DOI: 10.53469/jcmp.2025.07(12).20

Discussion on the Clinical Combined Acupuncture Therapy for Hirayama's Disease

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Abstract: ***Hirayama disease (HD) is a self-limiting, benign motor neuron disorder that typically manifests during adolescence, clinically characterized by muscular weakness and atrophy affecting either a single limb or the asymmetric distal segments of the upper extremities. Currently, the etiology of HD is recognized as multifactorial and remains incompletely elucidated. Owing to its low clinical prevalence and heterogeneous manifestations, this condition is frequently susceptible to misdiagnosis and underdiagnosis in clinical settings. In Western medical practice, the diagnosis of HD is established through a comprehensive evaluation integrating clinical features, imaging findings, and neuroelectrophysiological studies, with conventional treatment primarily involving cervical collar immobilization or surgical intervention. From the perspective of traditional Chinese medicine (TCM), HD is categorized as "wei syndrome" [1]. The core therapeutic principles emphasize warming yang and unblocking meridians, activating local qi in the affected channels, regulating qi and blood circulation, and restoring the dynamic balance between yin and yang. This therapeutic strategy is designed to nourish the local meridians and musculature, thereby alleviating the associated clinical symptoms.***

Keywords: Hirayama disease, Amyotrophy, Wei syndrome, Acupuncture**.**

1. Introduction

Hirayama disease, often abbreviated as HD, was first identified and officially reported by a team of researchers led by the Japanese scholar Hirayama back in 1959 [2]. It is classified as a self-limiting disorder, which means its progression will stop on its own over time without continuous worsening. This condition mainly affects young people, specifically adolescents and young adults who are between 15 and 25 years old. What's more, HD shows a clear regional pattern in its occurrence. It is much more common in Asian countries, with Japan being one of the regions where the disease prevalence is notably higher. Another striking feature of HD is its obvious gender bias. Clinical data and case statistics from studies [3-4] have consistently shown that male patients make up a large majority of all diagnosed cases. In terms of clinical symptoms, the main problems patients face are progressive muscular atrophy and weakness in the distal parts of their upper extremities — that is, the lower arms and hands. These areas are exactly controlled by the C7–T1 segments of the spinal cord. Besides these core symptoms, some patients also experience temporary weakness in their affected limbs when exposed to cold temperatures. They may also notice tremors when they straighten their fingers. Because of these specific traits, most patients say their symptoms get worse when they stay in cold environments, and they feel better once the surrounding temperature rises [5]. The main purpose of this article is to review the disease's underlying causes, common diagnostic methods, and the current development of acupuncture therapy for HD. In doing so, it can help readers build a more thorough and all-round understanding of this rare neurological disorder.

2. Etiology and Pathogenesis of HD

Even today, the root causes of Hirayama disease (HD) are still complex and not fully understood. What's more, this disease is quite rare in clinical practice, which makes it harder for doctors to study its origins deeply. There are several main ideas about how HD develops, and three of them stand out: compression of the cervical spinal cord (the part of the spine in the neck) [6] Adolescence is a period of rapid growth and development in the human body, during which individuals experience significant height growth. At this stage, there is asynchrony in growth rhythms between the spinal cord and the dura mater, which leads to relative shortening of the cervical posterior roots. If the growth rate of the cervical posterior roots lags behind the growth rate of the arms, this results in relative shortening of the C5-T4 cervical posterior roots. Therefore, when the patient performs neck flexion (or "a neck flexion movement"), the relatively shortened cervical posterior roots will pull the lower cervical cord anteriorly, eventually resulting in anterior vertebral compression of the spinal cord.

Spinal cord dynamic factors [7–8], Frequent neck flexion movements or prolonged maintenance of a flexed neck posture in patients can cause the dura mater to compress the lower cervical spinal cord posteriorly, thereby impairing local blood circulation. If this condition persists chronically, the anterior horn of the spinal cord will undergo degeneration and necrosis due to ischemia and hypoxia, subsequently inducing neurogenic injury to the muscles it innervates, and ultimately resulting in abnormalities in motor nerve conduction.

