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

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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 3cases, 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 1 D 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 ptatus (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 [illegible]-[illegible]/WHO [illegible]-[illegible]), 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 4I-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 timozolomide chemotherapy. 15 months after shunt, MR images revealed tumor shrinkage (Fig 5L-Q).

Case 21 (post-operative multiloculated hydrocephalus)

A 22-year-old female was admitted in our hospital with the radiological diagnosis of central neurocytoma in the bilateral ventricles and obstructive hydrocephalus. She 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 purple-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 19-year-old male was admitted to our hospital with a mass in the pineal region and obstructive hydrocephalus. He complained of intermittent headache for half a year. Neurological examination revealed Parinaud syndrome. CT revealed a lesion with mixed density and calcification (Fig. 7A). MR images revealed a lesion with iso-T1 long-T2 signals and homogeneous enhancement (Fig. 7B-D).

The patient underwent a right frontal trans-callosal approach for total removal of the tumor (Fig. 7E-G). The tumor was found to be a grey-red mass with less defined borders in the pineal region. Postoperative pathology revealed the diagnosis of pineal parenchymal tumor of intermediate differentiation (WHO grade III). The patient was discharged 13 days after surgery. Ten days later, the patient was re-admitted to our hospital for sudden headache, nausea and vomiting. An emergency CT scan revealed the right fronto-tempo-parietal subdural effusion (Fig. 7H), and an emergency drainage was performed (Fig. 7I). Nine days after drainage, the patient still had intermittent headache and cognitive difficulties, CT scan revealed unchanged subdural effusion with hydrocephalus (Fig. 7J). The patient underwent bilateral Y-shaped VPS with programmable and anti-siphon shunt, one draining the right subdural effusion and the other one draining the left ventricular frontal horn (Fig. 7K). Five days later, the patient had a fever, and the CSF analysis revealed increased WBC count. Combined vancomycin and meropenem were used and the patient was discharged for radiotherapy with improved KPS. At the last follow-up 8 months after the primary surgery, the patient had been back to job (Fig. 7L-P).

Case 17 (sequential Y-shaped hydrocephalus)

A 52-year-old Male was admitted in our hospital for headache and vomiting. 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. And 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 chordoid 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 pelludidum 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 fissue 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 incurrence 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 incidence[3]. 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 Monra 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. hycephalus	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	Yes (6 months)	None	1.0 (death)	0

			involving Foramen of Monro								
18	42, F	Meningioma	Pineal region	Yes	No	Yes	Yes	Yes (1 months)	None	17.0 (alive)	90
19	52, M	Pineal parenchymal tumor of intermediate differentiation	Pineal region	Yes	Yes	Yes	Yes	Yes (6 months)	None	14.5 (alive)	50
20	20, M	Pineal parenchymal tumor of intermediate differentiation	Pineal region	Yes	No	Yes	Yes	Yes (2 months)	Radiotherapy	17.0 (alive)	100
21	23, F	Centrocytoma	Bilateral and the 3rd ventricle involving Foramen of Monro	Yes	No	Yes	Yes	Yes (3 months)	None	16.0 (alive)	70
22	68, M	Pituitary adenoma	Sellar region involving the 3rd ventricle	Yes	Yes	No	No	No	None	12.0 (alive)	90
23	43, F	Craniopharyngioma	Sellar region involving the 3rd ventricle	Yes	Yes	Yes	No	No	Surgery	8.0 (alive)	90
24	24, F	Diffuse midline glioma	Bilateral thalamus and hypothalamus	Yes	Yes	No	No	No	Stereotactic biopsy, radio- and chemo-therap	8.0 (death)	0
25	44, M	Craniopharyngioma	Sellar region involving the 3rd ventricle	Yes	Yes	No	No	No	Stereotactic aspirin and inner radiotherapy	8.0 (alive)	90
26	17, F	Diffuse midline glioma	Bilateral thalamus, hypothalamus and midbrain	Yes	Yes	No	No	No	Stereotactic biopsy, radio- and chemo-therap	8.0 (alive)	70
27	29, M	Malignant teratoma	Pineal region	Yes	Yes	No	No	No	None	0.5 (death)	0
28	27, F	Epidermoid cyst	Right temporal lobe	No	No	Yes	Yes	Yes (1 months)	None	6.0 (alive)	90

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)	Epidermoid 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 (6.3)
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

	OS	
Parameters	**Chi-square**	**P value**
Age (>40/≤40)	3.910	0.048
Gender (Male/Female)	0.032	0.858
Tumor grade (WHO □/WHO □-□)	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

