CSF Fistula

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Abstract: CSF surrounds the brain and spinal cord, which are contained within the dura mater. Any rift in dura can prompt CSF leakage/fistula. Common sites for CSF fistulas are the nose and ears, known as CSF rhinorrhea and CSF otorrhea, respectively. The etiology of CSF fistula includes neurotrauma, cranial as well as spinal surgery, chronic raised intracranial pressure (ICP), and a spontaneously occurring CSF fistula. Diagnosis and evaluation including 'defect' localization are the main challenge. CT or MR cisternograms assist in the diagnosis and localization of a CSF fistula. An untreated fistula can cause death and morbidity through meningoencephalitis. Treatment options include conservative and surgical treatment. Surgical treatment depends on the site and size of a fistula, associated pathologies, and the surgeon's preferences, which can be endoscopy or microsurgery. In this chapter, the etiology, clinical presentation, evaluation, and treatments of CSF fistulae, especially CSF rhinorrhea, CSF otorrhea, and spinal CSF fistula, are discussed briefly.

Abbreviations

CSF	cerebrospinal fluid	CT	computed tomography
ESS	endoscopic sinus surgery	FESS	functional endoscopic sinus surgery
HCP	hydrocephalus	ICP	intracranial pressure
MRI	magnetic resonance imaging	SSCD	superior semicircular canal dehiscence

1. Introduction

CSF leaks or fistulas happen when CSF escapes via a small rip or crack in the dura mater, which protects the CNS and retains the CSF in place. CSF might leak out through the epidermis, nasal mucosa, or external ear canal due to a rupture or hole. When CSF is lost, the previously cushioned brain sags inside the skull, resulting in a headache, and the intracranial pressure (ICP) within the skull decreases, resulting in intracranial hypotension. CSF fistulas can form anywhere along the spinal column (spinal CSF fistulae) or in the brain (cranial CSF fistulas, i.e., CSF rhinorrhea and CSF otorrhea).

2. CSF Rhinorrhea

The entry of CSF into the nose due to improper connection between the subarachnoid space and the nasal or perinasal sinus mucosa is known as CSF rhinorrhea (Figure 1) (Sumaily 2017).

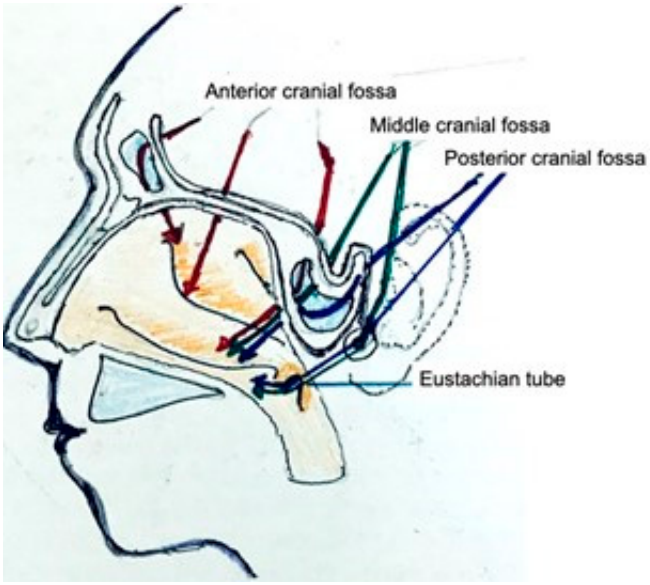

Figure 1. Schematic drawing showing different pathways of CSF rhinorrhea from intracranial fossae to nasal cavity. Source: Figure by authors.

2.1. Etiology

2.1.1. Traumatic

CSF leaks are most commonly caused by trauma (80–90%), caused by either a head injury or iatrogenic factors (Abuabara 2007; Bell et al. 2004; Platt and Parnes 2009; Bumm et al. 2009). Traumatic etiologies (blunt as well as penetrating cranio-facial injuries) account for more than eighty percent of all cases of CSF leaks, with young males being the most commonly affected (Figures 2–6).

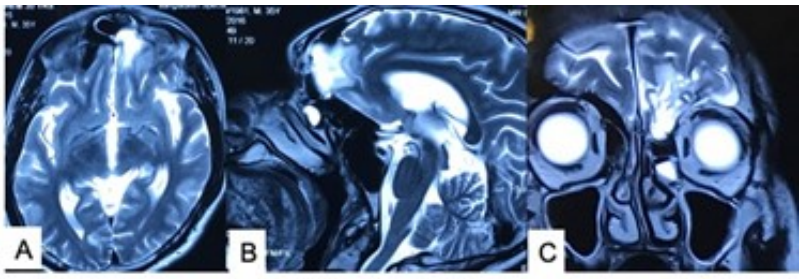

Figure 2. (**A**–**C**) MRI of the brain T2W images showing post-traumatic CSF rhinorrhea through posterior wall of the left frontal sinus. Source: Figure by authors.

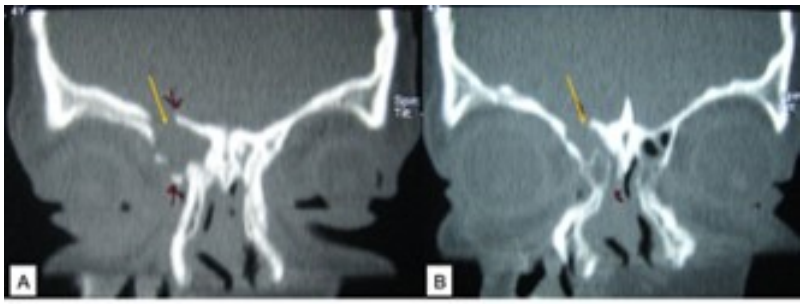

Figure 3. (**A**,**B**) CT scan of anterior skull base (coronal images) in bony window, showing traumatic fractures resulting in a CSF fistula through the ethmoid sinus (arrow marked). Source: Figure by authors.

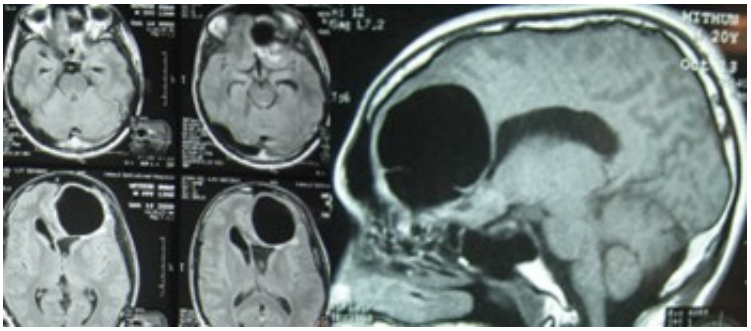

Figure 4. MRI of the brain: axial FLAIR images (left side) and T1W sagittal image (right side) showing post-head injury frontal tension pneumocephalus (with CSF rhinorrhea through ethmoid sinus). Source: Figure by authors.

