Endoscopic Third Ventriculostomy

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Introduction

Endoscopic third ventriculostomy (ETV) is a minimally invasive procedure indicated for the treatment of hydrocephalus. An ETV is performed by fenestrating the floor of the third ventricle, thus creating a passage between the third ventricle and the prepontine cisterns.

Hydrocephalus is commonly treated with cerebrospinal fluid (CSF) diversion with shunt placement. Shunts are effective and potentially life-saving, but they have a number of inherent problems. Shunt infection occurs in approximately 5% to 8% of cases. An infection necessitates multiple surgeries, prolonged hospitalization, and potentially significant morbidity. Shunts can also fail. The risk of shunt failure requiring subsequent surgery in children is approximately 50% within 2 years of implantation.[1] These and other concerns encouraged the exploration of other treatment options, including ETV.

Success in performing an ETV largely depends on appropriate patient selection. In general, appropriate candidates have symptomatic obstructive hydrocephalus. The ETV Success Score (ETVSS) is an extremely useful tool for estimating the procedure’s chance of success. “Success” is defined as the patient not requiring further surgery within 6 months of shunt placement (Table 1). The ETVSS was derived from a retrospective analysis of patients aged 19 years or younger. The utility of the scale has been confirmed in multiple subsequent studies.

Table 1. The ETV Success Score*

Score	Age	Etiology	Previous Shunt

0	<1 mo	Postinfection	Yes
10	1 to <6 mo	—	No
20	—	Myelomeningocele, intraventricular hemorrhage, nontectal brain tumor	—
30	6 mo to <1 y	Aqueductal stenosis, tectal tumor, other	—
40	1 to <10 y	—	—
50	≥10 y	—	—

* Calculated as age score + etiology score + previous shunt score. The ETVSS is equivalent to the chance of having a successful ETV without failure 6 months after the shunt-placement procedure. (Adapted from reference 2.)

Advantages

The risk of infection with ETV is much lower than that with shunt insertion. The risk of infection after an uncomplicated ETV is approximately 1%.[3]

The duration of effect after a successful ETV is significantly better than that after shunt placement. The risk of shunt malfunction is highest in the first 2 years after insertion, and the risk of failure continues over time. In contrast, the survival curve after an ETV flattens. The risk of failure of an ETV is highest in the first 6 months after the procedure but drops significantly after 1 year.[4] In other words, a patient who does not require further surgery in the first year after an ETV is unlikely to ever require further surgery.

Disadvantages and Complications

Surgeons performing ETV should be experienced and comfortable with endoscopic techniques. In large series, the overall rate of complications is approximately 8.5%. The risk of permanent morbidity is around 3%.

Complications include diabetes insipidus, memory loss, hemiparesis, hypothalamic dysfunction, seizures, and CSF leak, among others.

The most dreaded risk is injury to the deep vascular structures, including the basilar artery or its perforators, or the internal cerebral vein. The risk of hemorrhage severe enough to abort the procedure is approximately 4%. The risk of severe hemorrhage causing stroke or death is less than 1% in experienced hands.[3]

Preoperative Considerations

The anatomy of the ventricles and surrounding structures must be analyzed carefully. Anatomic variants, such as an enlarged massa intermedia (common in patients with spina bifida) can make access to the floor of the third ventricle challenging. Tumors can distort the ventricles or obscure normal anatomy. Ideally, there is space between the dorsum sella and the basilar artery. A basilar artery situated tightly next to the bone is a relative contraindication for ETV. The trajectory to the floor should be planned, and any other procedures should also be considered.

