

CSF Rhinorrhea

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Definition

Disruption of the arachnoid and dura mater
coupled with an osseous defect

PLUS

A CSF pressure gradient that is either
continuously or intermittently greater than
the tensile strength of the disrupted tissue

Aetiology

Classification is via etiology

- Meningoencephaloceles
 - Congenital
 - Acquired
- Spontaneous
- Traumatic
 - Blunt
 - Penetrating
- Iatrogenic

Aetiology

- Accidental Trauma-80% of all CSF rhinorrhea
- Procedure related-16% of all CSF rhinorrhea.
- Non-traumatic-4% of all CSF rhinorrhea.

Aetiology

Traumatic

■ Accidental

- Usually anterior skull base fracture:
 - sphenoid
 - fovea ethmoidalis
 - Posterior table of frontal sinus (Lothrop or osteoplastic flap)
- Can Be:
 - Immediate leak of CSF (Majority within 48hrs)
 - Delayed – 95% present within 3 months
- Conservative Mx – most seal with 10 days
- Persistent leaks (>10 days) should be closed (bone spicule)

Aetiology

Iatrogenic (50% may have a delayed presentation)

- Complication of neurosurgical procedures
- Complication of rhinologic procedures
 - Sinus surgery
 - Septoplasty
 - Other combined skull base procedures
- Frequently seen on:
 - lateral wall of the olfactory fossa (thin, 1-0.1mm thick, perforated by ant ethm art, medial limit of frontal sinus dissection)
 - fovea ethmoidalis\
 - Ethmoid skull base 35%, Cribriform 27%, sphenoid 18.9%

Etiology

Non-traumatic

- Spontaneous:
 - Cribriform plate (dilatation of the dural sheath around olfactory fibers – small prolapse of dura causing a leak)
 - lateral wall sphenoid sinus
 - Well pneumatized sphenoids into the clinoid process
 - Sphenoid is in close contact with temporal lobe and MCF
 - Arachnoid granulation tissue sacs erode the bone
 - Prolapse of dura and arachnoid with leak
 - May be associated with BIC hypertension

Aetiology

- Non-Traumatic cont...
- Elevated intracranial pressure
 - Intracranial neoplasm
 - Hydrocephalus
 - Non-communicating
 - Obstructive
 - Benign intracranial hypertension
- Normal intracranial pressure
 - Congenital anomaly
 - Skull base neoplasm
 - Nasopharyngeal carcinoma
 - Sinonasal malignancy
 - Skull base erosive process
 - Sinus mucocele and Osteomyelitis
 - Idiopathic



Aetiology

- Meningoencephaloceles
 - Spontaneous (congenital or acquired)
 - Previous traumatic event
- Congenital = 1st few years of life
- Consists of meninges and dura containing CSF with a variable amount of brain prolapsing through a funnel shaped defect of the skull base
- Brain tissue = non-functioning – remove it

Physiology

- CSF produced by choroid plexus (0.35ml/min).
- CSF circulates from ventricles through foramina Luschka and Magendie to subarachnoid space.
- Total CSF volume is 140mL (90 – 150 recycled 3-5x/day)
 - 20 mL (ventricles)
 - 50 mL (intracranial subarachnoid space)
 - 70 mL (paraspinal subarachnoid space).
- CSF flows into the subarachnoid space and is absorbed along the cerebral convexities by arachnoid villi
- Normal CSF pressure is 5-15cm H₂O in the lumbar cistern
- Neurological symptoms occur at 15-20 cm H₂O

Physiology

- CSF pressure maintained by relative balance between CSF secretion (choroid plexus) and CSF resorption (arachnoid villi 1 way valve requiring a pressure gradient of 1.5-7cm H₂O).
- CSF resorption rate plays major role in determining CSF pressure.
- CSF rhinorrhea requires
 - disruption of barriers that normally separate the contents of the subarachnoid space from the nose and paranasal sinuses
 - Pressure gradient to produce flow of CSF.

Pathophysiology

- Immediate Leaks
 - Dural tear, bony defect
- Delayed traumatic leak
 - Slowly intact dural layer has herniated through a bony defect finally tearing the dura and causing a leak
 - Dural layer was always torn but leak occurs after haematoma dissolves

Hubbard JL Spontaneous cerebrospinal fluid rhinorrhea: evolving concepts in diagnosis and surgical management based on the Mayo Clinic experience from 1970 through 1981. *Neurosurgery*. Mar 1985

Lindstrom DR, Toohill RJ, Loehrl TA, Smith TL. Management of cerebrospinal fluid rhinorrhea: the Medical College of Wisconsin experience. *Laryngoscope*. Jun 2004

Pathophysiology

- Spontaneous Idiopathic Leaks
 - Increased ICP (mean csf = 32.5)
 - Impaired absorption of CSF by Arach Villi
 - Pulsatile force exerted to weak points e.g. Crib Plate
 - Ant Cranial fossa is subject to wide variations in icp
 - Arterial
 - Respiratory fluctuations
 - Primary non-traumatic
 - Focal atrophy
 - Rupture of arachnoid projections (accompany olfactory nerve)
 - Persistence of olfactory lumen

Pathophysiology

- Spontaneous Idiopathic Leaks
 - Obese, middle aged women with spontaneous clear rhinorrhea
 - Pressure headaches
 - Pulsatile tinnitus
 - Balance disturbance
 - Visual disturbance



Pathophysiology

- The most common anatomic sites of CSF leaks are the areas of congenital weakness of the anterior cranial fossa and areas related to the type of surgery performed.
 - 39% cribriform plate and air cells of the ethmoid sinus
 - 15% fistula extends to the frontal sinus
 - 15% sella turcica and sphenoid sinus.
- Common sites of injury secondary to endoscopic sinus surgery include the
 - lateral lamella of the cribriform plate
 - posterior ethmoid roof near the anterior and medial sphenoid wall.
 - Rare: middle or posterior cranial fossa and can reach the nasal cavity by way of the middle ear and eustachian tube.

