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Chapter 8

Presentation of the Success Rate of ETV in Distinct Indication Cases of Hydrocephalus

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Additional information is available at the end of the chapter

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Abstract

Endoscopic third ventriculostomy (ETV) is an endoscopic fenestration between the floor of the third ventricle and subarachnoid space. It is the procedure of choice for obstructive hydrocephalus (HC). The indication includes obstructive HC caused by aqueduct stenosis, tumors, brain infarction, cystic lesions, hematoma, postinfectious and posthemorrhagic HC, malformation of the fourth ventricle, and further uncommon indications. In this chapter, surgical techniques and the success rate of ETV in distinct indications will be presented and discussed. The overall success rate of ETV is reported at 60–90%. The outcome of the procedure depends highly on the underlying pathology and age. A very favorable outcome is reported in case of aqueduct stenosis (67–93.5%). High success rate is observed in case of cerebellar infarction (86%), tumors (56–81%), and intraventricular cysts (56–95%). In case of intraventricular hemorrhage (43–73%), infection (60–64%), anatomical aberration (21–80%), and communicating HC (65–72%), a significantly inferior success rate is reported. It is well known that ETV has a lower success rate in children (68–71%) compared to adults (70–90%). The overall high clinical success rate in short-term and long-term follow-up confirms that ETV is the gold standard for treatment of occlusive HC. It is effective, safe, and simple.

Keywords: third ventriculostomy, hydrocephalus, neuroendoscopy, success rate, indication

1. Introduction

1.1. Classification of hydrocephalus

Walter Dandy classified hydrocephalus as "communicating" or "noncommunicating" type based on examinations with ventricle and lumbar puncture in 1913. He considered hydrocephalus as "noncommunicating" or "obstructive" if after the injection of a colored solution in the lateral

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ventricle, the dye did not appear in the spinal fluid removed by lumbar puncture. In contrast, if this solution appeared, hydrocephalus was classified as "communicating" [1]. This classification was simple and useful to understand hydrocephalus, but first was found inadequate to understand the pathophysiology [2]. The proposed classification is hydrocephalus without obstruction (true communicating hydrocephalus) in case of choroid plexus papilloma and with obstruction at the level of venous outflow (skull base anomalies and congenital heart disease), arachnoid granulations (hemorrhage and infection), basal cisterns (hemorrhage and infection), outlets of fourth ventricle (infection and Chiari malformation), Sylvian aqueduct (congenital anomalies and tumor compression), and foramen of Monro (congenital anomalies, tumor, and postshunt ventricular asymmetry) [2]. Nonetheless, the classic nomenclature remained in clinical practice due to its simplicity.

1.2. History of neuroendoscopy

The first neuroendoscopic procedure was introduced in 1910, when L'Espinasse performed a cauterization of plexus choroideus with a cystoscope. The other pioneer in neuroendoscopy was Walter Dandy. He published his results in 1913. Clarifying the physiology, Dandy performed plexectomy to cease the cerebrospinal fluid (CSF) production as a treatment for communicating hydrocephalus. In case of obstruction, he created an internal bypass for communication between the third ventricle and cortical subarachnoid space [2, 3].

The first endoscopic third ventriculostomy (ETV) was performed by William Mixter in 1923. This successful technique later served as a model for modern neuroendoscopy. He performed a perforation of the third ventricle floor by an intraventricular approach through the foramen of Monro. The invention of Hopkins optics, cold light sources, and digital cameras was a great development in the history of endoscopy. Following technical development and clinical application, neuroendoscopy spread over the world [3].

1.3. Endoscopic third ventriculostomy nowadays

For obstructive hydrocephalus, endoscopic third ventriculostomy is the procedure of choice. This is an endoscopic fenestration between the floor of third ventricle and subarachnoid space. The CSF flow can be restored and balanced out [4]. This procedure is considered effective, safe, and simple. The complication rate is lower as it has a lower infection rate; there is no foreign material and no overdrainage compared with shunt [4].

Nowadays, the use and role of endoscopic ventriculostomy expand continuously. It remains challenging to estimate and define the success of the procedure [5], even though many studies have overall focused on this topic [5–14].

2. Indications

In general, endoscopic third ventriculostomy is the gold-standard treatment for noncommunicating hydrocephalus. The indication for ETV includes all cases with obstruction between

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the third ventricle and the subarachnoid space with preserved CSF absorption from subarachnoid space into the venous system [2, 14, 15]. In case of obstruction at the level of Pacchionian granulation and venous outflow, performing ETV is definitely not recommended [14].

2.1. Aqueduct stenosis

One of most common etiology of obstructive hydrocephalus is the aqueduct stenosis [16, 17] (**Figures 1** and **2**). The obstruction can be congenital or acquired. In 75% of cases, it is idiopathic. In remaining cases, it can be caused by infections, hemorrhages, or malformation of the central nervous system or can be related to genetic factors [16]. The first and best treatment option for this is ETV. Ventriculostomy has a good outcome regardless of etiology of the stenosis, whether it is congenital, acquired, or tumor-related obstruction [14]. However, the success rate is different in certain cases of the aqueduct stenosis. It depends on the etiology, age, clinical, and radiological characteristics [13]. In certain cases, as an alternative treatment or even simultaneous, aqueductoplasty may be performed [7, 18]. The result of aqueductoplasty is comparable with the success of ETV. As alternative treatment, it may be useful in case of thick and tough floor of the third ventricle preventing the hypothalamus or vessel injuries [18].

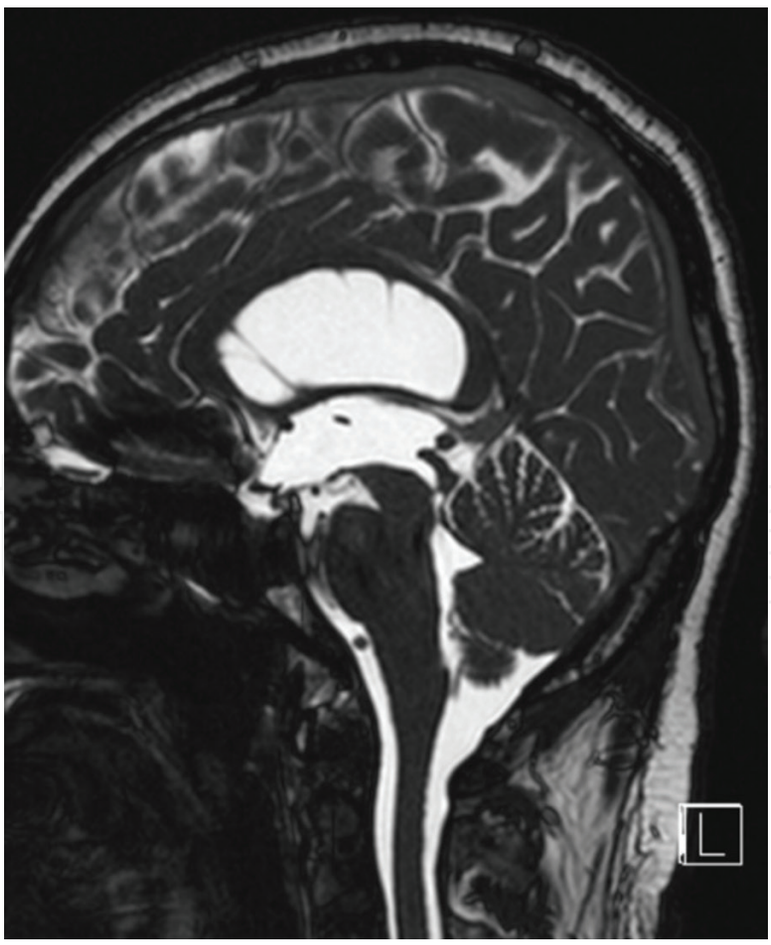

Figure 1. Aqueduct stenosis (MRI scan TRUFI sequence).

