LETTER TO THE EDITOR

To The Editor

Transtentorial Fluctuations and Atypical Parkinsonism After Ventriculo-Peritoneal Shunting

Keywords: Aqueductal stenosis, Hydrocephalus, Ventriculo-peritoneal shunt, Intracranial hypertension, Intracranial hypotension, Atypical Parkinsonism

Parkinsonism and oculomotor abnormalities have been reported in patients with aqueductal stenosis and hydrocephalus after treatment with ventriculo-peritoneal shunt (VPS). The exact pathophysiology is unknown, but it likely results from mechanical damage due to dorsal midbrain structures and their connections.[1–8] Here, we describe two patients who developed progressively worsening transtentorial pressure fluctuations, Parkinsonism, and oculomotor abnormalities after VPS.

A 35-year-old man underwent VPS placement after aqueductal stenosis and hydrocephalus were incidentally discovered upon CT brain for head trauma. One year later, he developed progressively more severe fluctuations between intracranial hypertension (headache, altered consciousness, and hydrocephalus) and intracranial hypotension (orthostatic headache with slit-like lateral ventricles). Any attempt to recalibrate VPS settings to treat hypertension resulted in a progressively faster return to hypotension. Similarly, any attempt to recalibrate VPS settings to treat hypotension resulted in a progressively faster return to hypertension. Three years later, he presented with diplopia and tremor. Two weeks after changing VPS to treat hydrocephalus, examination revealed eyelid retraction, supranuclear gaze palsy, and right-sided predominant Parkinsonism (MDS-UPDRS-III=39) (Supplementary Video, segment 1). Three weeks later, he presented with severe orthostatic headaches. Ocular findings were unchanged, but Parkinsonism worsened to include drooling, motor blocks, and severe right-hand tremor (MDS-UPDRS-III=51) (Supplementary Video, segment 2). CT brain revealed slit-like lateral ventricles and significant upwards displacement of the midbrain. Carbidopa/levodopa 187.5/750 mg/day was started with moderate improvement (MDS-UPDRS-III=33) (Supplementary Video, segment 3). Transtentorial pressure fluctuations were resolved with endoscopic third ventriculostomy, but Parkinsonism and oculomotor abnormalities persisted. Case 1 is summarized in Figure 1.

A 26-year-old man with a pineal tumor underwent VPS placement for aqueductal stenosis and hydrocephalus. Six months later, he developed transtentorial pressure fluctuations and mild, right-sided predominant Parkinsonism after VPS externalization for biopsy of the pineal tumor. Similar to Case 1, these fluctuations were aggravated by any attempt to recalibrate VPS settings (Figure 2). Ten days later, Parkinsonism worsened despite carbidopa/levodopa 75/300 mg/day (MDS-UPDRS-III=59) and he developed slowed vertical fast-phase ocular movements (Supplementary Video, segment 4). Transtentorial fluctuations stabilized after pineal biopsy and VPS internalization, but Parkinsonism and oculomotor abnormalities persisted.

Parkinsonism and oculomotor abnormalities were likely secondary to midbrain displacement, stretching, and compression due to fluctuating transtentorial pressures. External CSF drainage from the supratentorial compartment in patients with aqueductal stenosis could create a pressure gradient with the infratentorial compartment that predisposes to significant midbrain displacement through the tentorium, as well as third ventricle expansion and contraction. As previously reported, downstream frontal lobe dysfunction might be associated with levodopa-resistant Parkinsonism in these patients.[5] Remarkably, therapeutic attempts to recalibrate VPS settings were associated with progressive worsening in transtentorial pressure fluctuations and further midbrain displacement in the opposite direction. In Case 1, the fluctuations became clinically evident 1 year after VPS placement for congenital aqueductal stenosis. Midbrain damage was clinically apparent 2 years later and continued to progress until fluctuations were stabilized by endoscopic third ventriculostomy. In Case 2, worsening transtentorial pressure fluctuations and midbrain damage were triggered by VPS externalization for pineal tumor biopsy. Clinical progression continued until fluctuations were stabilized by VPS internalization. Dorsal midbrain compression by the pineal tumor caused aqueductal stenosis in this patient and could have contributed to the midbrain syndrome as well. Similar to previously reported cases,[3,5–7] Parkinsonism and oculomotor abnormalities stabilized but persisted after transtentorial fluctuations resolved. This persistence might reflect irreversible damage to dorsal midbrain structures and their connections. As opposed to VPS, endoscopic third ventriculostomy may decrease the risk of creating or abruptly changing the supratentorial/infratentorial pressure gradient with subsequent midbrain damage in these patients.

In conclusion, some patients with aqueductal stenosis and hydrocephalus may develop transtentorial pressure fluctuations with midbrain displacement, compression, and shearing leading to Parkinsonism and oculomotor abnormalities after VPS placement. Recalibration of VPS settings in these cases could worsen the pressure fluctuations and provoke further midbrain injury despite subsequent stabilization of intracranial pressure.

