Hydrocephalus II

Shunt Dysfunction and Neuroendoscopy

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Shunt Dysfunction and Infection

Shunt Dysfunction and Infection

Infection

Skin breakdown over hardware

Mechanical failure

- Undershunting
 - Separation of shunt components, fractures, migration of hardware
- Overshunting
 - Subdural hematoma

These account for majority of shunt problems

Shunt Dysfunction and Infection Epidemiology

25~60% of patients

• 17% in 1st yr after insertion in peds

• Higher risk:

- Preemies
- Children age <6 mos or weight <3 kg at time of shunt insertion

Undershunting

Undershunting Etiology

Blockage within system

- Choroid plexus
- Glial adhesions
- Build-up of proteinaceous accretions, blood, cells (inflammatory or tumor)
- Ventricular end most common site
- Disconnection, kinking, or breakage of system
 - With age, silicone elastomers calcify, break down, & become more rigid & fragile which may promote subcutaneous attachments
 - Barium impregnation may accelerate process
 - Tube fracture often occurs near clavicle, likely due to \uparrow motion there

Undershunting Evaluation

- History
 - Symptoms of active hydrocephalus
 - Reason for initial insertion of shunt
 - Date & reason of last revision
 - Type of hardware
- Physical
 - Signs of active hydrocephalus
 - For children, plot head circumference on graph of normal curves
 - Before sutures close, head circumferences crossing growth curves
 - Swelling along shunt tubing from CSF dissecting along shunt tract
 - Ability of shunt reservoir to pump & refill
 - May exacerbate obstruction, esp if shunt is occluded by ependyma due to overshunting initially
- In children presenting only w/ N/V, esp those w/ cerebral palsy & feeding G-tubes, R/O GE reflux

Undershunting Evaluation

o Imaging

- Shunt series plain X-rays
 - For VP shunt: AP & lateral skull & "low" CXR and/or AXR)
 - R/O disconnection, break, or migration of tip
 - Disconnected shunt may continue to function by CSF flow thru a fibrous tract
 - Various hardware may be radioluscent & can mimic disconnection
- U/S
 - Maybe useful in neonates w/ open fontanelles
- CT head
- MRI
- Radionuclide shunt-o-gram
 - Assess shunt function using radionuclide, iodinated contrast
- o Shunt tap
 - If infection suspected
- Surgical exploration of shunt
 - May be the only means to definitively prove / disprove functioning of various shunt components
 - Even when infection not suspected, CSF & removed hardware should be cultured



Shunt revision

Pumping the Shunt Reservoir (PS Medical Valve)

- To assess flow thru proximal catheter, distal port is depressed first (to occlude run-off into distal catheter), then dome is depressed
 - Reservoir refills promptly = shunt patent proximally
 - N refill time ~15-30 secs
- To assess flow thru distal catheter, proximal port is depressed first, then dome is depressed
 - Reservoir depresses w/ little resistance = shunt patent distally
- Sensitivity 19%, specificity 81% of identifying shunt malfunction

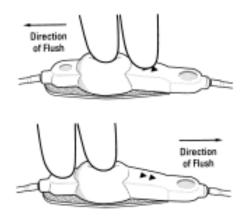


Figure 2. Digital compression of a delta valve (PS Medical Corporation, Goleta, California) requires occluding flow in one direction before compressing the central chamber. (Reprinted from the Johnson & Johnson Catalog from Johnson & Johnson Professional, Inc. with permission.)

From: Naradzay JFX et al. J Emerg Med. 1999; 17(2):311-322.

