INVITED EDITORIAL

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Recurrent spinal CSF-venous fistulas

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Spinal CSF-venous fistulas (CVF) are aberrent connections between the spinal subarachnoid space and the paraspinal veins and/or epidural venous plexus that allow unregulated egress of CSF into the venous circulation, effectively reducing CSF volume and causing the broad variety of clinical and radiographic manifestations of spontaneous intracranial hypotension.

In 2024, we mark the 10th anniversary of the first description of spinal CSF-venous fistulas as a distinct type of spontaneous spinal CSF leak [1] and celebrate remarkable advances in our understanding of this complex disorder that now constitutes the majority of CSF leaks encountered at our referral centre. The diagnosis and management of CVF today would in many ways be unrecognisable to experts in this field only a decade ago.

Diagnosis of CVF begins with the clinical history (new daily persistent headache with orthostatic, Valsalva manoeuvre, or second-half-of-the-day worsening is the most common but certainly not the only presentation), that is supported by signs of intracranial CSF volume depletion on brain MRI, and is further suggested by meningeal nerve root sleeve diverticula and no extradural fluid collection on spinal MRI. Finally, advanced myelography performed in the lateral decubitus position, most often digital subtraction myelography or dynamic CT myelography, must be performed with impeccable technique, and a bit of good luck, to capture the fleeting opacification of fistulised spinal veins in order to localise the CVF and provide a target for treatment. Most patients have a single CVF, but two or occasionally even more CVF may be diagnosed simultaneously [2, 3]. The recent introduction of photon counting detector CT has increased the yield of decubitus dynamic CT myelography for CVF, but there is still no technique that detects every CVF [4].

The treatment of CVF is an area of intense research, although neither randomised trials nor head-to-head comparisons of techniques have yet been published. Case series have shown clinical and radiographic improvement in patients with CVF with open spine surgery (ligation of the nerve root, dural sleeve, and associated veins or placement of an aneurysm clip on the neurovascular bundle) [5, 6] and transvenous embolisation of the paraspinal and foraminal veins with Onyx liquid embolic agent [7, 8]. At least short-term success has also been reported with percutaneous administration of blood and fibrin glue [9], but other series have shown dismal odds of enduring relief after blood patching for CVF [10, 11]. In this issue of PJNNS, Zayat et al. add to the short list of publications describing recurrent CVF and their treatment [12].

When patients report persistent or recurrent symptoms after treatment of CVF, the differential diagnoses include incompletely treated CVF, opening of a new CVF, and headache due to another cause. Zayat et al. describe 10 patients with ongoing or relapsed symptoms among 42 treated for CVF. Of these 10 patients requiring retreatment, four were determined to have been initially treated successfully (transvenous embolisation in three, fibrin patching in one) because repeat myelography did not show their original CVF but did show a new CVF at a new level [12]. Recurrence was ipsilateral in every case. Defining the success of prior treatment as disappearance of the initial CVF on repeat myelography is a fraught definition because the sensitivity of CT myelography or digital subtraction myelography for CVF is probably no higher than 75% [2, 3]. Yet that nuance is less important than the main message: that patients may develop CVF at new levels after treatment of initial CVF, meaning that repeat diagnostic testing can be fruitful [12].

Also of interest in Zayat's series are the five patients with residual symptoms whose 'recurrence' was suspected to be at the same level as the original fistula. In other words, a primary treatment failure. A previous, larger study by Brinjikji et al. showed a similar likelihood for recurrent CVF to occur at or near the initial level: of 100 patients treated initially, 17 required retreatment, seven at the same level and five within two

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levels above or below the original CVF [8]. A similar pattern was seen in a 2021 report by Malinzak et al., which included four patients whose CVF were treated surgically and then recurred ipsilaterally at the same or within three levels [13].

That CVF recurrence is likely to occur very near the original CVF has several implications. It may suggest an underlying focal dural weakness that predisposes to CVF formation. It may suggest a loco-regional change in CSF and/or venous fluid dynamics that promotes CVF formation by raising CSF pressure or lowering venous pressure, which could blow or suck open, respectively, a connection between nerve root sleeve and vein. Indeed, the Brinjikji et al. series found rebound intracranial hypertension after transvenous embolisation to be a risk factor for CVF recurrence [8].

Despite major advances in the diagnosis and treatment of CVF in the 10 years since their initial description, we still have much to learn. I commend Zayat et al. for reinforcing the message that recurrence of symptoms or brain MRI signs of CSF volume depletion should prompt consideration of repeat myelography for possible new CVF.

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