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Miyazaki syndrome: Cervical myelo/radiculopathy caused by overshunting. A systematic review

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ABSTRACT

Objectives: Miyazaki syndrome is a cervical myelopathy or radiculopathy caused by cervical epidural venous congestion, due to cerebrospinal fluid over-drainage by an implanted ventricular shunt. The complex pathophysiology includes CSF pressure-changes consistent with the Monro-Kellie doctrine and a non-functional Starling resistor, leading to spinal epidural venous plexus enlargement and dilation. This venous congestion may be significant enough to exert compression on the spinal cord or nerve roots. The typical clinical and imaging findings together with a history of ventricular CSF shunting may establish the diagnosis, proven by a successful treatment. The aim of treatment is the abrogation of CSF over-drainage. The eligible interventions may be the followings: the increase of the opening-pressure of the valve system by the insertion of a new programmable valve if necessary, closing or removing the shunt.
Aim: We want to call attention to this rare iatrogenic condition with potentially severe consequences.
Patients and Methods: We perform a systematic literature-review and present our five cases.
Results: Once recognized in time, Miyazaki syndrome can be well taken care of.
Conclusions: Patients with chronic ventricular shunt need monitoring for CSF over-drainage to recognise potential complications such as cervical myelopathy or radiculopathy.

1. Introduction

Over-drainage is a well-known complication affecting patients with implanted ventricular (ventriculo-peritoneal - VP, or ventriculo-atrial - VA) shunts [5,24].

Rarely, overshunting may lead to cervical cord signs called “Miyazaki-syndrome”, (MiY) [3,8] or “overshunting-associated myelopathy” [2,13,16,32]. In these cases, overshunting of cerebrospinal fluid (CSF) results in cervical epidural venous plexus dilation and venous engorgement leading to cervical cord or nerve root compression. Typically, there is no orthostatic headache. CSF over-drainage due to lumbo-peritoneal shunt may have similar complications, but these cases are not referred to as MiY respecting the original definition [2,3,5,8,13,16,32].

To our knowledge, there are just a few published MiY cases and the pathomechanism is unclear. We aim to call attention to this rare condition of practical significance.

2. Methods and materials

We performed a systematic review of the literature according to PRISMA (Preferred Reporting Items for Systematic reviews and Meta-Analyses) guidelines (Table 1). We searched "Miyazaki syndrome", OR "over-shunting myelopathy", OR "cervical myelopathy, veins, hypotension shunt" OR, "myelopathy, cervical veins" in PubMed and Google Scholar online electronic databases. We collected those cases meeting the following inclusion criteria: implanted ventricular shunt, enlargement of the epidural venous plexus with clinical signs of cervical spine or nerve root compression. We have reviewed our cases as well. The Institutional Ethics Committee approved our study and we obtained a written informed consent from our patients. We collected data of both literature- and own cases on patient age and gender, indication of shunt-implantation, shunt-type and, the onset of cervical symptoms following the last shunt surgery, clinical signs, radiology findings, treatment and outcome.

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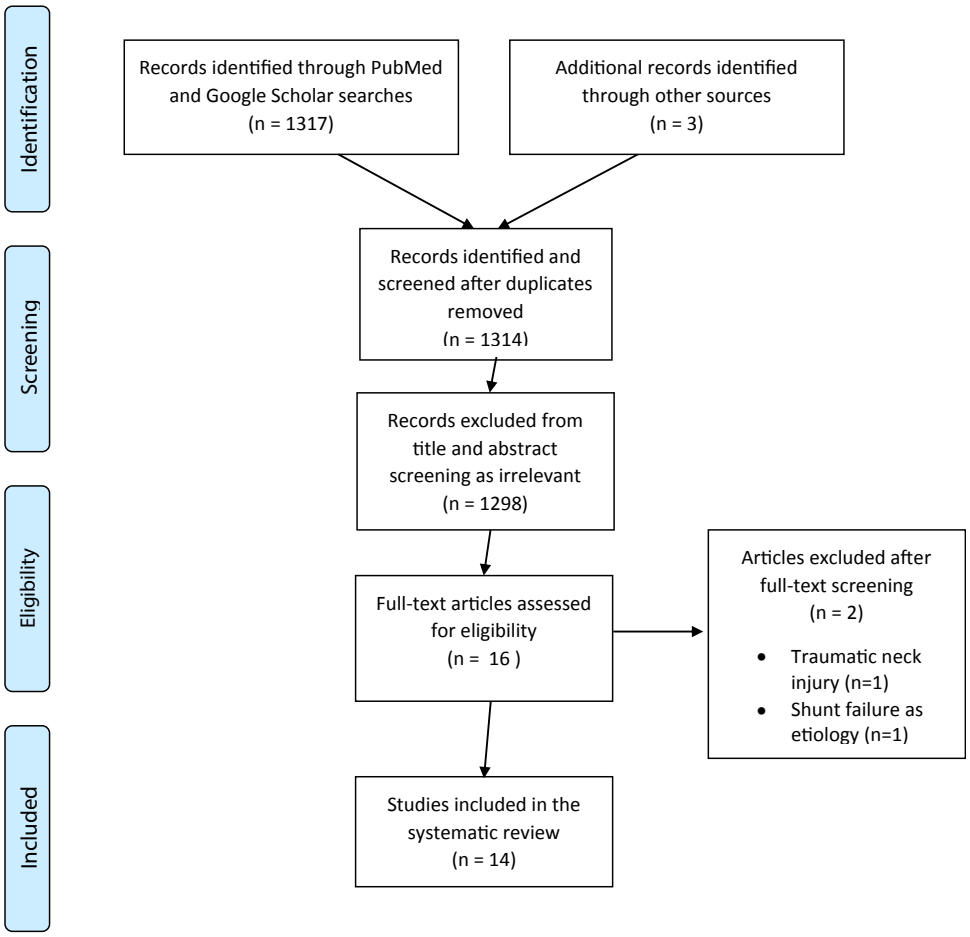
Table 1
PRISMA 2009 Flow Diagram.