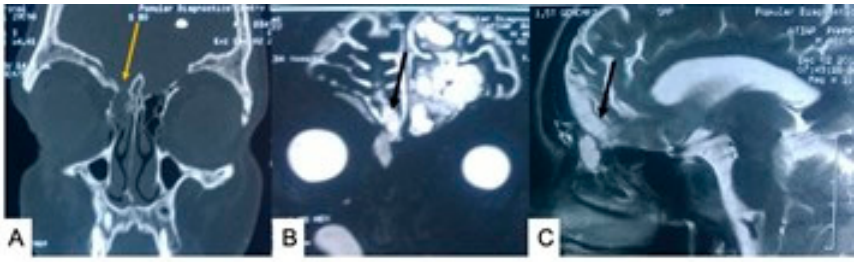

Figure 5. (**A**) CT scan coronal image of bony window showing traumatic fractures (right side) resulting in CSF fistula. (**B**,**C**) MR cisternograms (with a coronal image and a sagittal image, respectively) showing CSF tract through the same defect seen in CT. Source: Figure by auyhors.

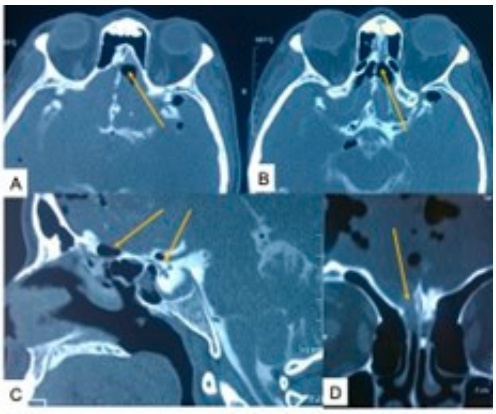

Figure 6. CT cisternogram of a patient with traumatic CSF rhinorrhea: (**A**,**B**) axial images; (**C**) sagittal image; and (**D**) coronal image showing pneumocephalus and a bony defect at the left olfactory fossa and tuberculum sella resulting in a CSF fistula. Source: Figure by authors.

In 12–30% of instances, basilar skull fractures occur, with the anterior cranial fossa being the most prevalently affected area. The dura has greater adhesion to the base of the skull at this level, so traumas can quickly impact it (Patrascu et al. 2017).

CSF leaks are classed as immediate or delayed in terms of occurrence latency: 60% appear in the third or fourth day following the occurrence, 70% appear within the first seven days, and 95% appear within the first three months (Patrascu et al. 2017).

Iatrogenic injury accounts for 16% of traumatic CSF fistula cases. Basically, any surgical procedure conducted around the skull base can cause iatrogenic CSF leaks (Patrascu et al. 2017). The most common causes of iatrogenic CSF leaks in recent decades have been neurosurgical operations (craniotomies and the removal of hypophyseal tumors or suprasellar tumors). FESS (functional endoscopic sinus surgery) has now become the leading etiology of iatrogenic CSF fistulas, accounting for less than 1% of all surgical endoscopic operations. Because of the thickness of the bone of the anterior skull base, the cribriform plate (80%) is the commonest site of lesions observed during endoscopic sinus surgery (ESS) procedures. The frontal sinus (8%), the sphenoid sinus (4%), and the posterior fovea ethmoidalis are other common sites of injury. Iatrogenic CSF fistulas usually occur in just 50% of patients within seven days after surgery, unlike traumatic leakage. The majority of the time, when the leakage occurs, the patients have already been discharged. When there is a possibility of a CSF leak, it is critical to tell patients about the commonest symptoms (Patrascu et al. 2017).

2.1.2. Spontaneous

Spontaneous leaks can occur as a result of elevated intracranial pressure (Figures 7–12) or when intracranial pressure is normal (Schlosser et al. 2006; Schlosser and Bolger 2003; Lopatin et al. 2003; Banks et al. 2009). In forty-five percent of non-traumatic CSF fistulas, leaking at high pressure seems to be implicated (Patrascu et al. 2017). Many causes of raised ICP have been identified in the literature, although the idiopathic mechanism remains the most common. CSF pressure variations affect the anterior cranial base dura, potentially leading to dural deficiency in bone floor defects due to a variety of reasons (respiration and artery pressure fluctuations and Valsalva-like actions during nasal blowing).

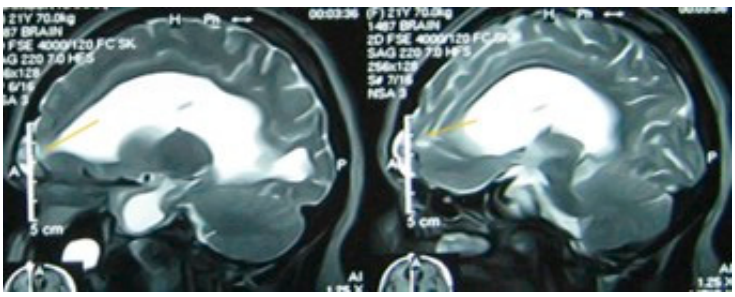

Figure 7. MRI of the brain (T2W sagittal images) showing spontaneous trans-frontal sinus CSF fistula due to chronic increased intracranial pressure (hydrocephalus). Source: Figure by authors.

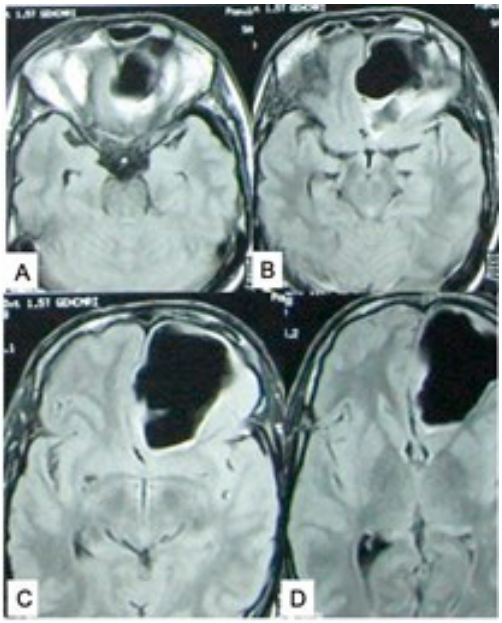

Figure 8. (**A**–**D**) MRI of the brain (axial FLAIR images) showing frontal tension pneumocephalus in a spontaneous CSF fistula (through left frontal sinus) case. Source: Figure by authors.