Caused by the obstruction of microcirculation in the neck, poor blood flow in the tiny blood vessels of the neck (called impaired cervical microcirculation). To be more specific, two research teams—led by Hashimoto et al. [9] and Gourie-Devi et al. [10]—have shared a similar view. They believe that doing intense physical exercise for a long time or doing heavy manual labor may put constant, low-level stress on the neck area. Over time, this stress can slow down the blood flow in the small blood vessels around the cervical spinal cord. Many experts think this poor microcirculation is one of the key reasons why people with HD develop muscle atrophy in their upper limbs.

Racial genetic factors [11] are also one of them. The incidence of HD is notably high in Japan and India, ranking among the

highest globally. Familial cases have only been reported in Japan, while those in other countries are mainly sporadic. Since HD was first reported in 1959, there have been only 7 reports of hereditary juvenile distal upper limb muscular atrophy (i. e., HD-related hereditary cases), and each family harbors only 2 affected individuals. Among these, 5 families involve affected brothers, 3 involve affected father-son pairs, and 2 involve affected mother-child pairs. The currently hypothesized genetic patterns include autosomal recessive inheritance, autosomal dominant inheritance, and X-linked inheritance; additionally, its pathogenesis may also be associated with specific gene mutations.

3. Diagnosis and Treatment of HD

Currently, the diagnosis of HD doesn't rely on a single test—instead, it mainly depends on three key components that work together: clinical manifestations (what doctors observe in patients), imaging examinations (like X-rays or MRIs), and neuroelectrophysiological tests (tests that check nerve and muscle function). Let's break down each part to make it easier to understand.

First, let's talk about clinical manifestations. HD typically starts during adolescence—usually when kids are between 10 and 20 years old, a time of rapid growth. The most obvious signs are muscular atrophy (muscle wasting) and weakness in the distal parts of one or both upper limbs. "Distal parts" here mean the hands, wrists, and forearms, rather than the upper arms or shoulders. Importantly, these symptoms are often asymmetric—meaning one arm is usually more affected than the other. For example, a patient might notice that their right hand is weaker than the left, or that the muscles in their left forearm look smaller than the right. These issues directly impact patients' daily lives: simple tasks like putting on clothes, buttoning a shirt, holding a pen to write, or even lifting a cup can become difficult because their hands and wrists lack strength.

Next is imaging examinations. When doctors do imaging tests on HD patients, they often find that the cervical spine (the part of the spine in the neck) has an increased range of motion when bending forward. In other words, when the patient tilts their head down, their neck can bend more than usual. This finding is closely linked to cervical instability—meaning the neck vertebrae aren't as stable as they should be—and it's also common to see a straightened or reversed curve of the cervical spine. Normally, the cervical spine has a gentle "C" shape that curves forward, but in HD patients, this curve might be straight or even curve backward. Notably, a definitive diagnosis (a sure diagnosis) can't be made with regular imaging alone—doctors need specific imaging of the cervical spine when the patient is in a flexed position (when they're bending their neck forward) [12-13]. This position helps doctors clearly see if the spinal cord is being compressed, which is a key sign of HD.

Then there are neuroelectrophysiological tests, which check how well nerves and muscles communicate. For HD patients, some tests show that the ulnar and median nerves—two major nerves that control movement and sensation in the hands and forearms—have a slower conduction speed. This means electrical signals travel more slowly along these nerves than normal. Additionally, the amplitude (strength) of the compound muscle action potential is reduced. Think of this as the "electrical signal" that tells muscles to contract—if the signal is weaker, the muscle can't contract as strongly. Singh et al. [14] also found that electromyography (EMG for short, a test that records muscle electrical activity) of the affected limb shows denervation (when the nerve supply to the muscle is lost) combined with muscular atrophy. What's even more notable is that over 90% of patients have this same finding in the corresponding muscle groups on the opposite side—even if that arm doesn't show obvious weakness or wasting yet. Plus, cold environments (like cold weather or touching something icy) can make these electrical abnormalities worse, which is why EMG is such a valuable tool for diagnosing HD—it can pick up signs that other tests might miss.