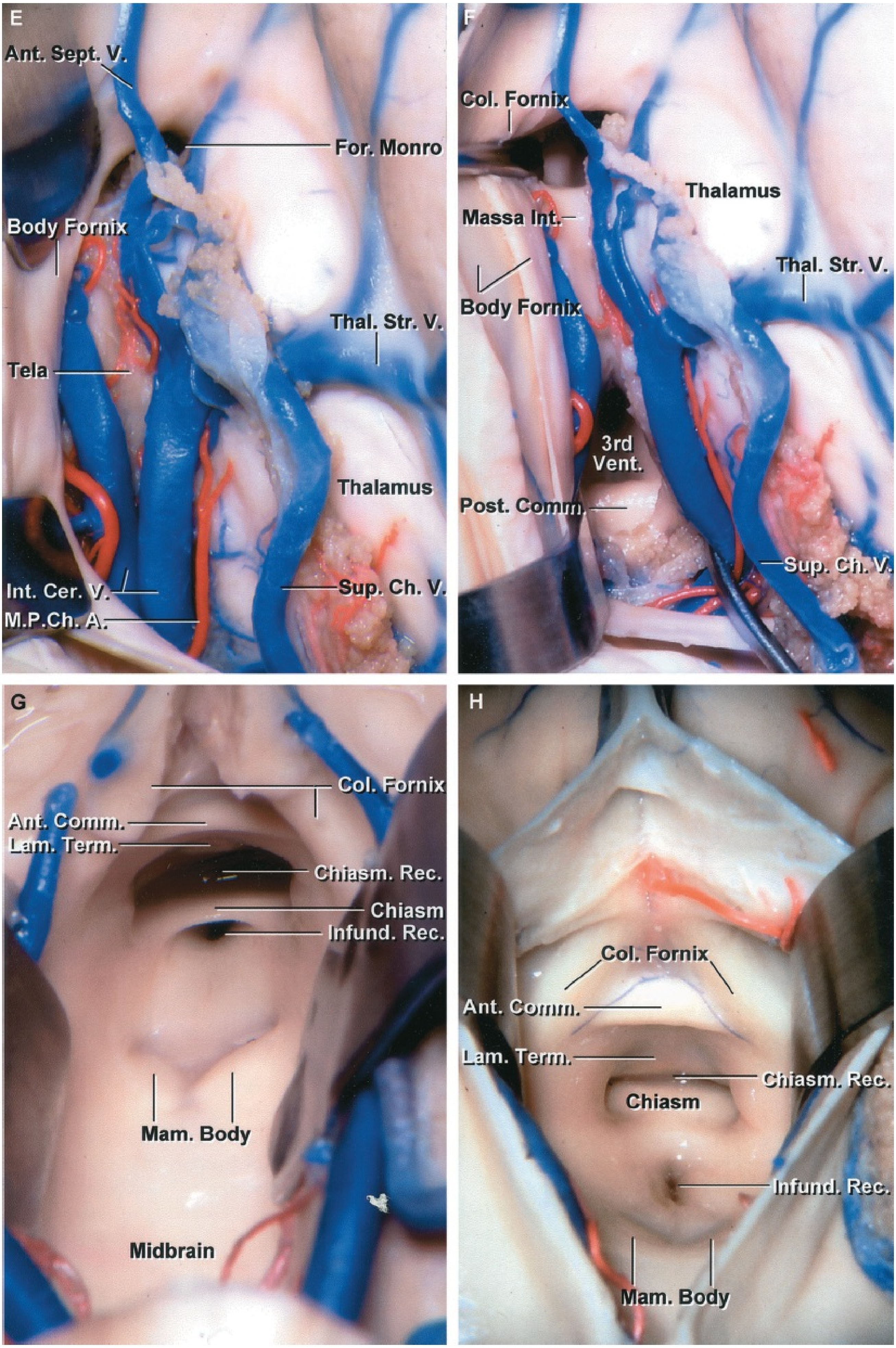
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Figure 1: The relevant anatomy of the floor of the third ventricle and location of the fenestration are noted. The mamillary bodies are identified. The infundibular recess appears as a red blush. The puncture site for the floor of the third ventricle is just anterior to the mammillary bodies.

