discontinued.
- Untethered cases from childhood may return later in life, particularly during the adolescent growth spurt.
- Among adults, surgical release is generally effective and good for pain elevation, but bladder function return is poor.

4. Cranio-Spinal and Other Anomalies

4.1. Chiari Malformation

Chiari malformations are a group of four different forms of anomalies of the hindbrain that are unrelated to one another. Types 1 and 2 are the most common Chiari malformations. The remaining types are made up of a small number of examples.

4.1.1. Chiari Type 1 Malformation

- Elongated, peg-shaped cerebellar tonsils, also known as primary cerebellar ectopia (Spillane et al. 1957), and adult Chiari malformation (as it is commonly identified in the 2nd/third decade of life).
- Extends below FM into the upper cervical spinal canal (Figures 13 and 14).
- Unlike Chiari malformation-2, the medulla oblangata is not shifted downward, the brainstem is unaffected, the lower cranial nerves are not lengthened, and the higher cervical spinal nerves do not run upward.
- Syringomyelia affects 30–70% of patients.
- Hydrocephalus has been linked to Chiari type 1 malformation patients.

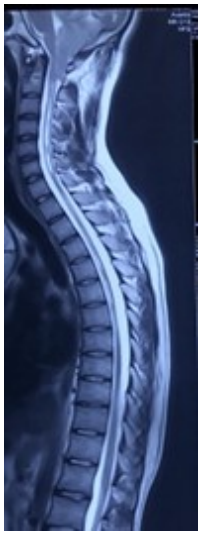

Figure 13. MRI of spine and cranio-vertebral junction (CVJ) showing type 1 Chiari malformation with syringomyelia. Source: Figure by authors.

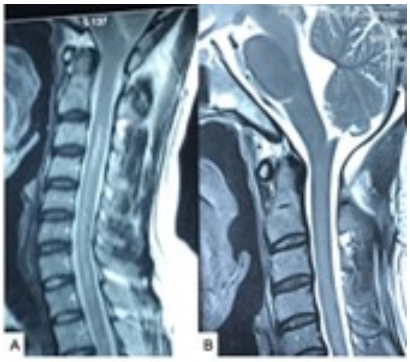

Figure 14. (**A**) preoperative MRI of cranio-vertebral junction (CVJ) and cervical spine (sagittal views) showing type 1 Chiari malformation with syringomyelia. (**B**) Post-operative MRI after three months of operation. Source: Figure by authors.

Associations

Chiari type 1 is usually associated with the following:

1. Base-of-the-skull and CV junction anomalies:
   (i) An underdeveloped occipital bone;
   (ii) Smaller and shallow posterior fossa;
   (iii) Platybasia;
   (iv) Basilar invagination;
   (v) Hypermobility of CVJ.
2. Spine-related abnormality:
   (i) Klippel–Feil syndrome;
   (ii) Occipitalization of atlas;
   (iii) Retroflexion of odontoid process;
   (iv) Scoliosis.
3. Meninges-related abnormalities
   (i) Low-lying tentorium;
   (ii) Thickened arachnoid/scar/fibrosis around brainstem and tonsils near to the foramen magnum;
   (iii) Veils of arachnoid that obstruct fourth-ventricular outflow.
4. Brain-related abnormalities
   (i) Space-occupying lesion in posterior fossa;
   (ii) Intracranial-space-occupying lesion;
   (iii) Craniosynostosis (especially cases involving all sutures).
5. Ventricle- and cistern-related abnormalities
   (i) Hydrocephalus;
   (ii) Elongated fourth ventricle;
   (iii) Retrocerebellar CSF spaces are obliterated or diminished.
6. Following LP shunt or multiple LP (acquired Chiari 1 malformation)

Symptoms

(i) This condition has an adult onset, and the average age at presentation is 41 years. There is a slight female preponderance.
(ii) The most common symptom is headache (sub-occipital HA, often detected via neck extension or valsalva).
(iii) Weakness of limb(s) and/or unsteadiness.
(iv) Loss of temperature sensation.
(v) Painless burns.

Signs

Three prime patterns of clustering of signs:

1. Compression syndrome of the foramen magnum: ataxia, sensory, and corticospinal neuro-deficits; Babinski sign; and lower-cranial-nerve palsies.

2. Central cord syndrome: hand atrophy, upper limb weakness, lower-limb weakness, dissociated sensory loss, and hyperactive deep tendon reflexes.
3. Cerebellar syndrome: ataxia, downbeat nystagmus during vertical movement, and dysarthria.

