Endoscopic third ventriculostomy for refractory low pressure hydrocephalus – 2 case reports and literature review

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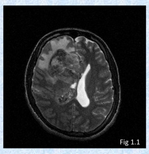
Background:

Low-pressure hydrocephalus (LPH) remains a poorly-understood neurosurgical problem which leads to significant clinical challenge in daily practice. Previous literature suggested prolonged external ventricular drainage (EVD) at sub-zero pressure to reverse ventriculomegaly before CSF shunting procedure. However, it is not always clinically feasible for prolonged sub-zero drainage. Furthermore, there may be persistent ventricular dilatation after CSF shunting procedure. Some studies suggested alternative/ additional treatment which include ETV.

Case reports

Case 1
23/M. Right frontal glioblastoma with partial excision done; complicated by hydrocephalus with VP shunt inserted.



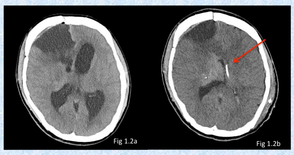


Fig 1.1
Pre-operative MRI showing a large right frontal tumor, later histologically confirmed to be glioblastoma

Fig 1.2
Post-operation CT brain showed hydrocephalus (Fig 1.2a). Left parietal VP shunt inserted, and ventricular size decrease significantly after VP shunt (Fig 1.2b; red arrow showing the shunt tip in-situ)

Patient admitted for decrease in consciousness; GCS E2M5V1
VP shunt revision was done twice; there was no significant clinical improvement; CT brain still showed persistent ventriculomegaly. Shunt tipping confirmed the patency and very low pressure. The shunt was externalised and drained 25cm below ear level. GCS gradually improved.

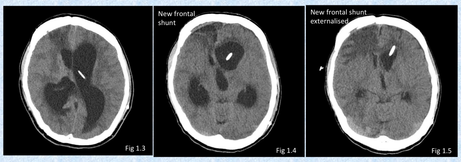


Fig 1.3
Patient admitted for decrease GCS (8/15). CT brain showed hydrocephalus.

Fig 1.4
After revision to a right frontal VP shunt (with medium Pudenz valve), GCS of the patient was still fluctuating. CT brain still showed persistent hydrocephalus

Fig 1.5
Right frontal shunt was externalised and drained 25cm below ear-level. There is significant clinical improvement and CT brain showed that ventricular size much improved.

Valve-less VP shunt system was inserted. However there was persistent hydrocephalus and there is clinical deterioration.

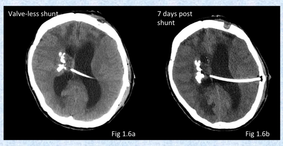


Fig 1.6
Left frontal catheter was removed. New left parietal valve-less VP shunt inserted.
Despite a valve-less system, there was persistent ventriculomegaly. Clinical patient has no improvement.

Valve-less VP shunt was externalised, ETV and new right frontal VP shunt with programmable valve were done. The externalised valve-less VP shunt was ligation 1 week after ETV. GCS gradually improved to E4M6V3. Patient subsequently died of glioblastoma disease progression 3 months after ETV.

Fig 1.7 post-ETV

The valve-less shunt was externalised. ETV was done. New left frontal VP shunt with also performed (with programmable valve) (Fig 1.7a; Red arrow showing the new left frontal shunt)

Externalised VP shunt was clamped and subsequently ligated. CT brain showed much improved ventricular size. Patient also gradually improved clinically.

CT brain (2 months after ETV) showed there is no further ventriculomegaly.

Case 2
M/42. History of childhood meningitis. AML with chemotherapy since late 2017.
Admitted for decrease in consciousness; GCS E3M6V4; LP suggested TB meningitis.

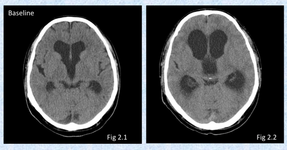


Fig 2.1
Patient has history of childhood meningitis. There was some baseline dilatation of the ventricle.

Fig 2.2
Patient was admitted for decrease in GCS. CT brain showed communicating hydrocephalus.

VP shunt with medium Pudenz valve was inserted. However, patient's GCS dropped to 12. CT brain showed persistent hydrocephalus. VP shunt revision was done. Intra-operatively all catheter confirmed to be patent. The valve was changed to low-pressure valve.

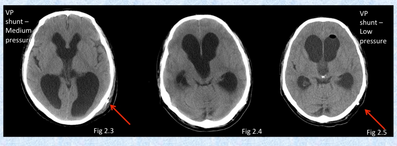


Fig 2.3
VP shunt with fixed medium pressure valve inserted. Intraventricular catheter not shown. Red arrow points to the distal catheter

Fig 2.4
Initially there is clinical improvement. But 1 week after VP shunt insertion, he developed hydrocephalus again with GCS dropped to 12.

Fig 2.5
VP revision was done. The patency was confirmed intra-operatively. The valve was changed to a fixed low-pressure valve

There was no significant clinical and radiological improvement. ETV was attempted to avoid prolonged EVD drainage. His GCS gradual improved to his pre-morbid state.

In view of his immuno-compromised state, prolonged sub-zero EVD drainage was avoided.
Instead, ETV was performed. Patient has gradual clinical improvement. CT brain on post-op day 5 showed resolution of hydrocephalus. (Fig 2.6)
Reassessment CT brain 2 months post-op showed ventricular size similar to baseline. (Fig 2.7)

2 months later he was admitted for decrease GCS again. CT brain showed hydrocephalus. Shunt tapping confirmed to patency. Ventricular endoscopy showed new fibrous membrane covering the ETV. Reopening achieved. Patient gradually improved after 2nd ETV

Patient was admitted again for decrease in GCS 2 months later. CT brain on admission showed hydrocephalus. (Fig 2.6)
Bedside shunt tapping confirmed the patency.
2nd ETV was attempted. Intraoperatively there is new fibrous membrane covering the initial ETV. Reopening of the ETV achieved. Post-operatively patient has clinical and radiological improvement. (Fig 2.9). He was discharged to rehabilitation centre.
Latest reassessment CT brain showed ventricular size similar to baseline. (Fig 2.10)

Literature review

LPH is a rare clinical diagnosis; characterized by neurological decline and ventriculomegaly that persists despite low intracranial pressure.
There is no stardard treatment of LPH yet.
Pang and Altschuler treated 12 patients with LPS with prolonged sub-zero EVD drainage, followed by definitive shunting surgery, with a low-pressure valve.
Since then, many literatures reported patients with refractory LPS despite the above-mentioned method. Additional management including positioning, neck-wrapping, intermittent valve pressure, valve-less shunt and active pumping negative-pressure shunt system were reported in literatures.
Limited studies reported use of ETV in patients with LPH; yet all studies provided evidence of its usefulness in patient with LPH. The latest study in 2016 showed that, four patients who were initially refractory to sub-zero drainage, were able to have shunting surgery shortly after ETV.
One patient did not require shunting after ETV.

Conclusion

For patient with refractory low pressure hydrocephalus, EVT can be considered as an addition/alternative treatment.

Reference:
1. Pang D, Altschuler E. Low-pressure hydrocephalic state and viscoelastic alterations in the brain. Neurosurgery. 1994 Oct;35(4):643-55; discussion 655-6.
2. Foster KA, Deibert CP, Choi PA, Gardner PA, Tyler-Kabara EC, Engh JA. Endoscopic third ventriculostomy as adjunctive therapy in the treatment of low-pressure hydrocephalus in adults. Surg Neurol Int 10-Mar-2016;7:26