History

- CSF leak is:
 - clear
 - watery discharge
 - usually unilateral
 - sweet
- Diagnosis is made more easily in patients with recent trauma or surgery than in others.
- Delayed fistulas are difficult to diagnose and can occur years after the trauma or operation (caution misdiagnosis of allergic and vasomotor rhinitis).
- Reservoir sign: Headache relieved by drainage of CSF. Drainage may be intermittent as the fluid accumulates in 1 of the paranasal sinuses and drains externally with changes in head position
- Headache and visual disturbances suggests increased intracranial pressure.

History

- Associated symptoms can assist in localizing the leak.
 - Anosmia (present in 60% of individuals with posttraumatic rhinorrhea) = injury in the olfactory area and anterior fossa, especially when it is unilateral.
 - Visual fields = Optic nerve suggests a lesion in the region of tuberculum sellae, sphenoid sinus, or posterior ethmoid cells.
 - Recurrent meningitis (pneumococcal meningitis), evaluate for a defect that exposes the intracranial space to the upper airway regardless of the presence or absence of CSF rhinorrhea.

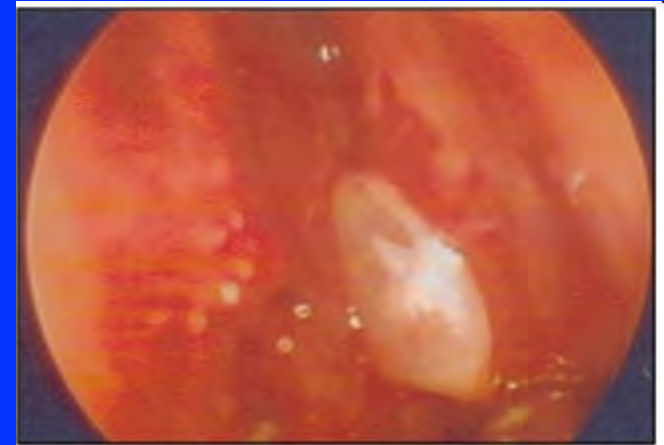


Fig 3. Operative endoscopic view. Fistula site is identified with the CSF coming out from the ethmoid roof.

Physical Examination

- Complete Examination including
 - rhinologic
 - otologic
 - head and neck
 - neurologic
- Endoscopy
 - encephalocele
 - meningocele.
 - Drainage of CSF can often be elicited on endoscopy
 - Valsalva maneuver or by compressing both jugular veins (Queckenstedt-Stookey test).
 - Change in head position can elicit a reservoir sign.

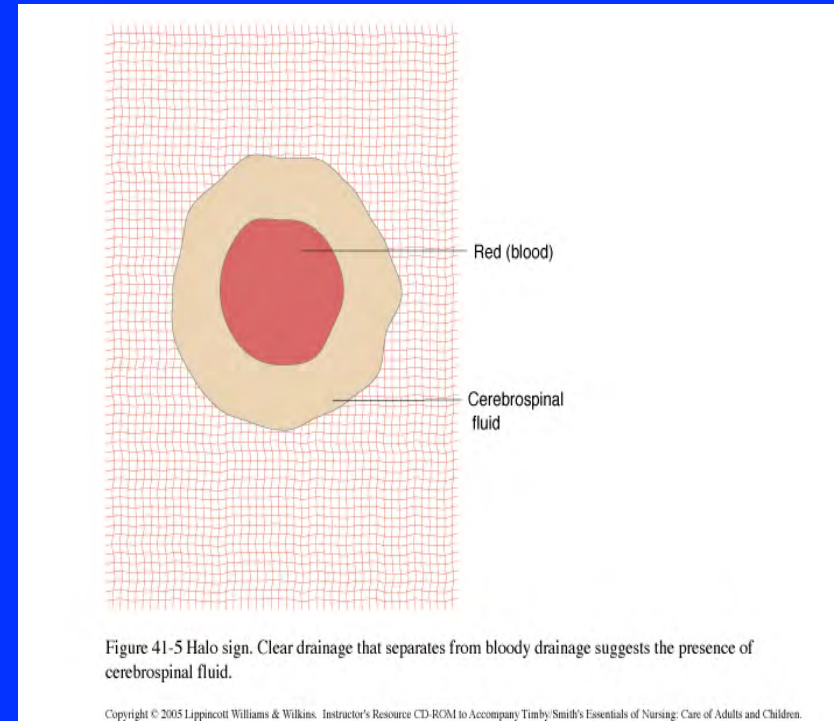


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Physical Examination

- Head trauma
 - mixture of blood and CSF may make the diagnosis difficult.
 - CSF separates from blood when it is placed on filter paper, and it produces a clinically detectable sign: ring sign, double-ring sign, or halo sign. However, the presence of a ring sign is not exclusive to CSF and can lead to false-positive results.



