Y-shaped Ventriculo-peritoneal Shunt for Adult Complicated Hydrocephalus: Report of 28 Illustrative Cases

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Research

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Abstract

Objective

To investigate the effectiveness of Y-shaped ventriculo-peritoneal shunt (VPS) (one abdominal tip connected with double or triple ventricular tips by one or two Y-shaped connectors) in the treatment of adult complicated hydrocephalus, we analyzed the long-term outcome and reported some illustrative cases.

Methods

Among 1,100 VPS surgeries between 2012 and 2017 in neurosurgery of Beijing Tiantan Hospital, twenty-eight (2.5%) adult patients with multiloculated hydrocephalus treated with Y-shaped shunt were analyzed.

Results

Nineteen patients underwent Y-shaped VPS (bilateral frontal horn shunt) before or without tumor resection/stereotactic biopsy, 7 patients underwent Y-shaped VPS after tumor resection (5 bilateral and 2 triple shunts), and the other 2 patients underwent sequential bilateral VPS (unilateral VPS with additional contralateral ventricular tip by Y-shaped connector 6 months later). The one-year and two-year hydrocephalus-free survival rate for was both 88.7%. The one-year and two-year overall survival rate was both 66.7%. Cox regression confirmed that the OS is correlated with tumor grades.

Conclusions

Y-shaped VPS strategy is an easy and reliable option for multiloculated hydrocephalus, which can be used as the first choice for some indications.

Introduction

Hydrocephalus is one of the complications after neurosurgical manipulation, hemorrhage or ventriculitis. Most can be easily resolved by endoscopy or unilateral ventriculo-peritoneal shunt (VPS). However, hydrocephalus in some cases, such as multiloculated hydrocephalus (MLH) [1], extra-ventricular hydrocephalus, and hydrocephalus with temporal horn entrapment, cannot be easily resolved by endoscopy or unilateral VPS. Most MLH cases have been reported in the pediatrics [2]. Such complicated hydrocephalus can be occasionally developed in adult patients with intracranial tumors involving bilateral Foramen of Monro or an isolated fourth ventricle obstruction [3]. Although cerebrospinal fluid pathway can be restored by tumor resection in most cases, hydrocephalus may still exist in some cases due to brain edema or adhesion after surgical manipulation. Besides, some patients with tumor involving bilateral foramen of Monro cannot accept tumor resection due to the high surgical mortality and morbidity. Therefore, it is necessary to resolve such complicated hydrocephalus before or after tumor resection.

A consensus of surgical strategy for complicated hydrocephalus has not yet been reported. Current treatment methods included microsurgical fenestration of separate compartments by endoscopy or craniotomy, shunt surgery with multiple catheters placed in the compartments, or combinations of these modalities [1]. Each method has its advantage and disadvantage due to the individual anatomic complexity and CSF hydrodynamics in each case. From 2012 to 2017 in our hospital, 28 adult patients with complicated hydrocephalus were effectively treated with Y-shaped VPS (one abdominal tip connected with double or triple ventricular tips by one or more Y-shaped connectors). In some cases with diffuse midline glioma involving bilateral thalamus, or hydrocephalus with temporal horn entrapment, Y-shaped shunt strategy was easy, reliable and preferred as first choice. In this study, we reported the long-term outcome of Y-shaped VPS in 28 illustrative cases and proposed the optimal indication for such shunt strategy.

Materials And Methods

Patient characteristics and record of clinical and radiological material

Hydrocephalus was defined as abnormal accumulation of cerebrospinal fluid in the ventricles or cavities of the brain. The clinical, radiological data and operation records of 28 adult patients were retrospectively reviewed. The recorded information included patient age, gender, pathological or clinical diagnosis, tumor location and size, surgical outcomes, complications and survivals. Overall survival (OS) was defined as the duration between VPS and death or the last follow-up, and the hydrocephalus-free survival (HFS) was defined as the duration between VPS and shunt dysfunction or the last follow-up. Y-shaped connector (Medtronic) was shown in Figure 1D. Double or triple ventricular tips were connected by one or two Y-shaped connectors, as was illustrated in Figure 1D and 1E. The studies involving human participants were reviewed and approved by Ethics Committee of Beijing Tiantan Hospital. Written informed consent to participate in this study was provided by their own or by the participants' legal guardian/next of kin.

Statistical analysis

To select parameters associated with long hydrocephalus-free and overall survival, the Kaplan-Meier survival analysis and Cox regression model were used. All the statistical analyses were performed using R version 4.0.2 software (https://www.r-project.org). Statistical significance was defined as a two tailed \( p < 0.05 \).