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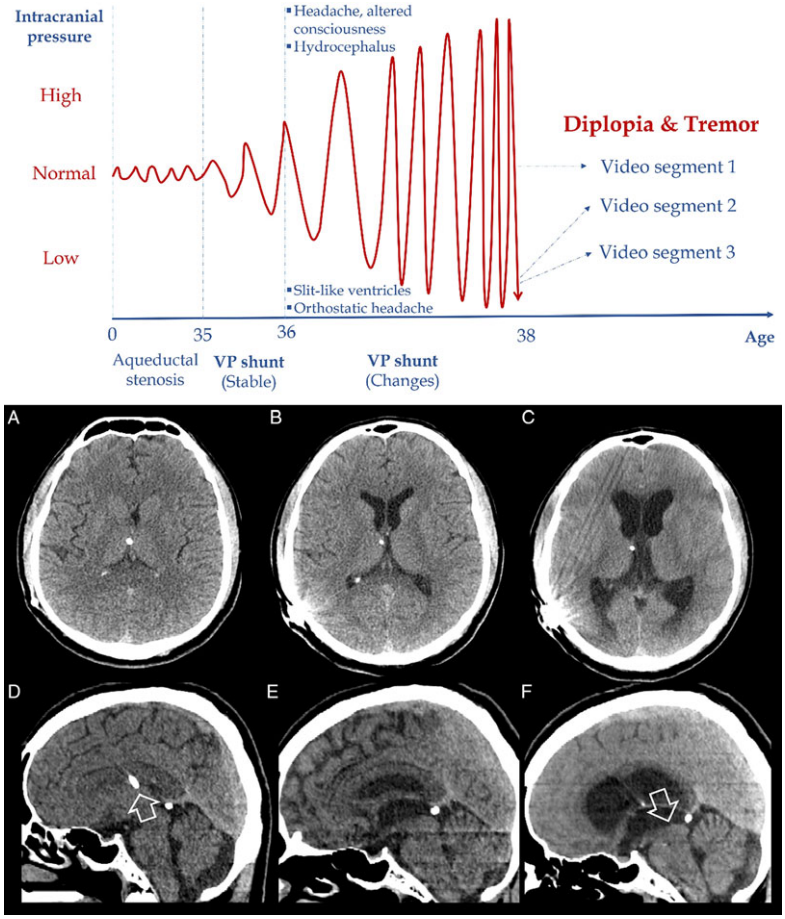

Figure 1: *Upper section: Timeline illustrating the progressive intracranial pressure fluctuations in Case 1. After this patient with congenital aqueductal stenosis was treated with ventriculo-peritoneal shunt (VPS), he developed progressively worsening transtentorial pressure fluctuations that led to alternating intracranial hypertension and hypotension. Fluctuations worsened with each therapeutic attempt to modify VPS settings and he eventually developed diplopia and tremor (see Supplementary Video, Segments 1–3). Lower section: Sequential axial and sagittal brain CT images corresponding to Case 1. (A) and (D) demonstrates transtentorial midbrain stretching and displacement toward the supratentorial compartment during an episode of intracranial hypotension. (C) and (F) demonstrates transtentorial midbrain compression and displacement toward the infratentorial compartment during an episode of intracranial hypertension. (B) and (E) were obtained between episodes of intracranial hypertension and hypotension.*

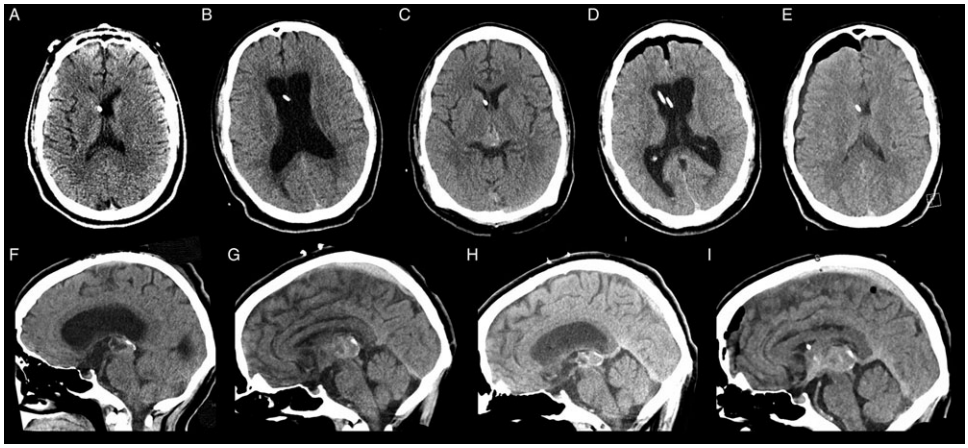

Figure 2: *Sequential axial and sagittal brain CT images corresponding to Case 2. (A) shows this patient's baseline, prior to the externalization of his VPS. (B) and (F) show worsening hydrocephalus (day 3). (C) and (G) shows resolved hydrocephalus (day 4), with stretching and displacement of the pineal mass and midbrain toward the supratentorial compartment. (D) and (H) shows interval worsening of hydrocephalus and pneumocephalus (day 6), with compression and displacement of the pineal mass and midbrain toward the infratentorial compartment. Є and (I) shows slit-like ventricles, subdural hygroma, and persistent pneumocephalus on day 10, with stretching and displacement of the pineal mass and midbrain toward the supratentorial compartment.*

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Disclosures

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Statement of Authorship

DSS: Project conception and execution, writing of the first draft of the manuscript. JM: Project conception and execution, review, and critique of the manuscript. CS: Project conception and organization, review, and critique of the manuscript. KJL: Project conception, organization and execution, review, and critique of the manuscript.

Supplementary Material

To view supplementary material for this article, please visit https://doi.org/10.1017/cjn.2020.228.

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