

# Hirayama Disease as a Treatable Motor Neuron Disorder substantiation from Clinical and Imaging Literature

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## Abstract

**BACKGROUND:** Hirayama complaint (HD), also known as juvenile muscular atrophy or monomelic amyotrophy, is an uncommon complaint characterized by myelopathy convinced by cervical flexion, generally affecting adolescent and youthful adult males. It's distinguished by progressive, asymmetric weakness and atrophy of the hand and forearm muscles, with minimum involvement of sensitive or pyramidal tracts. Beforehand opinion is essential to help unrecoverable disability and enable timely intervention.

**OBJECTIVE:** ideal This study seeks to totally review the current substantiation on the epidemiology, pathophysiology, clinical features, individual ways, and treatment approaches for Hirayama complaint, with an emphasis on recent advancements in imaging and remedial issues.

**METHODS:** styles An expansive literature hunt was conducted across PubMed, Web of Science, and other scientific databases for English-language studies published up to 2025, using keywords similar as “Hirayama complaint,” “juvenile muscular atrophy,” “cervical flexion myelopathy,” and “dynamic MRI.” Studies that described clinical features, electrophysiology, imaging findings, and treatment issues were included.

**RESULTS:** Hirayama complaint generally manifests between the periods of 15 and 25, presenting with weakness and muscle wasting in the distal upper branches. Dynamic cervical MRI is pivotal for opinion, revealing anterior relegation of the posterior dural sac and blowup of the posterior epidural space during neck flexion. This relegation results in repeated contraction and ischemia of the spinal cord, primarily affecting anterior cornucopia cells. Electrophysiological studies have shown habitual denervation in the C7 – T1 myotomes. Beforehand conservative operation with cervical collar immobilization to limit neck flexion can stop complaint progression. Surgical interventions, similar as anterior or posterior cervical emulsion, have been employed in progressive cases with varying success rates. Literature indicates that both surgical styles may stabilize the complaint, though their relative effectiveness remains uncertain. Recent data also explore imaging labels for prognosticating complaint inflexibility.

**CONCLUSION:** Hirayama complaint is a constantly overlooked but treatable cause of focal motor neuron dysfunction in youthful grown-ups. Feting crucial dynamic MRI features and enforcing suitable treatments can ameliorate clinical issues. farther exploration is necessary to upgrade surgical suggestions and prognostic models.( Ivysci)

**KEYWORDS :** Hirayama complaint, Juvenile muscular atrophy, Cervical flexion myelopathy Dynamic cervical MRI, Anterior cornucopia cell complaint

## INTRODUCTION

Hirayama complaint, a rare condition of cervical flexion myelopathy first discovered in Japan, is distributed as a lower motor neuron complaint that generally impacts the distal upper branches and is most common seen in adolescent males. The complaint medium involves unusual epidural dynamics during neck flexion, leading to contraction of the lower cervical cord and habitual ischemia of anterior cornucopia cells, setting it piecemeal from classical motor neuron complaint. Prompt clinical recognition and imaging are pivotal for opinion and precluding long-term disability. (Ivysci). Men are affected by this complaint five times more frequently than women (2). It's generally seen among youthful people who are in a period of rapid-fire growth. Its prevalence peaks at the age of 15 – 25 times (7,9). Clinical findings appear in the alternate or third decade and progress sluggishly (6). In this complaint, unilateral or asymmetric bilateral muscle weakness and atrophy, which progress sluggishly, do in the hands and forearm. The complaint is limited to upper extremity motor neuron involvement; lower extremity, sensitive, or bulbar involvement is n't observed (7,10). In some cases, muscle weakness becomes apparent in cold rainfall. Cases express that while weakness in fritters increases in cold rainfall, it gets normal in warm rainfall. thus, cases discover their complaint for the first time during downtime months. It is infrequently seen with hyperhidrosis and abnormal sympathetic skin responses (8). In 20 of the cases, weakness of the upper arm can be seen. While atrophy develops in the distal forearm and ulnar part, due to atrophy caused by lower cervical member damage, the brachioradialis muscle (oblique amyotrophy) innervated by C6 is generally recovered.

## METHODS

### Research Strategy:

The databases searched included PubMed and Web of Science through December 2025, with combinations of the keywords Hirayama complaint, juvenile muscular atrophy, cervical flexion myelopathy, dynamic MRI and surgery.

### Study Design and Protocol Registration :

This methodical review was conducted in agreement with the Preferred Reporting particulars for Methodical Reviews and Meta- Analyses (PRISMA) guidelines. This review aimed to synthesize the available substantiation on the clinical donation, individual features, radiological findings, and operation strategies for Hirayama complaint. A predefined protocol was established previous to the literature hunt.