Physical Examination

- Paradoxical rhinorrhea: when midline structures that act as separating barriers (e.g., crista galli, vomer) are dislocated. This dislocation allows CSF to flow to the opposite side and manifest at the contralateral naris.
- The clinical findings most frequently associated with CSF rhinorrhea are meningitis (30%) and pneumocephalus (30%).

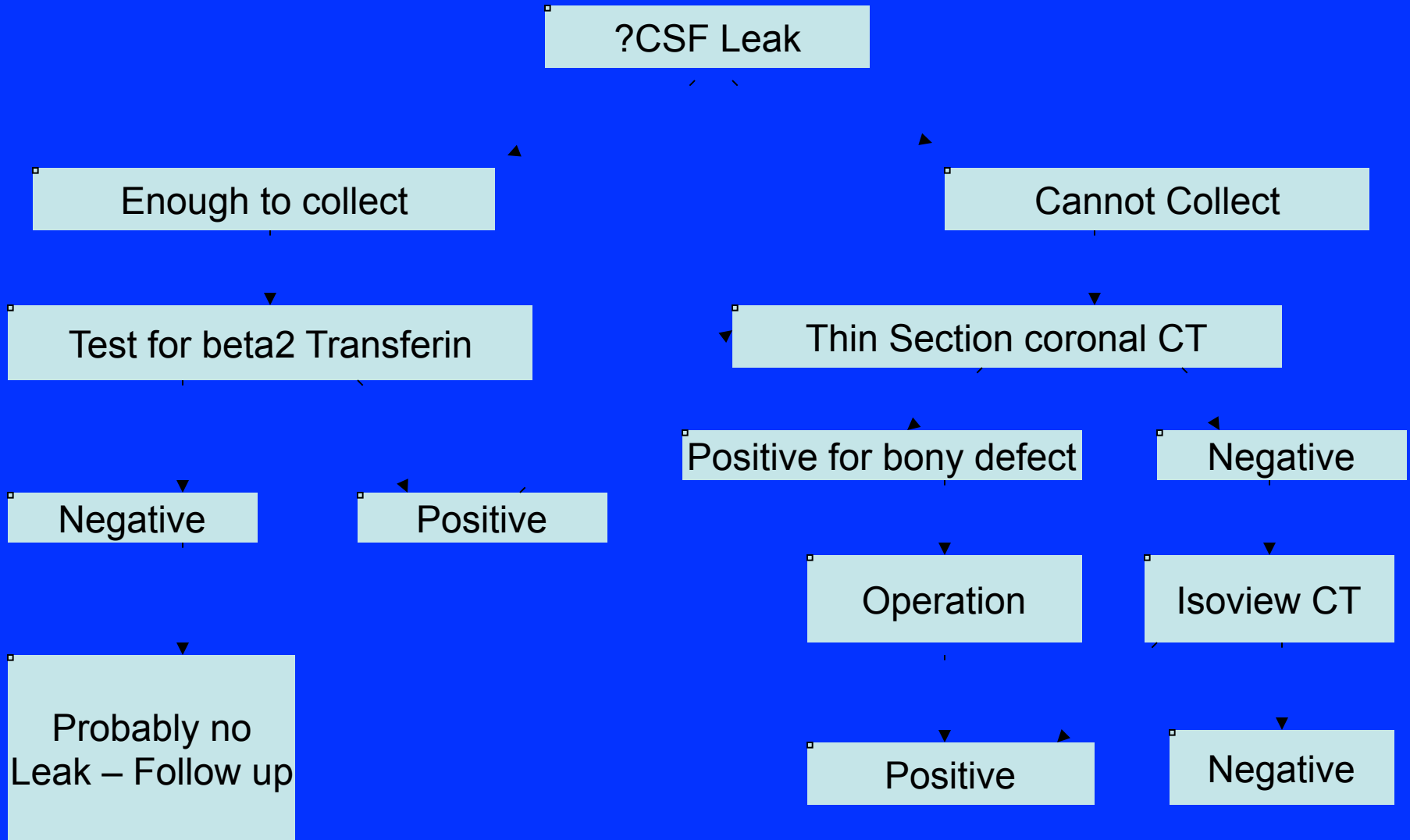
Management

- Depends on
 - cause
 - location
 - severity of the leak
- Traumatic: the interval between trauma and leak is important.
- The natural history of CSF leak depends on the etiology.
- Traumatic leaks often stop spontaneously.
 - 1 week in 70% of patients
 - 3 months in 20-30%
 - 6 months in most patients;
 - leakage rarely recurs.
- Nontraumatic leaks
 - third stop spontaneously, and they tend to persist for several years, with intermittent leakage.

Management

- Indications for Surgical Repair:
 - Large high-volume fistulas
 - Nontraumatic leaks
 - Prolonged leaks regardless of etiology
 - Recurrent leaks
- Indications for immediate surgical exploration and closure of dural fistulas
 - Open wounds that are connected to the dural defect
 - Closed head injuries with intracranial complications
 - Fistulas caused by and detected at operation, either intracranial or nasal
 - CSF leak associated with intracranial complications, such as meningitis.