Shunt Tap Indications

- To obtain CSF specimen
 - To evaluate for shunt infection
 - To obtain cells for cytology e.g. in PNET for malignant cells
 - To remove blood e.g. in intraventricular hemorrhage
- To evaluate shunt function
 - Measure pressures
 - Contrast studies: proximal injection of contrast (iodinated or radio-labelled). Distal injection of contrast
- As temporizing measure to allow function of distally occluded shunt
- To inject medication
 - Antibiotics for shunt infection or ventriculitis
 - Chemotherapy agents
- For catheters placed within tumor cyst (not a true shunt)
 - Periodic withdrawal of accumulated fluid
 - For injection of radioactive liquid (usually phosphorous) for ablation

Shunt Tap Technique

Step	Information Provided
Shave area Prep w/ providone iodine solution Use 25 gauge butterfly needle (ideally a non-coring needle) - needle should only be introduced into shunt components specifically designed to be tapped	
Insert needle into reservoir & look for spontaneous flow into butterfly tubing; measure pressure in manometer	Spontaneous flow = prox end not completely occluded CSF pressure = pressure of ventricular system (should be <15 cm in recumbent position)
Measure pressure w/ distal occluder pressed if present	↑ in pressure = some function of valve & distal shunt
If no spontaneous flow, try to aspirate CSF w/ syringe	If CSF easily aspirated, pressure seen by ventricular system may be near 0 If no CSF obtained or if difficult to aspirate, prox occlusion
Send CSF for C&S, gram stain, protein, glucose, cell count	Check for infection
Fill manometer w/ sterile saline, & occlude proximal (inlet) port	Measure forward transmission pressure (thru valve & peritoneal catheter in presence of shunt w/ proximal occluder) - should be < ventricular pressure
Repeat measurement after injecting 3-5 mL of saline	If peritoneal catheter is in loculated compartment, pressure will be ++higher after injection

Radionuclide Shunt-o-gram

- Position patient, shave hair over reservoir & prep
- Tap shunt insert 25 gauge butterfly needle into reservoir
 - Measure pressure & drain 2-3 mL of CSF (send 1 mL for C&S)
 - Inject radio-isotope (for VP shunt in adult, use 1 mCi of ^{99m}Tc pertechnetate in 1 mL of fluid) while occluding distal flow (by compressing valve or occluding ports)
 - Flush in isotope w/ remaining CSF
 - Patients w/ multiple ventricular catheters need to have each injected to verify patency of that limb
- o Imaging
 - Immediately image abdo w/ gamma camera to R/O direct injection into distal tubing
 - Image cranium to verify flow into ventricles (proximal patency)
 - If spontaneous flow into abdo not seen after 10 min, patient sat up & rescanned
 - If flow not seen after 10 min, then shunt pumped
 - Look for diffusion of isotope within abdo to R/O pseudocyst formation around catheter

Overshunting



 Rapid drainage of CSF
 CSF drainage that occurs when intraventricular pressure < ventricular valve pressure

• Comprises:

- True intracranial hypotension
- Slit ventricles & slit ventricle syndrome

Overshunting Epidemiology

Incidence 5~55%

 10~12% of long-term shunt patients, within ~6 yrs of initial shunting

 Commonly occurs in infants w/ initial shunt insertion at age <6 mos **Overshunting** Complications

Subdural hematomas
 Craniosynostosis & microcephaly

 Controversial

 Stenosis or occlusion of sylvian aqueduct

Overshunting

Intracranial Hypotension

- AKA low ICP syndrome
- Very rare
- Presentation
 - H/A's postural in nature worse when upright, relieved w/ recumbency
 - Not usually assoc w/, but may occur w/ lethargy, N/V, neuro findings (e.g. diplopia, upgaze palsy)
 - May sometimes resemble those of high ICP
- Etiology: siphoning effect, "true" overshunting
- CT head: ventricles may be slit-like or N in appearance
- \circ Sometimes necessary to document drop in ICP (supine \rightarrow upright) for diagnosis
- Patients may also dev shunt occlusion may be difficult to distinguish from slit ventricle syndrome

Overshunting Slit Ventricles

- Totally collapsed lateral ventricles
- May be seen on CT head in 3~80% of patients after shunting
- Most asymptomatic
- Patients may occasionally present w/ symptoms unrelated to shunt (e.g. migraine)



Fig. 1. Radiological slit ventricles.

From: Olson S. Pediatr Neurosurg. 2004; 40:264-269.