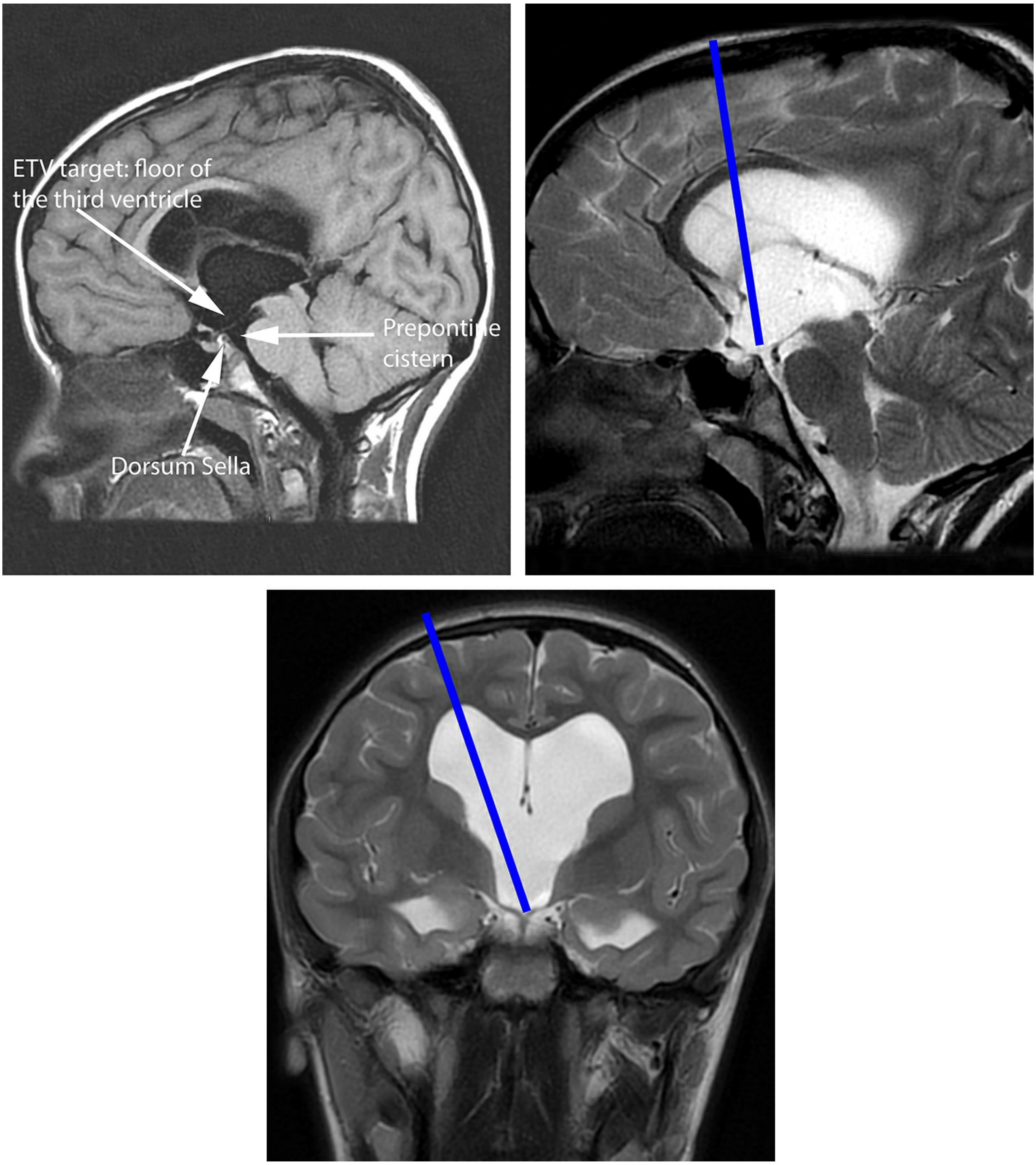

Figure 2: (Top Left) Sagittal T1-weighted MRI of a patient considered for

ETV. Relevant anatomy is labeled. (Top Right and Bottom) Sagittal and coronal T2-weighted MRI demonstrating the endoscope trajectory, shown in both views as a blue line. The entry point into the skull, foramen of Monro, and target area of the floor of the third ventricle should be aligned. In this case, the lateral and third ventricles are dilated, as is the foramen of Monro. The procedure is much easier if the foramen is dilated and there is room to maneuver the endoscope. There is also adequate space between the dorsum sella and the basilar artery. This relationship should always be studied before surgery.