When it comes to treatment, the primary options currently are conservative therapy (non-surgical treatment) and surgical intervention (surgery). Notably, researchers Shan Gui, Lian Yang et al. [15] made an important discovery: an imbalance in the strength of the deep cervical muscles (the muscles deep in the neck that support the spine) leads to excessive cervical flexion (too much bending of the neck forward), which in turn compresses the spinal cord. This is likely a key contributing factor to HD, so correcting this cervical sagittal imbalance (the uneven alignment of the neck in a front-to-back direction) could be a potential therapeutic target for treatment.

For conservative therapy, it's most suitable for patients with a disease duration of 4 years or less and who have had progressive symptoms (worsening symptoms) over the past 6 months. These patients can wear a neck brace—a device that wraps around the neck to limit excessive neck flexion. By preventing the neck from bending too far forward, the brace reduces pressure on the spinal cord. They can also take neurotrophic drugs, such as methylcobalamin (a form of vitamin B12) and other B vitamins. These drugs help support nerve repair and maintain normal nerve function, which can slow down muscle wasting and weakness.

For surgical intervention, it's recommended for patients who don't respond to neck brace therapy (meaning their symptoms don't get better or keep getting worse) or who have a prolonged disease course (the disease has lasted for a long time). Common surgical procedures include posterior cervical decompression combined with duraplasty [16-17], among others. Simply put, this surgery involves relieving pressure on the spinal cord from the back of the neck (posterior cervical decompression) and expanding the protective covering around the spinal cord (duraplasty) to give the spinal cord more space, reducing compression and preventing further nerve damage.

4. TCM Perspective on HD

In Traditional Chinese Medicine (TCM), there is no direct equivalent disease name for "Hirayama Disease (HD)". However, based on its clinical manifestations, HD can be categorized under the TCM concept of "wei syndrome"—a condition characterized by limb weakness, often accompanied by muscular atrophy and impaired voluntary movement. Notably, TCM practitioners have had a profound understanding of "wei syndrome" for over 2, 000 years.

Classic TCM texts such as *"Nei Jing", "Lei Jing", and "Jing Yue Quan Shu"* all documented this condition. Its onset is frequently associated with disorders of the five zang-organs, or induced by internal organ heat, emotional disturbances, kidney deficiency, or damp-heat. The core pathogenesis lies in the failure of meridians, tendons, bones, and muscles to receive adequate nourishment from qi, blood, and body fluids. Subsequent TCM scholars further elaborated that deficiencies of yin, yang, qi, blood, or body fluids, as well as internal stagnation of phlegm-dampness, blood stasis, or food retention, can all trigger "wei syndrome", In modern clinical practice, Fei Zhen typically presents with a combination of deficiency and excess syndromes, with the three most common TCM patterns being lung-stomach fluid exhaustion, liver-kidney depletion, and damp-heat infiltration. Treatment adheres to the fundamental TCM principle of "Bian Zheng Lun Zhi", tailored to the patient's specific condition.

5. Clinical Acses of Acupuncture Treatment for HD

Lin Lu, Anqi Wang, et al. [18] proposed that damp-heat and blood stasis obstructing the meridians, which leads to malnutrition of the affected limb, is the primary pathogenesis of HD. They adopted a combined therapy of pricking bloodletting, body acupuncture, and abdominal acupuncture: Pricking bloodletting was applied at the bilateral Jiaji (EX-B2) and Baxie (EX-UE9) of the affected limb to unblock local qi and blood circulation; Quchi (LI11) was selected to regulate the qi of the Large Intestine Meridian; Geshu (BL17) and Weishu (BL21) were used to treat blood disorders and nourish the skin, muscles, tendons and bones; body acupuncture included Hegu (LI4), Taichong (LR3) and Yanglingquan (GB34) together with local acupoints of the affected limb to relax tendons, unblock meridians and facilitate rehabilitation; abdominal acupuncture adopted Zhongwan (CV12), Qihai (CV6) and other acupoints to invigorate yang qi. After treatment, the motor function of the affected limb was significantly improved, enabling patients to resume normal daily activities with no recurrence during follow-up.