Overshunting Slit Ventricle Syndrome

- AKA non-compliant ventricle syndrome
- o <12% of shunted patients</p>
- 6~22% of children w/ radiological slit ventricles & H/A's
- o Triad:
 - Intermittent clinical features of shunt obstruction w/ distinct asymptomatic intervals
 - Slit-like appearance of ventricles on CT scan
 - Slow refill of shunt reservoir

Overshunting Slit Ventricle Syndrome

- Pathophysiology: small ventricles predispose to catheter obstruction, pressure then rises & only when ventricles marginally dilate does catheter begin to function again
 - Theories likely multifactorial
 - Ventricular pressure intimately related to ICP & when CSF pressure drops uncoupling occurs $\rightarrow \uparrow$ venous congestion & \uparrow brain elastance
 - \uparrow pressure w/ subependymal flow can cause subependymal & periventricular gliosis w/ \uparrow ventricular wall stiffness
 - Intraventricular pressure would need to be higher than usual to obtain ventricular dilatation
 - (Law of Laplace P=2T/R: pressure required to expand a large container < pressure required to expand small container)
 - Matsumoto et al. (1986) animal models
 - Low pressure valves in neonates lead to overshunting w/ radiological slit ventricles, development of microcephaly & craniosynostosis
 - Predisposes to ventricular catheter obstruction & prevents ventricles from expanding in response

Overshunting Evaluation

• Clinical exam

- Deep sunken fontanelle
- Overriding parietal bones
- Rapid decline in head circumference to microcephalic range
- Valve slow to refill after compression
- Monitoring CSF pressure
 - Lumbar drain
 - Shunt tap measure pressures w/ postural changes
 - -ve pressure when upright
 - Pressure spikes during sleep
- CT head
 - Slit like ventricles
 - May show evidence of transpendymal CSF flow
 - Possible SDH / ICH
- o Shunt-o-gram

Overshunting

Treatment of Slit Ventricle Syndrome

• Try to categorize patient

- If possible, then implement specific tx
- Otherwise: tx empirically as intracranial hypotension, then move onto other methods for tx failure
- Problems related to overshunting may be reduced by utilizing LP shunts for communicating hydrocephalus & reserving ventricular shunts for obstructive HCP
- VP shunts may also be more likely to overdrain than VA shunts b/c longer tubing resulting in greater siphoning effect

Overshunting

Treatment of Slit Ventricle Syndrome

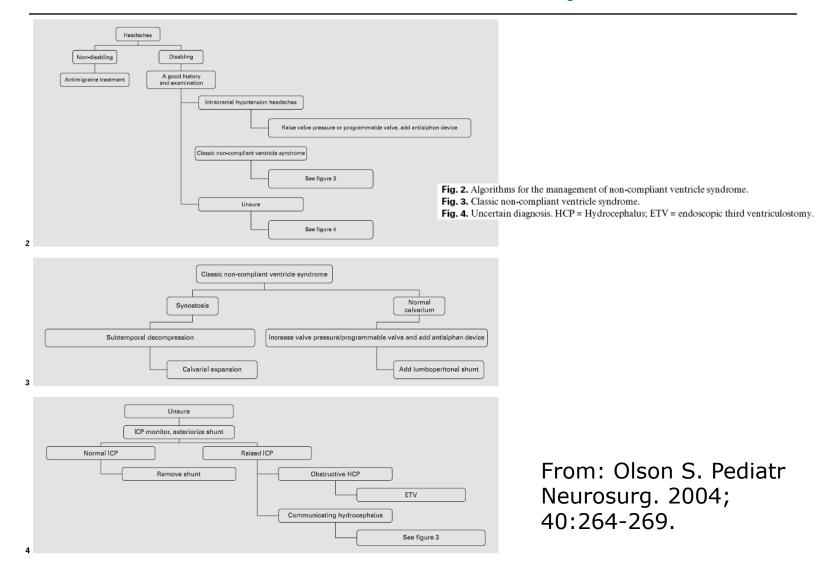
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- Intracranial hypotension
 - Postural H/A usually self-limited
 - Symptoms persistent after >3 days bed-rest & analgesics
 - Trial w/ tight abdo binder
 - Valve should be checked for proper closing pressure: if low, replace w/ higher pressure valve; if not low, antisiphon device +/- high pressure valve
 - Patients w/ long-standing overshunting may not tolerate efforts to return intraventricular pressure to N levels
- Asymptomatic slit ventricles
 - Prophylactic upgrading to higher pressure valve or insertion of antisiphon device now largely abandoned
 - May be appropriate at time of shunt revision when done for other reasons