CSF Leak Algorithm



PRACTICAL Stuff

- Use Beta-2 transferrin
- High-resolution fine cut CT looking for dehiscence of the anterior skull base
- High Resolution T2 weighted MRI if leaking at the time
- Encephaloceles should always have an MRI
- If u cant see a leak on scan, endoscopically look at the cribriform plate (PJ' s rule)
- Intrathecal Fluorescein – more later

Laboratory Ix

- Immunoelectrophoretic identification of CSF-specific marker proteins is the current standard for identifying CSF.
 - **Beta2-transferrin** assay is the most widely used test and is considered the criterion standard for diagnosis of CSF rhinorrhea.
 - Beta2-transferrin is a protein that is highly specific for human CSF
 - serum of newborns
 - vitreous humor
 - patients with liver disease.
 - high sensitivity and specificity
 - Rapid
 - Noninvasive.
 - 0.5 mL of fluid is necessary
 - difficulties in collection in intermittent, low-volume leaks.

Laboratory Ix

- Glucose Oxidase paper:
- Rapid but highly unreliable test
- Reducing substances present in the lacrimal-gland secretions and nasal mucus may cause false-positive results.
- Glucose, at a concentration of 5 mg/dL, can lead to a positive result with this test.
- Active meningitis can lower the glucose level in the CSF and may lead to false-negative readings.
- Glucose testing can be used as a first-line diagnostic test in a patient with a history of craniofacial injury
- All positive results should be confirmed with the beta 2-transferrin test, which is more reliable than the glucose test.

Laboratory Ix

- Total protein analysis, a test of specific gravity, and salt-content tests are no longer performed because other tests provide better results.

Imaging Studies – CT Scanning

- CT scans can reveal:
 - Fracture site that underlies a traumatic leak, reveal an
 - Underlying anatomic or developmental abnormality (Nontraumatic leak)
 - Information on the brain parenchyma in the vicinity of the leak
- Thin-section (1-mm) coronal CT scan
 - small defects of the cranial base that are not visible on standard CT scans or reconstructed images.



Imaging Studies – CT Scanning

- Pneumocephalus on a scan suggests a large dural tear.
- Deviated crista galli is a radiological sign in patients presenting with primary CSF rhinorrhea; this finding supports a congenital bony dehiscence as the etiologic basis for this condition.
- A modified technique known as digital subtraction cisternography may be useful if CT cisternography does not reveal the leakage site.
- An air fluid level in the sphenoid sinus
 - leak is in the sphenoid sinus
 - leak in the area of the posterior cribriform plate or posterior fovea ethmoidalis can drain into the sphenoid sinus by way of its ostium

Imaging Studies CT Scanning

- Omnipaque CT cisternography or metrizamide CT cisternography
- Low morbidity rate
- most useful in the presence of severe leakage
- High contrast with the air in the paranasal sinus.



Imaging Studies CT Scanning

- Intrathecal Agents:
 - Metrizamide, a water-soluble nonionic tri-iodinated contrast agent
 - Iohexol (Omnipaque)
 - Depicts the precise location of CSF leakage in most patients with active leaks
 - Intermittent or slow leaks may not be identifiable.

Imaging Studies CT Scanning

- Adverse:
 - nausea, headaches, and acute organic psychosyndromes
 - Severe toxicities have been reported in patients undergoing myelography, which requires a dose of contrast agent higher than that used for routine MCTC.

Imaging Studies CT Scanning

- Some controversy exists regarding the usefulness of MCTC in accurately delineating the site of leak. One study demonstrated positive localization by means of MCTC in 76% of cases examined; 87% of active fistulas were identified. However, none of the inactive fistulas were identified.
- In patients with intermittent or slow leaks who are poor candidates for MCTC, a repeat study can be performed after the intracranial pressure (ICP) is increased by intrathecally infusing isotonic sodium chloride solution just before imaging to activate the leak. Some have reported a good result by using the Valsalva maneuver or coughing to increase ICP.

Imaging: Nuclear Med

- Generally regarded as safe to utilise
- Agents utilised:
 - radioactive iodine-131
 - radioactive iodinated serum albumin (RISA)
 - ytterbium-169
 - diethylenetriamine pentaacetic acid (DTPA)
 - indium-111 DTPA
 - technetium-99m human serum albumin
 - 99mTc pertechnetate