Yanjie Fan et al. [19] diagnosed HD by integrating the symptoms, onset characteristics, and examination findings of adolescent patients. They proposed that prolonged exposure to damp environments would induce spleen deficiency, dampness retention, qi dysfunction, and meridian malnutrition, which consequently gives rise to flaccidity syndrome. The therapeutic principles were set as invigorating the spleen, resolving dampness, warming yang, and relieving paralysis. Fire needle therapy was adopted in clinical practice: Baihui (GV20), Jianyu (LI15) of the affected limb, and bilateral Jiaji (EX-B2) at the C5-C7 cervical segment were selected as the acupoints. In addition, the transverse needling technique was applied, with Qianding (GV21) penetrated to Xuanli (GB6) and Houxi (SI3) penetrated to Hegu (LI4). This approach was designed to expand the stimulation scope, connect the meridians of the affected limb, improve cerebral microcirculation as well as qi and blood circulation of the affected limb, and finally achieved favorable clinical outcomes.

6. Modern Mechanism Research of Acupuncture

From a modern medical perspective, the principle of acupuncture therapy is easy to understand: doctors target the corresponding meridians of the lesion site in patients with Hirayama disease (HD) and apply needle stimulation. This stimulation can effectively activate the secretion of neurotransmitters in the affected area—simply put, it prompts the body to release chemical substances that transmit nerve signals.

These chemical substances further stimulate the body's free nerve endings (the nerve terminals that can perceive external stimuli), triggering axonal and segmental nerve reflexes, which then relay these signals to the cerebral cortex. Upon receiving the signals, the cerebral cortex releases regulatory nerve impulses and initiates the neuro-humoral dual regulatory mechanism—a regulatory mode where nerves and body fluids work together—driving the affected tissues to produce favorable adaptive responses.

This process not only effectively promotes the repair and regeneration of damaged nerves but also significantly enhances the compensatory function of intact nerves [20]. To a certain extent, it helps HD patients restore muscle function and alleviate clinical symptoms such as limb weakness and muscle atrophy caused by muscular wasting.

7. Discussion

In Traditional Chinese Medicine (TCM), there is no direct equivalent disease name for HD. That is to say, you cannot find a disease term in TCM classic literature that perfectly matches the definition and diagnosis of HD in modern medicine. However, when we look at the typical clinical manifestations of HD, this disease can be categorized under the broad TCM concept of "wei syndrome".

What exactly is "wei syndrome"? It is a condition that mainly manifests as limb weakness. In most cases, it is also often accompanied by symptoms like muscular atrophy, and patients may also have problems with impaired voluntary movement of the limbs. Notably, TCM practitioners have had a profound and systematic understanding of "wei syndrome" for more than 2, 000 years. Many classic TCM texts have recorded this condition in detail. For example, "Nei Jing" (Yellow Emperor's Internal Classic), "Lei Jing" (Classified Canon), and "Jing Yue Quan Shu" (Complete Works of Jing Yue) all have special discussions about its symptoms, causes, and treatment ideas.

According to TCM theory, the onset of "wei syndrome" is frequently associated with disorders of the five zang-organs (the heart, liver, spleen, lungs, and kidneys). It can also be induced by several factors, including internal organ heat caused by excessive fire in the body, long-term emotional disturbances such as anxiety and depression, kidney deficiency that comes with aging or chronic illness, or the accumulation of damp-heat in the meridians.

The core pathogenesis of this syndrome lies in a key problem: the meridians, tendons, bones, and muscles of the body fail to receive adequate nourishment from qi, blood, and body fluids. Without sufficient nourishment, these tissues and structures cannot maintain their normal functions, which then leads to weakness and atrophy of the limbs. Later on, subsequent TCM scholars further elaborated on the causes of "wei syndrome". They pointed out that besides the above factors, deficiencies of yin, yang, qi, blood, or body fluids can all trigger this condition. At the same time, internal stagnation of pathological substances such as phlegm-dampness, blood stasis, or food retention can also block the meridians and cause "wei syndrome".

In modern clinical practice, HD (which is classified as "wei syndrome" in TCM) typically presents with a combination of deficiency and excess syndromes. This means patients may have both weakened organ functions and blocked meridians at the same time. Among all the possible TCM patterns, the three most common ones are lung-stomach fluid exhaustion, liver-kidney depletion, and damp-heat infiltration. For the treatment of HD, TCM always adheres to its fundamental principle of "Bian Zheng Lun Zhi" (treatment based on syndrome differentiation). Doctors will formulate personalized treatment plans tailored to the patient's specific condition, rather than using a one-size-fits-all approach.

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