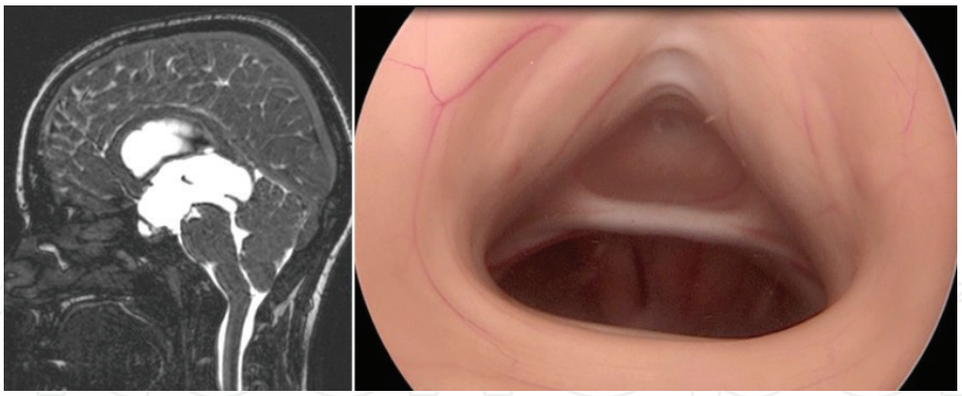

Figure 2. Aqueduct stenosis (left) MRI scan TRUFI sequence (right) endoscopic view.

2.2. Far distal obstruction

Obstruction distal to the fourth ventricle is a rare cause of hydrocephalus [19]. It can develop through intraventricular membranous obstruction [20, 21] or extraventricular compression [22] and in case of Dandy Walker syndrome [26] and Chiari malformation [23–25] (see below) [19]. Like all obstructive hydrocephalus, it may also be treated successfully with ventriculostomy, although an additional surgical technique with flexible endoscope is required (see below).

2.3. Tumors and cystic lesions

Endoscopic third ventriculostomy might be a surgical option in case of tumor- or cyst-related hydrocephalus, where the CSF flow disturbance is distal from the place of ventriculostomy (**Figures 3–5**). Therefore, in selected cases, it might be performed in case of tumor localization in the brainstem, posterior fossa, thalamus, pineal region, third and fourth ventricle, and other localization, e.g., cerebellopontine angle, frontal lobe, or diffuse growth pattern [27].

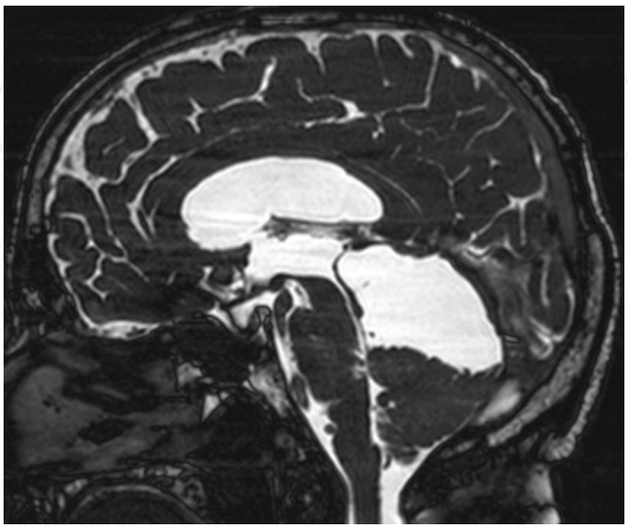

Figure 3. Arachnoid cyst with compression of the CSF pathway (MRI scan TRUFI sequence). In this case, a cystostomy and simultaneous ETV were performed.

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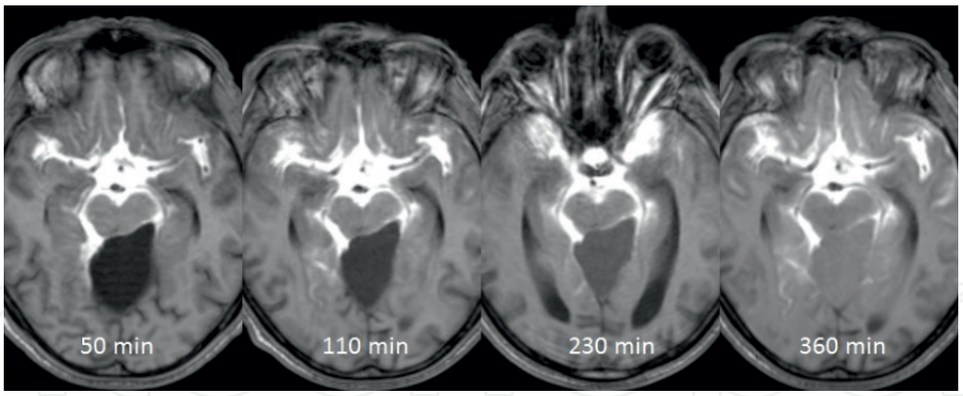

Figure 4. Arachnoid cyst with compression of the CSF pathway (cisternography). Same patient as in **Figure 3**.

In children, the posterior fossa is a common localization for various tumors (54–60% of brain tumors during childhood), and a hydrocephalus develops with high probability in these cases [13]. Frequent localizations are at the cerebellum, midline, brainstem, or cerebellopontine angle. Histologically the following tumors may occur: medulloblastoma, ependymoma, astrocytoma, ganglioglioma, cavernoma, primitive neuroectodermal tumor (PNET), meningioma, and others [28]. Gliomas in the midbrain and in the periaqueductal area are relatively rare and low grade. They show benign growth patterns, and malignant spreading is rare, although a suggested dissemination through CSF has already been reported [29]. The compression of the