Imaging: Nuclear Med

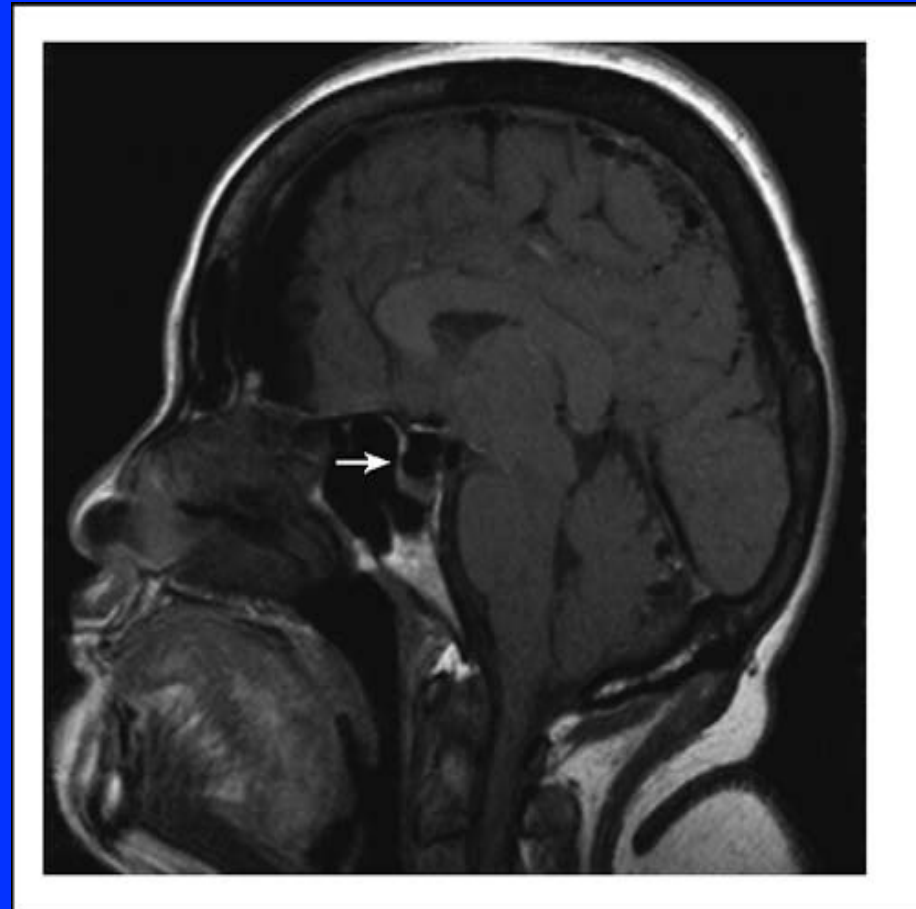
- Introduced either:
 - lumbar or suboccipital puncture then using serial scanning or scintiphotography
 - nasal pledgets in various high-risk areas then analyzed for the presence of the tracer.

Imaging: Nuclear Med

- Limitations:
 - They do not help in precisely identifying the location of the leak
 - The isotope is absorbed into the circulatory system and can contaminate extracranial tissue
 - Patient positioning can cause distal pledgets to incorrectly take up the isotope
 - Readings of radioactivity should be high to determine a true leak; borderline readings are not reliable. False-positive results are present in as many as 33% patients.

Imaging - MRI

- MRI typically is not recommended in the evaluation of a CSF leak because it does not demonstrate bony defects very well.
- Heavy T2-weighted image can reveal a brisk CSF leak.
- Encephalocele: Essential - Delineates extent
- Useful in Spontaneous idiopathic CSF Leaks as they have the highest rate of meningoencephaloceles
- Empty Sella Syndrome



Imaging: Overpressure radionuclide cisternography

- An undetected fistula can be elicited by increasing ICP.
- One group compared ORNC with MCTC. ORNC was more sensitive than MCTC at confirming the diagnosis of a leak, but MCTC was more helpful for localizing the site of the leak.

Imaging - X-ray

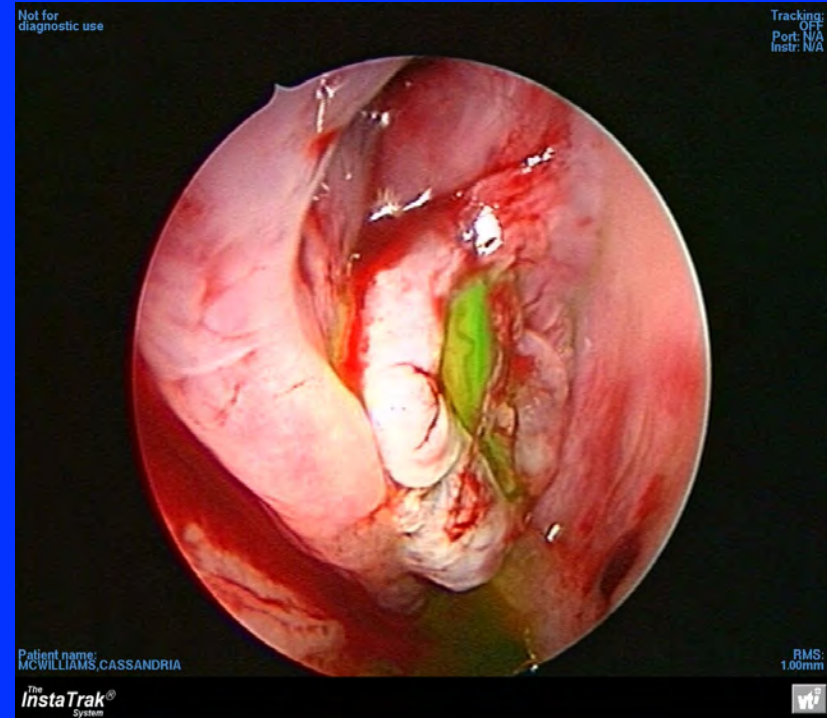
- In the absence of a history of trauma, plain radiographs are of little value
 - Fracture of the skull
 - Air-fluid level in the sinus
 - Aerocele in the cranial vault
- Pathognomonic sign of dural injury = Air in the subarachnoid space on a plain radiograph (large dural envelope tear)

Imaging: Pneumoencephalography

- This is an obsolete procedure that is also uncomfortable because patients are required to be in a head-hanging position.
- This study provides poor contrast with the paranasal sinuses and nasal airway.

