Cerebrospinal Fluid Rhinorrhoeas an Overview

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Cerebrospinal Fluid Rhinorrhoeas an Overview

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Abstract

This article discusses etiopathogenesis, clinical features and management of cerebrospinal fluid rhinorrhoea.

Cerebrospinal fluid rhinorrhoea is the leakage of cerebrospinal fluid from the subarachnoid space into the nasal cavity due to defect in both dura and bone.

Various causes of CSF rhinorrhoea include:

1. Traumatic
2. Iatrogenic
3. Idiopathic
4. Tumors

Physiology

CSF is formed primarily in the choroid plexus found in the lateral, third and fourth ventricles. Extra choroidal formation of CSF is from the paranchymal capillaries and from intra cellular water metabolism

CSF flows from its production sites in the two lateral ventricles through the foramina of monro into the third ventricle and to the fourth ventricle through the aqueduct of sylvius. Flow continues through the fourth ventricle, located in the brain stem and communicates with the cisterna magna through the midline foramina of Luschka from the cisterna magna. CSF flows into the subarachnoid space. CSF is absorbed into the cerebral venous system through the arachniod villai.

CSF is formed at the rate of 0.35ml/min,6 or 350-500ml / day and it varies with circadian rhythm. The total volume of CSF is turned over about three times a day. The normal CSF pressure is 5-156 mm Hg or 5-15 cm water in prone position and increases to 40 cm of water with movement into sitting position. CSF pressure varies with the time of the day, age of the patient, activity level, respiratory and cardiac cycles. Neurologic systems may develop when the pressure higher than 20cm of water is sustained.

Functions of CSF

- Physical support and buoyancy for the brain.

- Maintain safe intracranial pressure.
- Removal of byproducts of metabolism.
- Regulate the chemical environment of the brain.

Etiology and classification

CSF Rhinorrhoea is classified according to the etiology developed by omaya. He divided CSF Rhinorrhoea into traumatic and non-traumatic, subdividing the latter into non –traumatic with normal pressure and non traumatic with high pressure.

Classification of CSF Rhinorrhoea

Traumatic causes

- Accidental
- Surgical

Non traumatic causes

- High pressure leaks

Tumours

Hydrocephalus

- Normal pressure leaks

Congenital anomely

Spontaneous osteitis

osteomyelitis

Traumatic CSF Rhinorrhoa

Accidental Trauma

Accidental trauma in the most common etiology (80%) of CSF leaks. Leaks occur in 2-3% of patients with closed head injury and it 30% of patients with skull base fractures. CSF rhinorrhoea may occur directly through the anterior cranial fossa or indirectly from the middle or posterior fossa through the eustacian tube. Most frequent sites of CSF rhinorrhoea are Fovea ethmoidalis, Cribriform plate posterior wall of frontal sinus and Sphenoid sinus. Because the anterior cranial fossa dura adherent to the bone is easily torn by fractured bone edges and the point where the anterior ethmoidal artery enter the lateral lamella in the place of least resistance in the entire skull base that a
CSF fistula can occur.

In some patients avulsion of olfactory fibres from the cribiform plate by the shearing forces of a blunt trauma can rarely cause rhinorrhoea in the absence of fracture. Transverse fractures through the petrous bone cause CSF leak in to the middle ear and drain through the eustacian tube to the nasopharynx (Otorhinorrhoea). Rarely, a cranio orbital fracture together with the laceration of conjunctival sac may cause CSF to leak from the eye (Occulo Rhinorrhoea).

**Traumatic CSF rhinorrhoea**

Traumatic CSF rhinorrhoea usually begins within 48 hours of injury in 55% of the patients. The frequency increases to 70% by the end of first week. If the defect is small the bone may heal and in 60-70% of cases spontaneous healing occur within a week. Healing is unlikely to occur if

- The bone defect is large.
- Dura is penetrated by bony spikes
- Tissue healing is impaired by metabolic disease
- Increase in intra cranial pressure.