A relatively common scenario for ETV is a patient with obstructive hydrocephalus resulting from a pineal region mass for whom a biopsy may be warranted. In this scenario, the optimal trajectory to the tumor might differ from the trajectory to the ventricular floor. A trajectory to “split the difference” might be planned for a rigid endoscope. It is important to not compromise the effectiveness of either procedure by choosing a poor trajectory. Two burr holes with separate approaches can be considered. A flexible endoscope can facilitate this procedure. It is recommended that the ETV be performed before the biopsy. Pineal tumors can be vascular, and tumor biopsy might cause bleeding. This situation is rarely dangerous but can obscure the surgeon’s view.

ENDOSCOPIC THIRD VENTRICULOSTOMY

The patient is placed supine with the head slightly elevated (15°) and the neck slightly flexed. The head is generally kept in a neutral position. The height of the head should align with the operating surgeon’s elbow level for ergonomic ease. A screen displaying the navigation platform and the endoscope view should be in a natural sight line of the surgeons.

Figure 3: The operating room is organized with the head of the patient 180° away from the anesthesia team. Screens displaying neuronavigation and endoscope pictures should be in natural sight lines of the surgical team. The patient's head should be flexed approximately 15° and in the neutral position.

A foley catheter and arterial line are placed. There might be alterations in blood pressure and fluid balance during the procedure that would necessitate prompt intervention.

The patient's head is registered to the neuronavigation system. The incision is planned to optimize the trajectory to the floor of the third ventricle by lining up the entry point with the foramen of Monro and the floor. The incision should remain anterior to the coronal suture. The right side is preferred unless there is a compelling anatomic reason to approach from the left. Either a linear or curved incision can be used. A curved incision confers the advantage of allowing for future shunt placement at the area; a linear incision confers the advantage of allowing extension if a second trajectory is required.

Figure 4: (Top Left) The patient is registered to the neuronavigation system. (Top Right) The trajectory is planned. The navigation system is used to plot a line from an entry point on the scalp, through the foramen of Monro, and to the target floor of the third ventricle. The entry point should remain anterior to the coronal suture. (Bottom) The midline and the incision are marked. A linear incision confers the advantage of easy extension anteriorly or posteriorly, should the need for a new trajectory arise; a curved incision confers the advantage of allowing for subsequent frontal shunt placement, if needed.

The equipment should be checked and fully operational before skin incision. The quality of the endoscopic picture is evaluated, and the scope is “white balanced.” The focus is optimized. It is important to confirm the orientation of the picture with the endoscope. Both the focus and orientation can be checked by reading type on an object such as a suture pack. An irrigation system is attached to the endoscope; a balanced solution for irrigation is required to avoid local electrolyte imbalances that can affect hypothalamic function. All of the endoscopic instrumentation should be checked and organized before skin incision.

An external ventricular drain (EVD) catheter should be included in the equipment. A peel-away sheath is helpful for providing access to the ventricle and reducing trauma from passage of the scope. In this following, the Medtronic (Minneapolis, MN) Stealth AxiEM system is shown. The stylet from the system fits into the peel-away sheath to aid in cannulating the ventricle.

Figure 5: (Top Left) The endoscopic tools are organized and inspected before making the skin incision. We recommend using a peel-away sheath to cannulate the ventricle, which minimizes trauma from the endoscope passing into the ventricle and retains a potential path in case intraoperative bleeding impairs the surgeon’s vision. (Top Right and Bottom Left) The stylet from the navigation system fits into the peel-away sheath. (Bottom Right) The peel-away sheath is passed into the ventricle with neuronavigation. The sheath is passed until a “pop” is felt as it enters the ventricle. It is important to resist advancing the sheath after the tactile “pop.” The peel-away sheath also has measurements

marked. The surgeon should always confirm navigation with anatomy and refrain from advancing the sheath too deeply.

A high-speed drill is used to make a burr hole. If the skull is unusually thick, the inner table can be undercut with a bone curette or Kerrison rongeur to enable flexibility in the approach trajectory. It is important to allow plenty of room to angle and adjust the endoscope. The dura is visualized and widely coagulated. The dura is opened in a cruciate fashion as widely as the burr hole allows. The edges are coagulated to prevent bleeding during the procedure. The pia is then coagulated. The pia can be opened formally with an 11-blade scalpel. A small corticotomy is created. A peel-away sheath is then passed into the ventricle, ideally with neuronavigation assistance.