Results

Incidence and clinical features of complicated hydrocephalus

From 2012 to 2017, about 54,000 neurosurgical operations were performed in Beijing Tiantan Hospital, and VPS was performed in 1100 (2.0%) patients with hydrocephalus before or after tumor resection. Among VPS patients, 28 (2.5%) adult patients with complicated hydrocephalus were surgically treated with Y-
shaped VPS. Clinical data are summarized in table 1 and table 2. The age of onset ranged from 17 to 68 years; the mean age was 37±14 years (all means are expressed ± SD). These patients included 16 males and 12 females. Pathology was available in 16 cases. Tumors were diagnosed as craniopharyngioma in 8 cases, glioma in 7 cases, pituitary adenoma in 3 cases, colloid cyst in 2 cases, pineal parenchymal tumor of intermediate differentiation in 2 cases, malignant teratoma in 2 cases, central neurocytoma in 1 case, epidermoid cyst in 1 case, meningioma in 1 case, and pinealocytoma in 1 case.

Radiological features of complicated hydrocephalus

Tumor was located in the 3rd ventricle involving Foramen of Monro in 8 cases, 6 cases in the suprasellar region involving the 3rd ventricle, 4 cases in the sellar region involving the 3rd ventricle, 4 cases in the pineal region, 2 cases in the bilateral thalamus and hypothalamus, 1 case in the right ventricle, 1 case in the trigone of the left ventricle, 1 case in the right temporal lobe, and 1 case in bilateral ventricles and 3rd ventricle. Hydrocephalus before tumor resection/stereotactic biopsy was identified in 24 cases, including 23 bilateral and 1 unilateral. Hydrocephalus after tumor resection was in 9 cases, including 5 bilateral, 2 triple and 2 unilateral (contralateral hydrocephalus after unilateral VPS).

Surgical technique of Y-shaped shunt

Figure 1A-1C illustrates the position and the shape of skull incisions for bilateral frontal horn shunts. Two symmetrical incisions (3cm) are 2.5cm parallel to the midline, with the midpoint 2.5cm from the hairline. One incision (3cm) was on the parietal protuberance along with the temporal line. Figure 1D and 1E illustrates the main procedure of connecting two or three catheters to the Y-shaped connector, respectively. The procedure was as the following. a. puncture the entrapped ventricular apartments and subcutaneously guide the catheters to the incision on the parietal protuberance. b. abdominal skin incision and guide the peritoneal catheter to the incision on the parietal protuberance. c. connect the Y-shaped connector to the peritoneal catheter. d. then connect the Y-shaped connector to the ventricular catheters, avoiding folding of the catheters and connectors. e. make sure the smooth CSF drainage and place peritoneal catheter into the peritoneal cavity.

Y-shaped shunt for complicated hydrocephalus before or after tumor surgery

The treatment chart was shown in Fig 2. Of 24 cases with pre-op. hydrocephalus, 19 underwent Y-shaped VPS before or without tumor resection/stereotactic biopsy. All the ventricular tips were in the bilateral frontal horns. Tumor resection was performed in 12 cases, stereotactic biopsy in 4 cases, and no surgery in 12 cases. 7 patients underwent Y-shaped VPS after tumor resection, including 5 with double and 2 with triple ventricular tips. In 5 cases with double ventricular tips, bilateral frontal horns were punctured in 2 cases, frontal and temporal horns in 2 cases, frontal horn and subdural in 1 case. In 2 cases with triple ventricular tips, bilateral frontal horns with unilateral occipital horn were punctured in Case 6 and bilateral frontal horns with unilateral temporal horn were punctured in Case 7. Two patients underwent sequential Y-shaped VPS with duration of 6 months (Case 17 and 19). Case 17 without tumor resection suffered from contralateral hydrocephalus 6 months after unilateral VPS. Case 19 suffered from contralateral hydrocephalus 6 months after tumor resection and unilateral VPS. Contralateral ventricular tip was added to the original shunt by a Y-shaped connector and bilateral hydrocephalus was successfully resolved. Besides, programmable and anti-siphon shunt was chosen for 25 patients, and fixed shunt for 3 cases.

Surgical outcomes and complications

Patients’ status was improved in 25 cases and worsened in 3 cases. 4 patient received radiotherapy and chemotherapy, 1 received radiotherapy only, 2 received gamma knife, and 2 received stereotactic aspiration and inner radiotherapy. The other patients did not receive any other treatment except VPS. The Karnofsky performance status (KPS) at the last follow-up ranged from 50 to 100, with the median of 90. Patients were followed 0.5-58.0 months. 7 patients died of tumor progression or recurrence, 1 died of pulmonary infection, and 1 died of the shunt obstruction. VPS associated complications included intracranial infection and the shunt obstruction. VPS associated intracranial infection was found in one (3.6%) patient. After antibiotic treatment, the infection was cured and a revision surgery was not needed. Obstruction of the shunt was found in one (3.6%) patient and a revision surgery was performed. Inappropriate location was found in one (3.6%) patient.

