Progressive cranial nerve palsy following shunt placement in an isolated fourth ventricle

Case report

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Cranial nerve palsy is rarely seen after shunt placement in an isolated fourth ventricle. In the few reports of this complication, neuropathies are thought to be caused by catheter injury to the brainstem nuclei either during the initial cannulations or after shrinkage of the fourth ventricle. The authors treated a child who suffered from delayed, progressive palsies of the sixth, seventh, 10th, and 12th cranial nerves several weeks after undergoing ventriculoperitoneal shunt placement in the fourth ventricle. Magnetic resonance imaging revealed the catheter tip to be placed well away from the ventricular floor but the brainstem had severely shifted backward, suggesting that the pathogenesis of the neuropathies was traction on the affected cranial nerves. The authors postulated that the siphoning effect of the shunt caused rapid collapse of the fourth ventricle and while the cerebellar hemispheres were tented back by adhesions to the dura, the brainstem became the only mobile component in response to the suction forces. Neurological recovery occurred after surgical opening of the closed fourth ventricle and lysis of the basal cistern adhesions, which restored moderate ventricular volume and released the brainstem to its normal position.

KEY WORDS • brainstem shift • cranial neuropathy • isolated fourth ventricle • shunt • pediatric neurosurgery

Abbreviations used in this paper: CSF = cerebrospinal fluid; CT = computerized tomography; MR = magnetic resonance; VP = ventriculoperitoneal.
hand and leg weakness had greatly improved. There was no evidence of increased intracranial pressure. A CT scan demonstrated that all of the ventricles were now slit but that the catheter tip remained away from the brainstem (Fig 3).

By the 6th postoperative week, his right lateral rectus muscle was completely paralyzed. The left lateral rectus was paretic, and he had begun to manifest signs of progressive bilateral lower motor neuron–type seventh nerve palsy. Repeated imaging studies demonstrated no change from the previous studies. Despite high doses of dexamethasone, his facial weakness worsened, and 8 weeks after fourth ventricle VP shunt placement, he experienced complete right-sided vocal cord paralysis, a partial right hypoglossal palsy, and intermittent wheezing and choking while drinking. An MR image clearly revealed significant backward displacement of the brainstem toward the slit fourth ventricle (Fig. 4). In retrospect, previous MR images had demonstrated a gradually widening gap between the brainstem and the clivus.

**Second Operation.** We concluded that his multiple, successive cranial neuropathies were caused by nerve traction secondary to brainstem shifting and that the best and fastest method to resolve these problems without interrupting fourth ventricular CSF diversion would be to open the closed fourth ventricle and the subarachnoid pathways in the basal cisterns. Suboccipital craniectomy and C-1 laminectomy were performed. On opening the dura, we encountered dense arachnoid adhesions surrounding the entire spinomedullary junction, causing obvious craniospinal CSF blockage at the foramen magnum. The cerebellar hemispheres were firmly adherent to the suboccipital dura. The foramen of Magendie was closed over by opaque thick membranes. After performing wide resection of these membranes, the tip of the ventricular catheter was confirmed to be free from the floor of the fourth ventricle. We also found that both foramina of Luschka were also closed by thick membranes, which were also opened widely. Extensive lysing of subarachnoid adhesions was performed, and a small silastic tube was left between the fourth ventricle and the ventral cervical subarachnoid space. A wide decompressive duraplasty was then performed with a bovine pericardial graft.

**Second Postoperative Course.** Postoperative MR imaging revealed resumption of a more normal (ventral) position of the brainstem nearer to the clivus and a slightly larger fourth ventricle than the preoperative slit (Fig. 5). The child’s vocal cord and tongue paralysis recovered completely in 7 days. Within 2 weeks, the sixth and seventh nerve palsies began to improve. Six weeks later, his extraocular palsy was 80% resolved and his facial paralysis approximately 50% resolved.

**Discussion**

An isolated fourth ventricle is an uncommon but well-documented complication of long-term shunt placement for hydrocephalus. The incidence has been reported to be 2 to 3%. There is a strong association of isolated fourth ventricle with the postinfectious and posthemorrhagic types of hydrocephalus, and many of the patients had also undergone multiple shunt revisions and suffered infections. The assumption is that these inflammatory conditions not only lead to outflow blockage of the basal cisterns and the foramina of Magendie and Luschka but may also incite ependymal adhesions at the sylvian aqueduct, which would result in entrapment of CSF normally produced by the fourth ventricular choroid plexus. Oi and Matsumoto have postulated that slit ventricles resulting from shunt placement cause coaptation of the aqueductal wall and induce either a reversible aqueductal closure or, in some cases, a permanent proliferative obliteration of the aqueduct.