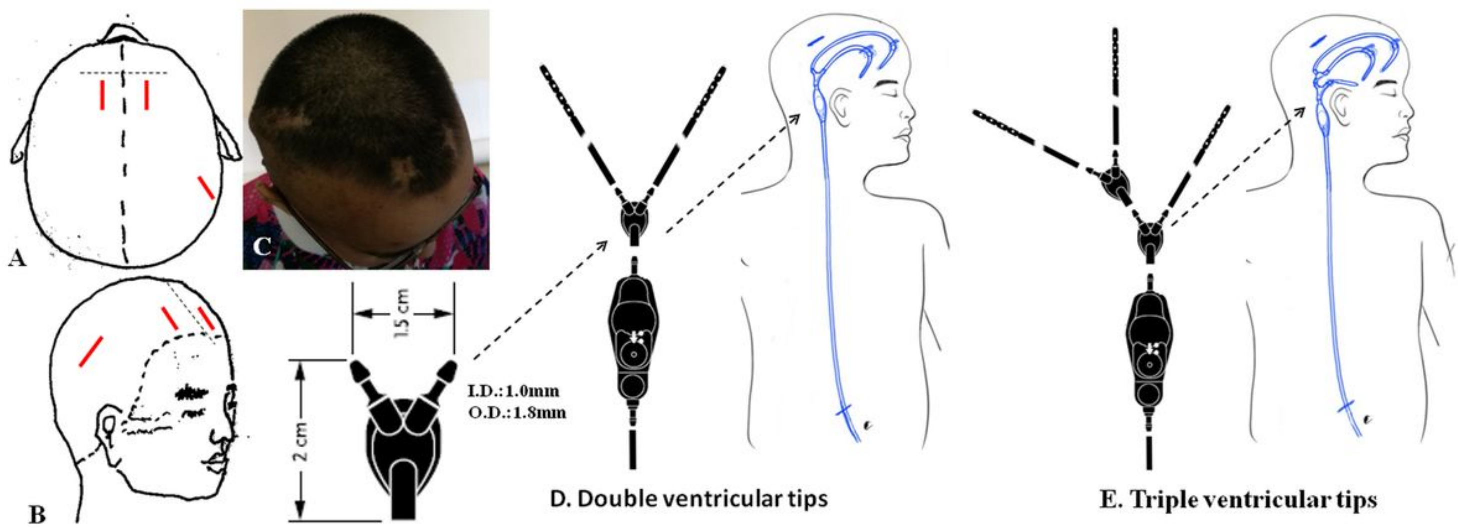
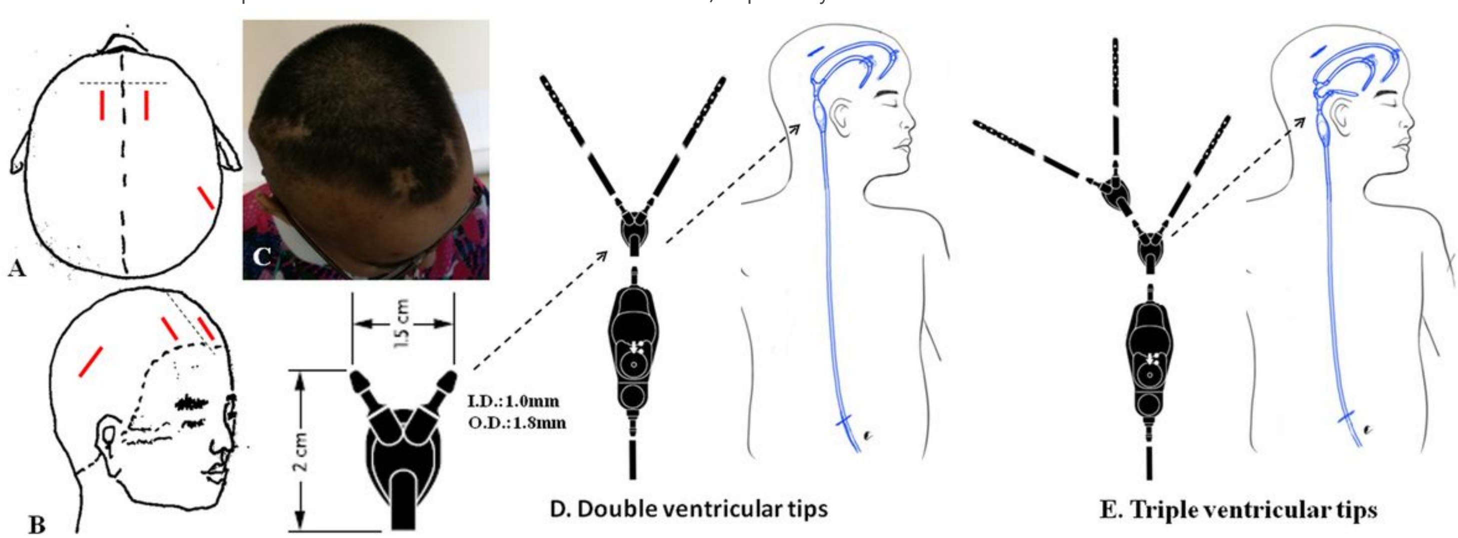