Clinical factors correlated with surgical outcome and survivals

Kaplan-Meier analysis of HFS and OS were shown in Figure 3A. The one-year and two-year HFS rate was both 88.7%. The one-year and two-year OS rate was both 66.7%. Patients’ age (>40/≤40), gender (Male/Female), tumor grade (WHO I/II), hydrocephalus before tumor resection (yes/no), VPS before tumor resection (yes/no), tumor resection (yes/no), hydrocephalus after tumor resection (yes/no), VPS associated intracranial infection (yes/no) were recorded and analyzed. Parameters significantly correlated with OS were identified using the log-rank test and Cox’s regression model (table 3). Univariate analyses revealed that elder patient age, hydrocephalus before tumor resection and lower tumor grade were significantly correlated with longer OS (Figure 3B). Multivariate analysis revealed only lower tumor grade was an independent prognostic factor for longer OS (OR: 0.147, 95%CI: 0.030-0.714, P=0.017).

Illustrative cases

Case 24 (diffuse midline glioma)

A 24-year old female was admitted in our neurosurgical emergency, who complained with continuous vertigo and vomit for 1 week. Neurological examination revealed optic disc edema. CT revealed bilateral thalamic hypo-density lesion occupying the Foramen of Monroe with hydrocephalus (Fig. 4A). MR images revealed hydrocephalus caused by hypothalamic and bilateral thalamic lesions with long T1 and long T2 signals as well as partial enhancement (Fig. 4B-F). Bilateral Y-shaped VPS was performed to resolve hydrocephalus with programmable and anti-siphon shunt (Fig 4G). Then, stereotactic biopsy was performed to elucidate the pathology as diffuse midline glioma (Figure 4H, WHO IV, IDH wild-type) with H3K27M (+++), IDH1/2 wild-type, MGMT promoter methylation (-), and EGFR mutation and amplification (-). Immunohistochemical staining showed GFAP (++), Oligo-2 (+++), ATRX (+), P53 (+), and Ki-67 (10%). The patients
underwent radiotherapy, concurrent and adjuvant temozolomide chemotherapy. MR images revealed tumor progression and spread 6 months after shunt (Fig 4H-M), and the patient died 2 months later.

**Case 26 (diffuse midline glioma)**

A 17-year old female was admitted in our emergency, who complained with continuous vertigo and vomit for 2 weeks. Neurological examination revealed optic disc edema. CT revealed a lesion with bilateral thalamic hypodensity occupying the Foramen of Monro with hydrocephalus (Fig. 5A-B). Bilateral Y-shaped VPS was performed to resolve hydrocephalus with programmable and anti-siphon shunt (Fig 5C). Post-VPS MR images revealed hypothalamic and bilateral thalamic lesions with long T1 and long T2 signals as well as mild enhancement (Fig. 5D-K). Then, stereotactic biopsy was performed to elucidate the pathology as diffuse midline glioma (WHO IV, IDH wild-type) with H3K27M (++) and MGMT promoter methylation (-). Immunohistochemical staining showed GFAP (+++), Oligo-2 (+), ATRX (+++), P53 (+++), and Ki-67 (2%). The patients underwent radiotherapy, concurrent and adjuvant temozolomide chemotherapy. 15 months after shunt, MR images revealed tumor shrinkage (Fig 5L-Q).

**Case 21 (post-operative multiloculated hydrocephalus)**

A 19-year-old male was admitted to our hospital with the radiological diagnosis of central neurocytoma in the bilateral ventricles and obstructive hydrocephalus. He complained of intermittent headache for 1 year, intensified headache with nausea and vomiting for 2 months. Neurological examination revealed optic disc edema. CT revealed a lesion with mixed density in the bilateral ventricles involving the Foramen of Monro (Fig. 6A). MR images revealed a lesion with mixed T1 and T2 signals as well as heterogeneous enhancement (Fig. 6B-D).

The patient underwent a right frontal trans-callosal approach for total removal of the tumor. During the operation, the tumor was found to be a grey-red soft mass in the bilateral ventricles. Postoperative pathology confirmed the diagnosis of central neurocytoma (WHO grade II). One week after surgery, the patient had intermittent fever, and the CSF analysis demonstrated intracranial infection. Combined Vancomycin and meropenem plus lumbar cistern drainage were used for treatment. About 5 weeks after the surgery, the patient still had cognitive difficulties, and an emergency CT scan revealed the right entrapped temporal horn (Fig. 6E). The patient underwent puncture and drainage of the trigone of the right lateral ventricle (Fig. 6F). However, about 9 weeks after the primary surgery, the MR scan showed the formation of an abscess in the surgical field (Fig. 6G). The abscess disappear after puncture and external drainage guided by the ultrasound (Fig. 6H). About 11 weeks after the primary surgery, the patient still had cognitive difficulties, and her left limbs were still weak. An emergency CT scan revealed bilateral hydrocephalus with right entrapped temporal horn (Fig. 6I-J). The patient underwent Y-shaped VPS with triple ventricular catheters and low-pressure anti-siphon shunt (Fig. 6K-L). After shunt, the patient was discharged for the rehabilitation therapy. The patient recovered well 1 year later (Fig. 6M-N).