Many isolated fourth ventricles are radiographically apparent but the patients are asymptomatic. Some of these patients, however, will present with truncal ataxia, poor mo-

**Fig. 1.** Preoperative MR images obtained 2 weeks before fourth VP shunt placement. **Left:** Sagittal T1-weighted image revealing an enormously distended fourth ventricle and a large cervical syrinx. The brainstem is pressed against the clivus. **Right:** Axial T1-weighted image revealing slit third and lateral ventricles and a functioning right VP shunt.
Progressive myelopathy due to cervicothoracic syringomyelia is an uncommon clinical concomitant in patients with an isolated fourth ventricle. In our case, the syrinx cavity showed nearly complete collapse only a few days after VP shunt placement, which suggests the origin of the syrinx is downward dissection of CSF through the opening of the central neural canal of the upper cervical cord at the obex. If the syrinx were generated by CSF being driven through the cord by intermittent high intraspinal subarachnoid pressure spikes associated with CSF blockage at the foramen magnum, it would not resolve with simple fourth ventricular CSF diversion. As Rekate pointed out, failure to spot a gaping obex at the time of craniotomy cannot exclude the possibility of patency of the central canal to pulsatile CSF dissection when the fourth ventricle is pressurized. The absence of CSF flow across the craniospinal junction in our case could have contributed to propagation of the hydromyelic cavitation.

The most widely used treatment for an isolated fourth ventricle is VP shunt placement. Progressive cranial nerve palsies have been reported. Postoperative cranial nerve palsies have been reported. Three of 12 patients treated by Lee, et al., suffered immediate sixth and seventh nerve palsies after shunt placement, which was attributed to direct injury to the brainstem at the time of cannulation. Eder, et al., suggested another mechanism for cranial neuropathies encountered in two of their five patients who underwent shunt placement. In these two cases, the tips of the catheters were initially located in the center of the ventricle shortly after insertion. The patients did not suffer from sixth and seventh nerve palsies until 1 to 6 weeks after surgery, when repeated MR imaging revealed a slit fourth ventricle and that the catheter tips appeared to be “touching” the floor of the ventricle. These authors thus presumed that the cranial nerve nuclei were injured by this anatomical shift of the catheter tip. Rekate also described two patients in whom the approximation of the catheter tip to the brainstem was similar after collapse of the shunted fourth ventricle and hypothesized the same mechanism for the delayed cranial neuropathies.

We are postulating a different pathogenesis for the delayed and progressive multiple cranial neuropathies in our patient and perhaps in some of the reported patients as well. The isolated fourth ventricle is essentially a closed box with soft, collapsible walls. When subjected to low or even subatmospheric pressure due to siphoning through the shunt, the fluid content will rapidly diminish to a minimum. Post–shunt placement imaging almost invariably reveals a slit fourth ventricle. The decompressed cerebellum quickly expands by taking CSF into its extracellular space, so that the reconstituted, thickened mantle will fill out some of the collapsing cavity; however, primarily there is significant inward shift of the ventricular walls toward the center. In our case, the brainstem retracted a considerable distance from the clivus, thereby likely exerting traction injury to the cranial nerves. Direct brainstem injury by the catheter is much less plausible because the catheter tip was never near the brainstem; 10th nerve palsy is not commonly seen with injury to the fourth ventricular floor, and successive involvement of multiple cranial nerves is a much more likely phenomenon of gradual nerve traction than direct nuclear damage.

Our theory prompted us to create venting outlets for the closed box by widely opening the occlusive membranes at the foramina of Magendie and Luschka so that the ingress of CSF could nullify the suction effect from the shunt and allow the brainstem to return to a more proximate position to the clivus. In addition, we lysed the subarachnoid adhesions, decompressed the craniovertebral junction, and restored normal craniospinal CSF dynamics; the intention was to stabilize further the pressure–volume relationship in the posterior fossa and to resolve the syringomyelia. Restoration of normal brainstem position and presumably relaxation of the cranial nerves prompted fairly rapid recovery of the cranial neuropathies.