A tactile “pop” is felt when the peel-away sheath enters the lateral ventricle. The inner stylet of the sheath is removed, and the endoscope is inserted. The peel-away sheath should be positioned such that it just barely passes the ventricular ependyma. Once the optimal depth is determined, the sheath is secured to the scalp with staples.

CSF can be collected with a syringe and a long angiocatheter, if needed, for analysis. Testing for tumor markers in the CSF might be required in the evaluation of patients with a pineal mass.

The surgeon must always maintain precise and strict control of the endoscope. All movements should be short, smooth, and controlled. There are a number of commercially available arms designed to hold the endoscope, which can be useful for situations in which a skilled assistant is not available. With 2 surgeons, one “drives” the scope, and the other manipulates the tools. The surgeon maneuvering the endoscope should always use both hands. The nondominant hand is placed on the skull, and the endoscope is maneuvered between the thumb and first finger. The dominant hand holds the camera with a pistol grip.

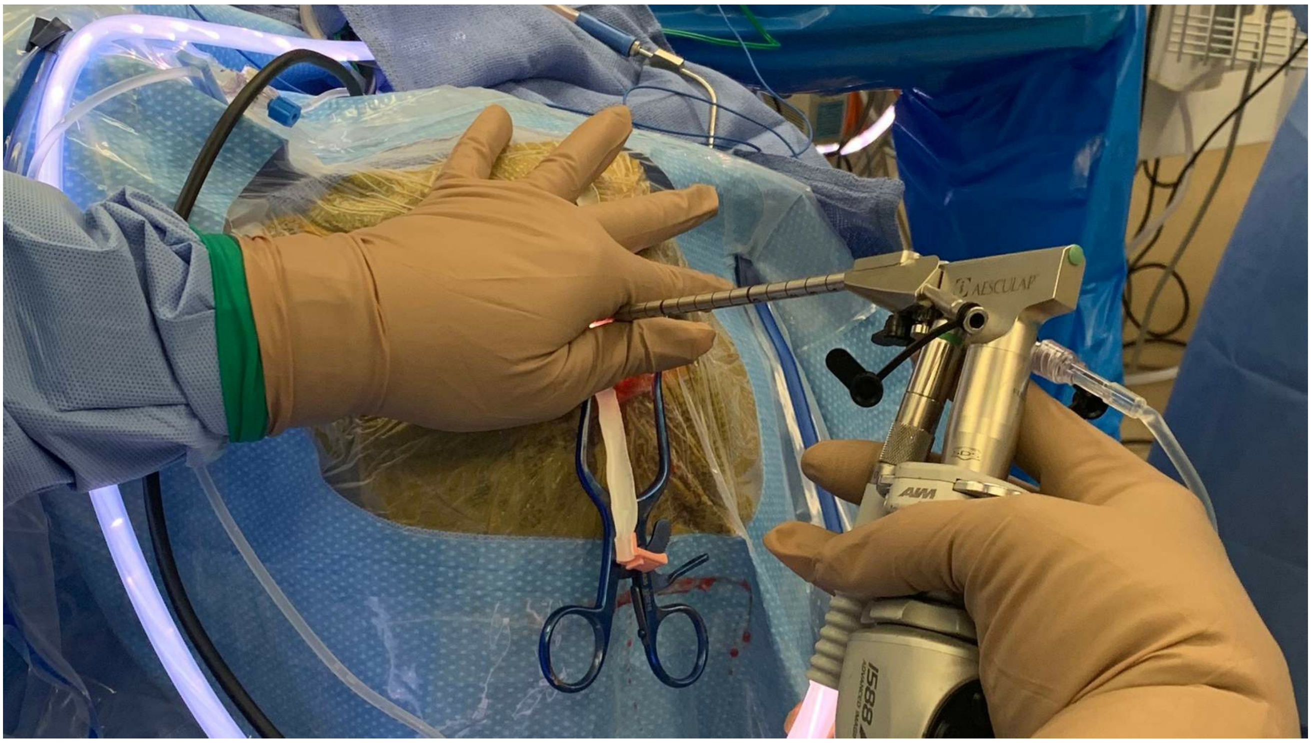

Figure 6: Strict control of the endoscope must be maintained at all times. The surgeon controlling the endoscope should use both hands. The nondominant hand is placed on the scalp, and the endoscope is passed through the thumb and first finger. The dominant hand maintains strict control with a pistol grip. The endoscope should be fully removed from the head before passing it from one surgeon to another.