3. Results

We identified 1317 papers in the online databases and three additional cases in references of papers. Excluding duplicate cases, 1314 studies remained. Based on title and abstract screening we found 1298 of them irrelevant for our review. Screening the full-text of the remaining sixteen studies, we found 14 of them meeting our inclusion criteria reporting on 15 patients (Table 2). We found five patients in our institutional database (Table 3).

In literature-cases, the indications for VP shunt placement were the followings: hydrocephalus related to subarachnoid haemorrhage, congenital hydrocephalus, postoperative intracranial pressure-increase, posterior fossa meningioma and arachnoid cyst. The mean age of patients was 43 years (17–77); eight males. In these published cases, an average of 12 years passed between shunt-insertion or last shunt-revision and the onset of cervical signs, lasting for 4 weeks to 7 years until the diagnosis of MiY. Motor and pyramidal signs ranging from brisk reflexes to spastic para-, or tetraparesis were the most frequent; upper limb sensory symptoms, ataxia and finger-clumsiness occurred. Two patients developed tetraplegia; their symptoms lasted the longest (3 and 7 years) without treatment, suggesting the progressive nature of MiY [2,8].

Headache and visual symptoms occurred in one patient [30]. Three patients had neck pain [7,11,22]. Mild cognitive impairment developed in one patient [31].

Magnetic resonance imaging (MRI) of the brain and spinal cord was performed in each case. All patients had a space-occupying enlargement of the epidural venous plexus and/or epidural veins narrowing the subarachnoid space, deforming the cervical cord and encroaching upon the exiting nerve roots. In one case, focal meningeal thickening might have contributed to the compression. Brain imaging revealed slit ventricles, midline shift, pituitary gland enlargement, low-lying cerebellum, subdural fluid collections, dilated dural sinuses and diffuse meningeal enhancement (Table 1). Bone thickening of the calvaria occurred as well [30].

In addition to the typical radiological presentation, low lumbar CSF pressure [19,34,35] and low intracranial pressure supported the diagnosis [8,31] in some cases. An improvement seen after increasing the opening pressure of an adjustable valve [2,22] had a diagnostic value, besides – and most importantly – being the aetiological treatment.

Four methods of resolving the CSF over-drainage were used: insertion of a programmable valve, [11,14,16,19,30,31,35] increasing its opening pressure [2,7], closing the shunt [8,23] and, shunt-removal [22].

3.1. Our patients

3.1.1. Patient 1

This now 38-year-old female patient underwent VP shunt-placement in infancy for hydrocephalus. Due to shunt insufficiency, we changed her ventricular shunts repeatedly. For treating a more recent failure, we inserted a new medium-pressure adjustable valve-system.

She presented with dizziness, upper limb paraesthesia and ataxia. The MRI revealed myelopathic signal changes in the upper cervical spinal cord. The epidural space/plexus enlarged ventral to the cervical cord, mainly at C2-C3 and dorsal to the clivus (Fig. 1).

Table 2
Literature cases.

Study Age year/ Gender	Indication of shunt implantation	Shunt type	Time-log of the onset of symptoms after last shunt implantation	Duration of cervical signs	Clinical signs	Radiology findings	Treatment	Outcome
Miyazaki et al., 53/M[9]	Subarachnoid hemorrhage induced hydrocephalus	Flow regulated valve; low pressure; no antisiphon device	1,25 years	7 months	bilateral palmar paresthaesia, hypalgesia, hypaesthesia; C2 dermatome hypalgesia; finger clumsiness; gait ataxia; Romberg sign; generalized hyperreflexia; deep sensory disturbance	Midline shift; epidural venous plexus enlargement; diffuse meningeal enhancement	Closing the shunt	Improvement
Matsumoto et al., 67/M[10]	Obstructive hydrocephalus caused by pilocytic astrocytoma	Programmable valve system	2 years	unknown	nuchal, shoulder pain; spastic ataxia	diffuse meningeal enhancement; extramedullary mass lesion	Shunt removal	Improvement
Wingerchuk et al., 72/F[11]	Posteror fossa meningioma	*no information*	23 years	4 years	spastic tetraparesis; generalized hyperreflexia; deep sensory disturbance	slit ventricles; dural enhancement; subdural fluid collections; epidural venous plexus enlargement	None	Worsening
Liu et al., 18/F[12]	Large porencephalic cyst	Distal slit-valve system	16 years	4 months	spastic tetraparesis; generalized hyperreflexia; bilateral clonus	enlarged epidural venous plexus	Shunt replacement with programmable valve	Improvement
Wolfe et al., 17/M[13]	Pilocytic astrocytoma resection cavity	VP shunt; no antisiphon device	15 years	unknown	spastic tetraparesis; generalized hyperreflexia; Babinski and Hoffman positive bilaterally	Diffuse meningeal enhancement; brain shift; jugular veins collapse bilaterally; epidural venous plexus enlargement	Valve replacement with programmable valve	Improvement
Ulrich et al., 17/M[14]	Obstructive hydrocephalus caused by retrocerebellar arachnoid cyst	VP shunt, replaced 6 years later (shunt failure)	14 years	2 years	progressive paraparesis	bone thickening of the calvarium; prominence of the hypophyseal gland; epidural venous plexus enlargement	Valve replacement with programmable valve	Slight improvement
Howard et al., 26/F[7]	Congenital hydrocephalus	VP shunt with 2 revisions, medium pressure	1/12 year	4 weeks	upper limb dysaesthesia; ataxia; generalized hyperreflexia; pyramidal signs	diffuse meningeal enhancement; pituitary gland enlargement; low-lying cerebellum; epidural venous plexus enlargement	Valve replacement with a programmable valve	Improvement
Cardoso et al., 32/M[15]	Communicating hydrocephalus	VP shunt (first non-programmable, then programmable)	10 years	few months	spastic tetraparesis	*epidural venous plexus enlargement*	Valve opening pressure was increased	Improvement
Caruso et al., 35/M[3]	Supra,- and infratentorial arachnoid cysts, agenesis of the corpus callosum	VP shunt, high pressure valve, no antisiphon device	19 years	7 years	progressive spastic tetraplegia	epidural venous plexus enlargement	Closing the shunt	Improvement
Dantas et al., 33/F[16]	Communicating hydrocehalus	VP shunt, high pressure valve	2 years	*unknown*	neck pain, headache, visual deficits, tetraparesis, paresthesias	epidural venous plexus enlargement	Valve replacement with programmable valve	Improvement
JM Ho et al., 64/F[17]	Subarachnoid hemorrhage induced hydrocephalus	VP shunt, medium pressure	14 years	12 months	spastic tetraparesis, gait ataxia, deep sensory disturbances, intention tremor	diffuse meningeal enhancement; epidural venous plexus enlargement	Valve replacement with programmable valve	Improvement
JM Ho et al., 22/F[17]	Congenital hydrocephalus caused by posterior fossa arachnoid cyst	VP shunt (shunt revision at age 17)	5 years	2 months	neck pain, "sunken eyes", numbness of palms, bilateral hand weakness, generalized hyperreflexia	diffuse meningeal enhancement; small ventricle size; epidural venous plexus enlargement	Valve replacement with programmable valve	Improvement
Martinus et al., 69/M[18]	Hydrocephalus after cerebellar cyst resection	VP shunt	38 years	few years	mild cognitive impairment, gait ataxia, spastic left upper extremity, Babinski positive	diffuse meningeal enhancement; pituitary gland enlargement; epidural venous plexus enlargement	Valve replacement with new valve	Improvement
Amano et al., 7/M[8]	Subarachnoid haemorrhage induced hydrocephalus	VP shunt; no anti-syphon device	10 years	3 years	Tetraplegia; generalized hyperreflexia; hypaesthesia, hypealgesia below C3; deep sensory disturbances; plantars going up	epidural venous plexus enlargement	Valve opening pressure was increased; laminectomy (spondylosis)	Improvement
			10 years		Paraesthesias of the limbs			Improvement