The onset of CSF may be delayed or recur after a period of cessation. Possible reasons for this are

- Lysis of blood clot at the fractured site
- Resolutions of soft tissue edema.
- Maturation and shrinkage of the dural scar
- Devascularisation and necrosis of the bone and soft tissue.
- Intermittent pulsations causing herniation of dura through a non-healed fractured site

**Surgical trauma**

Surgical procedures cause approximately 16% of CSF rhinorrhoea. With the development of new surgical procedures, the etiology has changed and now the incidents of iatrogenic CSF rhinorrhoea has increased. In the past, the majority of iatrogenic fistulas are due to neurologic surgeries and now the endoscopic sinus surgery is the leading cause for CSF rhinorrhoea (0.5-0.3%).

The two most common sites of skull base injury associated with FESS are lateral lamella of cribiform plate and posterior ethmoid roof near the face of the sphenoid. Injury to lateral lamella of cribiform plate can occur during an approach to anterior ethmoids or frontal recess or when resecting the middle turbinate close to the skull base. Posterior ethmoid defects occur typically in highly pneumatised maxillary sinus that extends superomedially causing decrease in posterior ethmoid pneumatisation.

The most common cause of rhinorrhoea following neurologic surgeries are transphenoidal management of pituitary tumours (0.5 – 15%), surgeries for acoustic neuroma (7-11%), cranio facial resection and transcranial approaches.

Post surgical traumatic CSF rhinorrhoea may be present during immediate post operative period in 50% of patients.

**Non-traumatic CSF rhinorrhoea**

Non-traumatic CSF rhinorrhoea is subdivided into high pressure and normal pressure leaks commonly occurs in adult females aged more than 30 years.

**High pressure CSF Rhinorrhoea**

In the high pressure leak the leakage of CSF is usually acting as a safety valve there by reducing the intra cranial pressure which commonly occur in cribiform plate and roof of ethmoid. In such cases closure of the defect will worsen the patient condition if the causative lesion is not treated. Increased CSF pressure results from tumours or hydrocephalus. Tumours may cause CSF rhinorrhoea either through direct erosion of bone or indirectly through pressure erosion. Treatment for the tumours such as surgery, irradiation or chemotherapy can cause devascularisation of the tumour bed and skull base defect.

**Normal pressure CSF Rhinorrhoea**

In non traumatic CSF rhinorrhoea occurs in normal pressure system probably results from normal physiologic increase in CSF pressure. The increase in CSF pressure generally is not able to erode the bone, the presence of sudden, increase in intracranial pressure caused by coughing or straining may be an important precipitating factor in the development of spontaneous CSF rhinorrhoea.

- Normal pressure leaks may result from congenital defects such as, preformed pathways, fistulas, meningoceles, meningoencephaloceles or encephoceles.
- The empty sella syndrome occurring because of an...
absent portion of the diaphragma sellae is classified as congenital. The empty sella syndrome occurring because of pituitary gland atrophy or degeneration is classified as spontaneous.

- Osteitis or osteomyelities of skull base bone may cause CSF rhinorrhoea infrequently.

Clinical features

- **CLEAR NON –STICKY FLUID DRAINING FROM THE NOSE** sometimes rarely confused with allergic or vasomotor rhinitis. In traumatic cases CSF is mixed with blood. Some patients may complain of salty taste (because of sodium content) of CSF or even a sweat taste (CSF has two thirds of Blood sugar). The amount of discharge may be continuous and copious in patients with non traumatic fistula then in patients with traumatic fistula for whom the leak is intermittent and scantly. CSF may accumulate in the sinuses when the patient is in supine position and then discharge through nostril when the head is flexed (Reservoir sign). If CSF leaks occur when the head is upright (or) tilted backwards suggests that the leak is through the cribriform plate, ethmoid roof or frontal sinus. Leakage occurs on tilting the head forward suggests that the leakage is from the sphenoid sinus or through the middle ear. CSF leak may be unilateral or bilateral but this does not always correspond to the site of the fistula.

- **HEAD ACHE**

  Head ache can be either high pressure or low pressure type. In high pressure head ache there will be steady increase in pain which is relieved by fluid drainage and in low pressure head ache the pain is less marked in recumbent position and are increased by the upright position which results from acute or chronic leak. In some patients headache can be caused by intra cranial air.