The relevant anatomic landmarks are identified. Do not assume that the ipsilateral ventricle has been entered, especially in a patient with smaller ventricles. The choroid plexus is usually the most easily identifiable landmark. One can follow this landmark to the foramen and identify the relevant veins. The thalamostriate vein is lateral to the choroid plexus and should be used to confirm which ventricle is in view.

The foramen of Monro lies in the venous angle between the septal vein (medial) and the thalamostriate vein (lateral), both join the internal cerebral vein. Between these veins lies the choroid plexus. The column of the fornix forms the anterior wall of the foramen, and the anterior pole of the thalamus forms the posterior wall.

The endoscope is advanced through the foramen to visualize the third ventricle. The floor proper extends from the optic chiasm to the inlet to the cerebral aqueduct. The lamina terminalis lies anteriorly and rostrally.

Immediately below the lamina terminalis, the optic chiasm will be apparent as a horizontal white or yellow band. Inferior to the chiasm is the infundibular recess, a brown or red dimple in the ependyma which gives rise to the pituitary stalk inferiorly. The infundibular recess is almost always visualized, is always midline, and serves as an anatomic landmark (see Fig 1 above).

Posterior to the infundibular recess lies the tuber cinereum, the thinnest part of the floor of the third ventricle formed by the ependyma on the ventricle side and arachnoid on the inferior side. Just posterior to the tuber cinereum are the mammillary bodies. In cases of extreme hydrocephalus, the floor might bulge downward into the interpeduncular cistern forming the premamillary recess. If the tuber cinereum is attenuated, the dorsum sellae and the basilar artery can be seen anteriorly and posteriorly, respectively.