**Case 20 (post-operative extra-ventricular hydrocephalus)**

A 52-year-old Male was admitted in our hospital for headache and vomit for 2 weeks. Radiological images revealed a mass in the third ventricle with left obstructive hydrocephalus (Fig 8A-D). Considering the risk of tumor resection, the patient refused tumor resection and received left frontal horn VPS to resolve hydrocephalus (Fig 8E). 6 months later, the patient was re-admitted in our hospital for headache and incontinence. MR and CT scan revealed right obstructive hydrocephalus (Fig 8F-I). To resolve the hydrocephalus, right frontal ventricular tip was added to the original shunt by a Y-shaped connector. The bilateral hydrocephalus was well resolved (Fig 8J). This is an illustrative case with sequential bilateral VPS with Y-shaped connector.

**Discussion**

Complicated hydrocephalus is defined as multiloculated ventricular compartments with or without extra-ventricular hydrocephalus due to surgical manipulation, ventriculitis, or tumor involving Foramen of Monro before or after tumor resection. Such rare cases cannot be easily resolved due to the complex of anatomy and CSF dynamics. The goal of treatment is to restore communication between isolated intraventricular compartments [2]. The existing evidence at case series level seems to favor neuro-endoscopy as the first line of treatments over traditional shunt surgery [3]. However, VPS is still the treatment of choice in communicating hydrocephalus or in patients after the failure of endoscopic fenestration [4]. Besides, the decision on what treatment to apply should be based on individual anatomy, underlying pathology, available techniques, and the experience of the treating surgeon [5]. In this study, we reported the long-term outcome of Y-shaped branched VPS for complicated hydrocephalus in adult patients.
According to the pathogenesis, complicated hydrocephalus can be divided into two major categories. One was caused by tumor occupying the Foramen of Monro without tumor resection. The other one was ventricular entrapment or extra-ventricular hydrocephalus due to surgical manipulation or ventriculitis. Emergent Y-shaped VPS before tumor resection is an optimal palliative treatment if tumor resection was not applicable or if the patient refused tumor resection to avoid high surgical mortality or morbidity, such as diffuse midline glioma from bilateral thalamus (Case 24 and Case 26) or choroid glioma in the third ventricle (Case 7 and Case 8). For the two adult patients with diffuse midline glioma [6], maximal safe resection was not feasible. Y-shaped shunt was chosen to resolve the hydrocephalus due to the occlusion of bilateral Foramen of Monro. Unilateral VPS with endoscopic septostomy of the septum pellucidum is an option for bilateral hydrocephalus. However, re-occlusion may occur due to tumor growth and/or ventricular shrinkage. Both patients received radiotherapy plus concurrent and adjuvant temozolomide chemotherapy. Case 24 with higher Ki-67 index (10%) died of tumor progression 6 months after VPS. Case 26 with lower Ki-67 index (2%) died of tumor progression 37.5 months after VPS. Chordoid glioma in the third ventricle is a rare disease with 32% mortality in the immediate postoperative period [7]. Gross-total resection was performed in Case 8. This patient suffered from multiloculated hydrocephalus after surgery and underwent Y-shaped VPS. However, she died of severe pulmonary infection 1 month after VPS. The overall survival of Case 7 was one year after VPS, who refused tumor resection and received radiotherapy and chemotherapy.

Malignant teratoma in the pineal region was also the indication for Y-shaped VPS [8], in spite of not involving bilateral Foramen of Monro (Case 12 and 27). Rapid growth of malignant teratoma would occlude the Foramen of Monro and caused bilateral hydrocephalus soon after unilateral VPS. So, Y-shaped VPS was recommended to prolong the HFS for patients with malignant teratoma in the pineal region. Besides, craniopharyngioma involving the third ventricle was also the indication for Y-shaped VPS (Case 1, 2, 10, 11, 16, 17, 23, 25). For acute hydrocephalus, Y-shaped VPS has its advantage in prolonging HFS and decreasing the medical costs. After resolving hydrocephalus, tumor resection for solid mass or stereotactic aspiration with inner radiotherapy for the cysts would be good options for safety.

Y-shaped shunt is indicative and reliable for post-operative multiloculated hydrocephalus with/without extra-ventricular hydrocephalus. Hydrocephalus after tumor resection is more complicated than that before tumor resection. Branched shunts were easier and reliable for hydrocephalus with extra-ventricular hydrocephalus (Case 20) or temporal horn entrapment (Case 8, 13, 18, 21, 28). While endoscopy fenestration is better for bilateral hydrocephalus due to the occlusion of foramen of Monro. For temporal entrapment, VPS has historically been recommended, while endoscopic fenestration of the choroidal tissue has varied success and is limited by technical feasibility and surgeon experience [5,9]. Compared with unilateral VPS, Y-shaped VPS has its advantage in prolonging HFS. And in successive bilateral hydrocephalus, Y-shaped VPS has its advantage in balancing the pump pressure and reducing complications compared with double separated VPS. When implementing Y-shaped VPS, choosing the ventricular catheter entry site was very important. Frontal entry was recommended for bilateral hydrocephalus without temporal horn entrapment [10]. Temporal or occipital entry is only suitable for the entrapment of temporal or occipital horn. For proper localization of the ventricular tips, neuro-navigation or intraoperative ultrasound can be helpful. Besides, pressure adjustable shunt was recommended.