(*continued on next page*)

Table 2 (*continued*)

Study Age year/ Gender	Indication of shunt implantation	Shunt type	Time-log of the onset of symptoms after last shunt implantation	Duration of cervical signs	Clinical signs	Radiology findings	Treatment	Outcome
Kovács A et al. 33/F[19]	Communicating hydrocephalus	pressure-adjustable VP Hakim valve system		a few months		Diffuse thickening and intense contrast enhancement of the dura mater together with dilation of the venous sinuses. Pituitary gland strongly convex. Marked dilation of the epidural venous system almost along the whole spine. Narrowing of the spinal canal and compression of the spinal cord with associated slight myelopathy	The opening pressure of the Hakim valve was increased. One month later, as the patient recovered, the valve was replaced and by further increasing the opening pressure, it was closed. Besides, an antisiphon device was built into the shunt system to prevent overshunting	

Table 3
Our Miyazaki syndrome patients.

Patient age/ gender	Indication of shunt implantation	VP Shunt type	Age at last shunt implantation	Time interval (years) between last shunt intervention and symptom onset	Clinical signs	MRI findings	Treatment
1. 38 / F	Infant hydrocephalus	adjustable valve	38	< 1	bilateral upper limb paresthesia, ataxia + dizziness	Enlarged epidural venous plexus, Myelopathy	Valve opening pressure set to high
2. 37 / M	Hydrocephalus due to aqueductal stenosis	non-adjustable valve	6	30	nausea, dizziness + ataxia	Enlarged epidural venous plexus, Myelopathy	Shunt replacement to adjustable valve system
3. 42 / F	Hydrocephalus due to aqueductal stenosis	adjustable valve	14	27	bilateral hand weakness and clumsiness	Enlarged epidural venous plexus, Nerve root compression	Shunt replacement to another adjustable valve system with higher opening pressure
4. 26 / F	Hydrocephalus due to aqueductal stenosis	adjustable valve	Childhood	Several years	spinal ataxia, neck pain, increased lower limbs jerks + headache, dizziness, nausea, galactorrhea	Enlarged epidural venous plexus, Slight cord deformity	Valve opening pressure set to high
5. 16 / M	Hydrocephalus due to meningitis	non-adjustable valve	2	14	hand muscle atrophy, polyneuropathy	Enlarged epidural venous plexus, Nerve root compression	Follow up(he improved with physiotherapy) 10 years later shunt replacement to adjustable valve system