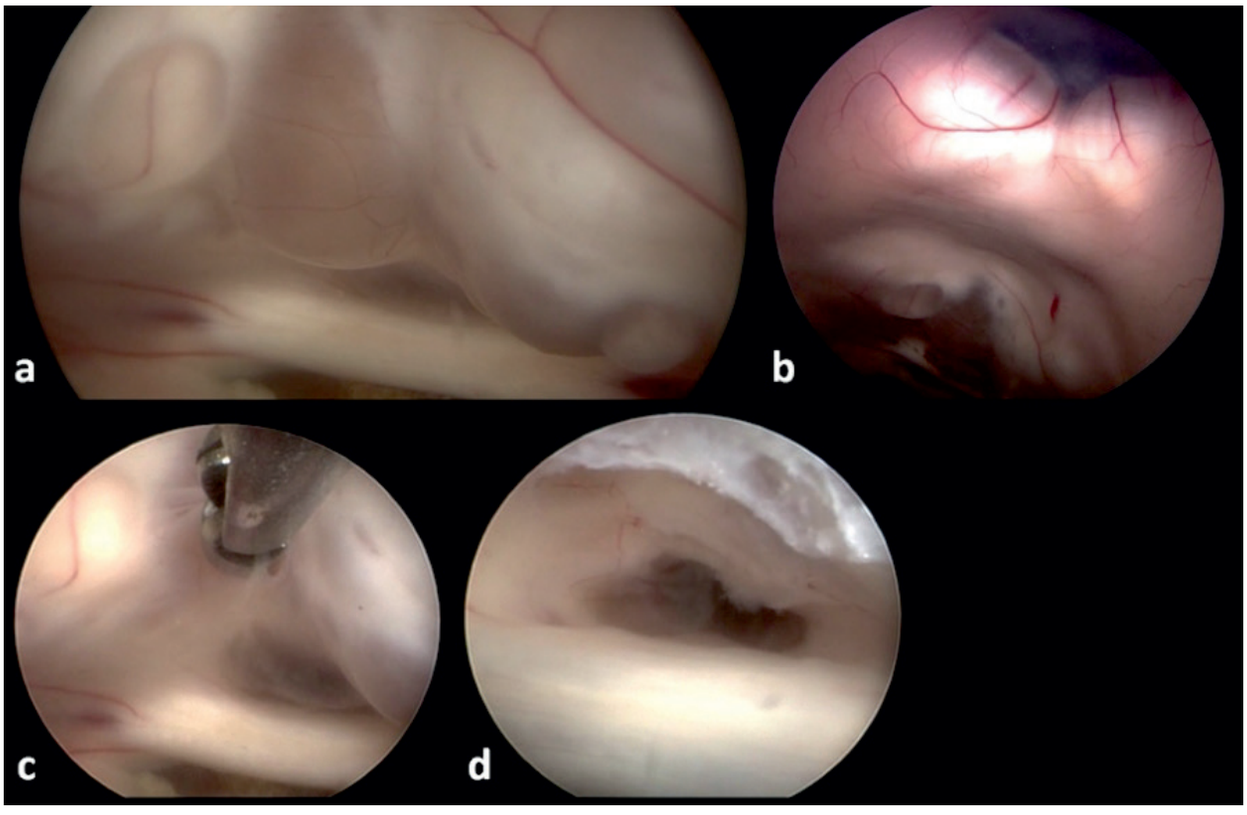

Figure 5. (a and b) Multiple cysts of the third ventricle with obstruction of the Sylvian aqueduct. ETV (see **Figure 9**) and cyst removal (c) were performed. (d) Free aqueduct.

fourth ventricle or the foramina of Magendie and Luschka leads to circulation disturbances and hydrocephalus [13]. ETV is recommended primarily for the treatment of children with midline posterior fossa tumor–related obstructions [30]. ETV can be combined with tumor biopsy, thus providing useful information for the diagnosis. In certain cases, even the tumor resection (e.g., third ventricle) might be performed [13, 30]. The resection alone can set the CSF pathway free, providing a normal CSF circulation. Therefore, performing ventriculostomy routinely is not recommended. After tumor removal, not only the causal problem of hydrocephalus might be solved. With performing the ETV an intraperitoneal spreading of the chemotherapeutic drugs through the shunt can be avoided [32]. The time of ETV in relation to the tumor resection is still under discussion [13].

Beside posterior fossa tumors, the indication expands to pineal tumors and third ventricle tumors [33]. The pathology of these is various. The pathology includes, but is not limited to glial tumors, ependymoma, pinealoblastoma, pinealocytoma, meningioma, germ cell tumors, primitive neuroectodermal tumor, teratoma, epidermoid, metastasis, and cysts [34, 35]. Colloid cysts, arachnoid cysts with extension in the ventricles or midline, and quadrigeminal cysts can be treated with endoscopic cystectomy, cystocisternostomy, cystoventriculostomy, ventriculocystostomy, and ventriculocystocysternostomy [36]. Even a combination with ventriculostomy may be performed [13].

2.4. Cerebellar infarction

Cerebellar infarction with space-occupying effect might lead to obstruction of the CSF pathway (**Figure 6**). The occurrence of stroke-related occlusive hydrocephalus is 10.9–27.2% of cerebellar stroke patients [37]. Deterioration of consciousness in case of cerebellar infarction can be caused by compression of the brainstem through increasing parenchymal edema or by hydrocephalus. A neurosurgical intervention is required in case of decline of consciousness [7, 37, 38]. For treatment of hydrocephalus, the alternative options are external ventricle drainage (EVD), shunt placement, and ETV. However, not every hydrocephalus caused by cerebellar infarction is a suitable candidate for ETV. In case of patients with severe deterioration of consciousness and/or brain stem compression, an adequate decompression should be performed [37]. In case of hydrocephalus, the high risk for infection in case of long-term ventricle drainage can be avoided with third ventriculostomy. The compression of prepontine cistern usually does not cause any surgical problems; the technique can be performed in the usual way [37]. Regarding the routine intracerebral pressure (ICP) monitoring in case of ETV patients, it is not recommended, as the level of the consciousness is an indicator for the success of ETV, and postoperative CT scan is performed to also control the ventricle size. There are many positive experiences concerning ETV procedures without routine ICP monitoring [12, 37].

2.5. Hemorrhage-related obstructive hydrocephalus

An obstruction of CSF pathway can occur in case of intraventricular or intraparenchymal bleeding with or without intraventricular extension, cerebellar hematoma, or subarachnoid hemorrhage [39, 40]. In case of intraventricular or intraparenchymal bleeding related to obstructive hydrocephalus, ETV can be an alternative to EVD placement (**Figure 7**). However,

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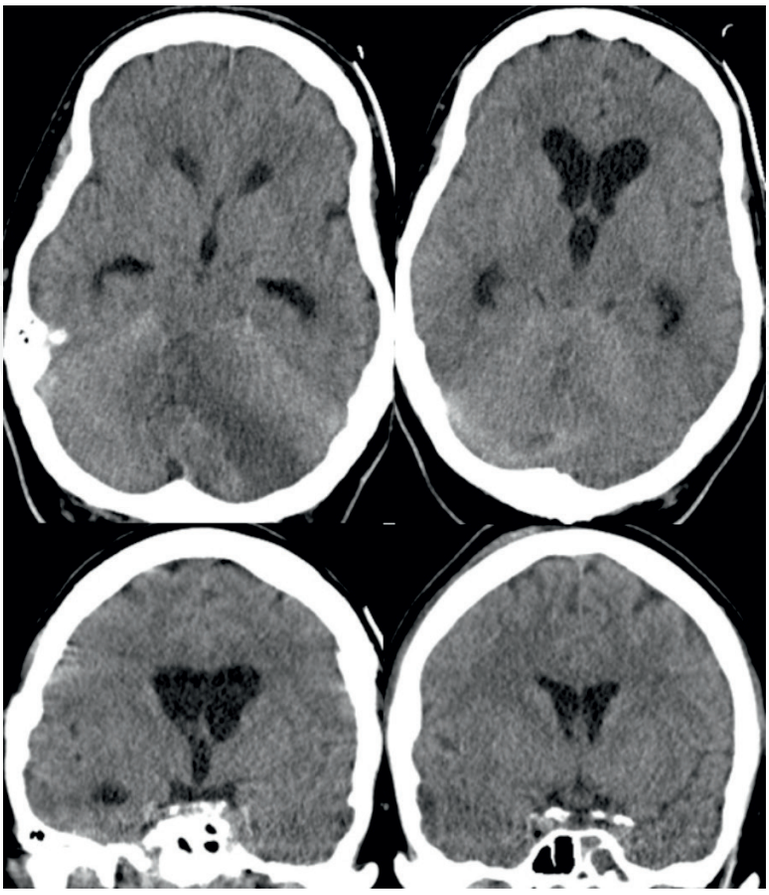

Figure 6. Obstructive hydrocephalus in case of cerebellar infarct (left above and below) preoperative CT scan—dilatation of temporal horn and frontal horn of lateral ventricle and third ventricle (right above and below) postoperative CT scan, reduced size of ventricle system.

only a small percentage of patients with the above-mentioned pathology are optimal candidates for ETV and its indication is rare [7, 39]. The gold standard for treatment of hemorrhage-related acute hydrocephalus remains EVD placement. Acute hydrocephalus can arise either by intraventricular hemorrhage or by intraparenchymal hematoma with space-occupying effect compressing the CSF pathway. There are advantages and disadvantages of preforming an ETV. The endoscopic procedure might reduce the risk of the drainage-related infections and reduce the numbers of surgical procedures, as the drainage has to be changed after 7–10 days. In most cases, it is required to remove the blood clot. With endoscopic procedure, the possibility is provided; however, this procedure can be time-consuming. In case of intraventricular bleeding, the endoscopic view is unclear. The visualization can be normalized through forced irrigation, but in case of a severe hemorrhage, the visual quality remains poor despite prolonged irrigation. The space-occupying intraparenchymal hemorrhage can displace the ventricles depending on the localization. Furthermore, the anatomy of the prepontine space and basilar artery might be distorted increasing the risk of basilar injury. In the acute phase, the floor of the third ventricle is thick in contrast to the subacute and chronic obstruction. This fact likewise causes a higher risk for basilar injury. These findings lead to technical challenges in the endoscopic procedure [39].