Figure 1

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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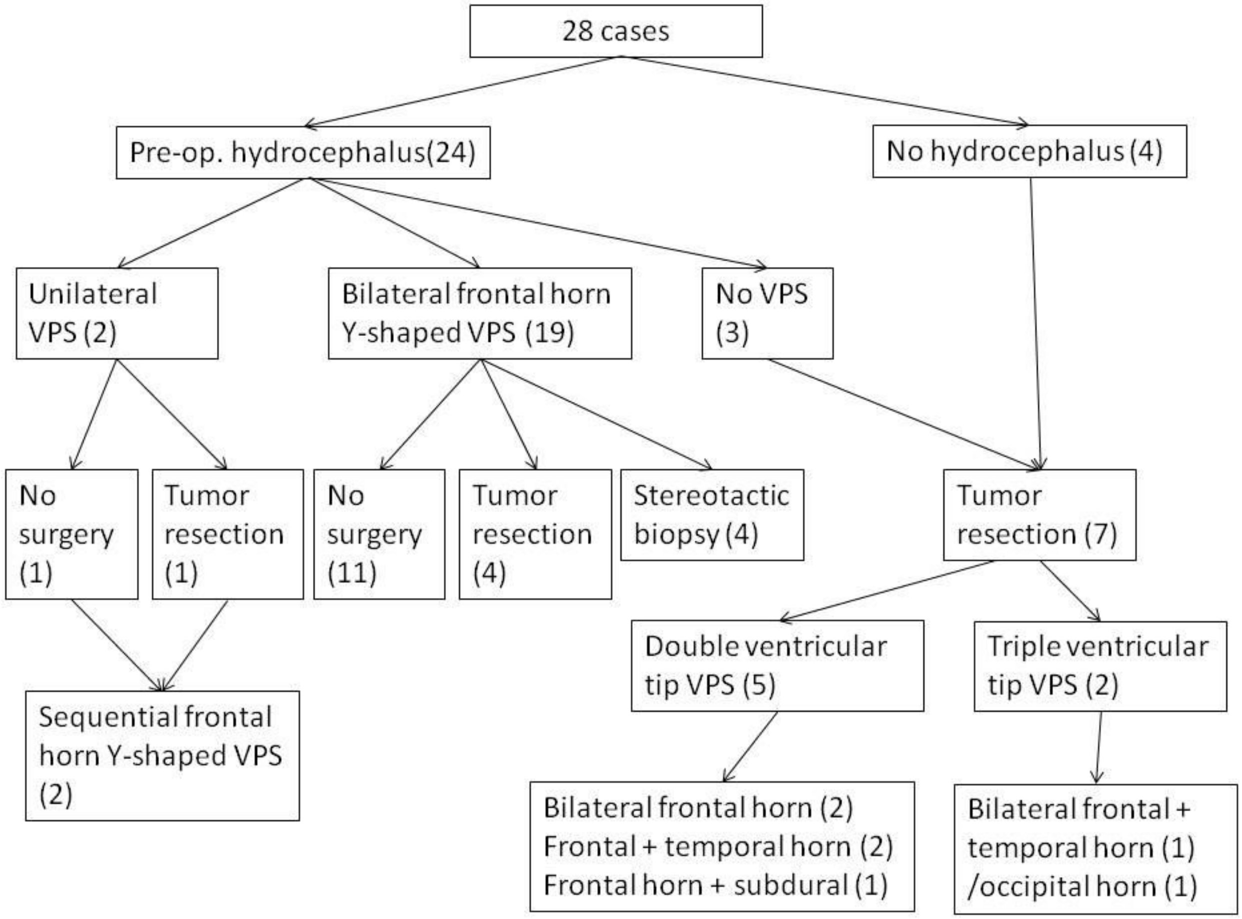
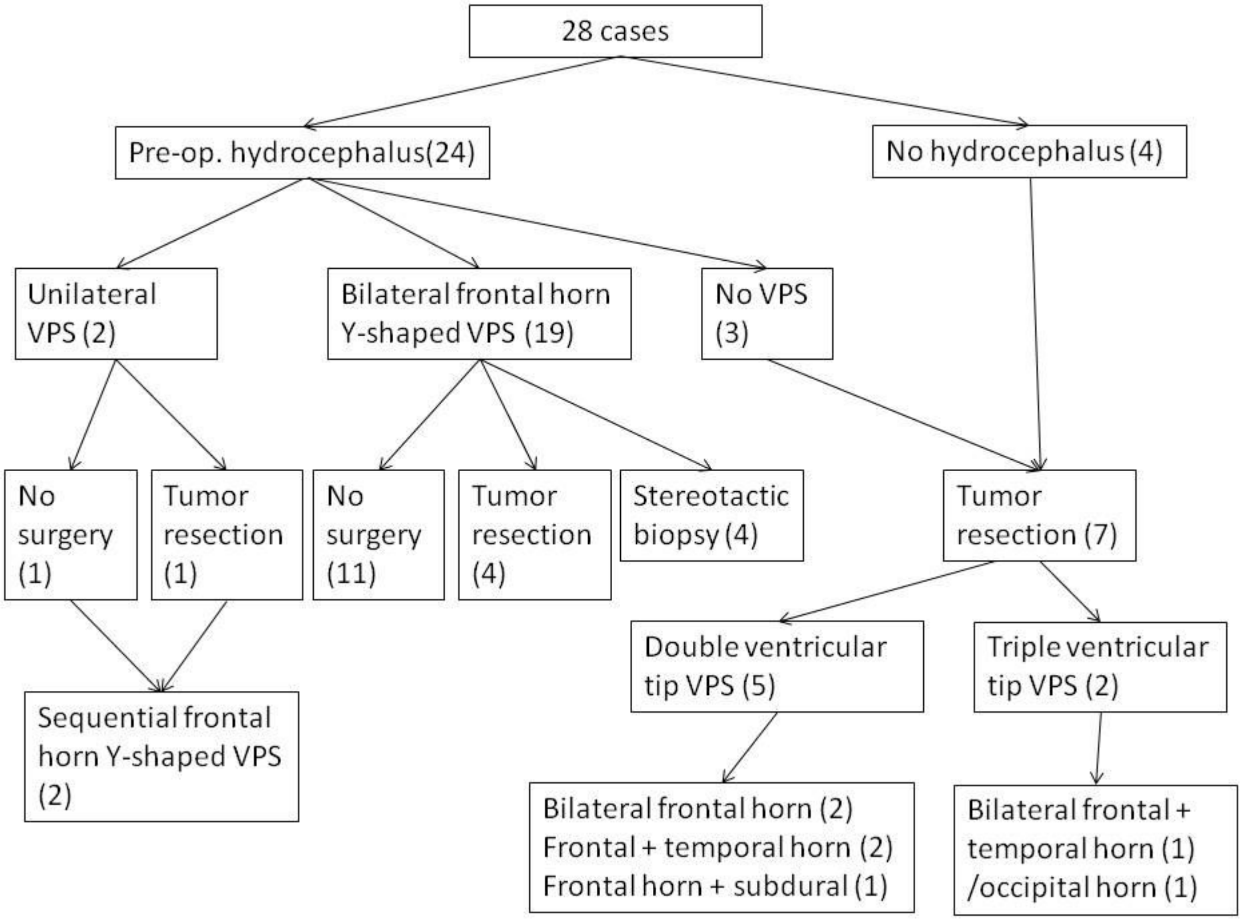


Figure 2

Treatment chart of 28 cases with complicated hydrocephalus.