To be believable, our postulate must be reconciled with the known fact that not everyone who undergoes shunt placement for an isolated fourth ventricle experiences cranial neuropathies.
Cranial nerve palsies and fourth ventricular shunt

Fig. 3. Contiguous axial CT scans obtained 3 weeks after fourth ventricular shunt placement revealing a slit fourth ventricle but the catheter tip is not touching the ventricular floor.

Fig. 4. Sagittal MR image obtained 8 weeks after fourth ventricular shunt placement revealing progressive backward retraction of the brainstem toward the slit fourth ventricle. Note the gap between the brainstem and the clivus is wider than that shown in Fig. 2 lower.

nial nerve palsy. In any system of hollow spheres with compressible walls, the change in volume is not governed alone by changes in pressure, but also by the compliance of the walls. In the case of an isolated fourth ventricle, the two wall components are the cerebellar hemispheres and the brainstem. The pliancy of the compressed (and thinned) cerebellum and its degree of adherence to the tentorium and suboccipital dura will ultimately determine how much the hemispheres collapse toward the center. The balance of the “suctioning” impetus will be borne by the viscoelastic
modulus of the brainstem. There is thus an expected variability in the amount of brainstem shift depending on the compliance and freedom of movement of the cerebellum. The unpredictability of symptoms is further explained by the unknown number of adhesions suspending the cranial nerves, which may augment the deleterious effect of traction.

We believe that backward shifting of the brainstem and cranial nerve traction injury occur more frequently than is generally recognized. Hirano, et al., presented one patient with delayed abducent palsy, which they thought was due to traction from ventricular shrinkage. Reexpansion of the ventricle by means of an on–off valve reversed the patient’s nerve palsy. In at least one patient reported by Eder, et al., sagittal MR imaging clearly demonstrated significant brainstem shifting that coincided with reduction of the fourth ventricle after shunt placement. In other instances, Eder, et al., and Rekate stipulated that the catheter tip was initially in the center of the fourth ventricle and “touched” the brainstem only after further ventricular shrinkage. Considering that the catheter is nonextendable, the only way its tip and the brainstem could meet in the interim would be backward movement of the brainstem. By implication, tension was thus placed on the cranial nerves.

To avoid these shunt-related complications, it has been suggested that the appropriate length of the fourth ventricular catheter be strictly predetermined and, most important, that a specific cannulation trajectory be adopted to place the catheter parallel instead of perpendicular to the ventricular floor. There have even been endorsements of CT-guided insertion of the catheter in a two-stage procedure. Adopting these measures will minimize the likelihood of direct catheter injury to the brainstem, but, if our theory were correct, would not prevent cranial nerve traction. To achieve the latter, a flow-regulated valve with antisiphon features may be used to maintain a positive intraventricular pressure. Intermittent fluctuations in fourth ventricular pressure, however, would still occur with changes in body posture, and shifting torque on the brainstem therefore could not be totally prevented. Alternatively, the closed cyst and surrounding subarachnoid pathways can be opened to allow CSF inflow into the fourth ventricle to avoid complete ventricular collapse. Inflow can be accomplished by performing an open craniotomy or by performing endoscopic fenestration through a limited suboccipital craniectomy.

We are certainly not advocating open posterior fossa craniectomy in every case of an entrapped fourth ventricle because progressive cranial neuropathies are an uncommon complication of routine fourth ventricular shunting. Open fenestration, however, is the most direct and immediate solution if brainstem shifting can be demonstrated and should be seriously considered. We also do not believe that combining the frontal and posterior catheters via a T-connector proximal to the valve will prevent brainstem shift because this does not reliably avoid siphoning of the fourth ventricle. Recently, Fritsch, et al., described performing endoscopic aqueductoplasty and placement of a stent from the side of the third ventricle to facilitate communication of the trapped fourth ventricle and the shunted lateral ventricle; this technique succeeded in eliminating the need for posterior fossa shunt placement in five patients.
patients tend to have slit lateral and third ventricles, conventional endoscopic technique can be difficult and fraught with risk for midbrain injuries. Perhaps there is a place for endoscopic aqueductoplasty and stenting by a posterior approach through the isolated fourth ventricle.

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References


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