Intractable hiccups: treatment by microvascular decompression of the vagus nerve

Case report

DENNIS L. JOHNSON, M.D.

Division of Neurosurgery, Children's Hospital, Penn State University School of Medicine, Milton S. Hershey Medical Center, Hershey, Pennsylvania

Idiopathic hiccups are usually managed with pharyngeal stimulation or a plethora of pharmacological agents. Hiccups that persist and prove intractable to these medical measures are treated by crush or ablation of the phrenic nerve, which denervates the major respiratory muscle. This is the first reported case of nondestructive microvascular decompression of the vagus nerve for the treatment of intractable idiopathic hiccups.

The vagus nerve was separated from the posterior inferior cerebellar artery by inserting a Teflon pledget between the nerve and vessel which eliminated the neurovascular contact. One year after the initial surgery, the hiccups recurred. The Teflon pledget had fallen out of place and the nerve was once again in contact with the artery. Once the contact was eliminated by wrapping the artery with a tuft of Teflon, the hiccups stopped. The patient has remained free of hiccups for 3 years.

It is concluded that patients with intractable idiopathic hiccups who fail medical therapy should be considered for microvascular decompression of the vagus nerve.

KEY WORDS • hiccups • microvascular decompression • vagus nerve

HICCUP is a sudden reflex contraction of the diaphragm which causes forceful inspiration. Hiccups may occur in clusters but usually last less than an hour or are responsive to any one of a myriad of folk remedies. Persistent hiccups lasting more than 48 hours are not only distressing to the patient but deserve thorough diagnostic evaluation. Hiccups associated with subphrenic abscess, mediastinal mass, or uremia are alleviated by treating the underlying condition. Intractable idiopathic hiccups are managed with pharyngeal stimulation, pharmacological agents, phrenic nerve block, and finally by surgical section or crush of the phrenic nerve.

The case of intractable hiccups described here was associated with a redundant loop of the posterior inferior cerebellar artery (PICA) which compressed the vagus nerve near its exit from the brain stem. This is the first reported case of intractable hiccups treated with microvascular decompression of the vagus nerve.

Case Report

This 16-year-old girl was knocked to the ground by a blow to the right side of the jaw during an altercation. She did not lose consciousness and sustained no fracture of the mandible but complained of intermittent right-sided retromastoid headaches. Hiccups first occurred 4 days after the injury; initially they subsided during sleep, but by the 3rd day they persisted during sleep also.

Examination. The patient did not complain of a sore throat, chest pain, dysphagia, abdominal pain, or vomiting. Neurological examination was normal. Chest x-ray films and computerized tomography (CT) of the brain with and without contrast enhancement were also normal. Trials of chlorpromazine and haloperidol had no therapeutic effect and caused incapacitating sedation. Carbamazepine, 1000 mg daily, was not beneficial. Valproic acid, 1500 mg daily, was effective and the hiccups recurred only when the patient neglected to take her medication; however, after 2 months this treatment also failed. Although intermittent hoarseness was noted by the mother, the patient's neurological examination remained normal. Sequential CT scans showed no abnormality. Cerebral angiography demonstrated an ectatic basilar junction displaced slightly to the right. The PICA came off the proximal basilar artery, looping against the side of the brain stem; impression of the
brain stem by this arterial loop was confirmed by magnetic resonance imaging (Fig. 1). Fluoroscopy of the diaphragm demonstrated intermittent spasm of the right hemidiaphragm, which coincided with hiccups. Brain-stem evoked potentials were normal.

**Operation.** The ninth, 10th, and 11th cranial nerve complex was approached through a right posterior fossa craniectomy performed with the patient in the sitting position. A branch of the PICA was found anterior to the nerve complex, pressing posteriorly against the rootlets of the vagus nerve at the root entry zone. Another arterial loop lay against the vagus and glossopharyngeal nerves near the jugular foramen. A thin 2 × 2-mm pledget of Teflon felt was placed between the vagus nerve and these two arterial loops.

**Postoperative Course.** No hiccups occurred after surgery, and the postoperative course was remarkable only for transient hoarseness. The hiccups recurred 1 year later and responded for a brief time to valproic acid, 1500 mg daily. After 3 months of continuous hiccups, the right ninth, 10th, and 11th cranial nerve complex was re-explored. The pledget of Teflon felt had become dislodged and lay free on the floor of the posterior fossa. Precisely when the pledget was displaced is not known. A tuft of Teflon was then wrapped around the loops of the PICA that pressed against the vagus nerve and was secured with a single 8-0 nylon suture. The hiccups once again ceased immediately, and the patient has remained free of hiccups for 3 years.

**Discussion**

**Pathophysiology of Hiccups**

Sudden inspiration terminated by glottic closure results in an audible “hic.” The rhythmicity and periodicity of hiccups suggests a self-perpetuating reflex arc. Although the respiratory musculature is primarily involved, hiccup is probably not a respiratory reflex but rather a primitive gastrointestinal reflex designed to bech excess amniotic fluid from the fetal stomach or dislodge food or debris from the esophagus, similar to the effector achieved with the Heimlich maneuver. The afferent limb of the reflex arc is formed by the vagus and phrenic nerves. The central connection is the reticular formation in the lower pons and medulla. The efferent limb is a barrage of motor impulses traveling through the visceral motor nuclei of the medulla, the vagus nerve, and the primary C-4 and C-5 spinal neurons which innervate the diaphragm via the phrenic nerve.