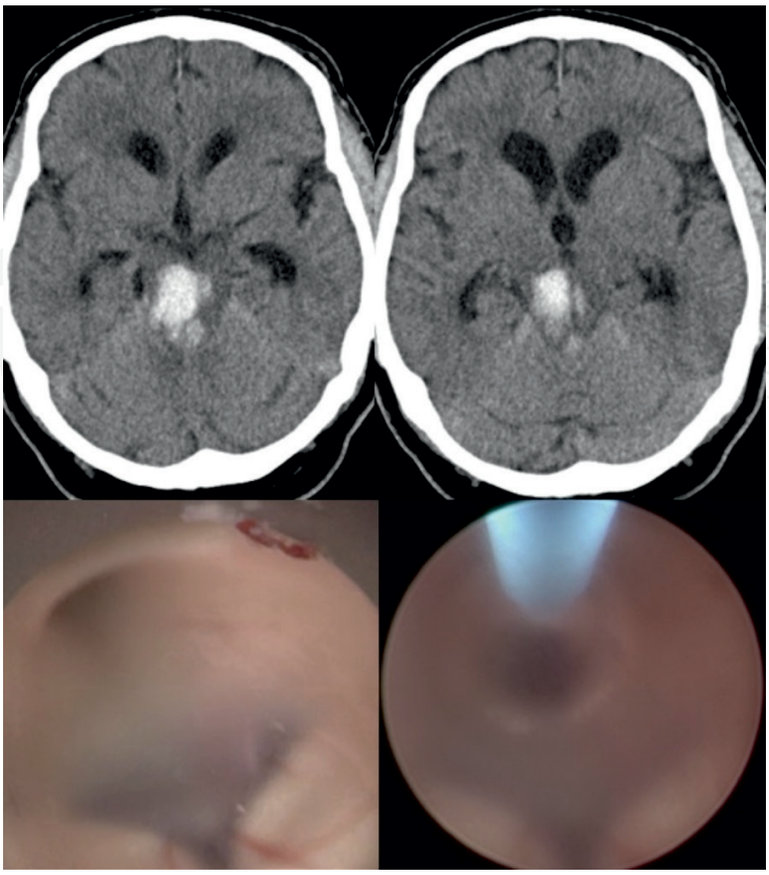

Figure 7. Intraventricular hemorrhage of forth ventricle with occlusive hydrocephalus. Obscured view due to the bleeding (above) and preoperative CT scan (left below) landmarks of third ventricle—infundibular recess, mammillary bodies (right below), balloon dilatation.

The clinical results are primarily influenced by the parenchymal damage. It is recommended to perform an ETV if the requirement of continuous ICP monitoring and liquor drainage for ICP reduction is excluded. In certain cases, where the CSF obstruction plays the dominant role and the hemorrhage does not have significant space-occupying effect, ETV might be the optimal option [39]. In these cases, the clinical condition can be dramatically improved. In all other cases, ETV should be considered if weaning the patients off the drainage is not successful within a week. As mentioned above, the standard treatment remains EVD placement. Thus, in case of doubt, EVD placement should be performed, even leading to an interruption of ventriculostomy [39].

2.6. Infection-related hydrocephalus

Despite the indication in postinfectious hydrocephalus (meningitis, ventriculitis, and shunt infection) with impaired CSF absorption, or ductal, foraminal obstruction is controversial, ETV might be also successful in certain cases. As consequence of the infection, cisternal scarring might occur, leading to higher failure rate [40]. Regarding the etiology in children, the occurring pathogen depends on the age. Hydrocephalus following prenatal infections may be caused by Toxoplasmosis or Cytomegalovirus. In neonatal period, the most frequently occurring bacteria

are Gram-negative bacteria during the first 14 days of age and Gram-positive bacteria after the first 2 weeks of age. Further pathogens may be Candida species at this age of life. In postnatal period, hydrocephalus may be caused by bacterial (such as Haemophilus influenzae type B), viral, or fungal (such as Cryptococcus) infections [41]. In adults, the infection can be bacterial (*Streptococcus pneumoniae*, *Neisseria meningitidis*, *Haemophilus influenzae*, *Listeria monocytogenes*, and group B Streptococcus), viral (most frequently Enteroviruses), or fungal (Cryptococcus species) [42].

The tuberculous meningitis–related hydrocephalus is discussed separately, because of peculiar pathological behavior. It is still a frequent disease in the developing countries [43]. As hematogenous dissemination from primary complex, tuberculous foci might be deposited in the cortex, meninges, choroidal plexus, or ventricular wall, and through rupture into the subarachnoid space, it leads to meningitis [44]. In acute phase, there is an acute inflammation in ventricles with ependymal infection and tuberculous exudate. Although the exudate can be washed out during ETV, ventriculostomy is more difficult due to acute inflammation and anatomical changes [14, 45]. In the chronic phase, the aqueduct can be obstructed by exudates, small tuberculomas, or infection of ependyma. In this phase, the procedure has a higher success compared to the acute phase; however, the floor of the third ventricle is thickened, which could complicate the surgery [40].

2.7. Congenital malformation–related hydrocephalus

Central nervous system (CNS) anomalies might lead to congenital hydrocephalus. The most common cause is the congenital aqueduct stenosis as mentioned above. It can be caused by infection, hemorrhage before birth, or tumors [16]. In this case, endoscopic ventriculostomy is the treatment of choice. Ventriculostomy might be also a surgical alternative in case of Chiari I and II malformation with fourth ventricle outlet obstruction, myelomeningocele, syringomyelia, and Dandy-Walker malformation–associated hydrocephalus [13, 19, 26, 46, 47]. The success of ETV in these cases is diverse.

Hydrocephalus develops in considerable number in cases with neural tube defect [47, 48]. The incidence of hydrocephalus in patients with meningomyelocele increases after closure the defect [48]. Performing the endoscopic procedure may be technically difficult, due to the anatomical variations (enlarged interthalamic adhesion, stenosis, or atresia of foramen of Monro and so on) [47]. Chiari malformation can be concurred either with communicating or with noncommunicating hydrocephalus. Although ETV may be a good alternative for shunt placement in these cases, the success depends highly on the optimal selection of cases for endoscopic procedure [25]. In case of obstructive hydrocephalus, there is a better chance for success. The majority of Dandy-Walker syndrome (DWS) is associated with hydrocephalus [26]. In this case, the primary treatment continues to be controversial. In general, the initial treatment remains the shunt placement. Endoscopic excision of obstructing membrane as initial procedure is not recommended because of the associated risks for morbidity and mortality. ETV may be an alternative for shunt placement in patients with frequent shunt malfunction. In DWS concurred with aqueduct stenosis, the combination of ventricle-cyst stenting and ETV may be performed [26].

2.8. Obstruction due to giant aneurysm

An obstructive hydrocephalus through giant aneurysm is rare, but it has been already reported in some cases [49–52]. Commonly, these cases are treated via shunt placement, although an aneurysm growing after shunt implantation has been already reported. The theory is that the decreased intracranial pressure after shunt placement reduces the tamponade effect, leading to aneurysm expansion and an increase in the risk for rupture [51].

In certain cases, an endoscopic ventriculostomy might be performed [49, 50]. Even though the anatomy is commonly changed as a consequence of the aneurysm (e.g., the third ventricle or aqueduct may be compressed and the anatomical structures as the floor of ventricle and mammillary bodies may be displaced), the landmarks can be identified and the surgery can be performed in the usual way [49].