- **HYPOSMIA OR ANOSMIA**

  Hyposmia or anosmia is due to olfactory nerve damage from fracture of the cribriform plate.

- **RECURRENT ATTACKS OF MENINGITIS**

  Infection alone may be the first sign of fistula without history of CSF Rhinorrhoea, most of the patients are belong to the delayed post traumatic group. Possible explanations for this are,

  - Age related shrinkage of brain previously plugging a defect.
  - Cerebral – dural scar that sealed the scar did not provide reliable barrier to infection.
  - Growing fractures of the ethmoid leading to the formation of a herniated encephalocele that stretched and ruptured as a result of intracranial pulsations.

**BEDSIDE TESTS**

**HALO SIGN OR TARGET SIGN**

A clear watery fluid leakage from the nose is likely to be CSF. If the fluid is mixed with blood or nasal discharge the presence of CSF is indicated by halo sign. The discharge is placed on filter paper CSF will migrate farther and form ring like pattern around the blood and mucus in the centre.

**HAND KERCHIEF TEST**

A wet hand kerchief that dries without stiffening suggestive of CSF leak.

**GLUCOSE OXIDASE TEST STRIPS**

The test strips are positive at a relatively low level of glucose. Reducing substances in the lacrimal gland secretions and nasal mucus may cause a positive reaction. Hence a negative test excludes the present of CSF but a positive result cannot be interpreted except in the presence of CSF infection.

**Laboratory tests**

**BETA – 2 TRANSFERRIN**

- Beta-2 transferrin is highly sensitive and specific in identifying fluid as CSF.
- Beta -2 transferrin is a polypeptide involved in ferrous iorn transport.
- It is produced by desialisation of the normal Beta -1 transferrin in CSF through cerebral neuraminidase. It is found only in CSF, perilymph and vitreous humour.
- Nasal secretions can be tested for the presence of this protein and less than 1 ml of fluid is required for diagnosis.
- False positive results are possible in patients with chronic liver disease, inborn errors of glycoprotein metabolism, genetic varients of transferrin, neuro psychiatric disease and rectal carcinoma . when these pathologic conditions are suspected sampling of venous blood should be sampled for comparison.

**BETA TRACE PROTEIN**

Beta trace protein is another brain specific protein produced mainly in the leptomeninges and choroid. It is the second most abundant protein in CSF after albumin. It can also found in serum and perilymph . Beta trace protein is a reliable marker for detection of CSF in nasal secretions and it is used most commonly in Europe .

**GLUCOSE CONCENTRATION**

Glucose more than 30mg /dl or two thirds of blood glucose in clear nasal fluid indicates the presence of CSF in the nasal discharge.
CHLORIDE CONCENTRATION
Chloride concentration more than 110 mg/l suggests that the fluid is most likely CSF.

Role of imaging

PLAIN RADIOGRAPHY
Plain radiography are of limited value but they may show skull base fractures, fluid in the paranasal sinuses and intracranial air.

PLAIN CT SCAN
Plain CT brain is recommended in cases of spontaneous CSF leak to exclude causes such as intra cranial mass or hydrocephalus

HIGH RESOLUTION CT ( HRCT )
HRCT provides thin sections (0.6-1mm) in both the axial and coronal planes. The axial images shows the posterior wall of frontal sinus and sphenoid sinuses. Coronal images shows the ethmoid complex, roof of sphenoid sinus and the tegmen of the middle ear. HRCT is able to identify even the smallest bone defect along with skull base with high sensibility. HRCT is independent on leak activity at the time of imaging.

CISTERNOGRAPHY (CTC)
In CTC an intrathecal injection of non ionic contrast medium in the lumbar region.In the presence of active leak CTC demonstrates movement of the contrast through the defect. The rate of detection is lower if no leak is present at the time of investigation. The site of leakage is indicated by bony dehiscence, contrast agent in the adjacent para nasal sinuses,distortion of subarachnoid space and brain herniation. CTC is of particular use when the frontal and sphenoid sinuses act as reservoirs. CTC is contra indicated for patients with active meningitis and increased intracranial pressure. Weakness of this technique includes its inability to detect an active leak at the time of study, adverse reactions, and increase exposure to radiation. Contrast agents such as iohexol and iopamidol have a lower incidence of side effects.