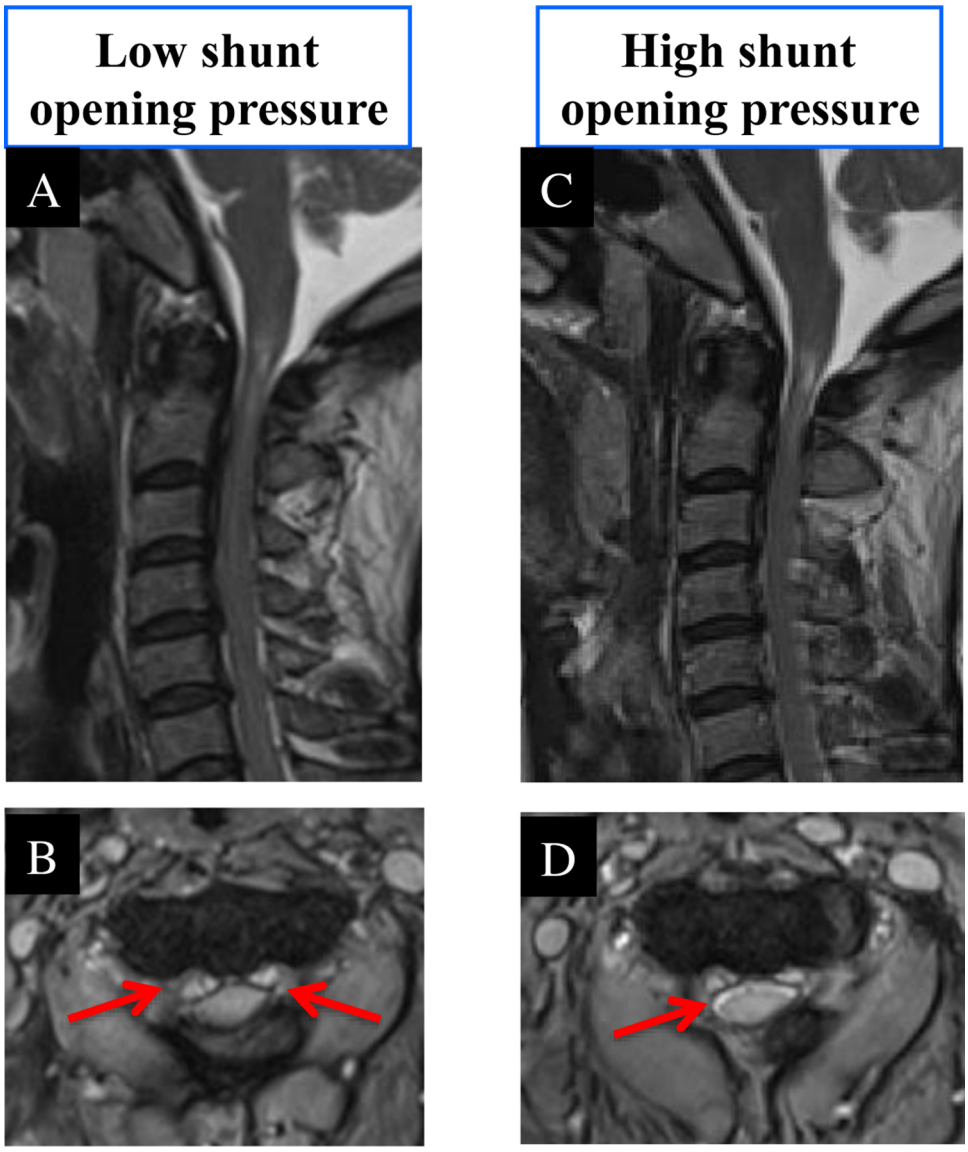


Fig. 1. Patient 1 with bilateral upper limb paraesthesia and ataxia. Note that the patient has also congenital spinal canal narrowing, aggravating the changes secondary to CSF hypotension. **A and B**: when the shunt opening pressure is low, there is significant dilation of the epidural venous plexus with complete effacement of CFS around the mildly deformed cord, well seen at the level of C2 on B (arrows). There is myelopathy at C1-2. **C and D**: when the shunt opening pressure is set to high, the dilation of the epidural venous plexus has decreased with associated reappearance of the peri-medullary subarachnoid space, best seen on the axial image, D (arrow). No significant interval change of the myelopathy, but clinically improved.

Imaging studies and clinical history suggested CSF hypotension secondary to shunt over-drainage. We raised the opening pressure of her valve (Sophysa SM8) from 110 to 170 mmH_2O and later to 200mmH_2O. During the 7-month follow-up, she has improved both clinically and radiologically (Fig. 1).

3.1.2. Patient 2

This 37-year-old man had a history of VP shunt implantation with a medium fixed-pressure valve (Pudenz) at age six for congenital aqueduct stenosis. Later, he developed nausea and dizziness with minimal CSF hypotension signs on MRI. More recently, he presented with ataxia. MRI of the cervical spine revealed the enlargement of the cervical epidural plexus narrowing the subarachnoid space with intra-medullary signal change. This suggested CSF hypotension caused by overshunting. We performed shunt revision introducing an adjustable valve (Sophysa SM8) with 170 mmH_2O opening pressure and a new catheter. In addition to the radiology regression of CSF hypotension signs, the patient has improved clinically, too, and when we raised the CSF opening pressure to maximum resistance, she has recovered (Fig. 2).

3.1.3. Patient 3

This 42-year-old female had bilateral VA shunt implantation at age 8 for resolving her hydrocephalus caused by congenital aqueduct stenosis. It was replaced by a VP shunt at age 14. She did well for long with some learning difficulties and unchanged dilated lateral ventricles (Fig. 3) and she developed bilateral hand weakness and clumsiness recently. Cervical MRI revealed bilateral cervical nerve root compression secondary to cervical vein engorgement and a dilated epidural venous plexus (Fig. 3).We replaced her Sophysa SM8-200 valve to Sophysa SM8-300, which was set at 300. Her radiological and clinical signs of CSF over-drainage have been improving.

3.1.4. Patient 4

This female patient underwent an endoscopic 3rd ventriculotomy, then an adjustable VP shunt implantation for congenital aqueduct stenosis and hydrocephalus in early childhood. At age 26, she presented with spinal ataxia, neck pains, brisk lower limb reflexes, headache, dizziness, nausea and galactorrhoea. Her VP shunt-valve was at a low opening pressure (50mmH_2O). MRI of the spine revealed an enhancing cervical epidural mass and thickening of the thoracic dorsal epidural fat with enhancing structures, and the MRI brain showed diffuse dural thickening. At this point, a chronic inflammatory granulomatous process involving the dura and epidural space was erroneously diagnosed, so she received steroids, resulting in a temporary clinical improvement. However, because there was no interval-change on follow-up MRI, we performed a C2-C4 laminectomy to decompress the spinal canal.