Diagnostic Procedures

- Intrathecal Fluorescein (0.1-0.2 mL of 5% solution mixed with 10 mL of CSF)
 - Most accurate method of localizing the site of the leak
 - Preoperatively and intraoperatively to localize and repair the causative defect
 - Transient neurologic complications have been reported with this technique. US Food and Drug Administration (FDA) has not approved the intrathecal administration of fluorescein.
- Optimal Nasal Endoscopy performed 20-30 minutes after an intrathecal injection of fluorescein



Diagnostic Procedures

- Look for fluorescent CSF that is visible with the light of the endoscope
- Blue-light filter on the endoscope can enhance visualization
- Trendelenberg, valsalva and patience intraoperatively
- Routine otologic examination can reveal fluorescence medial to the tympanic membrane, as can examination of the oral cavity and posterior pharyngeal wall.
- Pledgets can be placed in the nasal cavity and look for absorption of the contrast agent. Precise location of the stained pledget can point to the leak. This technique is typically useful in situations in which MCTC is not helpful and surgery is being considered.

Diagnostic Procedures

- The intrathecal injection of nonradiographic dyes and their recovery on cottonoids inserted inside the nasal cavity are of historic interest.
- Indigo carmine, methylene blue, and Evans blue dye has been abandoned because of neurologic toxicity (Paraplegia has been reported.) and poor localization of the leak.

Medical Management

- 1- to 2-week trial of bed rest with the patient in a head-up position.
- Coughing, sneezing, nose blowing, and heavy lifting – avoided
- Stool softeners or laxatives
- Lumbar drain or repeated lumbar puncture can be used to decrease ICP (removal of 150 mL of CSF/day).
 - Theoretical possibility of increased risk of meningitis
 - Pathology: decreased outflow of CSF may allow bacteria to enter the basal cisterns
 - Lumbar puncture should not be performed in patients with cerebral edema or increased ICP.

Medical Management

- Acetazolamide:
 - Increased ICP is problematic and contributes to the CSF leak
 - Diuretic
 - Decreases CSF production by as much as 48%.

Medical Management

- Prophylactic antibiotics
 - Controversial
- No prospective controlled study has been conducted to resolve this question
- Impression:
 - routine use of prophylactic antibiotics is not indicated and that this practice may select out resistant organisms
- In some high-risk groups, antibiotic therapy may be justifiable
 - Previous sinus operations or preexisting sinusitis
- Use of steroids and diuretics can be considered
- Disadvantage with the conservative approach is the risk of meningitis and brain abscess.

Operative Repair

- Defects generally involve the anterior skull base
- Intracranial
- Extracranial

Intracranial Repair

- Intracranial repair
 - Old standard method of repair
 - Leaks arising from an anterior defect can be approached from a frontal anterior fossa craniotomy.
 - middle fossa craniotomy or posterior fossa craniotomy (Rare) is required for leaks arising in those areas.



Intracranial Repair

- Different repair techniques include the use of:
 - free or pedicled periosteal or dural flaps,
 - muscle plugs
 - mobilized portions of the falx cerebri
 - fascia grafts
 - Flaps
 - Leaks arising from the sphenoid sinus are difficult to reach by means of an intracranial approach.

Intracranial Repair

- Advantages

- Ability to inspect the adjacent cerebral cortex
- Direct visualization of the dural defect
- Better ability to seal a leak in the presence of increased ICP
- When preoperative localization attempts fail to reveal the site of a leak, intracranial approach with blind repair has been successful. Cribriform and the sphenoid area, if necessary, are covered with the repair material.

- Disadvantages:

- Increased morbidity
- Increased risk of permanent anosmia
- Trauma related to brain retraction (hematoma, cognitive dysfunction, seizures, edema, hemorrhage)
- Prolonged hospital stays. Failure rates for this approach are 40% for the first attempt and 10% overall.

External Repair

- External approaches
 - Anterior osteoplastic flap through a bicoronal or eyebrow incision
 - External ethmoidectomy
 - Transethmoidal sphenoidotomy
 - Transseptal sphenoidotomy
 - Transantral approach



External Repair

- External Ethmoidectomy
 - Lynch incision
 - Soft tissues elevated off the lamina papyraccia and lacrimal bone
 - Ethmoid septae are exenterated
 - Single/multilayer closure with or without bone or artificial material
 - De-epithelialise the area around the leak to provide a surface for the graft to adhere to

External Repair

- Graft material include:
 - Fascia lata
 - Temporalis fascia
 - Septal or turbinate mucosa
 - Muscle, fat
 - Septal cartilage
- All of these materials can be placed by using the endoscopic technique as well.
- Alternatives to the less invasive, and probably equally effective, endoscopic techniques.
- For cribriform or fovea ethmoidalis leaks, a transnasal ethmoidectomy is performed
- Sphenoidal leaks, sphenoidotomy is performed as well; graft material is placed over the fistula

External Repair

- Success rates for the extracranial or external approach are 86-100%.
- Disadvantages include
 - Inability to treat concomitant intracranial abnormality
 - difficulty with frontal and sphenoid repair with prominent lateral extensions
 - Relative ineffectiveness of repairing high-pressure leaks from below.
- Despite these disadvantages, primary surgical repair of CSF rhinorrhea is usually extracranial unless exploring intracranially is necessary to repair other damage.