Evaluation

(i) MRI of the brain and cervical spine is the investigation of choice (Figures 13 and 14).
(ii) Traditionally tonsillar descend >5 mm is defined as pathologic, with 3–5 mm being borderline.
(iii) Most cases are associated with syringomyelia.

Indications for Surgery

Early surgery is advised for clinically symptomatic cases. Asymptomatic cases can be followed up and operated on when the patient becomes symptomatic. Symptomatic patients who are stable for years can be followed up, and surgery is recommended when the patient deteriorates.

Surgical Techniques

Posterior fossa decompression via sub-occipital craniectomy with augmented duroplasty with or without cervical laminectomy of C_1 (sometimes C_2 or C_2). The goal of surgery is to decompress the brainstem as well as reestablish normal CSF flow at the craniospinal junction.

Surgical Complications

Sleep apnea, respiratory depression, CSF leak, cerebellar herniation, vascular injuries (PICA), etc.

4.1.2. Chiari Type 2 Malformation

This condition is also known as Arnold–Chiari malformation

Pathology

Almost all neonatal Chari II patients have a myelomeningocele, which is caused by an in utero CSF leak in turn caused by open spinal dysraphism. Patients with Chiari II who do not have a myelomeningocele are considered to have had a smaller neural tube defect or have had the defect closed in utero.

Major Findings

(i) Caudally dislocated cervico-medullary junction, pons, medulla, and fourth ventricle.
(ii) Cerebellar tonsils are situated at or below the level of the foramen magnum.
(iii) Normal cervicomedullary junction flexure is replaced by a "kink-like" deformity.
(iv) Others: Beaking of tectum, hydrocephalus, bony abnormalities such as abnormalities of cervicomedullary junction, assimilation of C1, platybasia, basilar invagination (BI), Klippel–Feil deformity, etc.

Presentation

Onset is commonly in the neonatal stage and rare in adulthood. Neonates develop rapid neurological deterioration with profound brain stem dysfunction. Patients may present with the following symptoms (Pollack et al. 1992; Park et al. 1983):

1. Swallowing difficulties (neurogenic dysphagia);
2. Apneic spells, which are more common in neonates;
3. Stridor, which occurs as a result of vagus nerve paresis and is commonly transient but may progress to respiratory arrest;
4. Aspiration;
5. Arm weakness, which may result in quadriparesis;
6. Nystagmus, especially downbeat nystagmus.

Evaluation

Cranial and cervical MRI is the investigation of choice. The potential findings are as follows:

A medulla-situated "Z" bend deformity, a cerebellar peg, tectal fusion ("tectal beaking"), increased interthalamic adhesion, extension of the medulla, low levels of attachment of the tentorium, hydrocephalus, syringomyelia, a trapped fourth ventricle, and corpus callosum agenesis/dysgenesis.

Surgical Technique

If hydrocephalus has developed, CSF shunting is performed. Expeditious posterior fossa decompression should be performed if neurogenic dysphagia, stridor, or apneic events occur.

Decompression of the cerebellar tonsils via suboccipital craniectomy with cervical laminectomy with augmented duroplasty is used. In cases with a significant syringomyelic cavity, a syringo-subarachnoid shunt is also placed.

4.1.3. Other Chiari Malformations

Chiari type 0: Patients with syringohydromyelia but no hindbrain herniation who improve after posterior fossa decompression are classified as Chiari type-0 (Iskandar et al. 1998).

Chiari type 1.5: The whole cervicomedullary junction (including the obex) is located below the foramen magnum, and suboccipital decompression with or without duroplasty has little effect.

Chiari type 3: Chiari type 3 is a rare condition. The cerebellum dislocates below the foramen magnum, resulting in an occipital encephalocele. Some individuals have an occipital and high cervical encephalocele combined with herniation of the medulla, fourth ventricle, and entire cerebellum. According to certain sources, occipital encephaloceles are linked to cerebellar and medulla caudal displacement. The majority of cases have a poor prognosis since they are life-threatening.

Chiari type 4: Type 4 Chiari was originally identified as cerebellar hypoplasia without herniation (Chiari 1895).

4.1.4. Syringomyelia

Syringomyelia is a general term referring to a pathology in which a cyst or cavity is formed within the spinal cord (Figures 13–15).

