CEREBROSPINAL FLUID LEAKS

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Objective

- CSF Physiology
- History
- Physical
- Diagnostic Tests
- Localization Imaging Modalities
- Classification
- Repair
CSF Production

- Ultrafiltrate of serum
- Produced by choroid plexus at 20 mL/hr
- 140 mL total volume actively circulating
- Turned over 3 times daily
- CSF pressure range 5-15 cm H2O in adults
- Acts as a cushion for the brain as well metabolite transport
CSF Flow
Pathophysiology

- Production vs absorption
- Disruption of arachnoid, dura, bone, and sinus mucosa

- Etiology – sinus or neurosurgery, skull base trauma, infections or tumors eroding skull base, congenital skull base defects
Pathophysiology II

- Elevated ICP

- Persistent pressure exertion on structurally weak areas of the skull base may result in bony erosion and may result in a CSF leak [Benign intracranial hypertension (BIH or IIH)]
Classification of Cerebrospinal Fluid Rhinorrhea

**Traumatic**
- Accidental
- Immediate
- Delayed

**Surgical**
- Complication of neurosurgical procedures:
  - Transsphenoidal hypophysectomy
  - Frontal craniotomy
  - Other skull base procedures
- Complication of rhinologic procedures:
  - Sinus surgery
  - Septoplasty
  - Other combined skull base procedures

Cummings 5th Ed. Ch 54.
## Classification of Cerebrospinal Fluid Rhinorrhea

**Nontraumatic**
- Elevated Intracranial Pressure
- Intracranial neoplasm
- Hydrocephalus:
  - Noncommunicating
  - Obstructive
- Benign intracranial hypertension

**Normal Intracranial Pressure**
- Congenital anomaly
- Skull base neoplasm:
  - Nasopharyngeal carcinoma
  - Sinonasal malignancy
- Skull base erosive process:
  - Sinus mucocele
  - Osteomyelitis
  - Idiopathic

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*Cummings 5th Ed. Ch 54.*
Anatomy Quiz
Anatomy Quiz

Radiopaedia.org – Olfactory Fossa
Anatomy Quiz

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History/Presentation

- Unilateral clear rhinorrhea
  - Positional
  - May be intermittent
  - May be bilateral

- Salty taste (sometimes sweet or metallic)
- Recurrent meningitis
- History of sinus surgery, neurologic surgery.
- History of skull base trauma

American Rhinologic Society - CSF Leaks.
Differential Diagnosis

- Vasomotor Rhinitis
- Allergic Rhinitis
- Retained Sinus Irrigation
- CSF Otorrhea
Physical Examination

- Rhinoscopy – Anterior vs flexible vs rigid
  - Glistening mucosa
  - Active flowing clear fluid
- Lean forward
- Ocular exam - Abducen Palsy
- Fundoscopic Exam – Papilledema
Diagnostic Tests

- Halo Sign
- Glucose
- B₂ Transferrin
- B Trace Protein
Halo Sign

- Based on concept of capillary action
- CSF will travel farther than the blood so a halo results.
- This also happens with tears and mucus so it is not specific
Glucose

- Rhinorrhea is applied to glucose oxidase strips and color change is observed.

- High false positives – tears and mucus can produce change on the glucose oxidase strips as well.
B2 transferrin

- Protein electrophoresis performed on tears, CSF, serum and nasal mucus
- B2 transferrin protein band was identified in CSF only
- Gold standard for diagnosis of extracranial CSF
- Difficult to collect, must be kept cool, need a few ccs
B-trace Protein

- 2\textsuperscript{nd} most common protein in CSF (albumin 1\textsuperscript{st})
- Produced by meninges and choroid plexus
- Present in serum but very low level
- *Renal insufficiency may increase serum level of BTP
Localization

- Nasal Endoscopy with Intrathecal Fluorescein
- Radionuclide Cysternography
- CT Cysternography
- MRI Cysternography
Nasal Endoscopy with Intrathecal Fluorescein

- Introduced 1960
- Most popular
- Requires LP
- Lean forward, examine

Complications of intrathecal fluorescein – reports of grand mal seizures, death
  - Complications are isolated and occur at high dose
  - Keerl et al – low dose (<50mg) is unlikely to cause adverse events
  - Recommended dilution – 0.1 mL of 10% fluorescein (IV preparation) diluted in 10 cc of pts CSF
Intrathecal Fluoresceine

- Complications
  - Reports of grand mal seizures, death
- Complications are isolated and occur at high dose
- Keerl et al – low dose (<50mg) is unlikely to cause adverse events