Endoscopic Repair

- **Advantages**
 - Better field of vision
 - Enhanced illumination and magnified-angle visualization
 - Ability to clean the mucosa off the adjacent bone without increasing the size of the defect
 - Accurate positioning of the graft. McMains et al reported a highly successful case series of 92 patients with long-term follow-up, solidifying the role of endoscopic control of CSF rhinorrhea.
 - lowered morbidity
 - no anosmia
- No prospective comparative analysis of extracranial and intracranial techniques has been performed
- An overall success rate of 90-95% has been reported with endoscopic repair of CSF leaks.

Endoscopic Repair

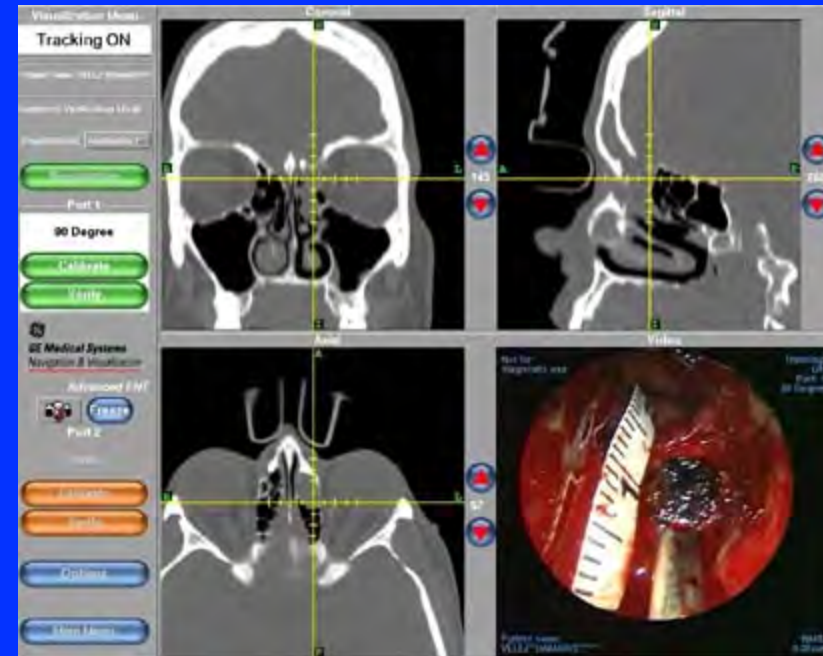
- Anatomic considerations
 - Accidental Trauma
 - Cribriform plate 23.1%
 - Ethmoid skull base 20.5%
 - Multiple sites 35.9%
 - Iatrogenic
 - Ethmoid skull base 35.1%
 - Cribriform 27%
 - Sphenoid 18.9%
 - Spontaneous
 - Cribriform and sphenoid



Figure 1. Fistula of right sphenoid sinus, drainage of CSF with fluorescein marker (green).

Endoscopic Repair

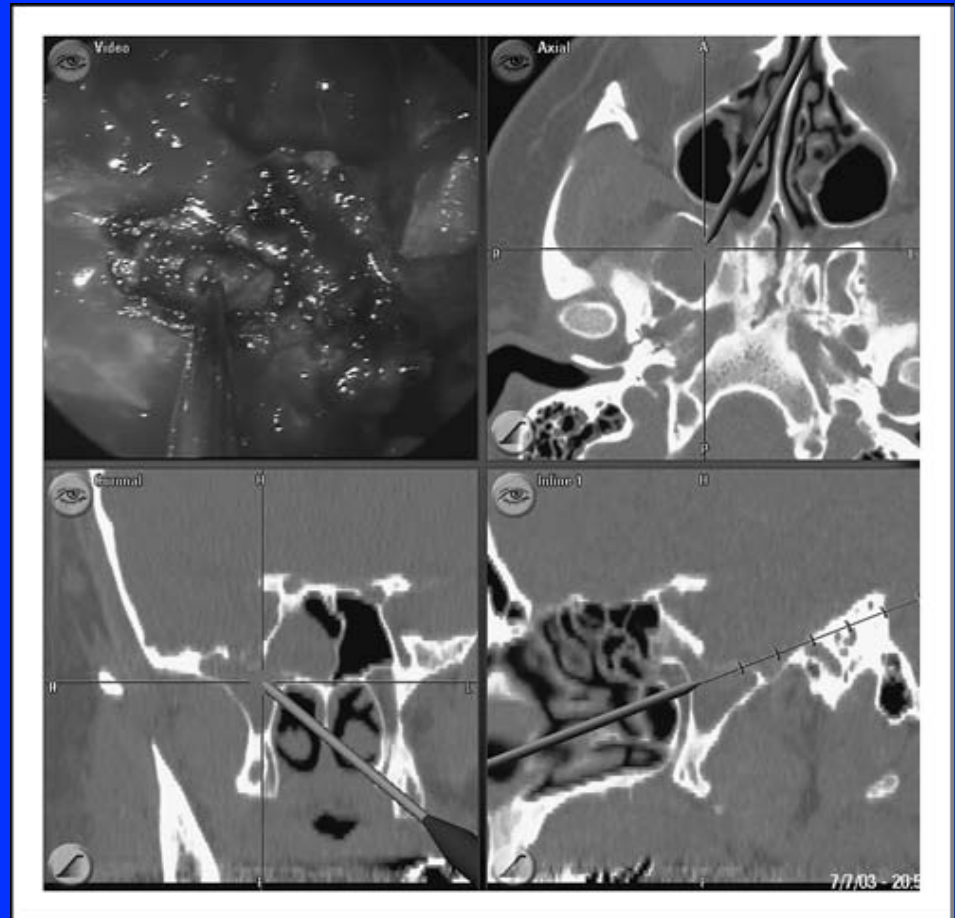
- Nose prepared with adequate vasoconstriction
- Ethmoidectomy +/- harvesting of graft
- Wide exposure over the leak with de-epithelialisation
- Mucosal graft with glue/ bath plug fat from the lobule fat of the greater trochanter etc