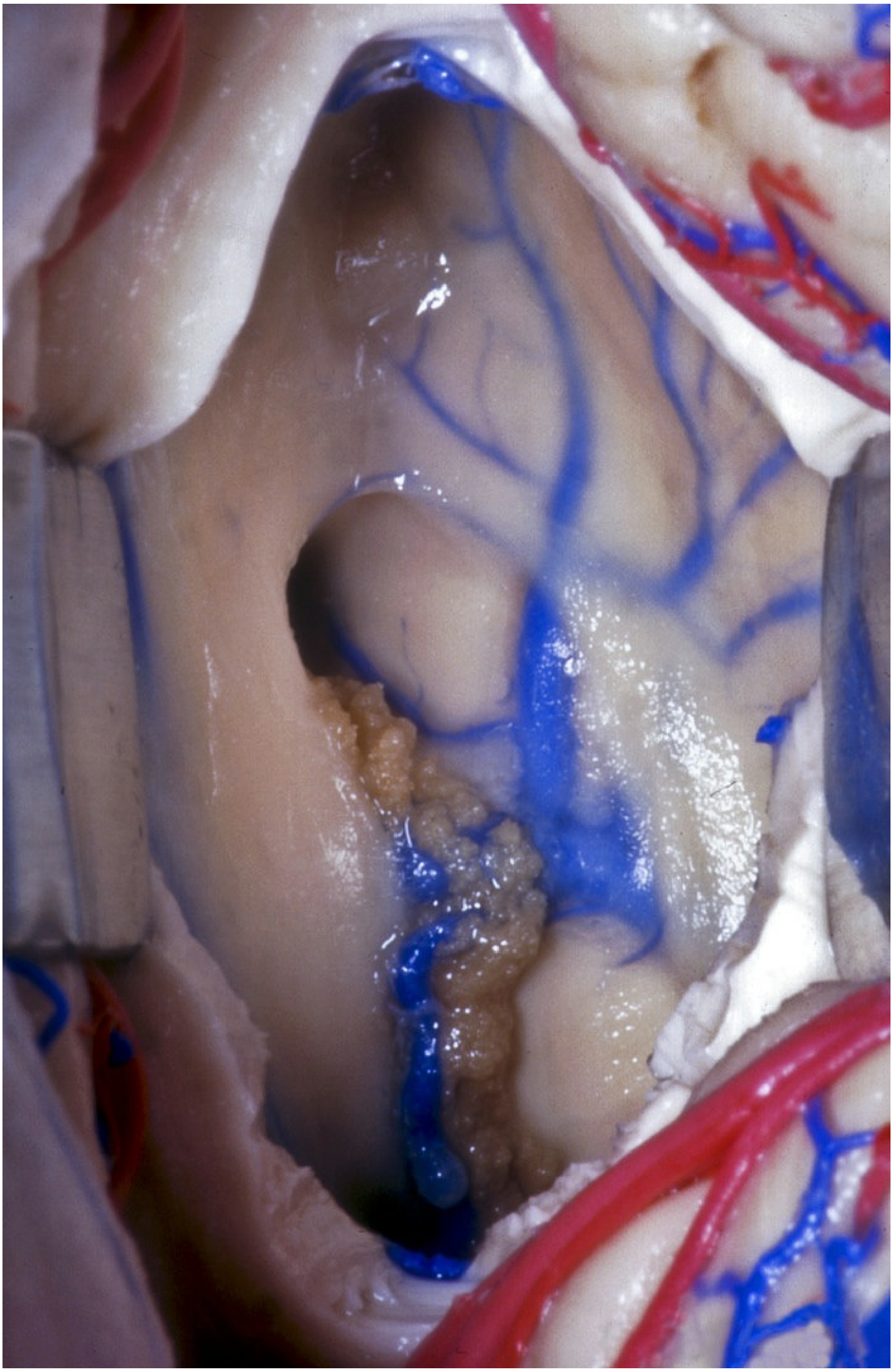

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Figure 7: The landmarks discussed above that ensure entry into the correct (right) ventricle are demonstrated.

The point of perforation of the floor is in the midline and just anterior to the mamillary bodies. Multiple techniques and tools for perforating the floor have been described. I use the stylet from a standard EVD kit or endoscopic grasping forceps to make the initial perforation. This stylet fits through the working channel of a standard rigid endoscope and has a blunt tip that protects against lacerating a perforating vessel below the floor.

Once perforated, the hole is enlarged. Multiple tools for this task have been described. A Fogarty balloon catheter can be inserted and inflated to widen the stoma. As an alternative, endoscopic grasping forceps can be inserted and spread. It is vital to avoid closing the grasping forceps after the floor has been breached so that a penetrating vessel on the other side is not inadvertently grabbed; the forceps should be inserted through the hole, opened, and then withdrawn without closing them until the tips are directly visualized.

There is no defined optimal size for the stoma. Ideally, the stoma is as large as possible without encroaching on the mamillary bodies, infundibular recess, or walls of the third ventricle. I make the hole big enough to easily pass the endoscope into the prepontine cistern. It is critical to confirm that the prepontine cistern has been entered. Two layers must be penetrated, the tuber cinereum and the membrane of Liliequist. One source of procedure failure is penetrating the floor (tuber cinereum) but not adequately traversing the membrane. An unobstructed view of the basilar artery confirms the appropriate location. The floor of the third ventricle should pulsate and flap freely after creation of the stoma.

Irrigation is minimized. Overzealous irrigation can cause hypothalamic damage from electrolyte imbalance or direct pressure. The floor of the third ventricle often bleeds slightly after stoma creation, but it will stop with gentle irrigation and does not require cautery.

Finally, the endoscope is withdrawn. The fornix should be inspected for any injury. Small bruises on the fornix are rarely of consequence but should be avoided. If any venous bleeding is visible, irrigate the space until hemostasis is achieved. Remove all instruments from the device channels. The peel-away sheath is removed with the endoscope, and the cerebral parenchyma is evaluated for any bleeding. We place an absorbable gelatin sponge over the cortical defect to prevent CSF leakage. A burr hole cover can be considered to prevent a cosmetically noticeable dip if the burr hole is near the hair line.