- Recommended dilution – 0.1 mL of 10% fluorescein (IV preparation) diluted in 10 cc of pts CSF infused over 30 minutes
Radionuclide Cysternography

- Various radiolabeled tracers such as:
  - Radioactive Iodine labeled serum albumin (RISA)
  - Diethylenetriamine/pentaacetic acid (DTPA)

- Requires LP
- Scintillation Camera detects radiolabeled tracer
- Intranasal pledgets placed in areas of concern and are assessed with gamma counter 12-24 hours later
- Elevated pledget: serum count ratio c/w leak
Radionuclide Cysternography

- Drawback
  - poor spatial visualization

Medscape. CSF Leak Imaging.
CT Cysternography

- Intrathecal administration of radiopaque contrast (metrizamide).
- Can detect ~80% of CSF Leaks

Cummings 5th Ed. Ch 54 CSF Rhinorrhea
CT Cysternography

- Advantages –
  - Great bone detail of sinuses
  - Thin cuts

- Drawbacks -
  - Requires active CSF flow
  - Poor soft tissue detail

*Clinical Endocrinology. 2000;52L43-49. Copyright 2000*
MR Cysternography

- Non-invasive
- T2 weighted images with fat suppression and video reversal
- CSF black
  - CSF in Sphenoid

Cummings 5th Ed. Ch 54 CSF Rhinorrhea
MR Cysternography

- **Advantages**
  - Non-invasive
  - Good soft tissue detail
    - Can differentiate inflammatory tissue from meningoencephalocele
  - Sensitivity ~87%

- **Drawbacks**
  - Time of acquisition
  - Thick slices may not show small skull base defects
CT and MRI

- Zapalac et al.
  - Recommend B2 transferrin for diagnosis and imaging with High Resolution CT and MRI for localization

- CT shows great bony detail of sinuses in thin slices to detect defect – no CSF marker

- MRI with excellent soft tissue anatomical detail also may detect meningoencephalocele and intracranial masses

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## Classification

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- Skull base erosive process:
  - Sinus mucocele
  - Osteomyelitis
  - Idiopathic

*Cummings 5th Ed. Ch 54.*
Classification

- Traumatic
  - Non surgical Trauma (Accidental Trauma)
  - Iatrogenic

- Non Traumatic
  - Elevated ICP
    - BIH
    - Intracranial Neoplasm
    - Hydrocephalus
  - Normal ICP
    - Congenital Anomaly
    - Skull base neoplasm
    - Skull base erosive proses
      - Mucocele
      - Osteomyelitis
      - Idiopathic
Traumatic - Accidental

- 80% due to non surgical trauma
  - 2% of all head traumas
  - 21-30% of all basilar skull fractures

- Timing
  - 50% seen within 2 days
  - 70% within first week
  - Almost all within 3 months*
    - Wound contraction, necrosis of bone edges, devascularization of tissues, resolving edema, etc.

- Vast majority resolve with conservative measures
Traumatic - Accidental

- Anterior skull base more likely than middle or posterior

- Most Common Sites
  - Sphenoid – 30%
  - Frontal – 30%
  - Ethmoid/Cribriform 23%
  - Temporal bone fracture – CSF flows into nasopharynx
Surgical Trauma (Iatrogenic)

- 16% of all CSF leaks

- Common Sites
  - Neurosurgery – 67% Sphenoid (Pituitary)
  - Otohns – FESS
    - Ethmoid / Cribiform – 80%
    - Frontal Sinus – 8%
    - Sphenoid Sinus – 4%
FESS traumatic leaks

Fig. 1. Magnified view of the ethmoid bone in the coronal plane with the most common sites of fracture labeled. CN, cranial nerve.

Prosser JD, 2011
Surgical Trauma (Iatrogenic)

- Repair immediately if noticed intraop
- If delayed diagnosis, trial of conservative management, then OR.
Classification

- **Traumatic**
  - Non surgical Trauma (Accidental Trauma)
  - Iatrogenic

- **Non Traumatic (Spontaneous)**
  - Elevated ICP
    - BIH
    - Intracranial Neoplasm
    - Hydrocephalus
  - Normal ICP
    - Congenital Anomaly
    - Skull base neoplasm
    - Skull base erosive process
      - Mucocele
      - Osteomyelitis
      - Idiopathic (Spontaneous)

*The term “spontaneous historically included many of etiologies listed in the “Non-Traumatic” category*
Spontaneous CSF Leaks