Must Read PJ's Book. Endoscopic sinus surgery.

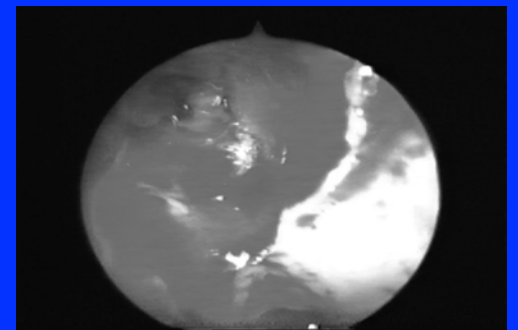
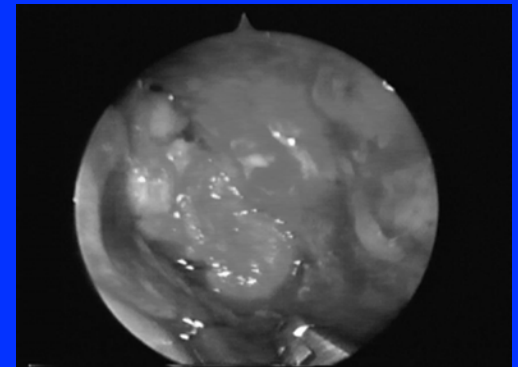
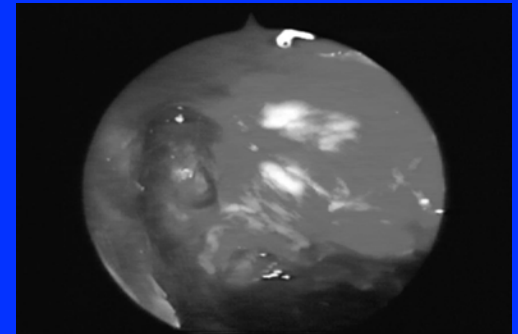
Endoscopic Repair

- Intra-op image guidance can provide tri-planar reconstructions of bony defects and aid in the localization and surgical repair of defects



Endoscopic Repair

- Intra-thecal fluorescein
 - Seizures, leg weakness, opisthotonus
 - 0.1ml 10% in 10 ml of CSF with slow intrathecal injection over 10 minutes
- Topical placement of fluorescein 5% pledgets with color change from yellow to green fluorescence



Adjunctive Care

- Adjuncts to extracranial endoscopic repair include:
 - Coupling repair with postoperative conservative care can increase success rates for CSF leak repair
 - bed rest
 - lumbar-drain placement
 - acetazolamide therapy
- The intraoperative techniques of free mucosal transfer, either from the middle turbinate or from the anterior sphenoid wall, can additionally support the classically described overlay or underlay repair techniques.
- No randomized control studies have demonstrated that use of any particular tissue adhesive and/or fibrin sealant increases success rates.
- Post repair packing, many packing materials have been used, with no clear benefit from any particular product
- PJ uses gelfoam, fibrin glue, gelfoam in a layered fashion

Operative Repair: Other Complications

- Meningitis is the most frequent and severe complication of a CSF leak
 - *Streptococcus pneumoniae*
 - *Haemophilus influenzae* are the most common pathogens.
 - The risk of meningitis during the first 3 weeks after trauma is estimated to be 10%.
 - The rate is 40% in nontraumatic rhinorrhea.

Operative Repair: Other Complications

- Meningitis caused by a persistent CSF leak is associated with a high mortality rate.
- Because of the relatively low rate of spontaneous closure, a conservative approach for these indications is not recommended.
- Spontaneous closure rates vary with the etiology; the recurrence rate after spontaneous closure was 7% in 1 study.
- The surgical mortality rate is 1-3% for intracranial procedures and is negligible for external procedures.
- The morbidity for intracranial approaches is clinically significant, with anosmia being the most common complication (10-25% of patients).

Conclusions

- CSF rhinorrhea can occur as a result of craniofacial injury, iatrogenic surgical trauma, or spontaneous causes.
- Several diagnostic tools are available to aid in diagnosis.
- In the case of early-onset traumatic fistula, a conservative approach can be used.
- Other causes require a more aggressive intervention.
- Endoscopic and external techniques have replaced invasive craniotomy.