Etiologies

A. Primary syringomyelia: the absence of an identifiable cause

B. Secondary syringomyelia: due to partial obstruction of the spinal subarachnoid space. Etiologies are:

1. Chiari I malformation: the commonest cause of syrinx.
2. Post-inflammatory: (a) postinfectious—meningitis; (b) chemical or other sterile inflammations.
3. Post-traumatic: (a) severe post-traumatic kyphotic deformity, (b) arachnoid scarring, and (c) severe trauma to the spinal cord and/or its coverings.
4. Postsurgical: many years after uneventful intradural tumor resection (e.g., neurofibromas).
5. Basilar arachnoiditis: (a) idiopathic and (b) postinfectious.
6. Basilar invagination with a narrow foramen magnum.
7. Dandy–Walker syndrome.

Presentations

1. Dissociative sensory loss: Pain and temperature sensations are lost, but touch as well as joint position awareness are preserved. Painless ulcerations from unnoticed injuries and/or burns are also exhibited.

2. Cervical and occipital pain are the most common. Dysthetic pain in the sensory loss distribution is also exhibited.

3. Hand and arm weakness due to lower motor neuron weakness.

4. Painless (neurogenic) arthropathies (Charcot's joints) characterized by a loss of pain and temperature sensation, particularly in the shoulder and neck.

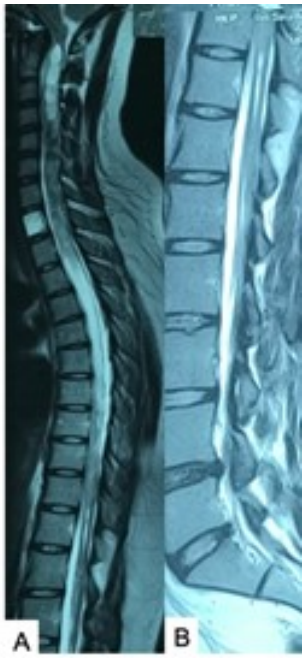

Figure 15. MRI of the spine (sagittal images): (**A**) CVJ and cervico-dorsal spine and (**B**) dorso-lumbar spine showing syringomyelia without an obvious etiology (possibly Chiari-0). Source: Figure by authors.

Evaluation

MRI

Treatment

Treatment of the underlying cause.

4.1.5. Split-Cord Malformation (SCM)

A split-cord malformation (SCM) is a type of concealed spinal dysraphism and tethered spinal cord condition that is rather uncommon. The majority of these instances appear in infancy, with neurocutaneous stigmata being a common early symptom. SCM accounts for almost one-third of all occurrences of spinal dysraphism, in which the spinal cord is separated into two equal or unequal halves across a section of its length (Figure 16). All double spinal cords, which seem to have a common embryological genesis, are classified as SCMs. Two hemicords, each one with its own central canal as well as the surrounding pia, are contained within a distinct dural tube and divided by a dural-sheathed stiff osseocartilaginous/bony median septum in Type I SCMs. Diastematomyelia is a term used to describe this condition, which involves spinal anomalies. Overlying skin abnormalities such as nevi, hypertrichosis, lipomas, dimples, or hemangiomas affect two-thirds of the population. The majority of these patients have an orthopedic foot abnormality. Tethering of the cord is the most prevalent cause of symptoms, which can typically be alleviated by untethering. The dura must be reconstructed as a single tube once the bony septum is removed. A nonrigid fibrous median septum separates two hemicords within a single dural tube in Type II SCM. Diplomyelia is a term that has been used to describe this condition. Nerve roots protrude from each hemicord. At the level of the split, there are usually no abnormalities in the spine, but there is usually spina bifida occulta in the lumbosacral region. One treatment consists of surgically untethering the chord. Surgical removal of the spur and detethering of the filum are also employed (Chowdhury et al. 2016; Hoffman 1992).

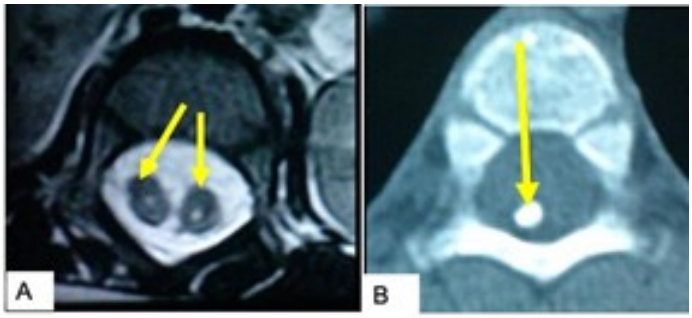

Figure 16. Split cord malformation (SCM). (**A**) MRI of the dorsal spine (axial view) diplomyelia and (**B**) CT scan of dorsal spine (axial view) showing bony septum. Source: Figure by authors.