MAGNETIC RESONANCE CISTERNOGRAPHY (MRC)
MRC is a non invasive technique that can detect CSF fistula in multiple planes which does not involve the use of contrast (or) spinal puncture. On the T2 weighted fast spin echo the CSF has a characteristic bright signal that can generally distinguished from inflammatory paranasal sinus secretions. MRC is consider positive if herniation of brain tissue or arachnoids through a bony defect and CSF signal in the paranasal sinuses continues with CSF in the sub – arachnoid space. MRC is superior to CTC in cases of

- Multiple dural defects
- Intermittent leakage
- Dural defect <1mm

RADIONUCLIDE CISTERNOMGRAM(RNC)
RNC is similar to CTC in that the radio active material most commonly TE-99 is injected intrathecally followed by gamma camera imaging in different positions. RNC is particularly useful in low volume or intermittent leaks. In such cases RNC is combined with endoscopic placement of nasal pledges that are placed in sphenoid recess ,middle meatus and olfactory cleft before starting the study. After imaging the blood samples are taken and the pledges are removed at the same time, the normal ratio (radionuclide count in pledges / radioneuclide count in blood sample) should be < 0.37, the pledge with highest count is assumed to have been nearest to the leak

INTRATHECAL FLUORESCIN
This technique is highly successful and accurate in diagnosing and localising an active CSF leak most commonly used as an adjacent to intraoperative localization of a skull base defect. 10ml of CSF is withdrawn by lumbar puncture is mixed with 0.2 to 0.5ml of 0.5% fluorescein and slowly injected through a lumbar drain. Fluorescein stained CSF can be seen as bright yellow or green. Use of a blue light filter makes the test sensitive upto 1 in 10 million. Side effects of this technique includes lower extremity weakness, numbness, generalized seizures, opisthotonus and cranial nerve deficits.

PET SCAN
PET scan has been used to demonstrate a leak in some difficult cases where the side and site of the fistula is not obvious. This is particularly useful in cases of CSF otorrhoea where it is not clear whether the leak is from posterior fossa or middle cranial fossa.

Management

Conservative Management
Most CSF leaks resulting from accidental and surgical trauma heal with conservative measures over a period of 7-10 days. Conservative management consists of
• Bed rest with head end elevation
• Avoidance of straining activities such as nose blowing, sneezing and coughing
• Use of laxatives and stool softeners to reduce straining

If the leak does not resolve within 3 days intermittent or continuous drainage of CSF may be tried for the next 4 days with removal of 150ml/day. Continuous CSF drainage is hazardous and should be used in caution. Over drainage can lead to intracranial aeroceles, severe brain displacement and coma. Intermittent drainage of 20-30ml over and 8 hour period into a closed system is safer.

A non-traumatic high pressure leaks caused by increased intracranial pressure will probably resolve if the intracranial pressure is normalized. Intra cranial pressure can be normalized by use of diuretics such as azetazolamide or with ventriculo peritoneal shunting. Leaks that do not resolve with normalization of intracranial pressure need surgical management. Normal pressure non traumatic leaks rarely close with conservative therapy and almost always require surgical exploration.

Antibiotic prophylaxis remains controversial. Untreated CSF rhinorrhoea has been associated with a 25% risk of meningitis. Risk of meningitis is greater with

• Delayed CSF leakage
• Longer duration of CSF leakage
• Concurrent infection

The arguments against antibiotic prophylaxis are

• The antibiotics commonly used penetrate CSF poorly. If the antibiotics are used a combination of cotrimazol which is bactericidal in CSF and amoxicillin or penicillin which are bactericidal in nasal mucosa is recommended.
• Antibiotics may promote resistant strains of organisms within the nasopharynx and consequently lead to infection with resistant or unusual organisms.