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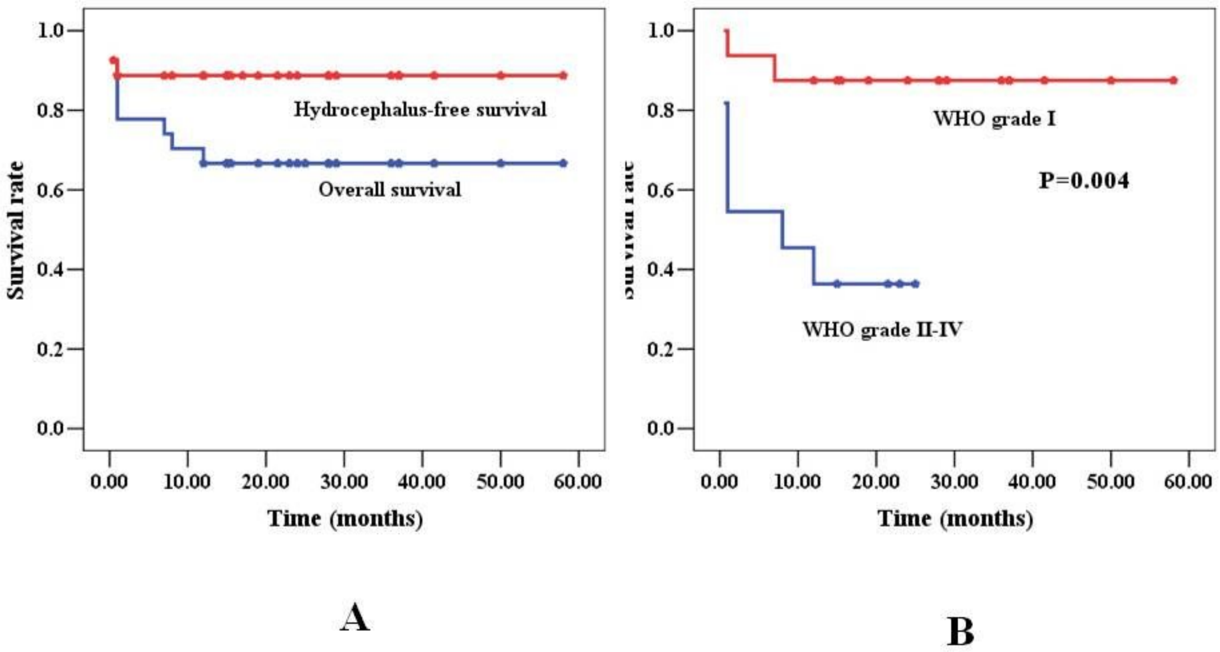
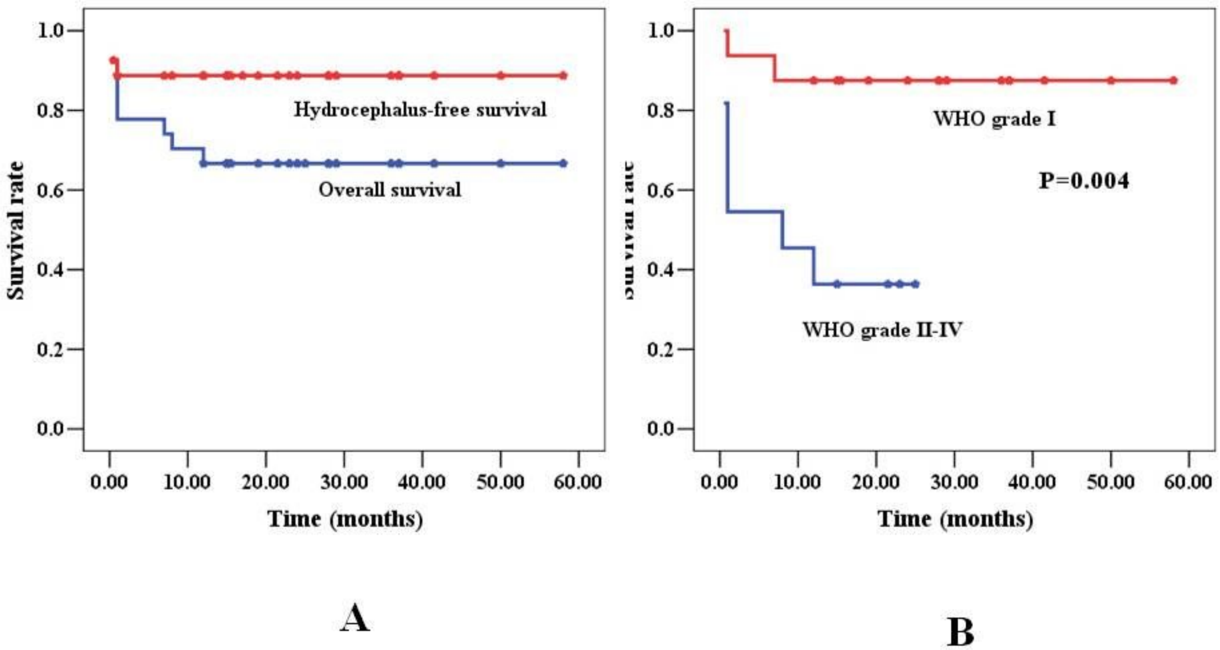


Figure 3

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).