The pressure applied to the thin bone areas of the anterior skull base (lateral lamella of the cribriform plate, the sphenoidal sinus lateral recess) determines ischemia via vascular pressure, bony thickening, and bone shaping, along with gap creation, regardless of the reasons for high intracranial pressure. The dura can herniate and create meningoceles through this opening, or the cerebral parenchyma can herniate through it if the breach is large enough (creating an encephalocele) (Woodworth et al. 2008). Patients with spontaneous CSF rhinorrhea showed several contemporaneous bone abnormalities in the skull base according to research by Lieberman et al. (Lieberman et al. 2015). According to the researchers, intracranial hypertension was discovered to be a decisive factor in the formation of these abnormalities. There have also been occurrences of CSF leaks linked to normal ICP, constituting 55% of spontaneous cases of CSF fistulas (Patrascu et al. 2017).

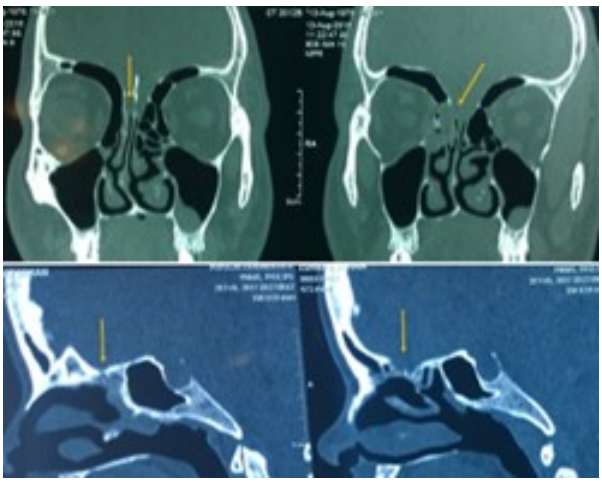

Figure 9. CT scan of head and skull base (bony window) (axial (**upper**) and sagittal (**lower**) images) showing bone gap in right olfactory fossa resulting in spontaneous CSF rhinorrhea through ethmoid sinus. Source: Figure by authors.

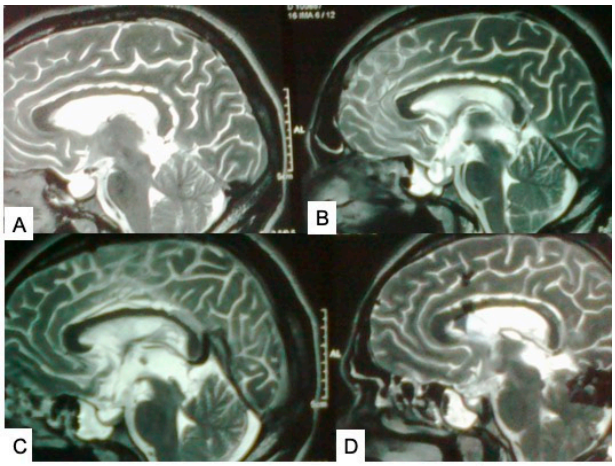

Figure 10. (**A**–**D**) MRI of the brain (T2W sagittal images) showing spontaneous CSF fistula through sphenoid sinus due to tuberculum sella and sella turcica defects. Source: Figure by auyhors.

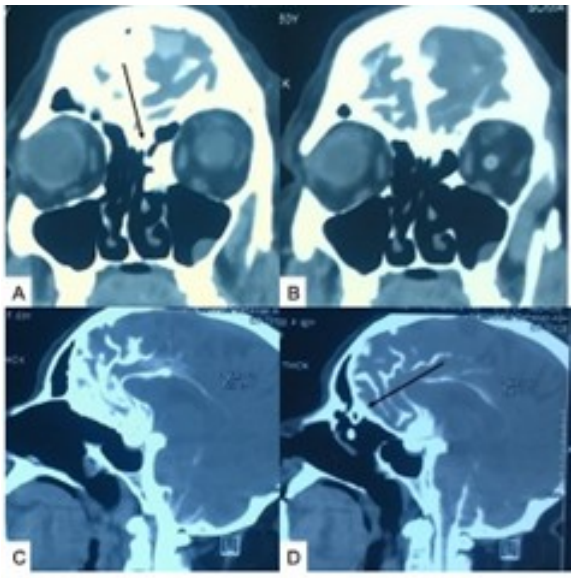

Figure 11. CT cisternogram ((**A**,**B**) coronal images and (**C**,**D**) sagittal images) showing spontaneous CSF fistula through left olfactory fossa defect. Source: Figure by authors.

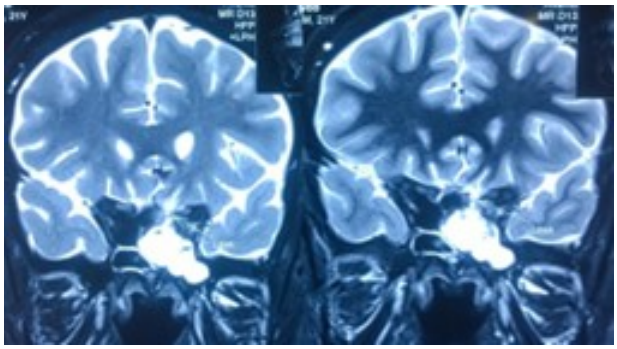

Figure 12. MRI of brain (T2W coronal images) showing spontaneous CSF fistula tract through left plannum sphenoidale and tuberculum sella into the spheno-ethmoidal sinus on left side. Source: Figure by author.

2.1.3. Congenital

Encephaloceles or a persistent craniopharyngeal canal can cause congenital CSF fistulae (Kim et al. 2000) (Figure 13). The capacity for meningeal herniation via the anterior cranial base may be determined by the presence of an anterior neuropore after delivery (meningo-encephaloceles). These are unusually linked to CSF rhinorrhea. Meningo-encephaloceles commonly emerge in the pediatric age group as a transilluminating tumoral mass in

the intranasal or extranasal cavity, which increases in size when a child cries (Furstenberg sign). All childhood intranasal tumors, particularly those emerging from the midline, should raise suspicion (Patrascu et al. 2017). Only once the surgeon have carried out imaging studies should he/she proceed with a biopsy (Woodworth et al. 2004). Another congenital lesion that can lead to CSF rhinorrhea is a persistent craniopharyngeal canal. Primary empty sellae syndrome, which is a congenital disease, can result in CSF leakage and is linked to brain tumors, hydrocephalus, and pseudotumor cerebri (Patrascu et al. 2017).