Minor bleeding of the intraventricular structures or choroid plexus will generally stop with irrigation. Severe bleeding might require cauterization. It is critical to cauterize only a directly visualized source. Indiscriminate cauterization can damage surrounding structures and increase hemorrhaging. If the patient develops bleeding that is unresponsive to irrigation, the EVD should be passed, off stylet, through the peel-away sheath, and the procedure should be aborted. An EVD can also be placed if there is doubt regarding the efficacy of the procedure in a patient with elevated intracranial pressure.

Postoperative Considerations

Patients should be closely monitored after the procedure for seizures, blood pressure lability, bradycardia, hyperthermia, and diabetes insipidus. It is not necessary to perform intracranial pressure monitoring in patients with primary obstructive hydrocephalus and no intraoperative complications. I leave an EVD only if there is significant intraoperative bleeding.

A postoperative computed tomography scan is optional. Perioperative medications vary among surgeons. Antibiotics are generally given for 24 hours. Some surgeons administer high-dose dexamethasone after the procedure to prevent inflammation and stoma closure, and some give antiseizure medications for 24 to 48 hours. Patients without any complications are generally discharged home in 1 to 2 days.

The need for postoperative imaging varies among patients. The ventricles generally do not collapse as much as they do with a CSF shunt. Any shrinking of the ventricles on long-term imaging is a good sign that the ETV is working. MRI can demonstrate perforation of the floor. Specifically, a 3-dimensional constructive interference in steady state (CISS) or fast imaging using steady-state acquisition (FIESTA) study can show patency of the stoma. Sagittal T2-weighted images will show a flow artifact if CSF is draining through the stoma.

Figure 8: Sagittal T2-weighted MRI of a patient who underwent ETV for obstructive hydrocephalus caused by a tectal glioma. The flow artifact is a dark plume crossing the floor of the third ventricle (arrow). Dynamic CINE flow images may also show flow across the ETV stoma.

Controversies

Given the inherent problems of shunt placement, people have attempted ETV in patients with a lower score on the ETVSS. ETV has been attempted in patients with normal-pressure hydrocephalus, although the results have been poor. ETV is technically challenging in patients with idiopathic intracranial hypertension (pseudotumor cerebri), given their generally smaller ventricles. Outcomes have varied.

ETV combined with choroid plexus cauterization (CPC) has been explored as an option in infants with spina bifida, posthemorrhagic hydrocephalus, congenital hydrocephalus, and postinfectious hydrocephalus. This procedure has worked well in developing nations. However, similar results have not been reproduced in the United States. A multicenter study performed by the Hydrocephalus Research Society found that the results of ETV/CPC were similar to those predicted by the ETVSS, bringing into question whether the CPC was beneficial.[5]

Pearls and Pitfalls

- Neuroendoscopy provides monocular vision inside small structures. Slow and gentle movements are safe and allow for depth perception.
- The etiology of hydrocephalus, clinical status, and results of imaging can all help determine which patients are ideal candidates for ETV. The ETVSS is a valuable tool for properly selecting patients for the procedure.
- Excessive manipulation while in the foramen can lead to damage of the anterior column of the fornix, which can result in a significant loss of quality of life.
- A key principle in neuroendoscopy is to always maintain the instrument in your field of view. To accomplish this goal, keep the endoscope behind the instrument tip, and advance the instrument and endoscope in small successive steps.
- Identify the relevant anatomy initially. Do not assume that the endoscope has entered the ipsilateral ventricle.
- Both the tuber cinereum and the membrane of Liliequist, when present, must be perforated. The best way to ensure entry into the prepontine cistern is to have a clear view of the basilar artery.

- Intraoperative bleeding generally stops with gentle irrigation. Aggressive cautery should be avoided. An EVD should be left in place if there is significant hemorrhaging.
- Unfavorable anatomic characteristics include small ventricles, a large massa intermedia, and a small space between the dorsum sella and basilar artery.

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