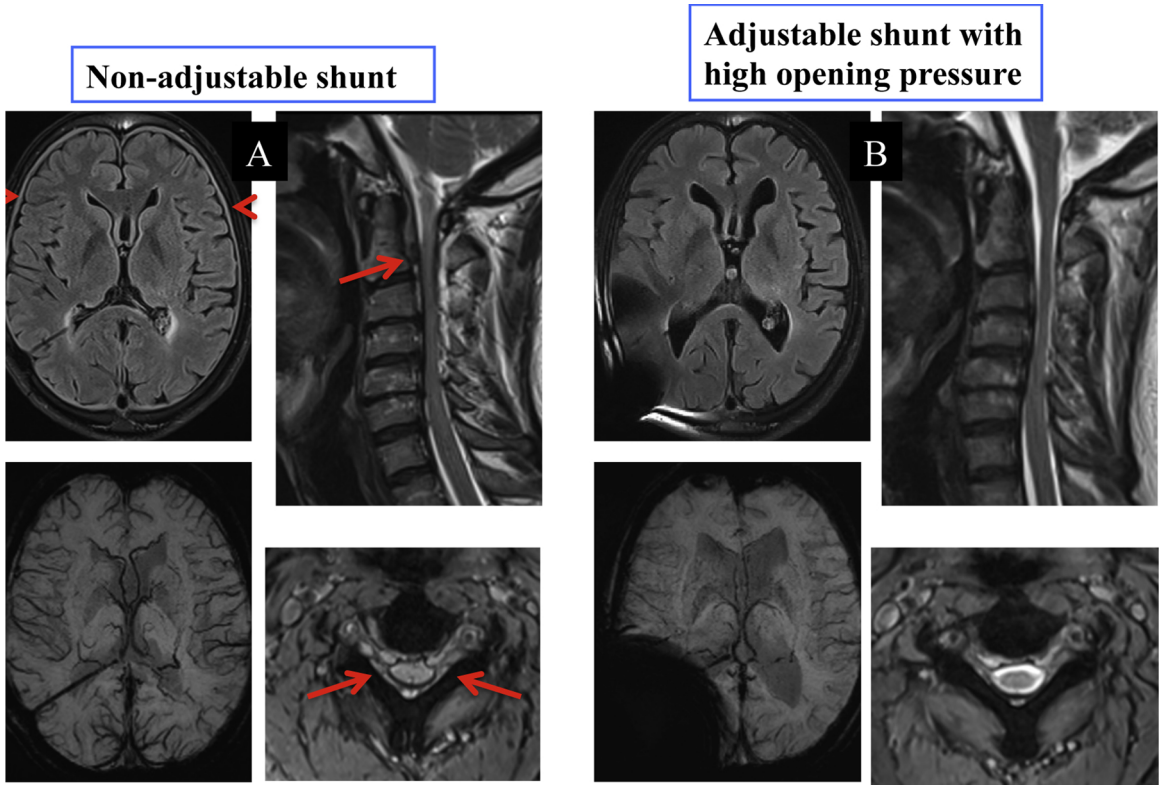


Fig. 2. Patient 2 with progressive ataxia. Axial FLAIR and SWI images of the brain as well as sagittal T2-weighted images of the cervical spine and axial T2-weighted images at C2/3. **Panel A**: with a non-adjustable shunt, there are signs of CSF hypotension with bilateral dural thickening on convexities (arrowheads), narrow ventricles and enlarged, dilated epidural venous plexus with associated obstruction of CSF spaces around the cord, particularly at C2/3 (arrows). Note the prominent veins on the SWI image due to generalized venous dilation. **Panel B:** After shunt revision with an adjustable valve system and opening pressure set to high, the ventricles are larger and there are no signs of CSF-hypotension: no diffuse intracranial dural thickening and the size of the cervical epidural venous plexus normalised with CSF again present around the cord. Note the obvious normalisation of cerebral veins on the SWI image. There is susceptibility artefact caused by the new shunt valve.

Reviewing her clinico-radiological data after surgery, we diagnosed CSF hypotension and MiY. After we had increased the shunt opening pressure, the epidural mass (consistent with an enhancing venous plexus) and the intracranial dural thickening disappeared (Figs. 4a-b) and her clinical symptoms improved.

3.1.5. Patient 5

The 16-year-old male patient had an old non-adjustable Pudenz shunt implanted in infancy for post-infectious hydrocephalus. His first shunt surgery occurred at age 8 months, then a shunt revision at age 2 years. At age 16, he developed right hand muscle atrophy and radiculopathy. Cervical spine MRI revealed narrow CSF spaces around the cord and a dilated epidural venous plexus encroaching on the nerve roots at several levels (Fig. 5). Physiotherapy resulted in some symptomatic improvement of his then unrecognised MiY. Ten years later, he presented with headache and double vision caused by Parinaud syndrome. CT brain showed dilated ventricles. When we replaced his old shunt by an adjustable valve system (Sophysa SM8) and the opening pressure was set to medium, he recovered.

4. Discussion

The typical symptom of intracranial hypotension is orthostatic headache [17]. It might be the consequence of the pressure or traction of pain-sensitive structures exerted by the descent of the brain [15,28]. CSF hypotension causing venous congestion and spinal cord compression (with no headache) is less known [1,6,9,25–27,29,33,12]. In these cases, an enlarged epidural venous plexus may compress the cord or nerve roots; focal meningeal thickening may contribute to the compression [22]. Martínez-Lage and colleagues reported on a patient with an inserted VP shunt, who had an incidental finding of cervical epidural venous plexus-engorgement after a road traffic accident [21]. Because the patient had no clinical signs of myelopathy or radiculopathy, one could consider this case a “silent” asymptomatic precursor of MiY frequently seen in radiology practice.