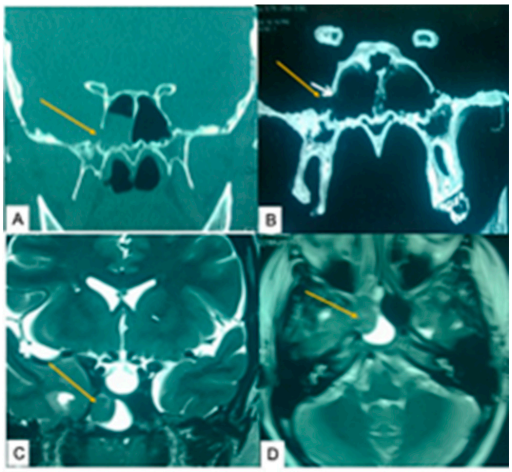

Figure 13. (**A**,**B**) CT scan of skull base (coronal section) showing defect in lateral wall of right sphenoid sinus resulting in a small temporal lobe encephalocele (pushing into the right sphenoid sinus) and CSF rhinorrhea (Sternberg's canal) (**C**,**D**). Source: Figure by authors.

2.1.4. Miscellaneous

Tumors, infection, mucoceles, and radiation can all induce erosion of the base of the skull. CSF rhinorrhea is rarely diagnosed as being caused by these factors. Locally malignant tumors, such as inverted papilloma and other neoplasms (nasopharyngeal carcinoma, osteomas), can also destroy bone in the anterior cranial base. Local inflammation and even dural rupture may result from the mass effect of bone disintegration. Even though tumors may not cause CSF rhinorrhea on their own, resection surgery frequently causes rapid leakage (Sumaily 2017).

2.2. Presentation

Unilateral clear and watery nasal discharge is the commonest indication of CSF rhinorrhea; however, this discharge might be mixed with blood if the trauma occurred recently. The volume of a nasal CSF leak may rise in the supine position. The collection of CSF in one paranasal sinus and its external flow through the nose due to positional changes of the head (the "reservoir sign") can cause intermittent leakage (Meco and Oberascher 2004). Patients may also taste salt. Although most people do not complain of a headache, the presence of a headache raises the risk of excessive ICP and intracranial tumor (Mokri et al. 2000; Mokri et al. 1998). It is crucial to note whether the headache goes away after the CSF leakage has subsided.

Other symptoms may assist one in pinpointing the leak's location in some circumstances. Anosmia (60% of post-traumatic CSF rhinorrhea cases) indicates that the anterior fossa and olfactory region have been injured (Mathias et al. 2016). Lesions of the tuberculum sellae, the posterior ethmoidal sinuses, or the sphenoidal sinus can cause optic nerve dysfunctions (Patrascu et al. 2017). Regardless of whether or not there is a CSF leak, individuals with recurrent meningitis should be investigated for abnormalities that expose the meninges to the upper airways (Bernal-Sprekelsen et al. 2005). Patients may experience nasal bleeding, red eyes, periorbital bruising, visual dysfunction, a loss of smell or cranial nerve palsies (most commonly I–III as well as V–VII), and acute-phase meningitis shortly after the neuro-trauma (Eljamel and Foy 1990; Cassano and Felippu 2009; Scholsem et al. 2008). Patients may develop recurrent nasal watery discharge, intermittent meningitis or cerebral abscesses, headaches, a sweet/salty taste in the nasopharynx, and hyposmia in the chronic phase after the trauma. The probability of the development of recurrent meningitis after trauma ranges from 12.5% to 50%, with a neurological problem incidence of 29.4% (Patrascu et al. 2017; Mokri et al. 1998). The ability to distinguish CSF rhinorrhea from other nasal discharges is still crucial in detecting CSF fistulas.

Patients with obvious CSF rhinorrhea after trauma have a clear diagnosis that just needs to be confirmed. The presence of hemorrhagic clots or nasal bleeding as a result of facial fractures, as well as the interval of CSF rhinorrhea, might also complicate the diagnosis (Yilmazlar et al. 2006).

2.3. Physical Examination

Complete rhinologic, otologic, ophthalmologic, and neurologic assessments are included in the physical examination. Encephaloceles or meningoceles can be detected with a physical examination and an intranasal endoscopy. One can detect the existence of a CSF leak by examining a patient when they perform the Valsalva maneuver or when both their internal jugular veins are squeezed simultaneously (Queckenstedt-Stookey test) (Patrascu et al. 2017). Physical examinations may not be definitive in many cases, especially for patients with intermittent CSF leakage (Bolger and Kennedy 1992). The "target sign", which means the ability of CSF to move and generate a bull's-eye stain on filter paper around the central bloody spot, may be seen in some situations. This classic indicator, on the other hand, has poor specificity and can be caused by saliva or tears (Dula and Fales 1993). Physical examination of patients with bilateral CSF rhinorrhea does not reveal anything about the defect's location. Paradoxically, CSF may leak into the contralateral nostril if the midline anatomical structures (crista galli, vomer) are damaged (Patrascu et al. 2017).

2.4. Laboratory Tests

The gold-standard beta-2-transferrin test is used to identify the presence of CSF. Beta-2-transferrin is a central nervous system protein generated by neuraminidase activity. Its presence is an indirect indication of CSF rhinorrhea because it is not generally detected in nasal discharge. An amount of 0.5 ml of nasal discharge must be collected for this test. The test is both sensitive (99%) and specific (99%) (Patrascu et al. 2017; Warnecke et al. 2004; Chan et al. 2004). Beta-trace protein, which is detected in the CSF (35-fold higher quantities than serum), cardiac muscle, and plasma, is another test option, but it has poorer specificity than beta-2-transferrin. Renal failure, multiple sclerosis, and intracranial malignancies can all cause its levels to rise (Patrascu et al. 2017).

2.5. Neuro-Imaging Methods

After verifying that a patient has CSF rhinorrhea, the next task is to pinpoint the exact location and choose the best treatment.

2.5.1. High-Resolution Computed Tomography (HRCT)

The imaging approach of choice for diagnosing skull base anomalies associated with CSF rhinorrhea is a high-resolution computed tomography examination with axial, coronal, and sagittal reconstructions (Patrascu et al. 2017). The slices should have a thickness of 1 mm (Lund et al. 1994). CT scans can detect skull base deformities caused by iatrogenic or unintentional damage, anatomic disorders such as hydrocephalus (HCP) or pneumocephalus, and tumoral masses (Naidich and Moran 1980). CT scans are advised in all cases of suspected bone abnormalities of the base of the skull.