Slit ventricle syndrome

- Patients actually suffering from intermittent high pressure
- Total shunt malfunction: revise shunt
- Intermittent occlusion:
 - If symptoms occur early after shunt insertion / revision, initial expectant management may be indicated since symptoms will spontaneously resolve in many
 - Revision of proximal shunt (may be difficult due to small size of ventricles): follow existing tract & insert longer / shorter length of tubing based on pre-op imaging studies vs. insertion of 2nd ventricular catheter (leave 1st one in place) / LP shunt
 - Upgrade valve / programmable valve
 - o Antisiphon device
 - Subtemporal decompression sometimes w/ dural incision → dilatation of temporal horns (evidence for \uparrow ICP) in most, but not all cases
 - Calvarial expansion
 - o 3rd ventriculostomy

Overshunting Treatment of Slit Ventricle Syndrome



Infection



Infection Rate

- Majority of data from retrospective reviews using data collected before 1990
 - 18~22% of shunted patients
 - 5~6% per procedure
 - 6.2% in 1st post-op month, 7.4% overall
 - 9% by 6 months, 19% by 10 yrs
- Education Committee of International Society of Pediatric Neurosurgeons sponsored cooperative study (1994) – 38 centers, 773 patients, >1 yr follow-up
 - 6.5% of patients after 1 yr



- Majority soon after placement of shunt
 - Casey et al. (1997)
 - 92% occur within 3 months of shunt placement

Infection Risk Factors

- Age neonates & very young children
 - Casey et al. (1997)
 - ↑ infection rate in age <6 months vs. older (19% vs. 7%)
 - Age-related changes in density & identity of bacteria populations on skin, ↑ susceptibility to pathogens due to relative immune deficiency in neonates (less IgG levels in age <6 months)
- Duration of shunt surgery
- Previous shunt failure

- Possible assoc risk factors:
 - Reason for shunt placement
 - Type of shunt
 - Educational level of surgeon
 - Presence of spinal dysraphism – few studies w/ enough statistical power
 - Ammirati & Raimondi et al. (1987): subgroup analyses
 - ↑ infection rate in myelomeningocele patients shunted in 1st wk of life vs. age >2 wks (50% vs. 24%)
 - Clinically stable children might benefit from delay in shunt placement

Infection Presentation

- Varies w/ age
- Infants: ↑ irritability, apnea
 & bradycardia in severe cares
- General:
 - H/A
 - Lethargy
 - N/V
 - Fever
 - Meningismus
 - Photopobia
 - Gait disturbances
 - Seizures
 - Visual disturbances (upward gaze palsy & papilledema)
 - Abdo pain, abdo fluid collection / pseudocyst
 - Erythema or edema along shunt tubing

• Varies w/ organism

- Gram –ve bacilli E. coli: acute presentation w/ severe abdo pain, septicemia
- S. epidermidis: more indolent
- S. aureus: usually assoc w/ erythema along shunt tract

- Ventricular-vascular shunts:
 - Subacute bacterial endocarditis
 - Shunt nephritis (immune complex deposition in renal glomeruli) – hematuria + proteinuria

Infection Evaluation

- History & physical
- o Imaging
 - X-ray shunt series R/O disconnection in shunt tubing, movement of distal catheter out of peritoneal space w/ growth of child
 - CT ependymal enhancement characteristic of ventriculitis
 - Difficult in children w/ slit ventricles or unusual baseline ventricular anatomy
 - U/S for neonates
 - Abdo U/S R/O fluid collection
- o Shunt tap
 - Opening pressure, function
 - CSF for glc, protein, cell count, gram stain, C&S
 - \downarrow glc, \uparrow protein, \uparrow cell count suggest bacterial infection
 - Generally, C&S +ve in <50% tested

Infection Prevention

- Langley et al. (1993) meta-analysis, 12 studies
 - Peri-op abx use reduced infection rate by ~50% (CI 95%)
- Haines & Walters (1994) meta-analysis, 8 studies
 - Same result
- Studies included in meta-analyses differed wrt specific type, duration, dosage of abx used, as well as timing of 1st dose
- Little evidence to suggest that abx prophylaxis before dental procedures & use of abximpregnated silastic shunts reduce infection rates