MiY has a complex, multifactorial pathophysiology. The Monro-Kellie doctrine may have an impact: [10] there is constant volume within the rigid skull and spine maintained by neural tissue, CSF and blood. If the volume of one of the components decreases, a compensatory increase of the others necessarily follows. In case of CSF-loss, the thin-walled venous system provides the first compensatory mechanism: dilated dural sinuses, pachy-meningial thickening with dilated blood vessels and engorgement of the spinal epidural venous plexuses evolve. If this compensation is insufficient, fluid may transudate from the dilated veins of the pachy-meninges resulting in thin subdural fluid collections. An insufficient Starling resistor mechanism may be another factor [3]. The starling mechanism ensures continuous flow within collapsible tubes, such as the bridging veins within the subarachnoid CSF [16]. It maintains the hydrostatic pressure-hierarchy of the fluid compartments within the skull: pressure (P) arterial > P venous > P CFS > P superior sagittal sinus (SSS). Normally, when taking an upright posture the pressure of the SSS falls, which – without a buffering mechanism – would result in overdraining and siphoning of the cerebral venous blood leading to intracranial hypotension. The Starling resistor is a site of compression at the junction between the bridging veins and the SSS, using even sphincter-like smooth muscles as additional tools narrowing these junctions [4,10]. When standing up, the pressure of both the SSS and CSF drops, but CSF pressure drops less, remaining

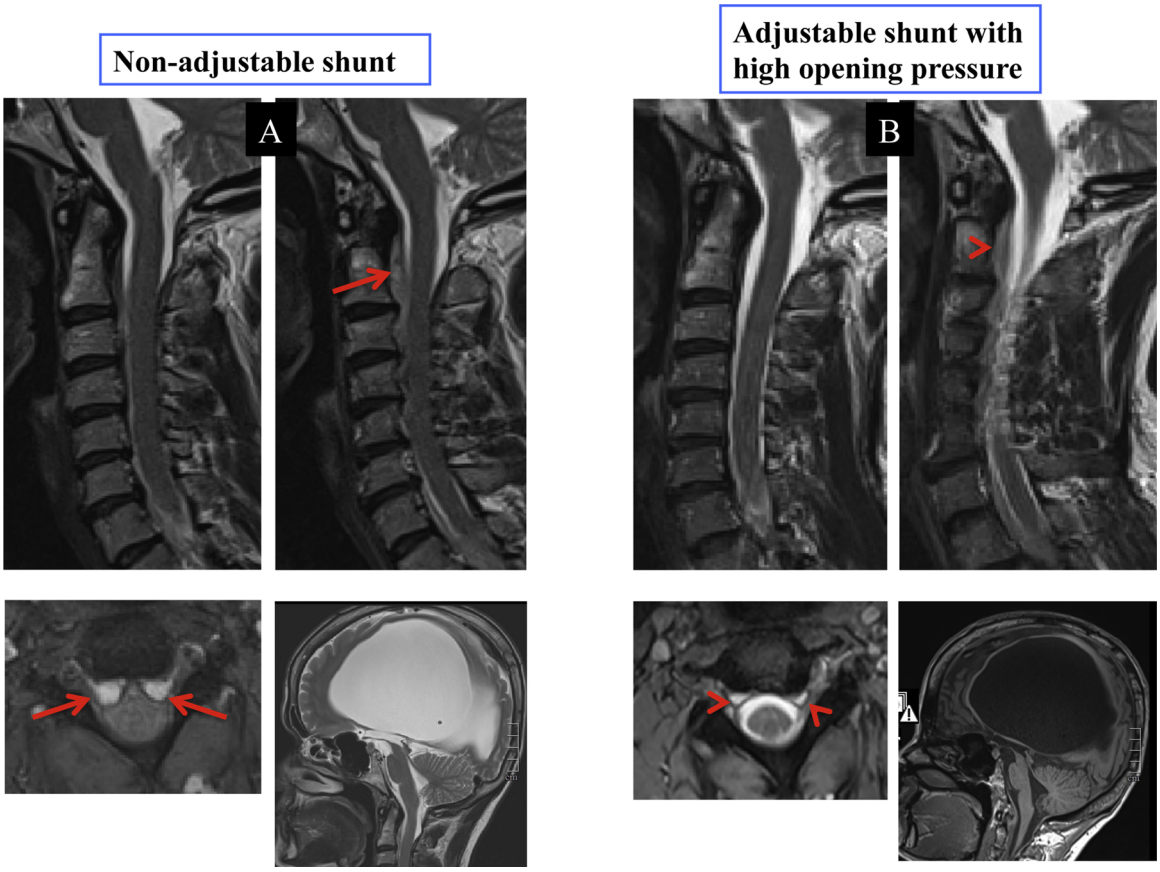


Fig. 3. Patient 3 with bilateral hand weakness and clumsiness **Panel A:** when the patient had a non-adjustable shunt with CSF over-drainage and hypotension, the sagittal midline, off-midline and axial T2-weighted images show very narrow CSF spaces around the cord and a significantly thickened epidural venous plexus at C2-3 (long arrows). There was chronic ventricular dilation. **Panel B:** When the shunt was replaced to an adjustable one with high opening pressure, CSF over-drainage was hampered and the signs of hypotension disappeared: there is normalization of the size of the epidural venous plexus (arrowheads) and CSF spaces are of normal appearance around the cord. Severe chronic dilation of the ventricles is unchanged.

higher. Thus, it can compress the bridging vein-junctions preventing venous over-drainage and maintaining venous pressure (higher than CSF pressure) and flow, proximal to the compression site [18,32]. In case of CSF diversion e.g. a VP shunt, the CSF-loss makes the Starling resistor non-functional: the decreased CSF volume cannot exert sufficient pressure to compress the cerebral bridging vein junctions resulting in venous over-drainage. A contributory factor may be the collapse of the internal jugular veins in upright posture, leading to alternative venous pathways: the cerebral venous blood outflows into the spinal epidural veins directly communicating with the cerebral venous system through the wide sub-occipital sinus. Therefore, in case of VP shunt CSF over-drainage, a significant enlargement of the epidural venous plexus and/or engorged epidural veins evolves that may compress the cord or nerve roots.