### Surgical Management

The surgical management of CSF Rhinorrhoea can be divided into intracranial and extra cranial approaches. Dandy described the first surgical repair through a bifrontal craniotomy in 1929. Dohlman was the first to document the first intracranial repair of CSF leak in 1948. In 1981 Wigand described closure of CSF leak using an endoscopic approach. Majority of traumatic CSF fistulas heal without surgical intervention. Patients who develop CSF rhinorrhoea, shortly after trauma do not need surgery to close the CSF fistula.

**Indications for early surgery are**

• Penetrating injury including gunshot wounds
• Anterior cranial fossa surgery indicated for other reasons such as intracranial hematoma or to repair compound facial fractures with accessible dural tears being treated at the same time.
• Meningitis once treated
• A large intracerebral aerocele
• Herniation of brain tissue through the nose
• Radiological appearances that indicate a low probability of natural dural repair.

**Delayed surgery is indicated for**

• Failed conservative management – CSF leak persisting beyond 10 days
• Recurrent or delayed CSF leak after 10 days
• Recurrent aerocele after 10 days
• Meningitis or abscess at any time after surgery.

When CSF rhinorrhoea results from surgery the dural injury should be repaired when it occurs. Post operative leaks will usually close with conservative management.

**Intra cranial repair** of CSF leak

Repair of anterior fossa fistulas can be approached by frontal anterior fossa craniotomy. Middle cranial fossa leaks from a petrous fracture are best approached through a subtemporal craniotomy. Posterior fossa leaks from the posterior petrous surface are often associated with hearing loss. If the hearing is lost these fistulas may be repaired through a Trans labyrinthine approach. If the hearing is intact, they should be approached via the posterior fossa. Leaks from the sphenoid sinus area are difficult to approach via the intracranial route.

Intra cranial surgery is indicated when

• operating for associated craniofacial injuries
• Large bone defects that may be difficult to repair endoscopically.
• The fistula site cannot be identified by endoscopic examination.
• Tumours with intracranial extension that are not amenable to endoscopic resection.

**Advantages of intracranial repair are**

• Improved exposure
• Ability to identify multiple defects
• Repair can also be done even under condition of increased intra cranial tension

**Disadvantages of intra cranial repair are**

• Increased morbidity
• Permanent anosmia
• Trauma related to brain retraction
• Increased hospital stay
Extra cranial repair of CSF leak

Extra cranial approach includes anterior osteoplastic approach via bicoronal or eyebrow incision, external ethmoidectomy, transethmoidal sphenoidectomy and transseptal sphenoidectomy have lower morbidity rates, higher success rates and no anosmia. They provide that best exposure of the sphenoid, parasellar and posterior ethmoids, cribriform plate, fovea ethmoidalis and fistulas in the posterior wall of frontal sinuses. Cerebral damage and the lateral extensions of the frontal and sphenoid sinuses cannot be assessed.

Disadvantages

- Facial scar
- Facial numbness
- Orbital complications

Endoscopic repair of CSF leak

Endoscopic approach to CSF fistula depends on the suspected site of the lesion. Intra-cranial lesions, comminuted fractures of the cranial base, fracture of posterior wall of frontal sinus are contraindications for endoscopic repair. Patients who have an active CSF tear during surgery will not require placement of fluorescein intrathecally. Fluorescein can help identify the site of small leak that is intermittent or that has recently stopped leaking. If blue-light filter is used on the light source even the smallest quantities of fluorescein can be visualised.