4.1.6. Arachnoid Cyst (Leptomeningeal Cysts)

Arachnoid cysts (ACs) are congenital anomalies that occur during embryonic development via the duplication of the arachnoid mater (intra-arachnoid cysts), and they contain CSF-like fluid. They do not communicate with the ventricular or subarachnoid CSF. They may have septations and are typically lined with meningothelial cells. ACs can also develop in the spinal canal. Their incidence is 0.5% in autopsy series. ACs constitute 1% of intracranial-space-occupying lesions. The male/female incidence ratio is 4:1. ACs are more common on the left side (Figure 17).

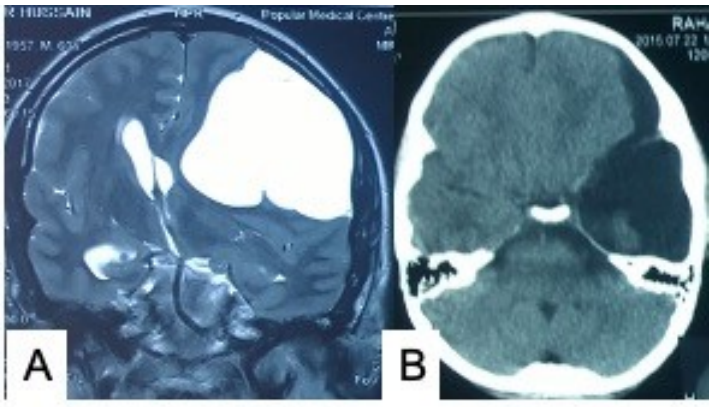

Figure 17. (**A**) MRI of the brain showing left-sided hemispheric symptomatic arachnoid cyst. (**B**) CT scan of the head showing a left temporal AC with hypoplastic temporal lobe. Source: Figure by authors.

There are two varieties of histological ACs:

- Simple arachnoid cysts: In these, the lining cells are capable of active CSF production;
- Complex ACs: Here, the lining may contain neuroglia, ependyma, and other tissue types.

Distributions of AC is (Sinha and Brown 2004):

Sylvian fissure, 49%;
Supracolicular, 10%;
Sellar and supra sellar, 9%;
Cerebral convexity, 4%;
Cerebellopontine angle, 11%;
Vermian, 9%;
Interhemispheric, 5%;
And clival, 3%.

Clinical Presentation

Most ACs are symptomless. Symptomatic ones commonly present in early childhood. The clinical presentation differs with the location of the arachnoid cyst and often appears mild in contrast to the size.

Clinical Features

- Features of intracranial hypertension;
- Sudden deterioration (due to hemorrhage);
- Location related focal signs and symptoms;
- Endocrine symptoms and visual impairment (suprasellar cysts);

- Seizures;
- As a focal swelling of the skull;
- Incidental finding.

Evaluation

- CT scan of the head
- MRI of the brain and spine: This combination is better than CT in distinguishing the CSF contained in ACs from the cystic tumor. It may exhibit cyst walls.
- Cisternograms and/or ventriculograms

Treatment

Incidental ACs: follow up.

Symptomatic ACs:

- Cysto-ventriculostomy (endoscopic/microscopic);
- Cysto-cisternostomy (endoscopic/microscopic);
- Excision/marsupialization;
- Cyst shunting (cysto-peritoneal) and/or CSF shunting.

Even after successful management, a part of the AC may remain because of the re-modeling of the bone and the great shift of the brain (Greenberg 2010; Chowdhury et al. 2018; VanDer Meche and Braakman 1983; Mayr et al. 1982; Pierre-Kahn et al. 1990; Altschuler et al. 1990; Hopf and Perneczky 1998; Sinha and Brown 2004).

Spinal Arachnoid Cysts (SACs)

A SAC nearly always occurs in the thoracic spine. A neurenteric cyst is a differential diagnosis for a ventral SAC. Most are extradural and can be associated with kyphosis, scoliosis, or spina bifida. Intradural spinal arachnoid cysts may be of congenital origin or may be caused by infection or trauma. They are commonly asymptomatic but can cause paraplegia. MRI of the spine is the most useful investigation. Symptomatic SACs need surgical (percutaneous/open) treatment (Greenberg 2010; Chowdhury et al. 2018; VanDer Meche and Braakman 1983; Mayr et al. 1982; Pierre-Kahn et al. 1990; Altschuler et al. 1990; Hopf and Perneczky 1998; Sinha and Brown 2004).

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