CT Cisternography

Additionally, a CT scan can be used with an intrathecal contrast material (iophendylate) to perform CT cisternography, a type of imaging. Although this procedure is more intrusive, it is more accurate in pinpointing the specific location of a bone deficiency and a CSF leak. When a patient has an active CSF fistula, CT cisternography has been shown to have a nearly 100% detection rate (Patrascu et al. 2017). This rate of detection is just 60% in the case of intermittent leaks (El Gammal and Brooks 1994). To conduct this research, pledgets must be placed in the anterior plate of the cribriform plate, the middle meatus, or the lateral sphenoethmoidal recess (Patrascu et al. 2017) under endoscopic control. Although this procedure does not offer data, the diagnosis of a fistula is certain when radioactivity is measured through the pledgets.

2.5.2. Magnetic Resonance Imaging (MRI)

Magnetic resonance imaging is another imaging technique employed in this regard, and it has high specificity for soft tissue lesions and CSF. Because of the hyperintense signal available via T2-weighted imaging that is

indicative of a CSF leak, MRI can distinguish a CSF leak from other intrasinusal fluid (Jones and Becker 2001). Intrathecal dye injection may be utilized with MRI to improve accuracy. This imaging method has worse sensitivity for detecting abnormalities of the skull base than CT and is significantly more expensive.

MRI Cisternography

Magnetic resonance imaging cisternography with Gadolinium contrast administered intrathecally is a technique whose efficacy has to be proven in more research. In situations of occult CSF fistulas in patients with recurrent meningitis who do not have CSF rhinorrhea, a diagnosis is considerably more challenging, and all neuro-imaging approaches must be balanced to arrive at the best therapeutic decision (Patrascu et al. 2017).

2.6. Perioperative Intrathecal Fluorescein

Many surgeons inject fluorescein intrathecally, both preoperatively as well as intraoperatively, to pinpoint the region where a CSF leak is taking place. An endoscope is used to assess the patient 30–60 min later. Fluorescein coloring can usually be seen without any filters in most circumstances. In the case of minor flaws, this fluid can only be detected with filters and black light. This technique has not been approved by the FDA since it has the potential to be neurotoxic in high dosages. This test's accuracy is determined by the area of the dural dehiscence, CSF volume, the time of examination following injection, and fluorescein turnover rate (Patrascu et al. 2017; Liu et al. 2009).

2.7. Treatment Options for CSF Rhinorrhea

The management of a CSF fistula can be split into two categories: conservative and surgical. Post-traumatic CSF fistulas are usually resolved with conservative care, although surgical therapy is suggested in the case of spontaneous CSF rhinorrhea. In the case of refractory CSF fistulas, one must use a combination of treatment options, including surveillance, decreasing CSF production, and surgery (intracranial or extracranial procedures) (Patrascu et al. 2017; Albert and Leibrock 2005).

2.7.1. Conservative Treatment

When it comes to post-traumatic leaks, conservative treatment is effective because there is a high chance of spontaneous resolution. In essence, conservative treatment entails bed rest (Gosal et al. 2015). To decrease CSF pressure at the interpeduncular cistern level, the patient has to lie in bed for 7 to 10 days with the head end of the bed raised 15–30 degrees (Patrascu et al. 2017). To reduce the strain and degree of intracranial pressure, stool softeners must be administered. The patient should not strain, blow their nose, or cough, nor should they carry anything heavy. It has been found that 75–80% of all post-traumatic CSF rhinorrhea cases can be resolved spontaneously in 7 days when treated with this method. Acetazolamide, for example, can reduce ICP by inhibiting the Na+/K+ ATP-ase activity that produces CSF (Patrascu et al. 2017).

Acetazolamide can thus be used as a supplement in the management of patients who have spontaneous CSF fistulas and high ICP (Carrion et al. 2001; Caballero et al. 2012). A lumbar drain is an additional tool that should be considered when the preceding management has failed for at least 5–7 days. Because it eliminates CSF pressure spikes, continuous draining is favored over intermittent drainage. To avoid side effects of over drainage like nausea, headaches, and vomiting, the suggested CSF drainage rate is around 10–15 cc/hour (Patrascu et al. 2017; Chan et al. 2004).

2.7.2. Surgical Treatment

Surgery is the only effective treatment option in some cases of CSF leakage. Indications of operation are listed in Box 1. The correct surgical treatment of CSF rhinorrhea is determined by the leak's etiology, location, and severity. The following are some general guidelines for dealing with CSF leaks: (1) treat HCP as well as meningitis before undertaking any surgical operations; (2) locate and extend the dural dehiscence; (3) dissect the bone and dural gap; (4) perform direct repair of the dural gap whenever possible; and (5) close the gap with grafts when direct dural repair is not possible (Patrascu et al. 2017).

Box 1. General indications for interventional repair (Patrascu et al. 2017).

1.	Repeated meningitis and persistent leaks despite nonsurgical treatment
2.	High-CSF-pressure fistulas and HCP
3.	Large pneumocephalus (with fluid levels higher than 2 mL) despite conservative treatment
4.	Imaging findings indicating a reduced chance of natural dural healing (bone destruction, skull base comminution, soft tissue coverage in the bony edges)
5.	Acute post-traumatic/postoperative fistulas that remain or reappear in the 10–13 days after conservative management has been conducted
6.	CSF fistulas and congenital cerebral dysplasia, especially after a single episode of meningitis.
7.	Intermittent/late fistulae

CT scans should be performed prior to surgery. A stereotactic guided approach is useful in locating the dural gap. For a successful outcome, a multidisciplinary team consisting of an otolaryngologist, an anesthesiologist, and a neurosurgeon is required. The treatment of iatrogenic CSF leaks should take place at the same time as the primary surgery.

Intracranial and extracranial interventions are used to address CSF leaks surgically.

Transcranial Approaches

In cases of comminuted or prolonged skull fractures, as well as hemorrhages in cranial fractures or contusions requiring a craniotomy, intracranial methods are used (Dodson et al. 1994). The fundamental benefit of this approach is that the dural defect may be seen directly.