- Persistent pulsatile pressure
- Pressure exerted on inherently weak portions of skull base
- Bony erosion develops
Spontaneous CSF Leak

- Patient Characteristics
  - Obesity – 82-92%
  - Middle-aged
  - Female – 70-80%
  - Recent weight gain

- Significant overlap in demographic, clinical, and radiographic characteristics between spontaneous leaks and Idiopathic Intracranial Hypertension (IIH)
Idiopathic Intracranial Hypertension

- Classic Presentation
  - Pressure Headache, pulsatile tinnitus, visual changes
  - CSF opening pressures >25 cm H₂O

- Factors associated with IIH
  - Female, obesity, reproductive age, recent weight gain

- 70% of pts with Spont CSF leak meet criteria for IIH
Spontaneous – Imaging Characteristics

- CT Imaging
  - CT – diffusely thin skull base, arachnoid pits
  - Pneumatization of lateral recess of sphenoid in 91% compared to 23-43% of normal patients. [Shetty PG 2000, Bolger WE 1991]
  - Most common sites – Lateral recess of sphenoid and ethmoid roof or cribriform plate
  - Multiple defects in 31% [Schlosser RJ 2002]
Spontaneous – Imaging Characteristics

- MRI
  - Mengingoencephalocele – in 50-100%
  - Sella visualization
Management

- Conservative Treatment
- Surgical Treatment
  - Transcranial
  - Extracranial
  - Transnasal Endoscopic
Conservative Measures

- Bed rest, elevated HOB 30 degrees
- Avoid
  - Coughing – rx antitussives
  - Straining – rx stool softeners
  - Sneezing
  - Nose blowing
  - Vomiting – rx anti emetics

- Prophylactic Antibiotics - controversial
Conservative Measures

- **Duration**
  - Study of 81 pts with traumatic CSF leaks (Yilmazler S 2006)
    - 3 days duration - 39% resolved
      - Temporal bone - 60%
      - Anterior skull base – 25%
    - 7 days duration – 85% resolved
      - Temporal bone significantly higher resolution rate than anterior skull base
Conservative Measures

- Indications
  - Non-Surgical Trauma
  - Delayed diagnosis of surgical trauma
  - Spontaneous Leaks – unlikely to be successful
Lumbar Drains

- Used if Conservative management fails
- Function to lower ICP and reduce flow through defect.
- Set drain at 10 mL per hour, monitor for headache

- Complications
  - Meningitis
  - Headache
  - Cellulitis
Surgical Repair

- Transcranial – via craniotomy, recurrence 27%
- Extracranial
- Transnasal
- Endoscopic Endonasal
Extracranial

- First described by Dohlman in 1948
  - Naso-orbital incision, dissection into sinus cavity.
  - Success rates 86-97%
  - Improved success rates, decreased morbidity
    - Avoids anosmia and brain retraction
  - Drawbacks – scar, numbness, orbital injury

- *Lateral aspects of frontal and sphenoid not reachable*
Transnasal

- Hirsch 1952 – closed two sphenoid sinus leaks
- Later, microscopes used
- Abandoned with advent of endoscopes
Endoscopic Endonasal Approach

- Wigand 1981 – initial description
- Now standard of care with 95% success rates

Basics
- Standard endoscopic techniques to approach leak
- Mucosa surrounding bone defect must be removed 0.5 cm on all sides
- Choose graft
Graft types

- Temporaliis fascia, Muscle plug
- Mucosal plug
- Fat
- Free cartilage (septal, conchal)
- Free bone (septum, calvarium, iliac crest)
- Dural substitute (Duragen)
- *20% shrinkage
- *Graft material does not affect outcome (Hegazy 2000)
Basics

- **Small defects**
  - Free mucosal or free fascial grafts in overlay technique

- **Large defects**
  - Free bone or cartilage graft in underlay with free mucosal overlay
    - Vs
    - Pedicled mucosal flap
Ethmoid Roof Repair

Sphenoid Repair

Figure 54-10. Layered reconstruction also provides an optimal barrier for sphenoid cerebrospinal fluid leaks, as shown in this illustration of such a repair of a midline sphenoid defect (after transsphenoidal hypophysectomy). Layers are as indicated in Figure 54-9. (Redrawn with permission from Lorenz RR, Dean Rt, Hurley DB, Chuang J, Citardi MJ. Endoscopic reconstruction of anterior and middle cranial fossa defects using acellular dermal allograft. Laryngoscope. 2003;113:496-501.)
After overlay placement...
- Apply fibrin glue
- Then absorbable nasal packing (GelFoam)
- Followed by non-absorbable nasal packing x5-7d