2.9. Further indications for ETV

Every surgical revision after shunt implantation is considered as shunt failure. Many studies established that endoscopic third ventriculostomy in shunt failures might be an effective treatment option (**Figure 8**). The presence of shunt failure does not exercise influence on the failure rate of ETV. However, the success seems to be diverse in certain etiology groups [14].

It was observed that hydrocephalus patients less than 65 years of age with idiopathic normal pressure, where the dominant symptom was gait disturbances and only minimal cognitive deficits could be observed, had a good success rate [14].

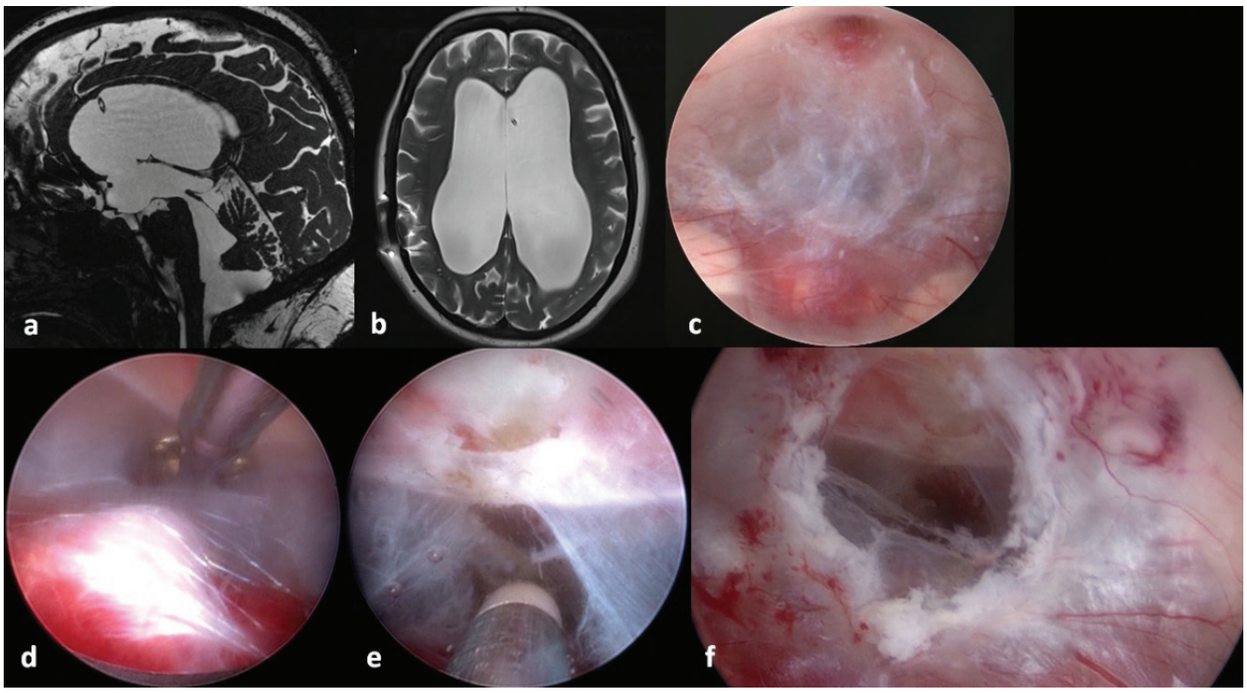

Figure 8. Hydrocephalus with shunt malfunction (a and b) preoperative MRI scan, (c) scarred thick floor of the third ventricle, (d) perforation of adhesions in the prepontine cistern with forceps, (e) with bipolar diathermy, and (f) view on the cisterns through the ventriculostoma.

3. Surgical technique

3.1. Neuroendoscopic equipment

For endoscopic ventricular surgery, either rigid or flexible endoscope can be used. Rigid endoscopes have a higher image quality and allow an easier insertion and handling of instruments. In contrast, the flexible one allows a flexible mobility of the scope, even though producing a lower image quality [53]. Endoscopic equipment for ETV includes various rigid rod lens Hopkins optics and instruments (scissors, hooks, puncture needles, forceps for tumor biopsy and grasping, bright cold Xenon light source, HD video camera system, irrigation device, and Fogarty balloon catheter) [7].

3.2. Surgical technique

The operation is performed under general anesthesia. The patient is placed in supine position, the head in 3-pin fixation and tilted slightly forward. The hair is shaved and the approach is marked. The standard placement of the burr hole is anterior to the coronal suture and 2 cm lateral to the midline. After skin disinfection and sterile draping of the operating field, the scalp is incised in a straight line, about 2–3 cm long. After placement of the burr hole (about 1 cm) and opening the dura, the operating sheath is introduced into the lateral ventricle and the trocar is inserted at about 5 cm depth of dura. The endoscope is fixed and the trocar is removed. The rigid 0° diagnostic optic is inserted for inspection and identification of the main landmarks. In the lateral ventricle, the fornix, the foramen of Monro, and the choroid plexus are identified (**Figures 9** and **10**). Under direct visual control, the endoscope is advanced through the foramen of Monro into the third ventricle. In the third ventricle, the main landmarks are the mammillary bodies and the infundibular recess. The diagnostic inspection is extended with

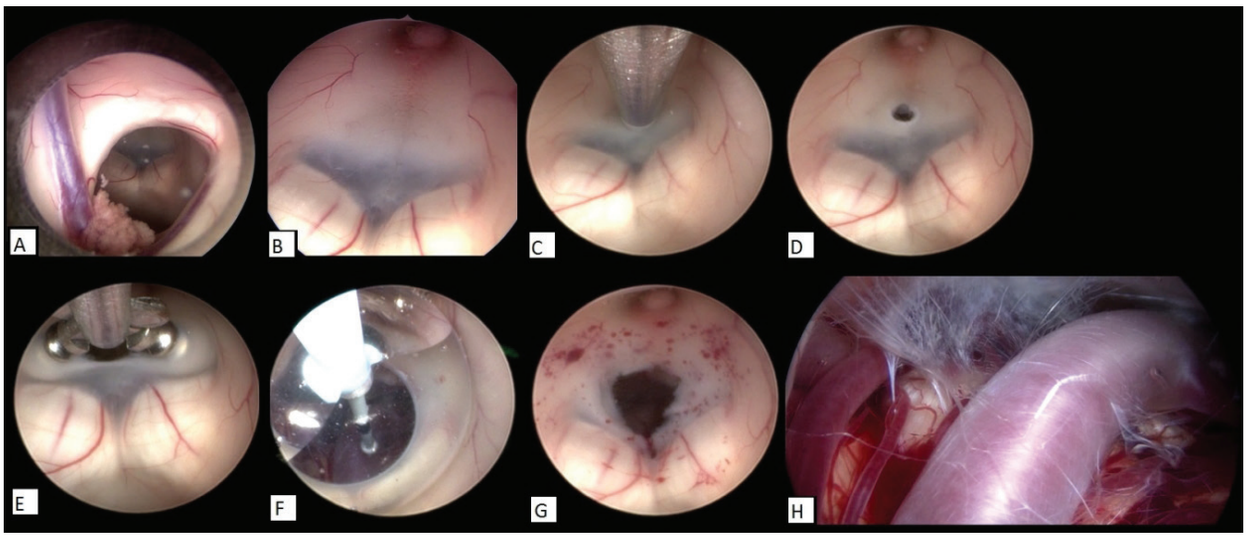

Figure 9. Steps of endoscopic third ventriculostomy. (A) View of foramen of Monro, plexus choroideus, anterior septal vein in lateral ventricle, (B) view of mammillary bodies, infundibular recess in the third ventricle, (C) ventriculostomy with bipolar diathermy, (D) ventriculostoma, (E) enlarging with perforation forceps, (F) dilatation with Fogarty balloon catheter, (G) expanded ventriculostoma, and (H) view of basilar artery.