Endoscopic Endonasal Surgery

Because of its low morbidity (no retraction to the brain or increased risk for anosmia) and wide range of surgical field vision, endoscopic endonasal surgery is now the preferred procedure for treating CSF fistulas. However, in order to insert the graft appropriately in this technique, one must pinpoint the exact location of the fistula (Hegazy et al. 2000; Lee et al. 2004; Lopatin et al. 2003; Marshall et al. 2001). Based on the site of the dural gap, several alternative endoscopic techniques have been devised. The major goal of endoscopic treatment is to expose the dural defect as much as possible, with intrathecal fluorescein being used in low-flow situations (Figure 14). If an encephalocele is discovered, it should be cautiously removed at once. To prevent mucocele formation, the neurosurgeon has to expose 0.2–0.5 cm of the bone around the gap and remove the remaining mucosa inside the defect before fixing it. This also accelerates osteogenesis and enhances graft acceptance (Hegazy et al. 2000; Elmorsy and Khafagy 2014). Various forms of grafts are employed in modern practice; however, their size should not exceed 30% of the defect diameter. Septal cartilage; bone from the septum, mastoid tip, or iliac crest; septal or turbinate mucosa; fascia (fascia lata or the temporal fascia); abdominal/thigh fat; pedicled (septal) flaps; or turbinate flaps are examples of grafting materials (Landeiro et al. 2004). It is worth noting that the pedicled flaps may discolor, fold, or compress. Many factors can impact the choice of graft type, including gap size and site, ICP, personal experience, and material availability (Wetmore et al. 1987). The overlay technique (directly over the gap), the underlay technique (in between the dura and the bone gap), and combination procedures are the three types of grafting techniques. Fibrin glue as well as autologous belly fat are also utilized to strengthen the transplant after it has been placed. As an overlay graft, a mucosal graft from the middle turbinate or septum can be used (Patrascu et al. 2017).

Fasciae (temporalis muscle or fascia lata) provide additional support that aids in sealing the defect.

When all the grafts are put in place, the fistula repair is conducted, using a sponge and non-absorbable nasal packing to put more pressure on the site. Neurosurgeons must carefully avoid obliterating the nearby sinus ostia. The size of the dural gap is an important parameter regarding the number of layers as well as the type of graft material when surgical planning is performed. During dural closure, one must apply the main rule —"watertight closure". When the defect is <2 mm, the graft type is not important for the success of the intervention. For fistula defects of 2–5 mm, it is advised to utilize overlay grafts (mucosal grafts or flaps), without any important dural lesions. If the fractured skull base is of the comminuted type, a composite graft should be used. Composite grafts or mucosa and bone grafts are the preferred treatment for defects larger than 5 mm. For the first 3–5 days after surgery, bed rest with the head end of the bed raised 15–30° is recommended. Antibiotics should be given, and blood pressure (BP) should be managed at a normal level (Patrascu et al. 2017; Jahrsdoerfer et al. 1981).

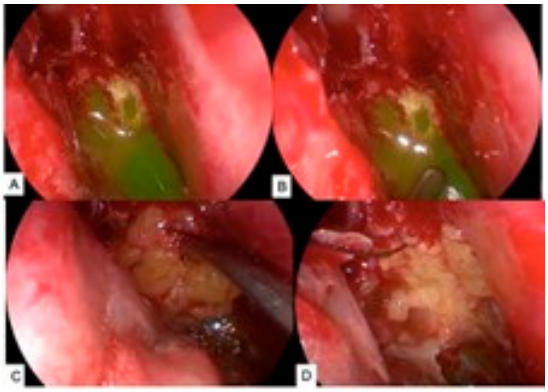

Figure 14. Endoscopic endonasal repair of CSF rhinorrhea (perioperative pictures), (**A**,**B**) showing a dural defect through which fluorescein (injected in the lumbar subarachnoid space)-stained CSF is coming out. (**C**,**D**) Pictures taken after the repair of the fistula with autogenous fat graft. Source: Figure by authors.

3. CSF Otorrhea

Even when CSF leaks through otologic structures, actual ear fluid leakage does not always occur. A rupture in the tympanic membrane or an abnormality in the external ear canal can cause CSF otorrhea; this is frequently the case when the leaking is caused by trauma or previous ear surgery. The CSF flows down the eustachian tube and manifests as a distinct form of CSF rhinorrhea if there is no such abnormality. It must be distinguished from a CSF leak from other parts of the skull base, such as the paranasal sinuses, in this case. It is also vital to realize that the fact that the leak is coming from the nose does not rule out the possibility of an otologic source. The majority of instances involve leaks as a postoperative consequence of base-of-the-skull surgery. According to reports, this problem occurs in 6–12% of such situations (Gacek and Leipzig 1979).

3.1. Congenital Cerebrospinal Fluid Leak

Problems in the otic capsule itself, aberrant patency of routes linked with the otic capsule, and defects far from the otic capsule are all possible congenital causes (Figures 15 and 16). Although congenital causes are more common in youngsters, they can manifest at any age and have even been observed among the elderly (Liao et al. 2016).

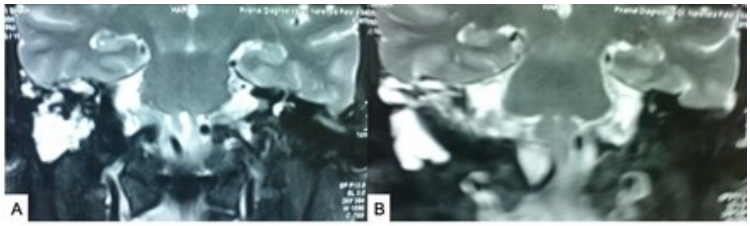

Figure 15. (**A**,**B**) MRI of brain and skull base (coronal T2W images) showing spontaneous CSF fistula (otorrhea) passing through right mastoid air sinus and middle ear cavity (tegmen tympani dehiscence). Source: Figure by authors.

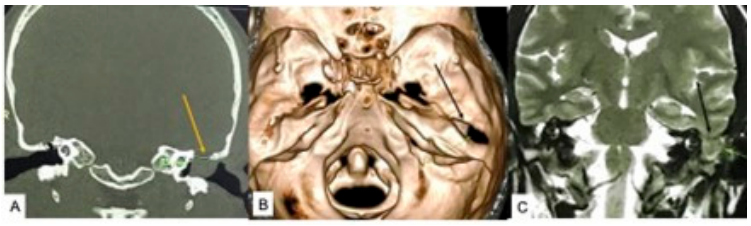

Figure 16. (**A**) CT scan of head coronal image in bony window; (**B**) CT scan 3D reconstruction of skull base showing left middle fossa bony gap on roof of middle ear cavity resulting encephalocele in middle ear cavity with spontaneous CSF otorrhea. (**C**) MRI T2W coronal image showing the encephalocele. Source: Figure by authors.

3.2. Acquired Cerebrospinal Fluid Leak

Temporal bone damage, surgery, or viral or neoplastic reasons can all induce acquired leaks (Jackson et al. 1997). Congenital spinal fluid leakage is significantly less prevalent than acquired spinal fluid leakage. The most prevalent etiology of acquired CSF fistulas is postoperative leakage following surgery. It is a well-known side effect of auditory schwannoma removal (Figure 17) and other skull base procedures. These CSF leaks are frequently visible in the first few days after surgery.