Postop care
- ICU x 24 hours neurochecks (hematoma, edema)
- Ceftriaxone
- Conservative measures, frequent debridements
- No strenuous activity x 6 weeks
- 90% successful primary repair, 96% in second attempts
Vascularized Flaps

- Pedicled mucosal flaps
  - Posteriorly based pedicled nasal septal flap is workhorse of extensively skull base recon
  - Based on posterior septal artery
Pedicled Nasoseptal Flap

Fig. 5. Axial view of the nasal septum demonstrating the posterior septal branch of the sphenopalatine artery.
Fig. 4. Endoscopic view of the posterior nasal cavity (A) with an artist’s representation of the same view (B). Dashed lines mark the location of cuts for the posteriorly based nasal septal flap. IT, inferior turbinate; SO, sphenoid os; SPF, sphenopalatine fossa; ST, superior turbinate.
Technique—Kassam et al 2008. All subsequent images depict reported technique.

**FIGURE 1.** The nasoseptal flap incisions at the anterior nasal cavity. Two parallel incisions are joined by a vertical incision anterior to the inferior turbinate.
FIGURE 2. The nasoseptal flap incisions at the right posterior nasal cavity. Two parallel incisions (dashed lines), one following the maxillary crest and the other 1 to 2 cm inferior to the olfactory cleft, are extended to reach the lateral nasal wall. The inferior incision follows the free edge of the posterior septum and then crosses the posterior choana. The inferior incision may be designed to include the mucoperiosteum of the floor of the nose. The superior incision extends laterally to cross the rostrum of the sphenoid sinus at the level of its natural ostium. A large middle antrostomy and exposure of the terminal internal maxillary artery are illustrated. This is only necessary if the flap will be stored in the maxillary sinus so as to approach the clival and paraclival areas.

FIGURE 3. Elevation of the nasoseptal flap following a subperichondrial and subperiosteal plane.
FIGURE 4. A, the nasoseptal flap is mobilized posteriorly after a posterior septectomy. B, the nasoseptal flap is “stored” at the nasopharynx. C, the nasoseptal flap is “stored” inside the maxillary sinus.
FIGURE 5. The nasoseptal flap is shown covering a defect at the planum sphenoidale.
55 Studies, 1778 CSF Leaks

- **Success**
  - Primary Repair (n=1326) – 90% success rate
  - Second attempts 96% successful

- **Sites of failure**
  - Sphenoid 48%  Ethmoid 40%  Frontal 15%

- **Site of Leak**
  - Ethmoid/crib plate – 53%  Sphenoid 30%
Patient I - DL

- Pt DL
- CC: runny nose
- HPI: 3 weeks right-sided rhinorrhea, positional. Salty taste with drip. Headaches, intermittent. CTH in ED showed possible sphenoid defect.

- ROS – as in HPI
- PMH – rheumatoid arthritis, obesity
- PSH- no neurologic or sinus surgery
Patient DL

- Physical examination
  - Slow steady drip of clear rhinorrhea from right nare when leaning forward
  - Neuro- alert and oriented

- Flexible Endoscopy
  - No visible flow of rhinorrhea when pt leaning forward
Patient DL

- Labs – Rhinorrhea sent for B2 Tranferrin +
- Imaging – CT Sinus and MRI Brain ordered

Impression: Possible spontaneous CSF Leak

Plan:
- Bedrest and elevate HOB
- No straining, coughing, emesis, or nose blowing.
- Pt advised of signs of meningitis for early detection.
Patient DL
Patient DL
Operation - DL

1. Right middle turbinectomy
2. Fluorescein at posterior cribiform plate near the sphenoid rostrum
3. Mucosa stripped around leak – bony defect identified
4. Dura pushed intracranially and Duragen underlay graft inserted
5. Septal cartilage inserted intracranially
6. Local mucosa flap to overlay cartilage with near-complete closure
7. Tisseal applied
8. Gelfoam

*Second leak identified along septum anteroinferior to this and repaired.
Patient AS

- 42 yo f with history of CRS underwent FESS in 2010 with subsequent CSF repair on Right. Presented for eval of poss CSF leak on left.

- Rhinorrhea from left – B2 transferrin inconclusive but suggestive
Pt AS
Patient AS
Pt NY

- Complicated history of CSF leak diagnosed after recurrent meningitis every 10 yrs. Multiple attempts at CSF leak repairs in past.
- Now with clear right sided rhinorrhea
References

- Images as cited within presentation via hyperlink.