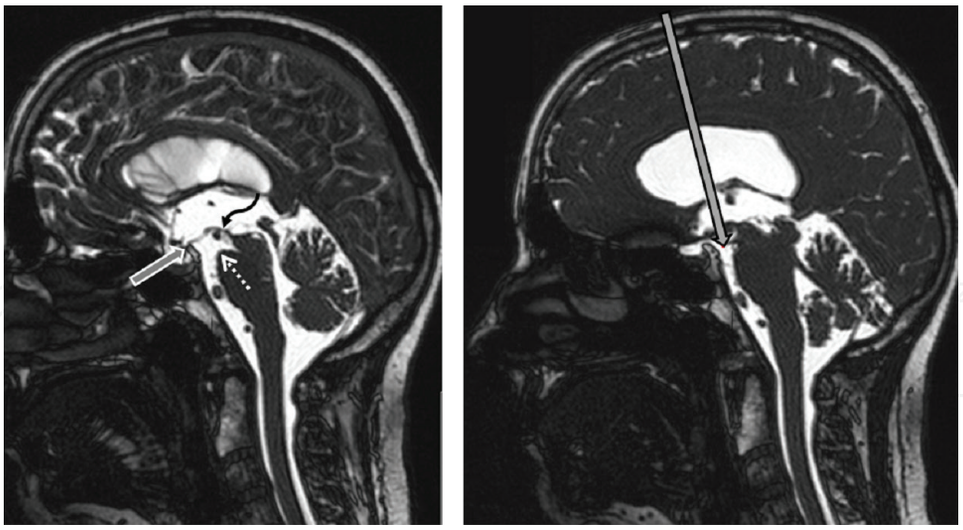

Figure 10. (Left) Landmarks on the MRI scan TRUFI sequence, (L) lateral ventricle, (III.) third ventricle. Infundibular recess (grey arrow), mammillary body (wavy arrow), and interpeduncular and prepontine cistern (dotted arrow). (Right) Entry point.

30°, 45°, and/or 70° optics to view the anterior and posterior parts of the ventricles if applicable. It can be useful in case of obstruction of the aqueduct or tumor. In the midline between the mammillary bodies and the infundibular recess, the perforation is performed. For fenestration, there are several possibilities. The perforation can be performed by sharp perforation with semisharp probe [54], by blunt perforation with the endoscope itself, the balloon catheter itself [8], by coagulation with monopolar or bipolar diathermy [7, 8, 54], and by laser perforation [54, 55], waterjet dissection [55], ventriculostomy forceps [7], or ultrasonic probe [56]. It is recommended to use a blunt perforation to avoid vascular injury [8, 55]. In case of thick ventricle floor, it is recommended to use cauterization with bipolar diathermy at low energy [7, 8], sharp perforation with semisharp catheter [54], or waterjet dissection [55]. However, laser perforation and monopolar diathermy should be avoided [8]. After the perforation by a rigid instrument, enlarging ventriculostomy is initially performed by a perforation forceps and subsequently by inflation of a Fogarty balloon catheter (4–7 mm). After ventriculostomy, an inspection is performed with the 0° diagnostic optic to identify the dorsum sellae and the tip of the basilar artery to ensure free CSF flow. If a Liliequist membrane or subarachnoid adhesions are present, they also should be perforated (**Figure 8**). While withdrawing the operating sheath, an active bleeding at the foramen of Monro or at the corticotomy should be ruled out. After removal of instruments, a gelatin sponge is inserted in the burr hole and the galea is tightly closed to avoid a subgaleal CSF accumulation and leakage. The skin is sutured with clamp or thread. As standard, no EVD is inserted (**Figure 1**) [7, 9].

In case of infants up to 2 years of age, the procedure is performed in the same way; the only exception is that the head is fixed with bandages running over the forehead [57].

In addition, a flexible endoscope can be used for transaqueductal inspection. Following ETV, the work sheath should be moved posteriorly to achieve a straight approach to the Sylvian aqueduct. If the size of diameter of the aqueduct allows it, the working optic can be withdrawn and

the fiberscope inserted. After forwarding the fiberscope through the aqueduct, the patency of outlets of the fourth ventricle can be inspected. The fiberscope is withdrawn and a 0° diagnostic optic inserted to check for contusion or bleeding during the withdrawal of the work sheath [19].

4. Success rate of distinct indications

In case of a successful third ventriculostomy, the symptoms mostly improve soon after surgery. Most of the patients, who suffered from deterioration of consciousness because of obstructive hydrocephalus, show a rapid improvement after the surgery [6]. Of course, it requires an adequate indication and it depends considerably on the underlying pathology [6, 7, 12] (**Table 1**).

Postoperative CT scan is routinely performed; however, the radiological findings alone do not count as an indicator of ETV success. It must be assessed in all cases with the clinical findings [6, 10, 58, 59]. After ETV, the intraventricular pressure does not decrease immediately in certain cases [10]. The ventricle size can increase, decrease, and remain unchanged after ventriculostomy [6]. In most cases, the ventricle size might correlate with the clinical outcome as early as 1 month after the surgery. In successful cases, a significant decrease can be seen, while it is not observable or not significant in ETV failures [58, 60]. Especially, decreasing of the third ventricle size is

Indication	Success rate (%)	References
Aqueduct stenosis	67–93.5	Vulcu et al. [6], Grunert et al. [9], Hellwig et al. [10], Hopf et al. [11], Moorthy et al. [14], Oertel et al. [63], Gangemi et al. [65]
Far distal obstruction	72–76	Oertel et al. [31], Mohanty et al. [26], Warf et al. [47]
Tumors	56–81	Vulcu et al. [6], Oertel et al. [7], Grunert et al. [9], Hellwig et al. [10], Hopf et al. [11]
Cysts	56–95	Vulcu et al. [6], Oertel et al. [7], Hopf et al. [11], Moorthy et al. [14], Ray et al. [27], MacArthur et al. [69]
Cerebellar infarction	86	Oertel et al. [7], Baldauf et al. [37]
Hemorrhage	43–73	Vulcu et al. [6], Oertel et al. [39], Grunert et al. [9], Siomin et al. [40], Roux et al. [73]
Infection	60–64	Mugamba et al. [13], Siomin et al. [40], Oertel et al. [63]
Tuberculous meningitis	60–83	Singh et al. [74], Bhagwati et al. [43]
Congenital malformations	21–80	Wu et al. [25], Mohanty et al. [26], Rei et al. [46], Warf et al. [47]
Giant aneurysm	No adequate data	
Further (e.g., noncommunicating HC)	65–72	Hellwig et al. [10], Moorthy et al. [14], Gangemi et al. [75]

Table 1. Success rate of ETV in distinct indication cases.

reliable [60]. The ventricle size decreases to smaller size as measured preoperatively but remains still bigger than in healthy patients. It shows presumably that the absorptive mechanisms do not work as well as in healthy patients and the successful ventriculostomy provides a compensated communicating hydrocephalus [61]. The postoperative examination can be supplemented by MRI scan. The presence of CSF flow void, which refers to signal loss in the MRI occurring with CSF, may indicate the success [58]. It is important to emphasize that even without any changes in the ventricle size, the patient can improve clinically as a result of successful ventriculostomy.

The overall success rate of ETV is reported at 60–90% [6–12, 62] (**Table 2**). The initial success rate in the early postoperative evaluation is higher than during long-term follow-ups [6, 63]. Different data regarding the success rate can be explained by the different definitions of ETV success and with the fact that lots of studies examined mixed population regarding the age, the underlying pathology, the time course of the follow-up, and the strategy of patient selection [6–8]. Based on the experience and on the data, it is recommended to define the clinical success of third ventriculostomy as needless of re-ETV or shunt placement 3 months after the surgery [6, 63]. However, there is no common agreement regarding the point of time when a ventriculostomy is considered completely functional. In contrast, according to a Canadian experience, the success strictly means "no further CSF diversion procedures" [64].