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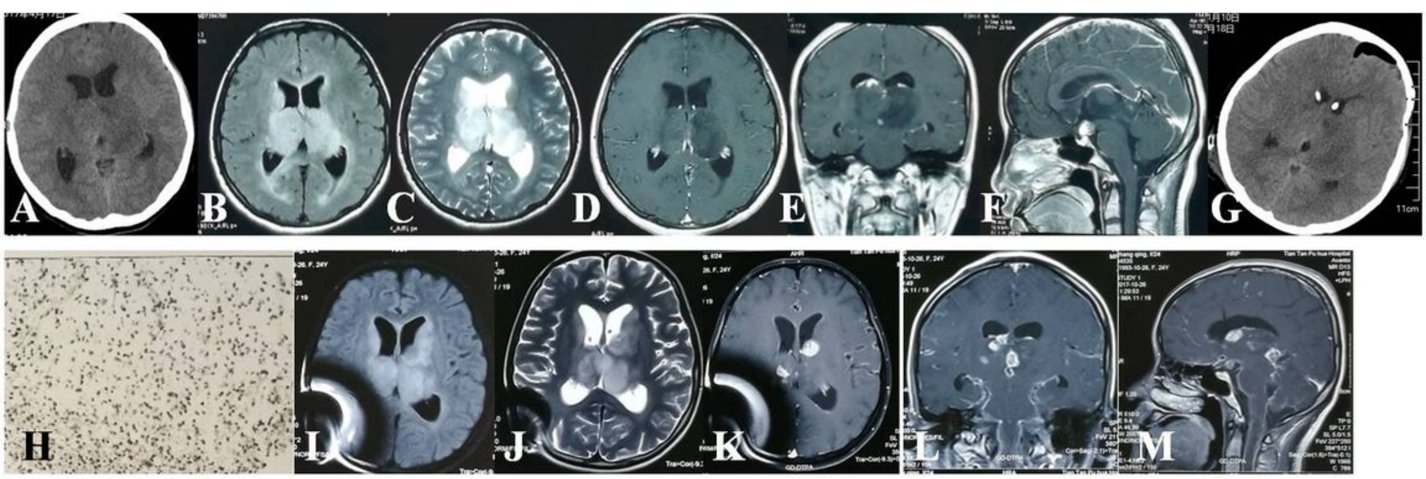
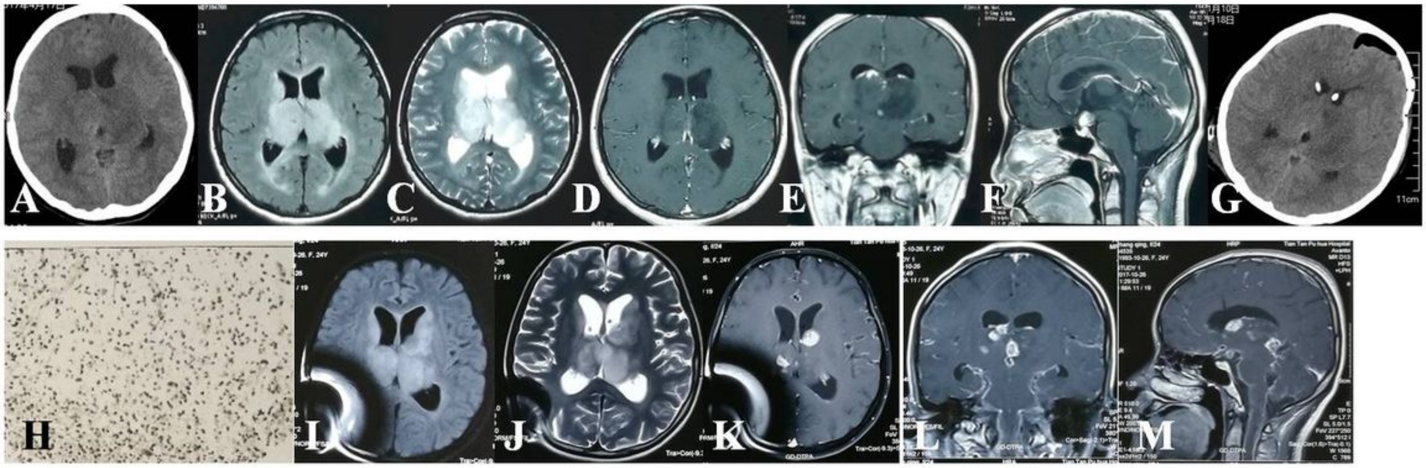


Figure 4

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).



