

## REVIEW ARTICLE

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# Understanding Hemifacial spasm: A Review Of An Uncommon Movement Disorder

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### Abstract:

*Hemifacial Spasm is a rare condition and often misdiagnosed even by neurologists. Although etiology, pathophysiology is clear, treatment is unsuccessful in many cases. In this review article we focus on etiology, pathophysiology, diagnosis and management of this infrequent movement disorder.*

**Keywords:** Hemifacial Spasm, Movement Disorder.

### Introduction:

Hemifacial spasm (HFS) is characterized by unilateral, paroxysmal, involuntary movement occurring in the muscles innervated by the ipsilateral facial nerve. The involuntary contractions usually begin in the orbicularis oculi muscle and gradually spread to the other muscles related to facial expression<sup>1</sup>. The majority of HFS cases occur unilaterally with an estimated 0.6% to 5% occurring bilaterally<sup>2</sup>. Some patients report worsening of spasms with fatigue, anxiety, and changes in position of the head (e.g., head to one side or the other on the pillow at night)<sup>3</sup>.

Although traditionally perceived as a benign illness, it can lead to increasing embarrassment and social withdrawal for the individual and in severe cases even functional blindness due to involuntary eye closure.

### Historical Perspective

F Schultze, in 1875, probably reported the first case of hemifacial spasm in literature, when he described a 56-year-old man with involuntary movements involving the left side of his face. The post-mortem revealed a giant aneurysm of the left vertebral artery compressing the left facial nerve<sup>4</sup>.

In 1888, Gowers elaborated on this syndrome further and described the classical features of this condition<sup>5</sup>.

The condition received its current terminology, when it was named as 'he´mispasme facial' by Babinski in 1905. Babinski at the same time also described another characteristic feature of this disease, thereafter known as 'the other Babinski sign' i.e. when orbicularis oculi contracts and the eye closes, the internal part of the frontalis contracts at the same time, and the eyebrow rises during eye occlusion."<sup>6</sup>. This typical feature distinguishes hemifacial spasm from blepharospasm in which this sign is absent.

### Epidemiology

HFS is prevalent in 9.8 per 100,000 persons<sup>7</sup>. The average age of onset for HFS is 44 years. Women and Asian populations have an increased susceptibility to HFS though valid prevalence data is scarce<sup>8,9,10</sup>. Worldwide estimates for the prevalence of HFS are 14.5 per 100,000 women and 7.4 per 100,000 men<sup>11,12</sup>. Families with HFS present with autosomal dominant inheritance and low penetrance although there have been only a few reported cases<sup>13</sup>. In addition, the genetic susceptibility is poorly defined as there is not a clear relationship between HFS and single-nucleotide polymorphisms in genes related to vascular compression<sup>14</sup>.

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## **Pathophysiology**

The accepted underlying pathophysiology of HFS suggests that the disease process is caused by facial nerve root entry zone myelin breakdown and ephaptic transmission, which is the passage of neural impulses through artificial chemical or chemical synapses. The root exit zone of the facial nerve is defined as the transition point between central (oligodendrocytes) and peripheral (Schwann) cell myelination<sup>15,16</sup>. This segment is sheathed by only an arachnoidal membrane and lacks both interfascicular connective tissue separating fibers and epineurium; these features increase this segment's vulnerability to compression<sup>16</sup>. Compared to similar disorders of the trigeminal, glossopharyngeal, and vagus nerves, a study correlated the length and volume of central myelin portions of these nerves with the incidence of the nerves' corresponding diseases<sup>17</sup>. One study suggested that the root exit zone was primarily involved in only 23% of its studied HFS patients whereas compression of a more proximal segment of the facial nerve when it emerges from the pontomedullary sulcus was implicated in 73%<sup>18</sup>.

Several theories have been put forward to explain how this compression of the facial nerve at its root exit/entry zone leads to hemifacial spasm.

One of them – the nerve origin hypothesis or the peripheral theory postulates that there is emphatic transmission of impulses between neighbouring neurons (i.e. coupling of adjacent nerve fibers due to local exchange of ions or local electric fields) leading to excessive or abnormal firing. Myelination is a natural inhibitor of ephaptic transmission and the demyelination due to local compression thus leads to hemifacial spasm<sup>19</sup>.

The other – the nuclear origin hypothesis or the central theory states that hemifacial spasm results from the hyperexcitability of the facial motor nucleus due to irritative feedback from peripheral lesions of the nerve<sup>20</sup>.

## **Etiology**

Hemifacial Spasm can either be primary or secondary. Primary HFS results from compression of the seventh nerve at the root exit zone in the

posterior cranial fossa by an aberrant or ectatic vessel, most commonly the superior cerebellar, the anterior inferior cerebellar or the vertebral artery<sup>21</sup>. The pattern of neurovascular compression can be divided into six different categories: (A) loop type, where the vascular loop itself creates the compression, (B) arachnoid type, where arachnoid trabeculae between the vessel and brainstem cause the vessel to tether to the nerve, (C) perforator type, where the perforating arteries from the compressing vessel tether the vessel to the brainstem, (D) branch type, where the nerve is caught between the compressing vessel and its branches, (E) sandwich type, where the nerve is sandwiched between two different vessels, and (F) tandem type, where one vessel compresses another vessel that compresses the nerve<sup>22</sup>.

Secondary HFS occurs with damage anywhere along the facial nerve from the internal auditory canal to the stylomastoid foramen<sup>23</sup>. Cases of secondary HFS have been linked to cerebellopontine angle (CPA) tumors and vascular malformations with other case linked to facial nerve trauma, demyelinating lesions, and vascular insults<sup>24</sup>. Collectively, these underlying issues of secondary HFS are thought to cause neural dysfunction and/or irritation of the facial nerve pathway<sup>25</sup>.