Most deaths were due to tumor progression instead of VPS. Multivariate analysis revealed only lower tumor grade to be an independent prognostic factor for longer OS (OR: 0.150, 95%CI: 0.031-0.728, P=0.019). Seven patients died of tumor progression or recurrence, 1 died of pulmonary infection, and 1 died of the shunt obstruction. The one-year and two-year HFS rates were both 77.4%. The one-year and two-year OS rates were both 65.3%. Therefore, this strategy for complicated hydrocephalus was reliable. The incidence of infection as a result of the shunt implantation in the literature varies widely from 0.17% to 13.2% [4]. Consistent with previous reports, the rate of infection in our study was 3.6%. Shunt dysfunction is another typical and common complication. Mechanical obstruction is the most common shunt complication with higher incidence3. Our results showed lower rate of obstruction (3.6%) and prolonged hydrocephalus-free patent.

There were limitations to this study. This was a retrospective review of a rare series of adult cases with complicated hydrocephalus. This series included a relatively small patient group. Results should be interpreted cautiously, and more and larger patient group study will be needed in the future.

Conclusions

Y-shaped VPS is indicated for hydrocephalus with occlusion of bilateral Monro foramen, extra-ventricular effusion or temporal/occipital horn entrapments. It is a reliable strategy for complicated hydrocephalus and can be used as the first choice for some indications, or as the salvage after the failure of endoscopic fenestration. Most patients can be benefit from this strategy with prolonged hydrocephalus-free survival, and the lower tumor grade was the only risk factor of longer overall survival.

Abbreviations

VPS: Ventriculo-peritoneal shunt; MLH: Multiloculated hydrocephalus; OS: Overall survival; HFS: Hydrocephalus-free survival; CSF: Cerebrospinal fluid; KPS: Karnofsky performance status

Declarations

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Authors’ contributions
XHR and CWY designed the study. XHR, CWY drafted the manuscript. CWY, XRL and YGW collected and interpreted the data. XHR carried out the statistical analysis. SL critically revised the manuscript. All authors read and approved the final manuscript.

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Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Ethics approval and consent to participate

Reviewed and approved by Ethics Committee of Beijing Tiantan Hospital, written informed consent to participate in this study was provided by their own or by the participants' legal guardian/next of kin.

Consent for publication

The patient gave their written informed consent for the publication of their data.

Competing interests

No potential conflicts of interest were disclosed for all authors.

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Tables

TABLE 1: Clinical, radiological, and pathological features of 28 patients with complicated hydrocephalus
| Case No. | Age (yrs), Sex | Pathological or clinical diagnosis | Location | Pre-op. hydrocephalus | Pre-op. V-P shunt | Tumor removal | Post-op. hydrocephalus | Post-op. V-P shunt | Other treatment | OS (months) | KPS last follow up |
|----------|----------------|----------------------------------|----------|-----------------------|------------------|---------------|-----------------------|------------------|----------------|-------------|------------------|
| 1        | 18, M          | Craniopharyngioma                | Sellar region involving the 3rd ventricle | Yes            | Yes                | No            | No                    | No               | Stereotactic aspirin and inner radiotherapy | 51.0 (alive) | 100              |
| 2        | 36, M          | Recurrent craniopharyngioma      | Sellar region involving the 3rd ventricle | Yes            | Yes                | No            | No                    | No               | Gamma knife     | 7.0 (death)  | 0                |
| 3        | 38, M          | Pituitary adenoma                | Sellar region involving the 3rd ventricle | Yes            | Yes                | Yes           | No                    | No               | None            | 43.0 (alive) | 90               |
| 4        | 20, F          | Glioma                           | The 3rd ventricle involving Foramen of Monro | Yes            | Yes                | No            | No                    | No               | None            | 1.0 (death) | 0                |
| 5        | 43, M          | Colloid cyst                      | The 3rd ventricle involving Foramen of Monro | Yes            | Yes                | No            | No                    | No               | None            | 34.5 (alive) | 90               |
| 6        | 21, M          | Gliosarcoma                      | The right ventricle                      | No             | No                 | Yes           | Yes                    | Yes (20 days) | None            | 1.0 (death) | 0                |
| 7        | 50, F          | Chordoid glioma                   | The 3rd ventricle involving Foramen of Monro | Yes            | Yes                | No            | No                    | No               | Radio- and chemo-therapy | 12.0 (death) | 0                |
| 8        | 35, F          | Chordoid glioma                   | The 3rd ventricle involving Foramen of Monro | No             | No                 | Yes           | Yes                    | Yes (1 month) | None            | 1.0 (death) | 0                |
| 9        | 57, F          | Pinealocytoma                     | The 3rd ventricle involving Foramen of Monro | Yes            | Yes                | No            | No                    | No               | Gamma knife     | 29.0 (alive) | 100              |
| 10       | 42, M          | Craniopharyngioma                | Sellar region involving the 3rd ventricle | Yes            | Yes                | Yes           | No                    | No               | None            | 30.0 (alive) | 100              |
| 11       | 49, M          | Craniopharyngioma                | Sellar region involving the 3rd ventricle | Yes            | Yes                | Yes           | No                    | No               | None            | 30.0 (alive) | 90               |
| 12       | 28, M          | Malignant teratoma                | The 3rd ventricle involving Foramen of Monro | Yes            | Yes                | No            | No                    | No               | Radio- and chemo-therapy | N/A         | N/A             |
| 13       | 18, M          | Glioblastoma                     | Trigone of the left ventricle            | No             | No                 | Yes           | Yes                    | Yes (20 days) | None            | 0.5 (death) | 0                |
| 14       | 54, F          | Pituitary adenoma                 | Sellar region involving the 3rd ventricle | Yes            | Yes                | No            | No                    | No               | surgery         | 21.0 (alive) | 50               |
| 15       | 34, M          | Colloid cyst                      | The 3rd ventricle involving Foramen of Monro | Yes            | Yes                | No            | No                    | No               | None            | 22.0 (death) | 90               |
| 16       | 49, F          | Craniopharyngioma                | Sellar region involving the 3rd ventricle | Yes            | Yes                | No            | No                    | No               | None            | 21.0 (alive) | 100              |
| 17       | 52, M          | Craniopharyngioma                | The 3rd ventricle                        | Yes            | Yes                | No            | Yes (6 months)         | None            | 1.0 (death) | 0                |
TABLE 2: Clinical characteristics in 28 patients with Y-shaped shunt