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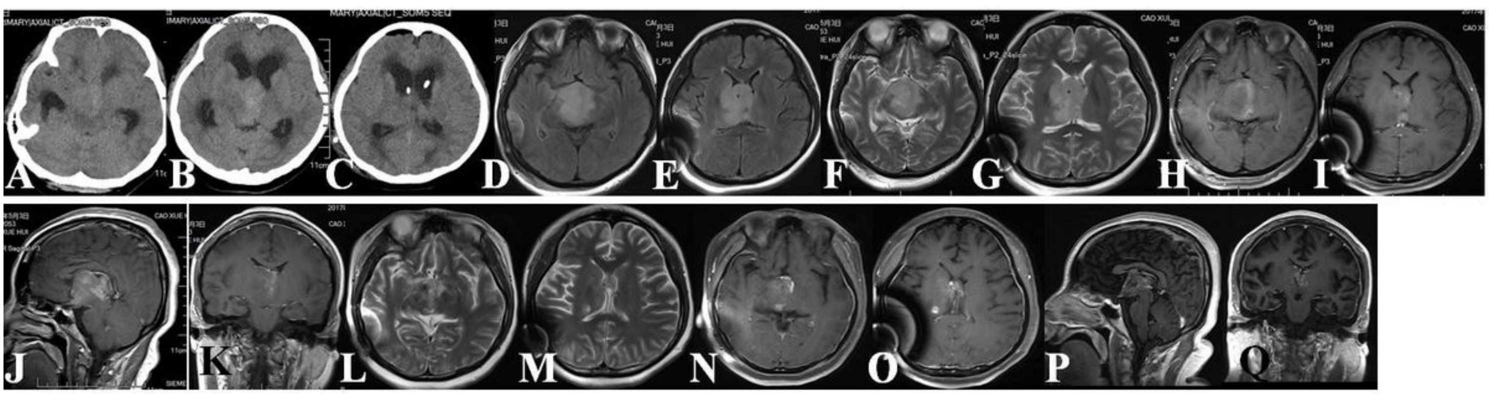
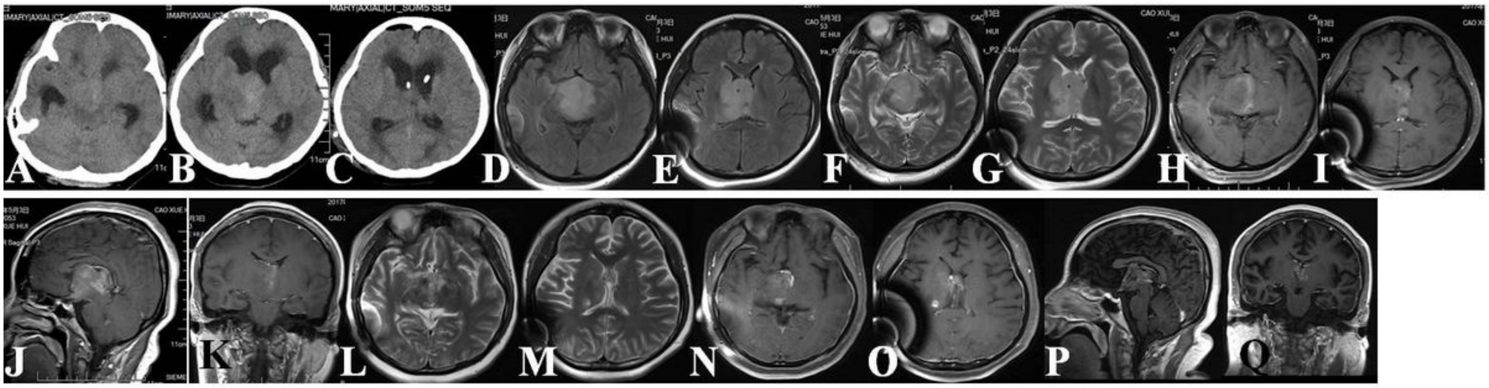


Figure 5

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).



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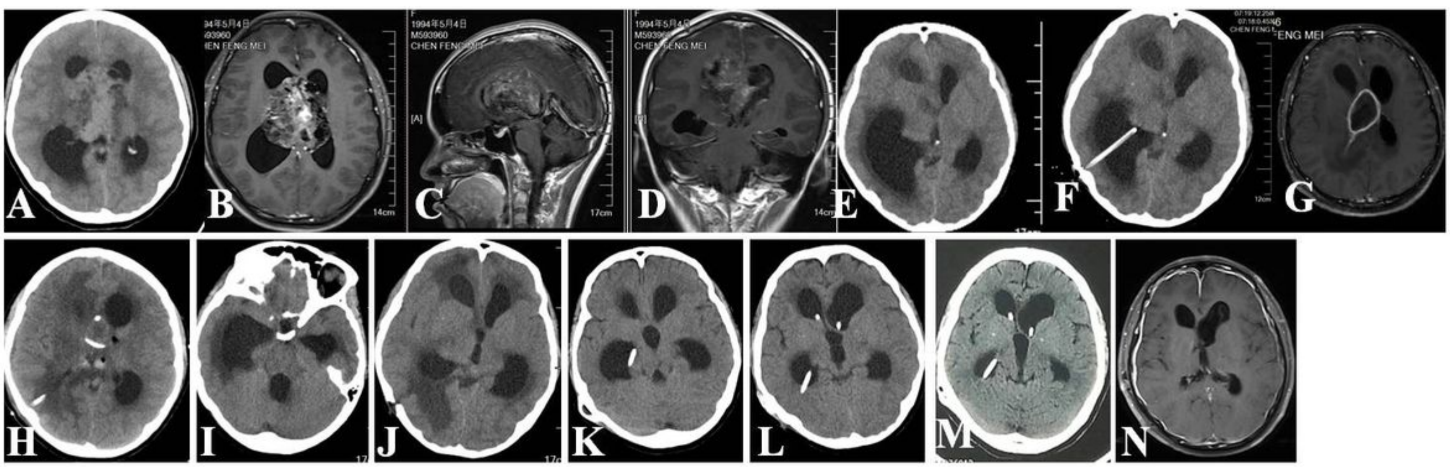
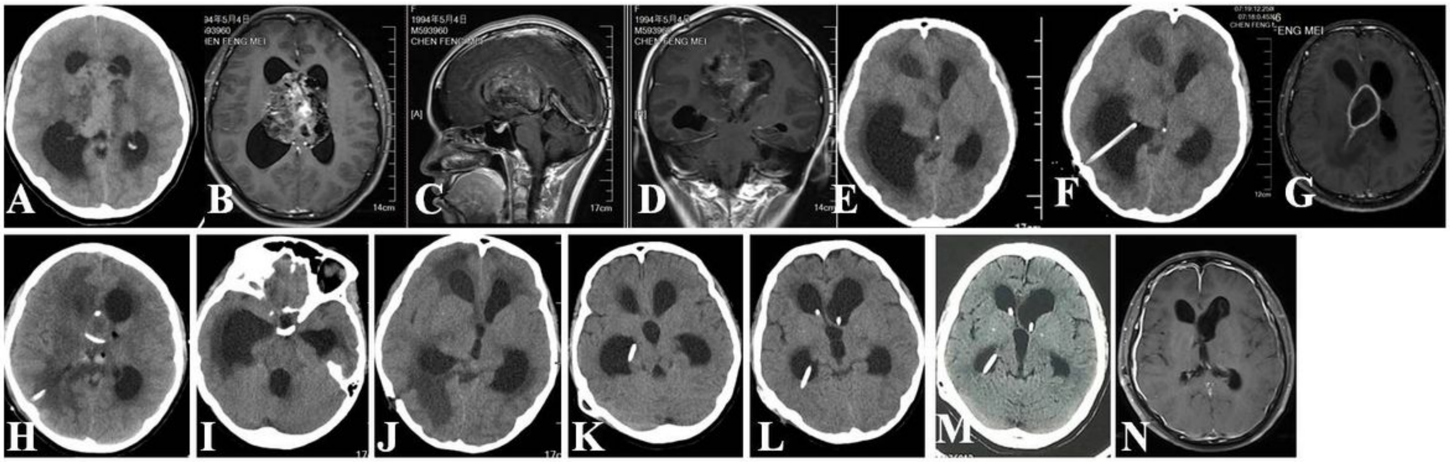