Infection Organisms

- Infection occurs via:
 - Bloodstream
 - Along shunt tubing from abdo source (generally assoc w/ bowel perforation)
 - Contamination of shunt material w/ skin flora @ time of surgery
- Most common: typical skin flora
- In most series: S. epidermidis > S. aureus (2:1)
 - Livni et al. (2004)
 - S. epidermidis secretes mucoid slime that enhances its ability to adhere to foreign bodies
 - Lower adherence rate for silicone vs. teflon
- Gram –ve bacteria: E. coli, Proteus, Klebsiella
 - From intestinal perforation
- Anaerobic diphtheroids, e.g. propionibacterium, can cause delayed infections
 - Difficult to assess & tx as C&S may remain -ve for >1 wk
- Fungal infections rare



- Infection = +ve C&S from CSF or from shunt hardware
- In most instances, only shunt hardware is C&S +ve while CSF remains -ve
 - Vanaclocha et al. (1996)
 - C&S positivity hardware vs. CSF (59% vs. 9%)
 - Suggests bacteria & other microorganisms favor adhesion to foreign materials over CSF

Infection Treatment

- Surgical removal of infected shunt or shunt externalization
- New shunt placed then or delayed until after course of abx
- May need EVD, lumbar drain, or intermittent LP's or ventricular taps as temporizing measure
- Abx course until CSF C&S –ve for minimum 72 hrs
- Drainage of abdo pseudocyst / abscess / wound may also be necessary
- IV vancomycin often used initially until sensitivities available

Infection Treatment

- Abx alone less effective than abx + surgery
 - Walters et al. (1984) retrospective review
 0 14% vs. 60%
 - Frame & McLaurin (1984) RCT
 - Removal of shunt + abx + interim EVD or ventricular taps vs. immediate surgical replacement of shunt + abx vs. abx only (IV & intraventricular abx given to all)
 - Higher cure rates @ 48 hrs, 1 & 4 months for surgery patients: 100% vs. 90% vs. 30%
 - Longer hospital stays for abx-only patients: 47
 vs. 33 vs. 25 days

Infection

Treatment: Antibiotics

Empiric abx

- IV vancomycin initially (CSF penetration ~18% conc of serum)
- Consider adding PO rifampin for increased coverage (10 mg/kg/day PO q12h)
- May change to IV nafcillin (unless penicillin allergic or MRSA +ve)
- Intraventricular injection of preservative-free abx

Abx for specific organisms

- S. aureus & S. epidermidis
 - If sensitive (MIC <1.0 µg/mL): IT gent + (IV nafcillin / cefazolin / cephalothin / cephapirin)
 - If resistant to nafcillin (i.e. MRSA) / cephalothin / cephapirin: PO rifampin + PO trimethoprim + IV & IT vancomycin
- Enterococcus: IV/IT ampicillin + IT gent (if intravascular shunt: add IV gent)
- Other streptococci: either antistreptococcal or above enteroccal regimen
- Aerobic GNB: based on susceptibilities; both beta-lactam & antipseudomonal aminoglycosides IV & IT indicated
- Corynebacterium sp. & Proprionibacterium sp. (diphtheroids)
 - If penicillin sensitive: use enterococcal regimen above
 - If penicillin resistant: IV + IT vancomycin

Infection **Treatment:** Antibiotics

Table 9. Antibiotics Used to Treat Shunt Infections		
	Children	Adults
Gram-positive organism*		
Vancomycin	10–15 mk/kg (q6h)	500 mg (q6h)
Nafcillin	Neonate: 25 mg/kg (q8h) Infant/child: 37.5 mg/kg (q6h)	250 mg-1 gm (q6h)
Rifampin	10–20 mg/kg up to 600 mg (qd IV or PO)	600 mg (qd IV or PO)
Gram-negative organism†		
Ceftriaxone	75–100 mg/kg (q6h)	1–2 gm (q24h)
Cefotaxime	Neonate: 50 mg/kg (q12h) Infant/child: 50 mg/kg (q8h)	1–2 gm (q8h)
Ciprofloxacin	± 0.011.7	400 mg IV
Chloramphenicol	4.25 mg/kg (q6h)	12.5 mg/kg (q6h)

Table O. Antibiation Handler Torot Chunt Info diana

Empiric coverage is administered for S. epidermidis infection. Intravenous administration is indicated unless otherwise specified.