| Patient | Age | Gender | Diagnosis                  | Location of Disease | Foramen of Monro | Sellar region involving the 3rd Ventricle | Right temporal lobe | Outcome | Months | Treatment                  |
|---------|-----|--------|----------------------------|---------------------|------------------|------------------------------------------|---------------------|---------|--------|---------------------------|
| 18      | 42  | F      | Meningioma                | Pineal region       | Yes              | No                                        | Yes                 | Yes     | 17.0 (alive) | None                     |
| 19      | 52  | M      | Pineal parenchymal tumor of intermediate differentiation | Pineal region | Yes | Yes | Yes | Yes | Yes (6 months) | None | 14.5 (alive) |
| 20      | 20  | M      | Pineal parenchymal tumor of intermediate differentiation | Pineal region | Yes | No | Yes | Yes | Yes (2 months) | Radiotherapy | 17.0 (alive) |
| 21      | 23  | F      | Centrocytoma              | Bilateral and the 3rd ventricle involving Foramen of Monro | Yes | No | Yes | Yes | Yes (3 months) | None | 16.0 (alive) |
| 22      | 68  | M      | Pituitary adenoma         | Sellar region involving the 3rd ventricle | Yes | Yes | No | No | No | None | 12.0 (alive) |
| 23      | 43  | F      | Craniopharyngioma         | Sellar region involving the 3rd ventricle | Yes | Yes | Yes | No | No | Surgery | 8.0 (alive) |
| 24      | 24  | F      | Diffuse midline glioma    | Bilateral thalamus and hypothalamus | Yes | Yes | No | No | No | Stereotactic biopsy, radio- and chemotherapy | 8.0 (death) |
| 25      | 44  | M      | Craniopharyngioma         | Sellar region involving the 3rd ventricle | Yes | Yes | No | No | No | Stereotactic aspirin and inner radiotherapy | 8.0 (alive) |
| 26      | 17  | F      | Diffuse midline glioma    | Bilateral thalamus, hypothalamus and midbrain | Yes | Yes | No | No | No | Stereotactic biopsy, radio- and chemotherapy | 8.0 (alive) |
| 27      | 29  | M      | Malignant teratoma        | Pineal region       | Yes | Yes | No | No | No | None | 0.5 (death) |
| 28      | 27  | F      | Epidermoid cyst           | Right temporal lobe | No | No | Yes | Yes | Yes (1 months) | None | 6.0 (alive) |