One may wonder how the soft and low-pressure venous engorgement can exert that severe compression, even sufficient to cause myelopathy. There might be a hidden blockage in the cervical venous flow contributing to the pressure-increase in the dilated venous plexus [20]. Such a coincidental venous blockage could explain the extreme rarity of MiY. On the other hand, a milder spinal epidural venous plexus-dilation occurs in the majority of spontaneous intracranial hypotension patients, further suggesting the primary role of CSF loss.

Our patients presented with dizziness, nausea, ataxia, bilateral upper limb weakness, clumsiness and paraesthesia. We detected an enlarged epidural venous plexus in each case. There was cord compression/deformity in three cases (1, 2 and 4) with myelopathic signal changes in patients 1 and 2. Radiculopathy was the main feature in patients 3 and 5 (Table 3). The radiology signs of intracranial hypotension caused by shunt overdainage are similar to those seen in intracranial hypotension of other aetiologies; e.g. dural thickening with contrast enhancement and dilated venous sinuses. Slit ventricles are typical in CSF hypotension, but dilated ventricles as in our patient 3 do not contradict to it, if there is no periventricular CSF imbibition or other associated typical hypertension signs. In such cases, the loss of brain mantle caused by early, long lasting hydrocephalus may lead to the persisting ventricular dilation despite low CSF pressure.

We felt that the radiology features, and in particular the spinal radiology signs of incracranial hypotension (primarily a dilated, thickened epidural plexus) associated with clinical myelo- or radiculopathy were sufficient to support the diagnosis of shunt overdrainge.

We have successfully increased the opening pressure of the adjustable valve system or replaced the non-adjustable valve by a programmable one setting them to medium and high pressures. Each technique was effective during 2–7 months follow-up.

5. Conclusions

Miyazaki syndrome is a rare cervical myelopathy caused by CSF volume loss and intracranial hypotension due to shunt overdrainage. The complex pathophysiology of myelopathy or radiculopathy involves a significantly dilated and thickened epidural venous plexus seen on MRI. The aim of treatment is abrogating CSF overdrainage by increasing the valve's opening pressure, replacing the valve to a higher-pressure one, closing/removing the shunt or implementing an antisiphon valve. Recognition of MRI findings is crucial to establish the diagnosis. Treatment may result in improvement or complete recovery.