CSF leaks at the ethmoid sinus or the lateral lamella of cribriform plate will require complete endoscopic anterior and posterior ethmoidectomy to gain wide exposure to skull base. CSF leaks at the cribriform plate approached directly through olfactory groove. Sphenoid sinus CSF tears can be approached in many ways including a trans – septo sphenoid approach, approach through the sphenoid - ethmoid recess or a transeptal approach. Defects located in the lateral recess of sphenoid sinus are difficult to access by the transeptal or transethmoid approaches and may require an endoscopic transpterygoid approach. Defects directly involve the frontal recess may require a combined approach using endoscopic and open techniques because the superior extent of the defect may be difficult to reach endoscopically and inferior posterior extension may be difficult to reach from an external approach. Once the tear is localised, the nasal or sinus mucosa around the site of the tear is removed for about 5mm to expose the bone around the defect this allows attachment of the free graft to the bone. Sinus mucosa continues to secrete mucus and may separate the graft from the recipient bed if the mucosa is not removed. When the appropriate mucosa is removed a diamond burr or curette can be used to abrade the recipient bed bone lightly and stimulate osteogenesis. If the dural defect is smaller than the bony defect the dural defect is enlarged to the size of bony defect for the adequate support of graft material with the underlying bone.

Graft materials

Historically, non vascularised graft such as pericranium, temporalis fascia, facia lata, muscle, fat, allograft or synthetic dura or surgical cellulose mesh were used. These graft carried high risk of necrosis, post operative CSF leaks and infection. Today dural closure is accomplished by auto graft such as temporalis fascia, facia lata, abdominal fat, septal mucoperichondrion and turbinate bone. Lypolised cadaver dura and bovine pericardium are also be used. These grafts are further supported by local or free vascularised tissue. The fibrin sealant provides a temporary watertight closure and creates an additional barrier to CSF leak.

Applying the graft.

The graft is applied using various techniques.

Onlay technique

The onlay technique is generally employed for defects located in the lamina cribrosa where the presence of olfactory nerve filaments make it difficult to dissect dura from the adjacent skull base. Cartilage or bony graft is placed on the extra cranial surface of the skull duraplasty is then completed with a second layer of free mucoperichondrium. This technique can also be used in lateral wall of extensively pneumatised sphenoid sinus.

Underlay technique

This is ideal for defects located in the fovea ethmoidalis. Graft material is positioned between the dura and the bone.

Bath plug technique

Once the defect has been prepared the skull base defect is measured. If the size of the defect is measured if the size of the defect is less then 12mm a fat plug is harvested from the ear lobe. If the defect is larger than 12mm, fat is obtained either from the region of the greater trochanter of the thigh or from the abdomen. The fat of the ear lobe is preferred because the fat globules are tightly bound and easy to work with. The fat plug should be the same diameter as the defect and 1.5 to 2cm long. A free mucosal graft is harvested from the lateral nasal wall (usually on the
opposite side of CSF leak)

A 4-0 vicryl is knotted through the one end of the fat and the suture passed down the length of fat plug. The fat plug is placed below the defect and a malleable frontal sinus probe in used to introduce the fat plug through the defect, once the fat plug has been safely introduced the plug is stabilised with the probe and the suture is gently pulled. The free mucosal graft is slide up the vicryl suture to cover the slightly protruding fat plug and skull base defect.

Cuff – link repair

This technique uses a double layer of lyophilised dura or fascia to sandwich the dural defect, taking advantage of the hydrostatic CSF pressure to seal the defect and stop the leak. This is a variation of bath plug technique to repair sellar and clival defects successfully.

Sandwich Technique

In sandwich technique the first layer of graft is inserted as an underlay, fibrin glue is then applied followed by a disc of septal cartilage designed to fit the bony defect this is followed by fibrin glue, the final layer of graft placed as overlay and the repair is supported with a vascularised graft.

Obliteration [ Tobacco pouch ] technique

This is an alternative procedure for defects located in the lateral recess of sphenoid sinus. Placement of graft is not possible in these cases because of the adjacent neuro vascular structures. After careful removal of the entire mucosa investing the sinus, the entire cavity is obliterated with fascia lata plus gel foam or abdominal fat and then sealed by mucoperichondral graft.

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Illustrations

Illustration 1

Figure showing CSF formation and circulation

Illustration 2

Figure showing routes of CSF leak
Illustration 3

Figure showing onlay technique of CSF leak repair

Illustration 4

Figure showing underlay technique of CSF leak repair
Illustration 5

Figure showing bath plug technique of sealing CSF leak

Illustration 6

Figure showing sandwich technique of CSF leak seal
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