TABLE 2: Clinical characteristics in 28 patients with Y-shaped shunt
| Characteristic                                | Value (%) | Characteristic                                | Value (%) |
|----------------------------------------------|-----------|----------------------------------------------|-----------|
| Age at diagnosis in yrs                      |           | Diagnosis                                   |           |
| Mean                                         | 37 ± 14   | Craniopharyngioma                           | 8 (28.6)  |
| Range                                        | 17-68     | Glioma                                      | 7 (25.0)  |
| Sex                                          |           | Pituitary adenoma                           | 3 (10.7)  |
| M                                            | 16 (57.1) | Colloid cyst                                 | 2 (7.1)   |
| F                                            | 12 (42.9) | Pineal parenchymal tumor of intermediate differentiation | 2 (12.5) |
| Hydrocephalus ahead of tumor resection       |           | Malignant teratoma                          | 2 (7.1)   |
| No                                           | 4 (14.3)  | Central neurocytoma                         | 1 (3.6)   |
| Bilateral                                    | 23 (82.1) | Epidemoid cyst                              | 1 (3.6)   |
| Unilateral                                   | 1 (3.6)   | Meningioma                                  | 1 (3.6)   |
| VPS ahead of tumor resection                 |           | Pinealocytoma                               | 1 (3.6)   |
| No                                           | 7 (25.0)  | Location                                    |           |
| Bilateral                                    | 19 (67.9) | The 3rd ventricle involving Foramen of Monro | 8 (28.6)  |
| Unilateral                                   | 2 (7.1)   | Suprasellar region involving the 3rd ventricle | 6 (21.4) |
| Tumor surgery                                |           | Pineal region                               | 4 (14.3)  |
| Resection                                    | 12 (42.9) | Sellar region involving the 3rd ventricle   | 4 (14.3)  |
| Stereotactic biopsy                          | 4 (14.3)  | Bilateral thalamus and hypothalamus         | 2 (7.1)   |
| No                                           | 12 (42.9) | The right ventricle                         | 1 (3.6)   |
| Hydrocephalus after tumor resection          |           | Trigone of the left ventricle               | 1 (3.6)   |
| No                                           | 19 (67.9) | The right temporal lobe                     | 1 (3.6)   |
| Bilateral                                    | 5 (17.9)  | Bilateral ventricles and the 3rd ventricle  | 1 (3.6)   |
| Univariate                                   | 2 (7.1)   | Other treatment                             |           |
| Triple                                       | 2 (7.1)   | None                                        | 19 (67.9) |
| VPS after tumor resection                    |           | Radiotherapy and chemotherapy               | 4 (14.3)  |
| No                                           | 19 (67.9) | Gamma knife                                 | 2 (7.1)   |
| Bilateral                                    | 5 (17.9)  | Stereotactic aspiration and inner radiotherapy | 2 (7.1) |
| Unilateral                                   | 2 (7.1)   | Radiotherapy only                           | 1 (3.6)   |
| Triple                                       | 2 (7.1)   | Cause of death                              |           |
| Infection after shunt                        |           | Tumor progression                           | 5 (55.6)  |
| No                                           | 27 (96.4) | Tumor recurrence                            | 2 (22.2)  |
| Yes                                          | 1 (3.6)   | Pulmonary infection                         | 1 (11.1)  |
| Peri-operative outcomes                      |           | Obstruction of shunt                        | 1 (11.1)  |
| Improved                                     | 25 (89.3) | Overall survival (months)                   |           |
| Worsened                                     | 3 (10.7)  | Cases of death                              | 9         |
| KPS at last follow-up                       |           | Median                                       | N/A       |
| Median                                       | 90        | Range                                       | 0.5-58.0  |
| Range                                        | 50-100    | 1 year OS rate                              | 66.7%     |
| Shunt valves                                 |           | 2 year OS rate                              | 66.7%     |
| Programmable, anti-siphon                    | 25 (89.3) | 1 year hydrocephalus-free rate              | 88.7%     |
| Low-pressure, anti-siphon                    | 1 (3.6)   | 2 year hydrocephalus-free rate              | 88.7%     |
| Moderate, anti-siphon                        | 1 (3.6)   |                                             |           |
| Low-pressure, no anti-siphon                 | 1 (3.6)   |                                             |           |
Table 3. Log-rank analyses of parameters associated with OS

| Parameters                                      | OS        |
|------------------------------------------------|-----------|
| Parameters                                     | Chi-square| P value  |
| Age (>40/≤40)                                  | 3.910     | 0.048    |
| Gender (Male/Female)                           | 0.032     | 0.858    |
| Tumor grade (WHO I/WHO II)                     | 8.081     | 0.004    |
| Hydrocephalus ahead of tumor resection (yes/no)| 5.694     | 0.017    |
| VPS ahead of tumor resection (yes/no)          | 0.569     | 0.451    |
| Tumor resection (yes/no)                       | 1.104     | 0.293    |
| Hydrocephalus after tumor resection (yes/no)   | 0.968     | 0.325    |
| Infection after VPS (yes/no)                   | 0.427     | 0.513    |

Figures

Figure 1
The position and shape of the skull incision was shown in A-C. Y-shaped connector used for Y-shaped VPS was shown in D. The connection of Y-shaped connector for double or triple ventricular catheters was illustrated in D and E, respectively.
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Figure 2

Treatment chart of 28 cases with complicated hydrocephalus.
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Treatment chart of 28 cases with complicated hydrocephalus.
Figure 3

Patients' overall survival and hydrocephalus-free survival plots (A). Patients with WHO grade II had longer survival than those with WHO grade II-IV (B, P=0.004).
Figure 3

Patients' overall survival and hydrocephalus-free survival plots (A). Patients with WHO grade I had longer survival than those with WHO grade II-IV (B, P=0.004).