As many studies have established, the success rate of the procedure depends highly on the underlying pathology and the age of the patients [6, 7]. Furthermore, the outcome is influenced by the indications and the performing neurosurgeon [12]. An ETV Success Score (ETVSS) was developed by Kulkarni et al. to estimate a 6-month outcome taking age and etiology into account. The success score is recommended to be used while selecting the optimal candidate for ETV. Even though ETVSS was initially developed to predict the short-term success, the score likewise correlates with the long-term outcome. The score ranges between 0 and 90, where 0 means extremely poor chance for success and 90 means very good chance for success. Regarding the success rate, the patients might be divided into three groups: high ETVSS-Group ≥80, moderate ETVSS-Group 50–70, and Low ETVSS-Group ≤40. The optimal candidate for ETV seems to be the patient over 10 years of age with obstructive hydrocephalus as aqueduct stenosis or tectal tumor and without infection, cerebral hemorrhage, or previous shunting in case history [5].

4.1. Aqueduct stenosis

Endoscopic ventriculostomy has the most favorable outcome in case of benign aqueduct stenosis (67–93.5%) [6, 7, 9–11, 14, 63, 65]. This fact supports the assumption that the therapy of

Age group	Overall success rate (%)	References
Adults	60–90	Vulcu et al. [6], Schroeder et al. [8], Grunert et al. [9], Hellwig et al. [10], Hopf et al. [11], Sacko et al. [12], Siomin et al. [62]
Children > 2 years	66–71	Vulcu et al. [6], Grunert et al. [9], Baldauf et al. [80], Oertel et al. [81], Beems and Grothenius [82], Etus et al. [83]
Children < 2 years	22–67	

Table 2. Overall success rate considering the age groups.

choice is ETV. Nevertheless, the success rate in each case depends on the etiology, age, clinical findings, and radiological characteristics [13]. The presence of aqueduct obstruction with free prepontine cistern is associated with significantly better outcome of ETV [66]. Regarding the etiology, the acquired stenosis has a better success result than the congenital form [67]. The clinical manifestation of the congenital form appears earlier than the acquired one, which contributes to the lower success rate. In infants, the success of the procedure seems to depend on the age as well as on the etiology. Under the age of 1 year, ETV also has higher success rate in case of aqueduct stenosis than in case of other etiology, although significantly less than in older children [9, 68]. Regarding age, the success seems to be related not only to the age at the time of the surgery but also to the age at onset of the pathological changes [13]. The different range of success rate derives from the fact that the studies examine mixed population regarding age and pathology [16] .

4.2. Far distal obstruction

Obstruction distal to the fourth ventricle is a rare cause of HC, but in any case, ETV might be a successful treatment with 72–76% of postoperative success [19, 26, 47].

4.3. Tumor and cystic lesions

The intraventricular, pineal region, infratentorial tumor- and cyst-related obstructive hydrocephalus has a high success rate after third ventriculostomy (56–81%, in cysts over 90%) [6, 7, 9–11, 63]. The best candidates seem to be patients with aqueduct obstruction in case of tectal tumors, pineal region tumors or cysts, and third ventricle tumors [11, 15, 27]. Simultaneously performing tumor mass resection or biopsy is also possible in certain cases. The success of ventriculostomy with simultaneous tumor removal or biopsy is reported [7, 69]. Another option is to perform ventriculostomy before or after the tumor surgery. There are diverging opinions on the timing [14]. The preceding ventriculostomy may reduce the chance for postoperative hydrocephalus in case of posterior fossa tumors [70, 71]. In case of cysts, a cyst resection or a fenestration between the cyst and the ventricle simultaneously may be performed to ETV. The success rate in these cases reaches 56–95% [6, 7, 11, 14, 27, 69]. After tumor removal, the cause of obstruction can be eliminated restoring the normal CSF flow. Therefore, performing ETV routinely is not necessary [32]. In case of posterior fossa tumors, higher success is established if hydrocephalus persists after tumor resection [72]. On the other hand, there are studies where the rate in case of tumors is significantly lower (56%) [6]. The different data can be explained by the fact that many studies do not differentiate between benign space-occupying lesions and progressive tumors; however, there is a significant difference between the two groups regarding the outcome [11]. The success of ETV depends on the localization and the growth pattern of the tumor. A progressive tumor is more likely to close the ventriculostoma earlier than a benign lesion or cysts. The same applies to a lesion in the third ventricle compared to an infratentorial tumor, which has no direct connection to the new CSF flow diversion. Likewise, the duration of the symptoms seems to be an influencing factor [7]. Regarding the surgical technique, it can be performed in a relatively uncomplicated way, but the obscuration of the ventricle anatomy by the tumor might cause difficulties [10, 14].

4.4. Cerebellar infarction

A cerebellar ischemic stroke might lead to obstruction of CSF pathway through the parenchymal edema. The occlusion can be treated with EVD or ETV depending on the endoscopic experiences of the surgeon. The ideal management is still controversial. An endoscopic ventriculostomy might be recommended if no brainstem compression exists [37]. The overall success of ETV in this case is about 86% [7]. Under certain circumstances, this etiology seems to be ideal for ETV, probably because of the acute onset and the pure obstructive origin [7]. The main indicator for successful ETV seems to be the level of consciousness. In case of no improvement of deteriorated consciousness despite ventriculostomy or in case of brainstem compression, a suboccipital decompression is required [37].

4.5. Hemorrhage-related obstructive hydrocephalus

The optimal management in case of hemorrhage-related obstructive hydrocephalus is still controversial. The overall success rate is 43–73% [6, 9, 39, 40, 63, 73], which is significantly lower than in other etiologies mentioned above. In this case, a difference was also established regarding the outcome in various age groups. The younger population has a lower success rate than the adult group. Moreover, in combination with infection, the rate is about 23% [40]. Evaluating the success of ETV remains difficult in any case, because of the adverse prognoses and clinical status in case of extensive intraparenchymal or intraventricular hemorrhage [39].

4.6. Infection-related hydrocephalus

The overall success rate amounts for about 60–64% in case of obstructive hydrocephalus caused by infection [13, 40, 63], even though adults benefit more from the procedure than children [13, 40]. The success of ventriculostomy depends on whether a prepontine scarring exits. In many case, ETV cannot be performed, because of intraventricular, ependymal scarring and anatomical distortion. In contrast, an obstruction, especially of the Sylvian aqueduct in postinfectious hydrocephalus, promises a better outcome [45]. As mentioned above, infection combined with hemorrhage has a lower success rate [40].

In tuberculous meningitis, the exudate is deposited in basal cisterns leading to an obstruction at the level of Sylvian aqueduct, at the outlet of fourth ventricle, or in the subarachnoid space [14]. ETV success amounts for about 60–83% in this case [43, 74]. The difference of the success depends highly on the thickness of the ventricle floor and the presence of the exudate. In acute phase, the tuberculous exudates in ventricle system and subarachnoid space and the inflammation of ependyma may impede the surgery and lead to lower success rate [14].

4.7. Congenital malformation–related hydrocephalus

Although opinions vary widely about the role of ventriculostomy in congenital CNS malformation–related hydrocephalus, it may be a successful option in certain cases [13, 26, 46, 47]. The overall success rate in case of brain malformation–related hydrocephalus amounts to 21–80% [25, 26, 46, 47]. These various rates may be explained by the fact that most surgeries are performed in infancy, which considerably influences the outcome. Moreover, obstructive hydrocephalus

overall has a higher ETV success rate compared with noncommunicating hydrocephalus. In patients with meningomyelocele, the success may be higher if ETV is combined with choroid plexus cauterization [47]. In case of Chiari malformation type I and syringomyelia-related hydrocephalus, a shunt independency with a high rate of causing a high ETV success may achieved [14]. The reduction of the caliber, even a resolution of the syrinx, was observed [13, 14].

