Peter J. Jannetta proposed the neurovascular compression (NVC) theory in 1967. He, together with his mentor Robert W. Rand, had been developing a technique of selective partial sensory rhizotomy for the treatment of trigeminal neuralgia (TN) and building on the pioneering works of Dandy, Gardner, and others. With the assistance of John F. Alksne, the first of these operations was performed through a subtemporal-transtentorial approach and with the novel use of an operating microscope. As exposure was gained, Jannetta revealed the superior cerebellar artery impinging on the proximal trigeminal nerve root and quietly exclaimed, “That’s the cause of trigeminal neuralgia.” Similar observations were evident in the next four such rhizotomy surgeries, although Jannetta found no similar compression in a series of cadaveric dissections. Later that same year in the neurosurgery clinic, Jannetta was assessing a gentleman with hemifacial spasm (HFS) and had an epiphany: this was another cranial nerve hyperexcitability condition analogous to TN and was also likely caused by pulsatile compression upon the proximal cranial nerve root. He explained this to the patient who then consented to what would become Jannetta’s first microvascular decompression (MVD) surgery. Together with his co-surgeon Paul H. Crandall, he identified and alleviated NVC of the facial nerve root exit and achieved a cure of the HFS. Two months later Jannetta and Rand performed their first MVD for TN with equal success. The neurosurgery community, however, was slow to accept this new cure for an old disease. Jannetta persevered despite the naysayers, the critics, and those who questioned his credibility. He gradually presented a body of surgical and clinical follow-up evidence that convinced a critical mass of adopters that MVD was an effective treatment, and neurosurgeons worldwide began to replicate his excellent results.

Over the course of decades, NVC has become established as the cause of nearly all cases of classic or typical TN, HFS, and glossopharyngeal neuralgia (GPN) when strict diagnostic criteria are applied. These conditions are also amenable to high rates of cure by the atraumatic alleviation of culprit vascular compression via Jannetta’s MVD procedure. However, the NVC theory has also been invoked by some as a potential cause of other conditions such as non-episodic facial pain (e.g., type 2 TN), Bell’s palsy, tinnitus, vertigo, hypertension, and diabetes. Evidence of an association between these conditions and culprit vascular compression is less robust, and MVD surgery has not been generally accepted as effective in their treatment. There remains an appropriately great burden of proof when proposing an NVC etiology, just as Jannetta faced 50 years ago.

In the current article, Dr. Honey et al. posit that NVC is a responsible etiology for some selected cases of laryngospasm, a condition that they have termed “hemi-laryngopharyngeal spasm,” or HELPS. Their three patients, as well as a patient in a previously reported single case, all suffered from episodic stridor, choking, and coughing that sometimes led to loss of consciousness. All had some degree of temporary symptom improvement with unilateral laryngeal botulinum toxin (Botox) injections. The authors suggest that culprit vascular compression upon vagus nerve rootlets was evident in each case and that symptoms were relieved with MVD. They also provide a thoughtful discussion regarding the possible mechanism underlying the proposed pathophysiology and conclude that MVD may be considered as a possible treatment in selected patients with HELPS.

But there are several factors to consider before accepting this proposed NVC etiology and microsurgical treatment. First, the clinical manifestations of the proposed HELPS condition should be more clearly delineated. While the reported patients had some clinical improvement with unilateral Botox injections, there was no confirmation that...
the laryngospasm was restricted to one side. This unilateral- 
ality should be confirmed with laryngoscopy, ultrasound, 
and electromyography studies as bilateral hyperactivity of 
the laryngeal muscles would not support an NVC etiology. 
Also, the loss of consciousness the patients suffered should 
be thoroughly characterized, particularly as unilateral in- 
volvement would be unlikely to cause airway obstruction. 
Was there hypoxia due to airway compromise? Was there 
cardiovascular hypotension or bradycardiac rhythms, as 
are rarely seen in GPN? Was there a vasovagal syncope relat- 
ed to the severe anxiety described in all of their patients? 
Finally, relating to clinical manifestations, it is noteworthy 
that no patients with HELPS had pain symptoms of GPN 
and that the clinical manifestations of GPN do not include 
laryngospasm. If NVC of cranial nerve IX and X rootlets 
were an underlying etiology of HELPS, then one should 
expect a clinical overlap with GPN.

Second, compression of the vagus nerve by the implic- 
cated posterior inferior cerebellar artery (PICA) was not 
as impressive as typically seen in patients with GPN, in 
whom the culprit vessel usually impinges dramatically 
upon cranial nerves IX and X at their root entry/exit zone, 
often causing distortion of the brainstem and proximal 
nerve. The PICA has a natural course of its lateral med- 
ullary segment between the cranial nerve X rootlets and 
variably contacts the nerve. A simple and ubiquitous 
juxtaposition of the artery and nerve are not sufficient 
evidence to invoke the NVC etiology. It remains to be de- 
termined whether vagus nerve NVC is more prevalent in 
patients with HELPS than in control patients (or on the 
opposite side). This could be assessed in a blinded review 
of high-resolution MRI, as has been effectively done for 
typical TN, HFS, and GPN.

Finally, a curative effect of MVD surgery would strongly 
support the NVC theory. This has certainly been 
well established for conditions of classic or typical TN, 
HFS, and GPN. In each of these conditions, a cure can be 
achieved in the vast majority of patients with an atraumatic 
alleviation of vascular compression of the respective root 
entry/exit zone without the need to induce any degree of 
nerve injury. However, it should be noted that microsurgi- 
cal injury to these respective nerves in itself may alleviate 
the hyperactivity symptoms, as originally demonstrated by 
Dandy and Gardner. In the series presented by Honey et 
al., all patients unfortunately suffered a symptomatic va- 
gus nerve injury as a complication of MVD surgery. While 
these clinical symptoms improved, there was no docu- 
tmentation with laryngoscopy or other studies that vagus nerve 
function had normalized. Therefore, it remains to be prov- 
en whether an atraumatic MVD of the vagus nerve roots 
could be curative of HELPS or whether the laryngospasm 
was instead improved with a surgically induced nerve in- 
jury analogous to other denervation procedures. An im- 
portant implication of this differentiation of the treatment 
effect may be the introduction of less invasive vagal rhiz- 
otomy techniques such as Gamma Knife radiosurgery, as 
has already been successfully utilized in the treatment of 
TN and GPN.

Laryngospasm poses a complex clinical challenge, with 
several potential underlying causes and variable respons- 
es to treatments. The authors are to be congratulated in 
their efforts to assess a possible NVC etiology not previ- 
ously considered for a subset of patients with this condi- 
tion. However, there is a great burden of proof to establish 
evidence of cause and cure, just as there was 50 years ago 
when Jannetta first presented his NVC theory and MVD 
surgery. I trust Dr. Honey and colleagues will continue 
studies into this possible cause of HELPS and provide us 
with a definitive set of findings and recommendations in 
the future. While I am currently not convinced that laryn- 
gospasms are caused by vagal NVC, history has shown 
that such skepticism may sometimes be overturned by dili- 
gent clinical and scientific efforts.

https://thejns.org/doi/abs/10.3171/2018.3.JNS18385

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Disclosures

The author reports no conflict of interest.

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INCLUDE WHEN CITING

Published online July 20, 2018; DOI: 10.3171/2018.3.JNS18385.

Response

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We thank Dr. Kaufmann for his thoughtful critique of 
our paper. His editorial highlights concerns with 1) con- 
firming the diagnosis of HELPS, 2) the etiology of loss of