## **Diagnosis**

Diagnosis of hemifacial spasm is mainly a clinical one. All patients must be subjected to a detailed history and clinical examination to look for any subtle neurological deficits which would point to an underlying secondary cause for the condition. The “Babinski-2 sign,” “other Babinski sign,” or “brow-lift sign” is a physical exam maneuver that is positive when a patient lifts his/her eyebrow with ipsilateral eye closure, signaling the synchronized activity of the frontalis and orbicularis oculi muscle during HFS<sup>26,27,28</sup>. This technique has been shown in one study to have high sensitivity (86%), specificity (100%), and interrater reliability (92%) for HFS diagnosis<sup>29</sup>.

Electromyography (EMG), Magnetic Resonance Image (MRI), and computerized tomography (CT) are used to confirm the diagnosis and differentiate

primary from secondary HFS. EMG can also be useful to differentiate HFS from other abnormal facial movement disorders; in HFS, spontaneous, high-frequency synchronized firing is seen on EMG<sup>30</sup>. Additional diagnostic techniques such as a CT angiogram are useful for microsurgical planning<sup>31</sup>.

### **Treatment**

Hemifacial spasm is a chronic condition with progressively increasing spasms. However due to the low prevalence of the condition, not a lot of controlled clinical trials have been done to determine the best therapeutic modality for these patients. Although, botulinum toxin remains the most popular choice for therapy, other options including oral pharmacotherapy and microsurgical decompressive surgeries are also beneficial in selected cases.

### **Drugs**

A large number of drugs have been studied and found to be of some efficacy in hemifacial spasm. These include anticonvulsants such as carbamazepine, clonazepam, gabapentin and other drugs like baclofen, anticholinergics and haloperidol<sup>32</sup>. The biggest limitation of oral drugs is their inconsistent efficacy and large number of side effects including sedation, fatigue and exhaustion<sup>33</sup>.

### **Botulinum toxin therapy**

The standard medical treatment for HFS is botulinum neurotoxin (BoNT) injections. Having been used since the early 1980s, BoNT injections provide low-risk symptomatic relief in 85% of HFS patients, making it the treatment of choice for patients with high anesthetic risk and those who refuse surgery<sup>34</sup>.

Botulinum toxin is a potent biological toxin derived from the organism *Clostridium botulinum*(35). It acts on the presynaptic region of the neuromuscular junction and prevents the calcium-mediated release of acetylcholine at the nerve terminal preventing impulse generation downstream resulting in functional reversible paralysis of the supplied muscles. Based on target site of action there are various serotypes of Botulinum Toxin. The most commonly used

commercially available preparation is type A. The resulting muscular weakness due to the injected toxin lasts for somewhere between 3-6 months. A large number of trials have validated the successful outcomes of this therapy with improvements in as many as 75-100% of individuals with hemifacial spasm<sup>36,37</sup>. However, the injections must be repeated every 3 to 6 months. Tolerance can develop in some cases, but the treatment is generally well tolerated. Local complications of these injections include ptosis, blurred vision, and diplopia that may improve after days to weeks<sup>38</sup>.

### **Surgery**

With the advent of botulinum toxin therapy, the need for operative intervention has drastically gone down in cases of hemifacial spasm.

Microvascular decompression (MVD) provides a curative treatment with long term relief of symptoms by alleviating vascular compression of the facial nerve root. The underlying principle of MVD is to separate the nerve-vessel conflict rather than isolate it with prostheses; important intraoperative considerations include prompt identification of the neurovascular conflict site, sharp dissection of arachnoids for maximal nerve root visualization, and electrophysiological monitoring to distinguish offending vessels<sup>39</sup>. MVD has excellent results with long-term success rates between 83% and 97% of cases<sup>40</sup>.

An analysis of twenty-two papers representing 5,685 patients treated with MVD for HFS found that an average of 91.1% of patients had complete resolution of symptoms over a median 2.9-year follow-up period<sup>41</sup>. Even with a first-time MVD failure, patients in one study who elected for repeated MVDs had a cure rate of 85% and did not suffer a higher rate of complication with a mean follow-up of 54.48 months<sup>42</sup>. Another small study found no significant difference between elderly and young patients in cure rate (96.3% versus 89.4%) and complication rate<sup>43</sup>.

Resolution of HSF after MVD may take several months to several years with small percentage of patients who fail to improve. In these patients, failure to improve may be attributed to inadequate decompression of the offending vessel, presence

of a previously unidentified secondary offending vessel, or implant compression/migration against the facial nerve (44). Generally, complications of MVD are uncommon and generally transient (45). In some cases, MVD can result in serious complications, which are thought to be caused by facial nerve stretching during cerebellar retraction, iatrogenic injury to surrounding structures, or prosthesis compression (46). Overall, serious complications following MVD were reported in less than 1% of cases(47).

### Conclusion

The benign appearing facial twitches of hemifacial spasm are actually very bothersome to the individuals who suffer from the condition both in terms of functional blindness as well as social embarrassment. Early recognition of the condition, ruling out secondary causes and instituting appropriate therapy is therefore a necessity. Botulinum toxin therapy offers a simple, noninvasive therapy for this condition. Patients who do not respond to the toxin injections or prefer a permanent cure can be offered surgical options as well. Either treatment modality when instituted gives good benefits to the patient and leads to a significant improvement in their quality of life.

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#### Statements

The authors report no conflicts of interest related to this study.

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