* Combination vancomycin or nafcillin and rifampin is recommended.

† Third generation cephalosporin is recommended.

‡ Not recommended for children <18 years old.</p>

qd = total daily dose.

From: Naradzay JFX et al. J Emerg Med. 1999; 17(2):311-322.



- Extended hospital stays
- $\circ \uparrow$ long-term mortality risk
- $\circ \uparrow$ seizure risk
- Delayed developmental milestones
- Lower IQ & poor school performance

Neuroendoscopy

Neuroendoscopy History

- 1910 L'Espinase attempted fulguration of choroid plexus in 2 infants w/ hydrocephalus with cystoscope – 1 patient died post-op
- o 1922 Dandy similar
- 1923 Fay & Grant visualized & photographed interior of ventricles of child w/ hydrocephalus w/ cystoscope
- 1923 Mixter 1st successful 3rd ventriculostomy
- 1943 Putnam endoscopic choroid plexectomies by cauterization high failure & peri-op mortality rates
- o 1970 Scarff similar
- Decline in neuroendoscopy w/ advent of ventricular shunts & development of microsurgery
- Rediscovery of neuroendoscopy w/ advances in technology in 1970's
- 1990 Jones 50% shunt-free success rate for endoscopic 3rd ventriculostomy improved to 60% in subsequent series

Neuroendoscopy Technology

- Ventricular cannula
- o Endoscope:
 - Rod-lens scope (rigid)
 - Clearer images
 - Fiberscope (flexible)

- Other ports:
 - Electrocautery
 - Irrigation

 RL or NS
- o Camera
- Video monitor
- Light source
 - Halogen, mercury vapor, xenon

Neuroendoscopy Uses

- Hydrocephalus
 - Obstructive hydrocephalus from primary aqueductal stenosis or compressive periaqueductal mass lesions
 - Septum pellucidotomy or septostomy for isolated lateral ventricles
 - Fenestration of loculated ventricles
 - Marsupialization & fenestration of intracranial cysts
 - Aqueductoplasty

- Neurooncology
 - Biopsy & resection of intraventricular tumors
 - Resection of colloid cysts
 - Endonasal transsphenoidal hypophysectomy
- Spine surgery
 - Thoracoscopic sympathecotmy
 - Discectomy
 - Lumbar laminotomy
 - Resection of tumors & cysts
- Craniosynostosis

Endoscopic Third Ventriculostomy

- For tx of obstructive hydrocephalus caused by primary aqueductal stenosis or compressive periaqueductal mass lesions
- More physiologic tx of obstructive hydrocephalus by allowing egress of ventricular CSF directly into subarachnoid space, bypassing downstream stenosis
 - Opening made in the floor of 3rd ventricle
- Alternative to VP shunt which is assoc w/ frequent & multiple complications
 - Opportunity for patient to have shunt-free existence

Endoscopic Third Ventriculostomy Patient Selection

Symptoms & signs of hydrocephalus

• Features on MRI

- Enlarged lateral & 3rd ventricles, w/ N or small 4th ventricle
- Midsagittal section demonstrating adequate space between basilar artery & clivus under floor of 3rd ventricle