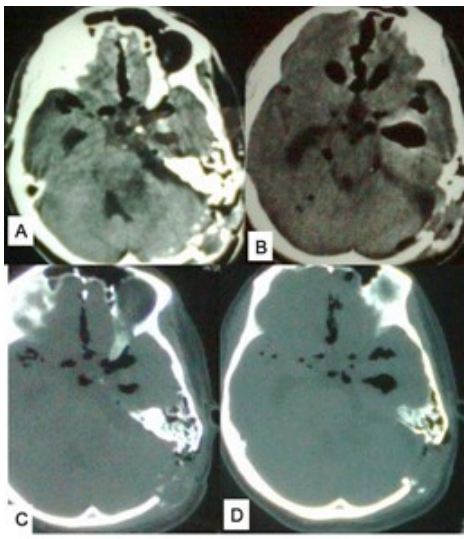

Figure 17. CT scan of head. (**A**,**B**) Axial images and (**C**,**D**) axial images in bony window showing extensive pneumocephalus after retro-sigmoid craniectomy for acoustic schwannoma where CSF rhinorrhea and otorrhea were developed through mastoid air sinus, middle ear cavity, and eustachian tube. Source: Figure by authors.

An acquired CSF leak can also be induced by mastoid surgery for chronic middle ear disease. If the dura is ruptured during surgery, the defect should be corrected as soon as feasible. Commonly, however, the dura is unharmed, but a deficiency in the tegmen's bony plate remains. The constant pulsations of the CSF thin the dura, precipitating the prolapse of the arachnoid and brain through this defect over time. This thin dura may shrink and break spontaneously, ending in a CSF fistula many years after the initial surgery.

Parts of the brain tissue, in addition to the dura, may prolapse through the dural deficiency, producing an encephalocele. Middle ear disorders, particularly cholesteatoma, can cause comparable issues even if surgery is not performed. Cholesteatoma can erode the tegmen plate, allowing dura or brain herniation to develop over time (Leung 2013).

3.3. Clinical Presentation

When there is a CSF leakage via the ear, the most common symptom presented is clear watery discharge from the external ear. But this is not always exhibited and only occurs when the eardrum or canal has been damaged in some way. If this is not the case, the CSF leakage could show up as a clear, watery nasal discharge. This discharge can be stationary or intermittent, and it may only be noticeable when the patient strains or leans forward. Some individuals report no discharge at all but instead a weird salty taste in the posterior part of the throat. CSF otorrhea is frequently accompanied by meningitis. It is found in 93% of children and 36% of adults experiencing an isolated CSF leak. Sensorineural hearing loss is another common complication of spontaneous leaks in children, occurring in 82% of cases. Seizures are another possible complication (Jahrsdoerfer et al. 1981; Gacek and Leipzig 1979).

3.4. Management

3.4.1. Surgical Treatment

A patient is at high risk of developing meningitis if CSF leaks through their ear structures. In most cases, surgery is required to correct this condition. A pressure dressing with a lumbar drain can often be used to repair

leaks that arise as a result of injury or recent skull base surgery. Surgery is advised for patients who have failed to improve following conservative treatment. CSF otorrhea is a broad term that refers to a variety of conditions. The majority of these leaks occur after surgery and are a well-known consequence of acoustic neurinoma surgery (Oh et al. 2019).

Some develop years after being precipitated by tympano-mastoid surgery or erosive pathologies such as cholesteatoma. Some may be caused by congenital anomalies in the inner ear and mastoid development.

Oh et al. discovered a link between CSF otorrhea and superior semicircular canal dehiscence (SSCD) in a retrospective study, showing that 21% of patients with CSF otorrhea had concomitant SSCD, while only 2% of controls had both (Oh et al. 2019; Brodie and Thompson 1997).

A compressive dressing as well as bed rest including head elevation are typically used to address otogenic CSF leaks caused by recent surgery or trauma. Eighty percent of the time, patients with CSF leaks caused by acoustic schwannoma surgery react to this treatment. This is also true for CSF leaks caused by temporal bone fractures, which usually close after 3–4 weeks of conservative treatment (Oliaei et al. 2012).

Surgical correction is the primary treatment for a spontaneous otogenic cerebrospinal fluid leakage. Surgery is reserved for individuals with postsurgical and traumatic leaks who have failed to respond to conservative management. The nature and location of the defect determine the correct surgical approach.

A transcanal technique can often be used to correct spontaneous CSF leaks for children with otic capsule anomalies (such as Mondini deformity). A stapedectomy is frequently performed, and the oval window is erased with soft tissue because patients rarely have the ability to hear.

In some situations of CSF leakage caused by a patent Hyrtl fissure, a transcanal technique can be used. A transmastoid technique is chosen in most situations concerning spontaneous leaking (Assietti et al. 1993).

3.4.2. Medical Therapy

Despite the fact that the presence of a CSF fistula puts a patient at a potential risk for meningitis, the use of preventive antibiotics is controversial. Many people feel that using antibiotics in the absence of an infection selects for resistant bacteria in the normal flora, making it more difficult to treat meningitis when it occurs. Antibiotics should not be applied until signs or symptoms of meningitis appear and a spinal tap confirms the diagnosis. Then, until cultures and sensitivities are returned and determined, respectively, broad-spectrum antibiotics should be used. According to a number of published studies, the risk of meningitis is greatly reduced when preventive antibiotics are administered in cases of post-traumatic CSF leaks, and their administration in this scenario is typically advised (Hanson 2020).

Medication is frequently used to reduce spinal fluid production. Diuretics (e.g., furosemide and hydrochlorothiazide), carbonic anhydrase (CA) inhibitors (e.g., acetazolamide), and steroids are examples of such drugs. These treatments are not utilized as the primary treatment for a CSF leak, but they are helpful supplements. If the output is low throughout the evaluation and diagnosis phase, they may make it difficult to find the fistula (Hanson 2020).

Continuous lumbar CSF drainage may be an effective complement to conservative treatment for a CSF leak caused by surgery or trauma. Uninterrupted CSF outflow reduces the ICP against the leak, allowing natural healing to take place. When the source of the leak is unknown, do not employ a spinal drain as it may obstruct localization and enable air entry into the cranial vault, resulting in pneumocephalus. A lumbar drain may aid healing following surgical repair in cases of spontaneous leakage (Hanson 2020).

Leaks originating in the posterior cerebral fossa anterior to the sigmoid sinus are particularly problematic as there is no arachnoid mesh in this part of the basal cistern. The leakage of CSF from this location is intense and widespread, and fascia alone is ineffective at controlling it. In most cases, a massive fat graft for the obliteration of the mastoid is required (Hanson 2020).