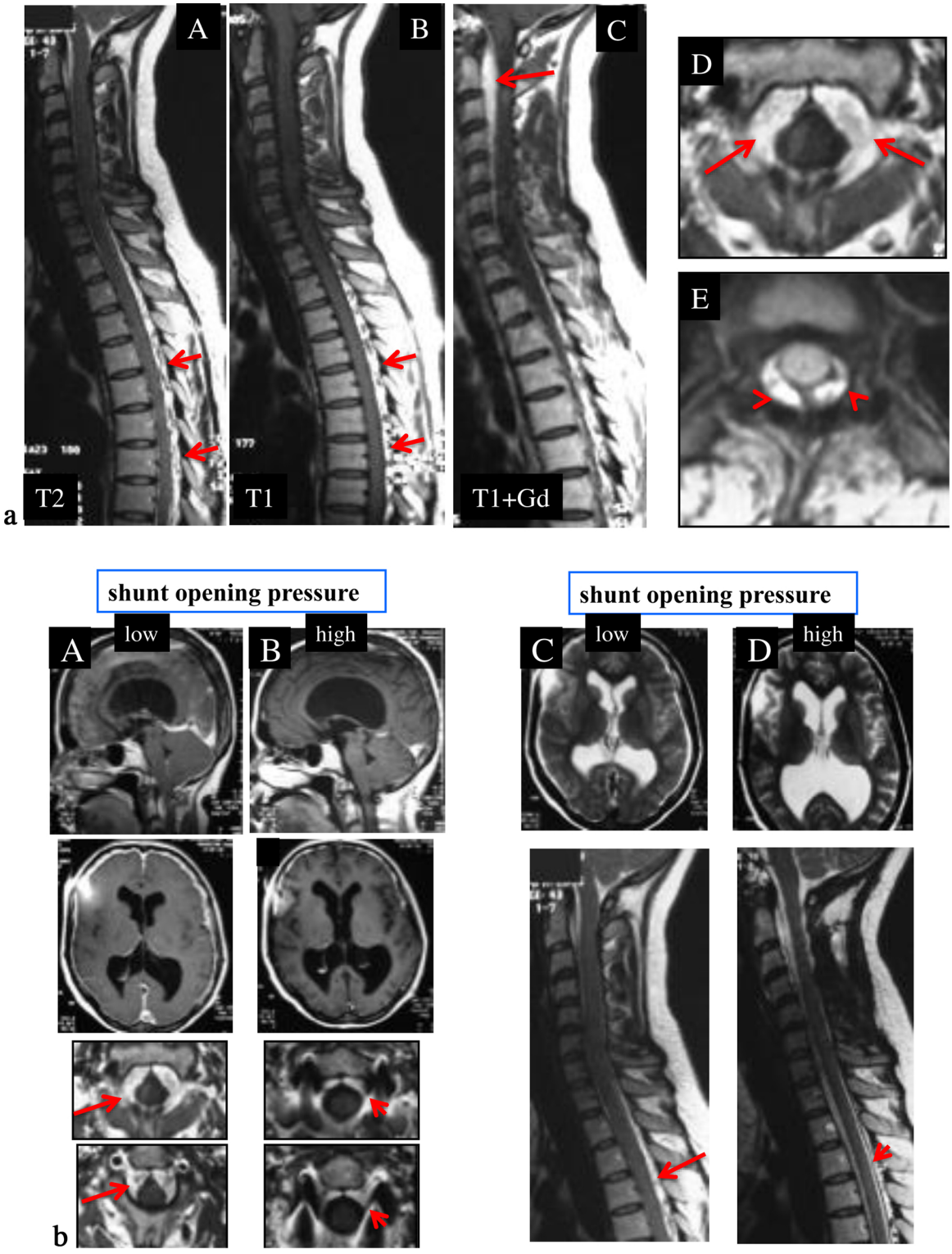


Fig. 4. a: Patient 4 with spinal ataxia and neck pain. Sagittal midline T2-weighted image (**A**) and T1-weighted image (**B**) show significantly narrow subarachnoid spaces with essentially no CSF seen around the cord. The dura is elevated from the ventral margins of the spinous processes in the thoracic spine by the expanded epidural space in which tortuous, dilated veins are present (short arrows). Sagittal off-midline **(C)** and axial contrast-enhanced T1-weighted (**D**) images demonstrate substantial thickening of the cervical epidural plexus, particularly at C2-C3 (long arrows). Axial T2-weighted image **(E)** at the mid-thoracic level also reveals the widened epidural space with dilated veins, significantly narrowed subarachnoid spaces and cord encroachment (arrowheads). b: Patient 4 with spinal ataxia and neck pain. Changes of signs of hypotension after increasing shunt opening pressure. **Panel A and C**: when shunt-opening pressure is set to low, there is easy over shunting resulting in relatively small ventricles with associated signs of hypotension, ie. significant diffuse thickening of the dura, thin subdural collection, dilated dural sinuses, narrowed sulci, basal cisterns and spinal subarachnoid spaces, sagging of the brain as well as substantial enlargement of the epidural venous plexus at C2/3 (long arrows) and widening of the dorsal epidural space in the upper thoracic spine (long arrow). **Panel B and D**: when shunt-opening pressure is set to high and CSF drainage is hampered, the ventricles are larger and there are no hypotension signs. Note the normalisation of the size of the cervical epidural venous plexus and the thoracic dorsal epidural space with CSF again around the cord and the reappearance of the dura (short arrows)!.

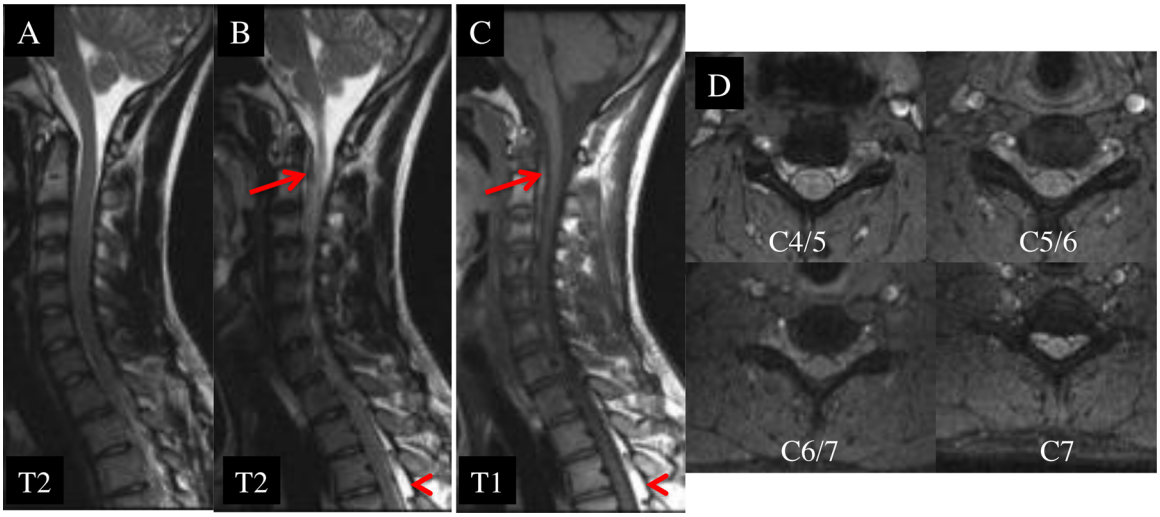


Fig. 5. Patient 5 with hand muscle atrophy and polyneuropathy on EMG.Sagittal T2-weighted (**A, B**) and T1-weighted images (**C**) show significantly narrowed spinal subarachnoid spaces with thickening of the upper cervical epidural venous plexus (arrows) and widening of the thoracic dorsal epidural space (arrowheads). Note, that there is even flow void within a dilated vein in the epidural plexus at C2 indicating prominent flow (B arrow). Axial T2-weighted images (**D**) at different levels reveal effaced subarachnoid spaces due to an enlarged epidural venous plexus with encroachment on the nerve roots.

Table 4
Spinal epidural conditions that may cause cord compression and myelopathy.

Unclassified	Neoplastic	Vascular	Infectious
Herniated intervertebral disc	Spinal benign tumours incl. haemangioma	Spinal epidural haematoma	Spinal epidural abscess
Epidural lipomatosis	Spinal malignant tumours	Vascular malformations	Vertebral osteitis/osteomyelitis
Epidural fluid collection			
Vertebral Paget's disease			Pyogenic discitis

Patients with chronic ventricular shunt need monitoring to recognise potential CSF overdrainage and its complications such as cervical myelopathy or radiculopathy. MiY needs to be considered in the differential diagnosis of epidural masses in ventricular shunt patients (Table 4). [13]

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