Figure 6

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).



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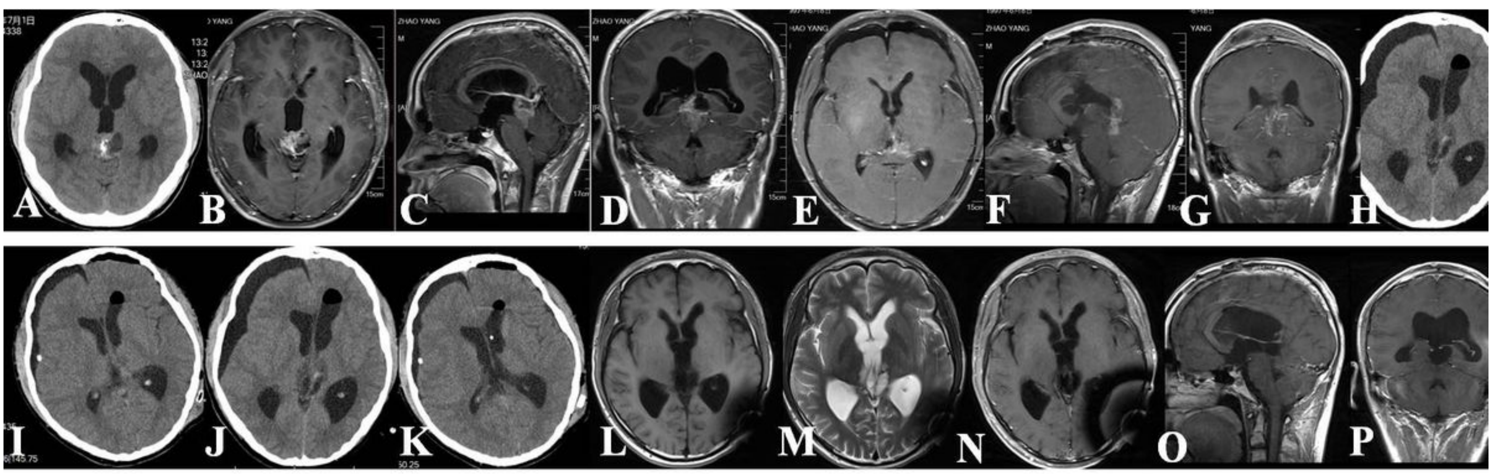
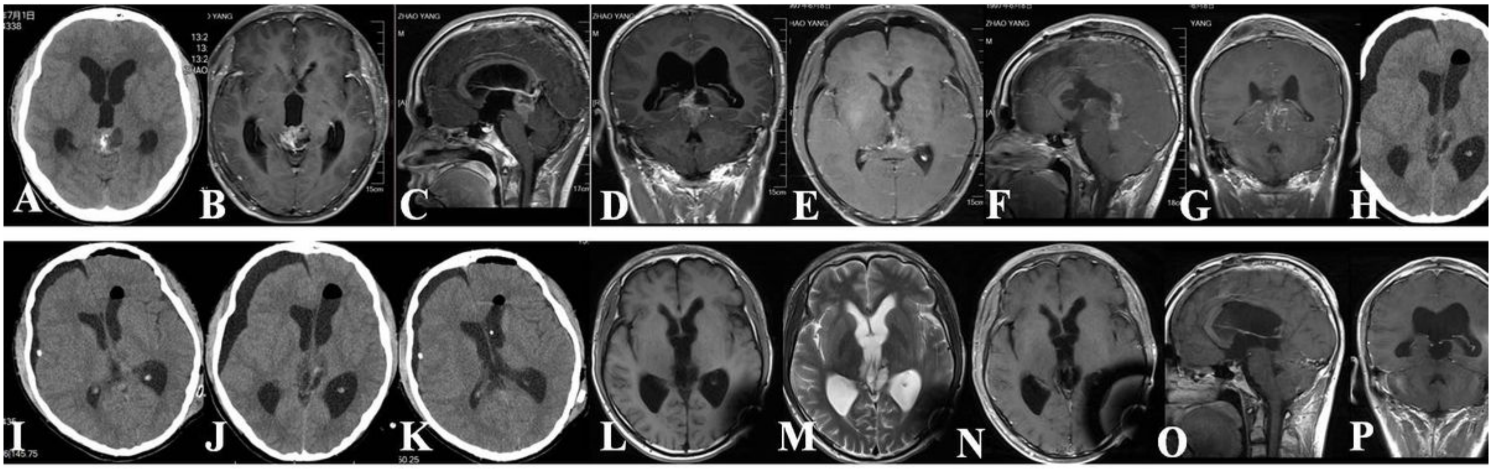


Figure 7

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).