Endoscopic Third Ventriculostomy Anatomy

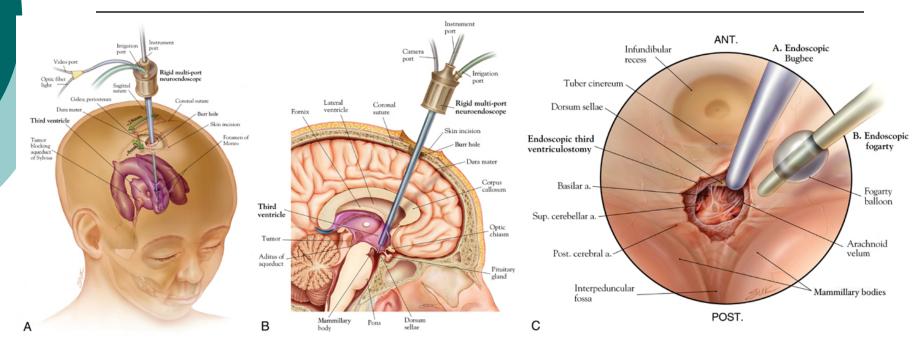


Fig. 1. Artist's illustrations demonstrating ETV. Oblique view (A) demonstrating typical location of the bur hole and trajectory; midsagittal view (B) demonstrating location of ventriculostomy; and magnified endoscopic view (C) of the floor of the third ventricle and site of ventriculostomy. a. = artery; ant. = anterior; post. = posterior; sup. = superior.

From: Li KW et al. Neurosurg Focus. 2005; 19(6):E1.

Endoscopic Third Ventriculostomy Anatomy

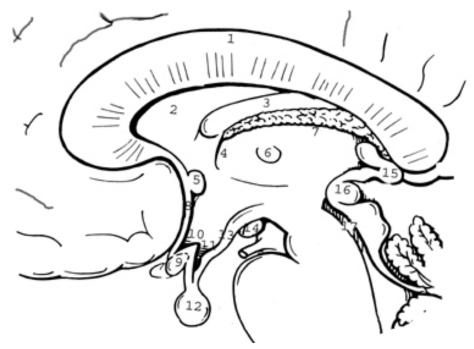


Fig. 2. Drawing showing a midsagittal view of the third ventricle. Anatomical features are designated by the following numbers: 1) corpus callosum; 2) septum pellucidum; 3) fornix; 4) foramen of Monro; 5) anterior commissure; 6) massa intermedia; 7) choroid plexus; 8) lamina terminalis; 9) optic chiasm; 10) optic recess; 11) infundibular recess; 12) pituitary gland; 13) tuber cinereum; 14) mammillary body; 15) pineal gland; 16) posterior commissure; 17) cerebral aqueduct.

From: Jallo GI et al. Neurosurg Focus. 2005; 19(6):E11.

Endoscopic Third Ventriculostomy Anatomy



: Endoscopic view through the right foramen of Monro.

From: Jallo GI et al. Neurosurg Focus. 2005; 19(6):E11.

Endoscopic Third Ventriculostomy Technique

- Burr hole at or just anterior to coronal suture, 2.5-3 cm lateral to midline
- Open dura in cruciate fashion, coagulate edges
- No. 14 Fr catheter used to cannulate lateral ventricle
 - Remove stylet
- Pass endoscope thru sheath to visualize lateral ventricle
- Identify Foramen of Monro & navigate scope into 3rd ventricle
 - Identify mamillary bodies & infundibular recess in floor
 - Sometimes basilar artery visible
- Puncture floor of 3rd ventricle & dilate opening
- On completion, remove scope & sheath
- Gelfoam plug in burr hole
- Close galea & skin

Endoscopic Third Ventriculostomy Post-op Care

• Observation in ICU x 1 day

o MRI

- CINE CSF flow thru opening in floor of 3rd ventricle
- Ax T2WI ("Poor man's CINE") flow void in floor of 3rd ventricle

Endoscopic Third Ventriculostomy Outcome

Overall success rate 50~90%

 Most failures occur soon after procedure

Reclosure of ventriculostomy ~22%

Longer follow-up studies necessary

Endoscopic Third Ventriculostomy Possible Complications

- Incidence 0~20%
- Bleeding
 - SAH injury to basilar artery
 - ICH
 - IVH bleeding from choroid plexus
 - SDH
- Injury to surrounding structures
 - Cranial nerve palsy CN III, VI
 - Fornix, caudate, thalamus, thalamostriate venous complex
 - Hypothalamic / pituitary dysfunction
 - Typically manifests as DI
 - Cardiac arrhythmias or resp arrest from manipulation or irritation of hypothalamus
- o Infection
- Mortality ~1%

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