Figure 4

CT revealed a lesion with bilateral thalamic hypodensity occupying the Foramen of Monro with hydrocephalus (A). MR images revealed hydrocephalus caused by hypothalamic and bilateral thalamic lesions with long T1 and long T2 signals as well as mild enhancement (B-F). Y-shaped VPS was performed to resolve hydrocephalus (G). Stereotactic biopsy confirmed the pathology as diffuse midline glioma (H, WHO IV, IDH wild-type) with H3K27M (+++) and MGMT promoter methylation (-). MR images revealed tumor progression and spread 6 months after VPS (I-M).
Figure 4
CT revealed a lesion with bilateral thalamic hypodensity occupying the Foramen of Monro with hydrocephalus (A). MR images revealed hydrocephalus caused by hypothalamic and bilateral thalamic lesions with long T1 and long T2 signals as well as mild enhancement (B-F). Y-shaped VPS was performed to resolve hydrocephalus (G). Stereotactic biopsy confirmed the pathology as diffuse midline glioma (H, WHO IV, IDH wild-type) with H3K27M (+++) and MGMT promoter methylation (-). MR images revealed tumor progression and spread 6 months after VPS (I-M).

Figure 5
CT revealed a lesion with bilateral thalamic hypodensity occupying the Foramen of Monro with hydrocephalus (Fig. 5A-B). Y-shaped VPS was performed to resolve hydrocephalus (Fig 5C). Post-VPS MR images revealed hypothalamic and bilateral thalamic lesions with long T1 and long T2 signals as well as mild enhancement (Fig. 5D-K). 7 months after shunt, MR images revealed tumor shrinkage after radio-chemical therapy (Fig 5L-Q).
Figure 5

CT revealed a lesion with bilateral thalamic hypodensity occupying the Foramen of Monro with hydrocephalus (Fig. 5A-B). Y-shaped VPS was performed to resolve hydrocephalus (Fig 5C). Post-VPS MR images revealed hypothalamic and bilateral thalamic lesions with long T1 and long T2 signals as well as mild enhancement (Fig. 5D-K). 7 months after shunt, MR images revealed tumor shrinkage after radio-chemical therapy (Fig 5L-Q).

Figure 6

CT shows a lesion in bilateral ventricles with hydrocephalus (A). The lesion in the ventricle shows mixed T1 and T2 signal with heterogeneous enhancement on the MR images (B-D). Post-operative CT shows enlargement of the temporal horn (E). The puncture and drainage was performed (F). Post-operative MR scans revealed the abscess formation in the surgical region (G). The abscess disappeared after the puncture and drainage (H). Multiple ventricular entrapments (I-J) were resolved by triple cavity VPS with two Y-shaped connectors (K-L). The patient recovered well and CT/MR shows resolution of hydrocephalus 1 year later (M-N).
Figure 6

CT shows a lesion in bilateral ventricles with hydrocephalus (A). The lesion in the ventricle shows mixed T1 and T2 signal with heterogeneous enhancement on the MR images (B-D). Post-operative CT shows enlargement of the temporal horn (E). The puncture and drainage was performed (F). Post-operative MR scans revealed the abscess formation in the surgical region (G). The abscess disappeared after the puncture and drainage (H). Multiple ventricular entrapments (I-J) were resolved by triple cavity VPS with two Y-shaped connectors (K-L). The patient recovered well and CT/MR shows resolution of hydrocephalus 1 year later (M-N).

Figure 7

CT scan shows high density signal with calcification in the pineal region, which caused hydrocephalus (A). MR images show a lesion with enhancement (B-D). Post-operative MR revealed total resection of the tumor (E-G). CT scan shows subdural effusion when the patient complained of headache and vomit 15 days after surgery (H). The puncture and drainage of subdural effusion was performed (I). The effusion was unchanged and hydrocephalus appeared after drainage for 9 days (J). Y-shaped VPS was performed, one tip in the left frontal horn and the other one in the right subdural effusion (K). MR images revealed the recovery of subdural effusion and hydrocephalus 8 months later (L-P).
Figure 7

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Figure 8

For the sequential case, radiological images revealed a mass in the third ventricle with obstructive hydrocephalus in the left ventricle (A-D). The patient received left frontal horn VPS to resolve the hydrocephalus (E). 6 months later, CT and MR scan revealed obstructive hydrocephalus in the right ventricular (F-I). To resolve right hydrocephalus, right frontal horn tip was added to the original shunt by a Y-shaped connector and the bilateral hydrocephalus was well resolved (J).
Figure 8

For the sequential case, radiological images revealed a mass in the third ventricle with obstructive hydrocephalus in the left ventricle (A-D). The patient received left frontal horn VPS to resolve the hydrocephalus (E). 6 months later, CT and MR scan revealed obstructive hydrocephalus in the right ventricular (F-I). To resolve right hydrocephalus, right frontal horn tip was added to the original shunt by a Y-shaped connector and the bilateral hydrocephalus was well resolved (J).