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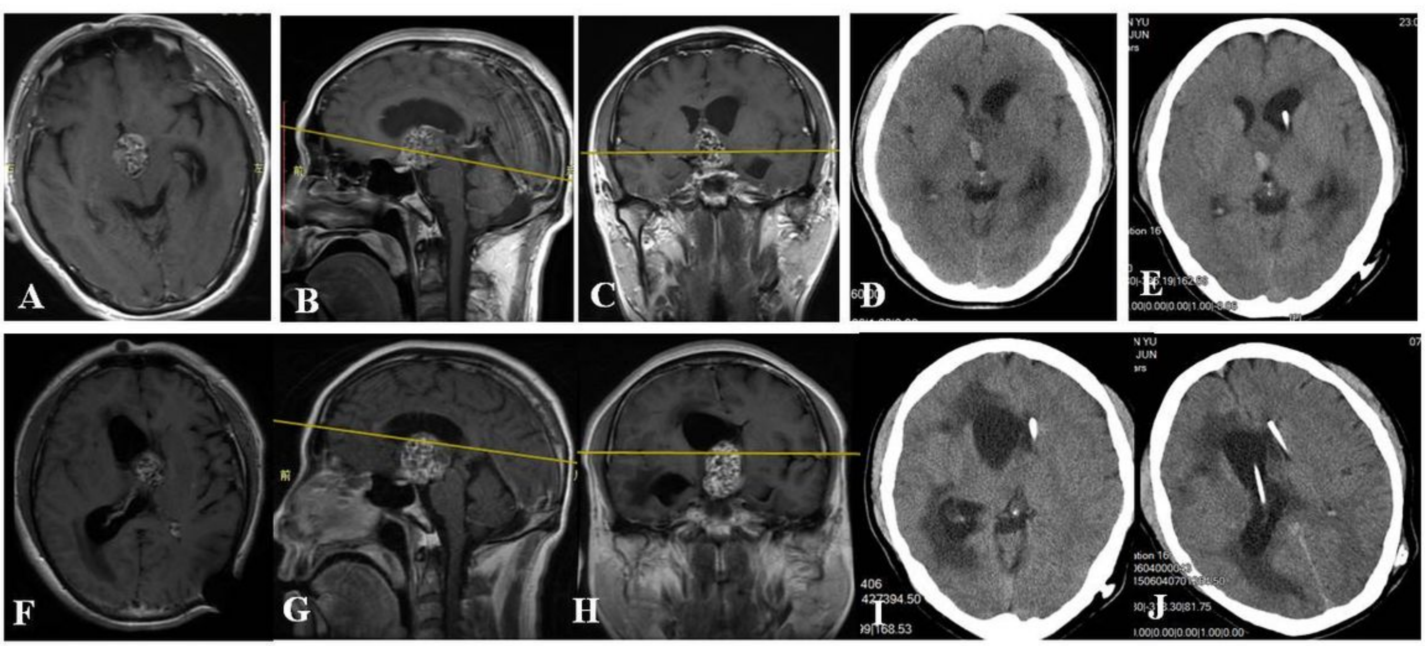
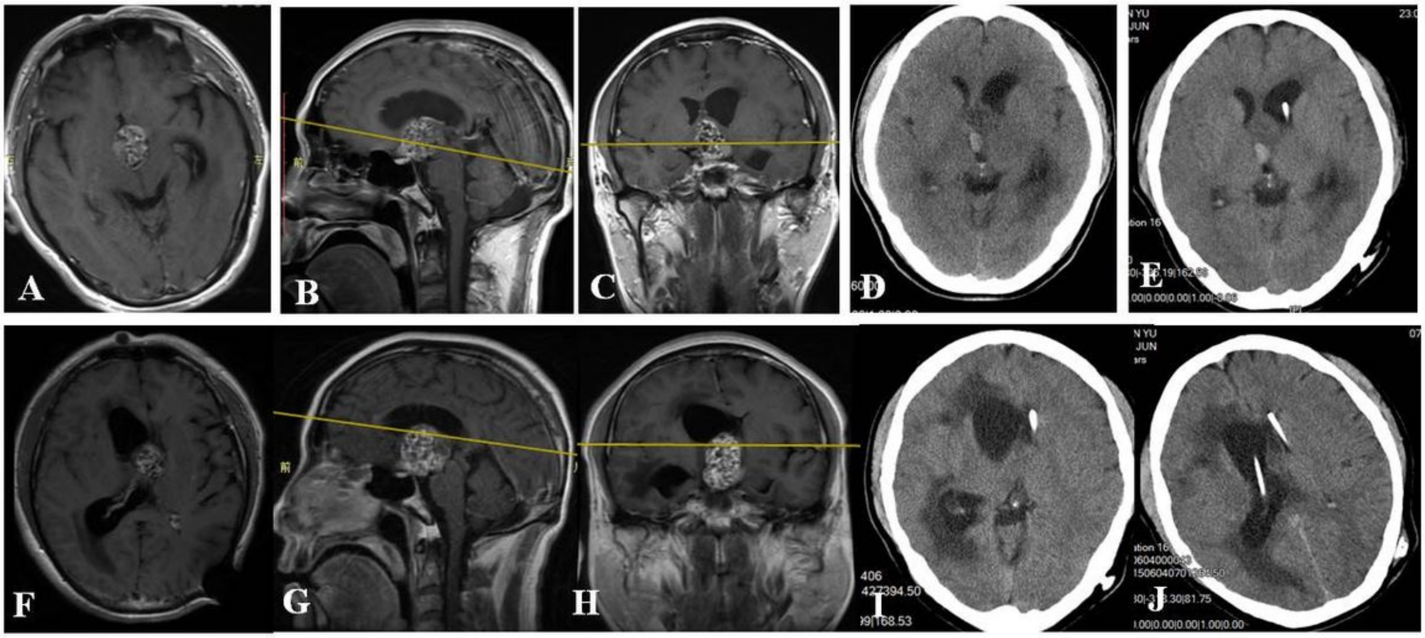


Figure 8

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).



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