The etiology of hiccups has been reviewed by several authors. Acquired intractable hiccups can be caused by a disease or lesion anywhere along the anatomical course of the reflex arc. Hiccups can be caused by cerebral lymph nodes enlarged by carcinoma, lymphoma, or tuberculosis. Within the chest cavity, carcinoma of the lung, myocardial infarction, pneumonia, pericarditis, and hiatal hernia are important causes of hiccups. Intra-abdominal processes include subdiaphragmatic abscess, peritonitis, gastrointestinal hemorrhage, abdominal neoplasm, pancreatitis, and cholecystitis. Hiccups have been reported with encephalitis, meningitis, and head trauma and with a variety of brain-stem and cervical spinal cord lesions: PICA thrombosis, brain-stem tumors, multiple sclerosis, and syringobulbia. Uremia, diabetes mellitus, and hyponatremia can also produce persistent hiccups. Treatment is directed at the underlying condition. Idiopathic hiccups usually resolve spontaneously or can be stopped by some form of pharyngeal stimulation. For persistent or intractable idiopathic hiccups, pharmacological suppression of the reflex has been achieved with a wide variety of agents. Hiccups refractory to pharmacological suppression are conventionally treated first with phrenic nerve blocks and, failing that, with phrenic nerve crush or section.

In a condition that has such a wide diversity of causes and remedies, treatment is often empiric but must be based on thorough diagnostic investigation. No conventional cause was found for this girl’s hiccups. Although refractory to medical treatment, her hiccups ceased after microvascular decompression and insulation of the vagus nerve. Moreover, when the hiccups recurred the insulating Teflon pledge was found to have slipped away from the nerve. When the nerve was reinsulated from the loops of PICA, the hiccups stopped.

**Microvascular Decompression**

Microvascular decompression has been popularized by Jannetta and coworkers1-4 for the treatment of tic douloureux, hemifacial spasm, tinnitus, vertigo, and glossopharyngeal neuralgia. Contact of pulsatile vascular structures on nerves, especially at the unmyelinated nerve entry zone, produces hyperactive dysfunction in the distribution of the nerve. The vulnerability of cranial nerves to hyperactive dysfunction at the point of entry to or exit from the brain-stem vascular compression is controversial. Walter Dandy5 first observed vessels associated with the trigeminal nerve in patients he operated on for tic douloureux. Sunderland6 elaborated...
Decompression of the vagus nerve for hiccups

on the neurovascular relationships at the brain stem but did not correlate them with neurologic dysfunction. Gardner and Miklos first suggested that compression of cranial nerves by adjacent vessels could cause hemifacial spasm and trigeminal neuralgia and that these clinical conditions could be relieved by vascular decompression. Following Jannetta's lead, the merits of microvascular decompression have been documented by many investigators. Others have questioned the pathophysiological basis of microvascular decompression and have not been convinced by the electrophysiological data in support of the procedure. In a study of 50 trigeminal nerve roots and their vascular relationships, Hardy and Rhoton found that 60% of the nerves were in contact with a vessel; 20 of the cadavers had bilateral vascular contacts. None of the individuals studied had suffered trigeminal neuralgia during their lifetime. In patients with hyperactive dysfunction syndromes, neurovascular compression is not always found. Among 526 patients collected from seven different series, 19% had no neurovascular compression. Moreover, about one-third of patients who undergo microvascular decompression continue to experience pain or dysfunction.

In a report of intractable hiccups associated with a posterior fossa arteriovenous malformation (AVM) indenting the right medulla, Laing et al. described remission of the hiccups for only 2 days after the AVM was resected. Relief was eventually achieved with bilateral phrenic nerve section. The patient was not reexplored for vascular compression, and no differential spasm of the diaphragm was reported. It is possible that neurovascular compression still existed even though the AVM had been removed.

In the present case the redundant vessel lay over the proximal vagus nerve, triggering the hiccup reflex through the afferent limb. Subarachnoid hemorrhage caused by the head trauma may have caused adhesions binding a pre-existing neurovascular relationship more firmly. At the first microvascular decompression procedure, the nerve was separated from the arterial loops by a Teflon pledget to eliminate the neurovascular contact. The vagus nerve was traumatized during the procedure, which accounts for the patient's transient hoarseness. Taarnhøj and Gardner and Miklos achieved quite acceptable results by simply traumatizing the trigeminal nerve root with a nerve hook. Trauma to the vagus nerve may account for relieving this child's hiccups; however, when the hiccups recurred, the pledget had slipped out of place, and the hiccups ceased when the nerve was reinserted.

Patients with intractable idiopathic hiccups who have unilateral rhythmic spasms of the diaphragm, revealed by fluoroscopy, and who fail medical therapy should be considered for microvascular decompression of the vagus nerve before phrenic nerve crush or section is attempted.

References

Manuscript received December 13, 1991.
Accepted in final form April 8, 1992.
Address reprint requests to: Dennis L. Johnson, M.D., Division of Neurosurgery, P.O. Box 850, Hershey Medical Center, Hershey, Pennsylvania 17033.