If a CSF leak is caused by a substantial (>1 cm) defect in the middle fossa floor, a combined middle fossa/transmastoid technique is the optimum solution. To determine the source of the leak, a mastoidectomy is performed initially. Attempting to minimize herniated brain tissue is not a good idea. Because these encephaloceles do not include functional brain tissue, they should be removed via bipolar cautery. Use the middle fossa method to fix the defect once it has been found. The middle fossa provides a great view of the defect as well as the possibility of using the lesion's intact bony margins to hold any repair material in place (Hanson 2020).

4. Spinal CSF Leaks

CSF leaks in the spinal axis, like their cranial counterparts, can be classified as traumatic or nontraumatic (or spontaneous).

4.1. Traumatic Spinal CSF Leaks

Iatrogenic injuries occurring from surgical, medicinal, or diagnostic operations are examples of traumatic spinal CSF leaks. Patients with a penetrating injury and cutaneous CSF fistula or with headaches after lumbar puncture both have simple diagnoses. Traumatic CSF fistulas have been described from the spinal subarachnoid area to the pleural space (Assietti et al. 1993; Sarwal et al. 1996). The presence of pleural fluid accumulations and symptoms of cerebral hypotension (severe positional headache) should indicate the diagnosis. A CSF leak is diagnosed by the presence of b2-tranferrin in pleural fluid.

4.2. Spontaneous Spinal CSF Leaks

The symptoms of spontaneous spinal CSF leaks are similar to those of spinal headache following a lumbar puncture; thus, the corresponding diagnoses should be easy to make. MR imaging of the brain is frequently requested as a result of severe headaches, nausea, and vomiting. Diffuse, significant dural enhancement on contrast-enhanced MR images can be misleading for meningitis or metastatic or inflammatory illnesses (Blank et al. 1997; Bruera et al. 2000; Christoforidis et al. 1998). In 60 to 70% of patients, subdural fluid accumulation suggestive of hygromas is reported. There is usually downward displacement of the cerebellar tonsils and compression of the basal cisterns (Lemole et al. 2001; Blank et al. 1997; Schievink et al. 1998; Thomson 1899).

Other warning signs include cranial neuropathies, such as uni- or bilateral sixth-nerve palsy, temporary visual problems, photophobia, hearing disturbances, facial numbness or paralysis, and stupor due to traction as well as the downward displacement of the brain stem (Berlit et al. 1994; Horton and Fishman 1994; Kosmorsky 1995; Schievink et al. 1998). The literature on spontaneous spinal CSF leaks is a little perplexing. Despite the fact that these symptoms are caused by spontaneous CSF leaks in the spine, patients are diagnosed with "spontaneous intracranial hypotension" (Lemole et al. 2001).

For patients with spontaneous spinal CSF leaks, radionucleotide cisternography is a rather common procedure used for analysis. The escape of CSF from the lumbar subarachnoid space causes rapid uptake of the tracer in the bloodstream, as indicated by the tracer's early presence in the kidneys and bladder in individuals with an active leak (Lemole et al. 2001; Schievink et al. 1998; Kadrie et al. 1976; Molins et al. 1990; Renowden et al. 1995). The location of a spinal CSF leak is occasionally recognized, but the leak is frequently below the study's resolution (Schievink et al. 1998; Kadrie et al. 1976).

If surgery is being considered, CT myelography is needed to locate the leak of CSF and may identify the underlying anatomic deficiency causing the leak, such as a meningeal diverticulum (Schievink et al. 1998). Myelography is frequently used to pinpoint the general location of a leak in preparation for CT scanning. If myelography fails to pinpoint the source of the leak, CT scanning along the complete spinal axis should be performed. Additional axial CSF collections may be visible via MR imaging of the spine, which can help locate the leak's source; however, there is little experience with using spinal MRI for individuals with a spontaneous spinal CSF fistula (Lemole et al. 2001; Matsumura et al. 2000).

4.3. Treatment of Spinal CSF Leaks

The treatment of spinal CSF leaks is similar to that used for cranial CSF fistulas. Resting in bed and a brief period of diversionary lumbar CSF drainage are often sufficient in the case of post-traumatic and postoperative CSF leakage. Although simple oversewing of the site can generally resolve postoperative leaks, patients should be monitored for indicators of ongoing leaking, such as positional headaches or the development of a pseudomeningocele. When conservative methods fail to resolve the fistula, surgical exploration should be explored (Lemole et al. 2001).

After surgery, a careful examination typically shows an undiscovered dural rupture that can be repaired using the direct suture method, fibrin glue administration, or both. The correct approach to treating spinal fluid leaks caused by penetrating trauma is determined by the location of the CSF leak, which is best determined via CT myelography. Some success has been found with percutaneous approaches for introducing fibrin glue to the leak location (Lemole et al. 2001; Hughes et al. 1997; Patel et al. 1996).

Fluid replacement and bed rest are the first steps in treating spontaneous spinal CSF fistulas to relive the symptoms of intracranial hypotension. Intravenous caffeine, glucocorticoids, mineralocorticoids, nonsteroidal anti-inflammatory drugs, and salt infusions are some of the medical treatments available. A lumbar epidural blood patch is a more direct and, in some authors opinion, more effective treatment. The findings reported by Szeinfeld et al. (Szeinfeld et al. 1986), revealing that blood injected into the lumbar epidural spaces extended upward and downward to involve eight or more spinal segments, may explain the success of this procedure. Fortunately, these conservative approaches will work for 60 to 70% of patients (Blank et al. 1997; Schievink et al. 1998; Inenaga et al. 2001). Surgical treatment should only be considered if symptoms persist after two sufficient blood patches have been applied.

The cervicothoracic junction or the thoracic spine are the sites of the majority of recorded leaks (Kamada et al. 2000). These leaks were caused by ruptured meningeal diverticula for which surgical confirmation was available. The surgical closure of leaky meningeal diverticula has been linked to positive results (Schievink et al. 1998; Inenaga et al. 2001). Epidural patching combined with fibrin glue has recently been utilized to treat spontaneous spinal CSF fistulas in patients who had previously failed to respond to epidural blood patches (Lemole et al. 2001).

Most meningeal diverticula have no recognized cause, and it is uncertain whether they're congenital or acquired. In some situations, however, an underlying weakening of the spinal meninges is likely to occur, predisposing the development of meningeal diverticula.

5. Conclusions

Anywhere along the craniospinal axis, CSF leaks can occur. Although cranial CSF leaks are most commonly caused by trauma, spontaneous occurrences are becoming more common, especially along the spinal column, where they present as spontaneous intracranial hypotension. Management options vary depending on the cause and degree of the CSF leak, but the general approach is to begin with conservative therapies and progress to more bodily invasive techniques if needed. Patients with substantial volumes of pneumocephalus, extensive base-of-skull fractures, or chronic CSF leakage may require immediate surgical intervention.

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