4.8. Obstruction due to giant aneurysm

Successful endoscopic ventriculostomy was reported in some giant aneurysm–related obstruction [49, 50]. The most feared complications are related to vascular injury leading to infarction, hemorrhage, or pseudoaneurysm development [49]. Although, as mentioned above, some successful ETV was reported, where the procedure seemed to be a good option for treatment of obstructive hydrocephalus, the experiences and reports in these cases are wanting.

4.9. Success in cases of further indications for ETV

In case of normal- and low-pressure hydrocephalus and malabsorptive hydrocephalus, the success rate amounts about 65–72% [10, 14, 75]. In patients less than 65 years of age and with communicating hydrocephalus, with only minimal cognitive deficits, where the dominant symptom is gait disturbance, the success rate is comparable with the rate after shunt placement [14].

The success in case of shunt malfunction or infection was observed in similar percentage of 67–80% [14, 76]. An exact indication for ETV in communicating hydrocephalus and shunt malfunction has not been defined yet, but as it can be seen, it may be an alternative in certain cases.

As mentioned earlier, patients with shunt malfunction might be treated successfully with ETV in certain cases as well. It is clear that a patient with aqueduct stenosis benefits more from ventriculostomy following shunt malfunction than patients with other etiologies. Consequently, ventriculostomy can be recommended in case of shunt malfunction if an obstruction exits in the CSF pathway. In contrast, patients without obstruction should be treated with shunt revision [77]. Following ventriculostomy, it is recommended to remove the shunt system to avoid the intermittent CSF diversion through the shunt [14].

4.10. Children

The debate is still open, whether the etiology or the age is the determining factor of ETV success in children. The overall success rate reaches about 66–71% in children [6, 7, 9]. In children under 2 years of age, having the chance for a success is significantly lower (22–67%), and infants under 6 months have the worst chance for restoration of the CSF circulation [6, 7, 9, 78–81]. While trying to predict the success using ETVSS, the age plays an important role [5]. The outcome has been examined in several studies regarding the underlying pathology in children, and the difference was also conspicuous between the various etiology groups [9, 78, 80, 81]. The final results correlate with data in adults. Patients with aqueduct stenosis overall have very favorable outcome [9, 78, 81]. Despite all these data, there are studies where no difference regarding the age [4] or etiology [82] was found. Nevertheless, based on the experiences and studies, the outcome seems to depend both on the underlying pathology and on the age [78, 83, 84].

5. Failure rate and complications

5.1. Complications and their prevention

Endoscopic ventriculostomy is considered less noninvasive and safe. The complications are rare and mostly related to the surgical procedure [85]. The average morbidity is 8.5% and mortality <1% [8, 11, 31, 85]. Permanent deficits are rare with an average value up to 2.38% [85], while the transient morbidity ranged up to 7.8% [8, 85].

The most feared complication is the vessel injury. Minor hemorrhage can be caused by small vessel injuries, for example in the cortex, brain parenchyma, choroid plexus, or margin of ventriculostoma during inflation of the Fogarty catheter. Minor hemorrhage can be managed with irrigation to achieve a tamponade effect [8, 55, 85]. In case of insufficient irrigation, coagulation with bipolar diathermy should be performed [8]. Major hemorrhage caused by injury of basilar artery and basilar perforating vessels, thalamostriate vein, or anterior septal vein injury can lead to uncontrollable hemorrhage and life-tethering situation with fatal outcome. Minor hemorrhage occurs in up to 16.5%, whereas major ones occur only in 0.49% [85]. These may occur for example due to false placement of instruments [7]. Brain nerve injury may occur in the same way [7, 85]. In many diseases such as developmental anomalies and tumors, the anatomy is distorted making the surgical technique difficult [85]. By using a rigid endoscope and correct position of fenestration, these complications can be avoided. Thorough inspection of the anatomical relationships on the preoperative MRI scan is essential.

Further complications may be epidural or subdural hematoma [8, 86–88], fornix contusion [8, 31], traumatic damage of thalamus and hypothalamus with endocrine and electrolyte disturbances [31, 85], hemodynamic alterations [6, 85], cerebral herniation syndrome in case of obstructed outflow channel or excessive irrigation [8, 85], CSF leakage [8, 85], abandoned procedure, or ETV failure (see below) [6, 12, 62, 63, 89]. The incidence of abandoned procedure ranges from 0 to 26% [8]. The most frequent causes are hemorrhage, complex anatomical circumstances, or inability to fenestrate the ventricle floor [8]. Postoperative infection may lead to meningitis, ventriculitis, brain abscess, or sepsis. Further nonspecific complication as thrombophlebitis, pneumonia, or wound infection may occur.

Summarizing, to prevent the complications, individual anatomical variations should be evaluated in the preoperative imaging. The relationship between the third ventricle floor and basilar artery should be thoroughly considered. The adequate technique with rigid instrument should be used and one has to consider the correct energy intensity and sources. As mentioned above, it is recommended to use low energy (maximum 10 W) and to avoid laser and monopolar diathermy. The correct placement of fenestration is essential; ideally, it is placed halfway between the mammillary bodies and infundibular recess in the midline. However, individual variations should be considered. After the perforation, the interpeduncular and prepontine cistern should be inspected and adhesions or the Liliequist membrane should be perforated.

5.2. Failure rate and re-ETV

Re-occlusion of the ventriculostoma is rare. Kulkarni et al. have reported that ETV has an initial higher failure rate in contrast to shunt placement, although it gets lower over time [5]. Most failures (58–97%) occur within the first 3 months [6, 7, 12, 62]. Within the first year, ETV failure reaches 16–20% [6, 62]. However, it might also develop after years; therefore, a long-term follow-up after the surgical procedure is required [12, 62, 89]. It remains controversial, whether re-ETV or shunt placement is the optimal choice of treatment for ETV failure. The decision depends on the age, etiology of hydrocephalus, imaging finding, and the duration between ventriculostomy and failure [63, 90]. In certain cases, repetition of ventriculostomy as the first choice is recommended, as a similar long-term success of 51–89% is expected [7, 16, 62, 63, 89, 91–94]. Some authors differentiate early failure from late failure, but the distinction between the two definitions is various (7 days—6 months) [4, 63, 89, 91, 93]. Multifactorial etiology including malabsorptive component, complications including hemorrhage and infection or technical failure as inadequate size of stoma with insufficient CSF flow through the stoma, and unperforated Liliequist membrane could be responsible for early failure [4, 63, 89, 93, 94]. The ideal size of stoma is not defined, and it seems to depend highly on the anatomy, while in case of a size between 4 and 7 mm, failures were also reported [89]. To avoid an intermitting CSF diversion through the shunt, ligature or removal of the shunt system is recommended, as mentioned above. Late failure could be caused by secondary closure as tumor progression, gliosis, and developing of membranes and adhesions in the subarachnoid space [4, 6, 11, 89, 91]. In children, the absorption of CSF at the Pacchionian granulations is immature [95], which may play a role in ETV failure. Further cause for failure might be the multifactorial etiology in certain cases [4]. The success of re-ETV is found higher in case of late-repeat-ETV than in early ones [63, 91, 96], although there are some controversial results [62]. As indicated above, the failure rate decreases during the follow-up [63]. Regarding the age, the results are controversial, but in most studies, higher failure rates are reported when less than 6–24 months of age [4, 63, 89]. Regarding the pathology, the failure rate amounts 10–20% in aqueduct stenosis, while in other indication cases, it amounts up to 50% [14]. If no obstructive cause for failure is seen on the MRI scan, a shunt placement is considered [63] taking a multifactorial etiology including malabsorptive mechanism as cause of failure. Re-ETV is also not recommended in case of tumor progression–related closure [89]. There are no exact criteria defined for repetition of ETV; an individual evaluation in all cases is expressly recommended.

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