### Data Sources and Search Strategy :

A comprehensive literature hunt was performed using electronic databases, including PubMed, MEDLINE, Scopus, and Google Scholar. The hunt covered papers published from the commencement of the databases until the most recent update. Keywords and Medical Subject headlines (MeSH) terms used in colorful combinations included “ Hirayama complaint, ” “ monomelic amyotrophy, ” “ juvenile muscular atrophy of distal upper branch, ” “ cervical flexion myelopathy, ” and “ anterior cornucopia cell complaint. ” The reference lists of applicable papers were manually screened to identify fresh eligible studies.

### Study Selection:

Two independent pundits screened the titles and objectifications of all recaptured papers for their ap-

plicability. Full- textbook papers were latterly assessed for eligibility grounded on the predefined addition and rejection criteria. disagreement between the pundits were resolved through discussion and agreement.

#### **Eligibility Criteria-**

- Original studies, case series, or case reports describing cases with Hirayama complaint
- joker or womanish cases progressed  $\geq 10$  times
- Clinical features of progressive distal upper branch weakness with muscle wasting
- Absence of significant sensitive poverties
- opinion supported by MRI cervical chine, particularly flexion studies
- papers published in English

#### **Rejection Criteria**

- Studies with deficient clinical or radiological data
- Reviews, studies, letters without original case data
- Studies describing indispensable judgments similar as amyotrophic side sclerosis, myelopathy, or supplemental neuropathies
- Beast studies

**Data birth :** Data were singly uprooted using a standardized data birth form. The uprooted variables included author details, time of publication, country, study design, patient demographics, duration of symptoms, clinical instantiations, neurological findings, MRI characteristics, electrophysiological findings, treatment modalities and issues.

**Quality Assessment:** The methodological quality of the included studies was assessed using applicable appraisal tools. Case reports and case series were estimated using the Joanna Briggs Institute( JBI) Critical Appraisal Checklist. The threat of bias was qualitatively assessed.

**Data conflation:** Owing to the diversity in study designs and outgrowth measures, a qualitative narrative conflation was performed. The findings were epitomized under thematic headlines, including clinical donation, individual criteria, imaging features, and operation approaches. A quantitative meta-analysis SPINALs was n't performed because of the variability in the reported issues.

**Ethical Considerations:** As this study was grounded on preliminarily published data, ethical blessing and informed concurrence were n't needed for this study.

#### **Results:**

**Epidemiology and Clinical donation:** utmost cases do in males aged 15 – 25 times, with asymmetric hand and forearm weakness and atrophy. sensitive involvement is generally absent in these cases.( PubMed)

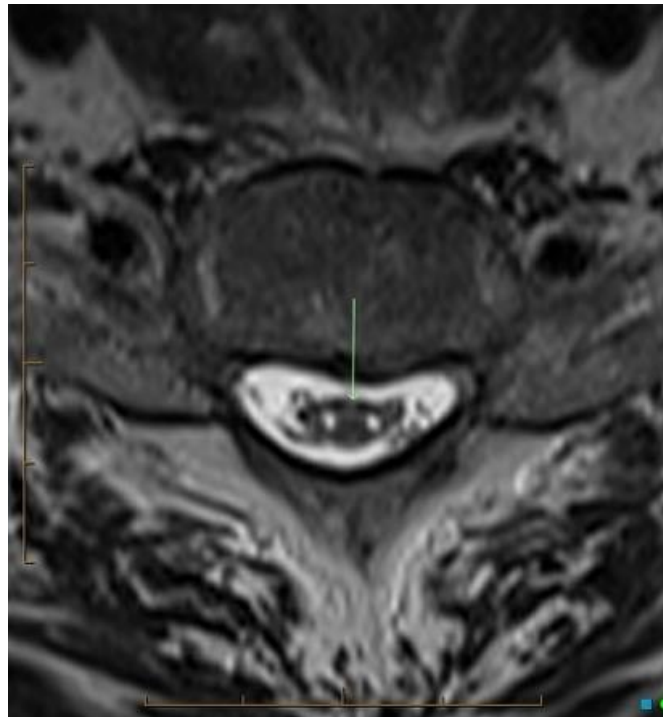
**Electrophysiological Findings :** EMG showed habitual denervation in the C7 – T1 myotomes, and sensitive studies were normal, supporting picky motor involvement.( Ivysci)

**Imaging Features:** Dynamic flexion MRI reveals anterior dural deportation and blowup of the posterior epidural space with venous plexus engorgement. MRI may reveal lower cervical cord atrophy and signal changes.( OUP Academic)



**THESE IMAGE SHOWN THE SPINAL CORD :**



**THESE IMAGE SHOWN IN CERVICAL SPINE MRI :****Discussion:**

Hirayama complaint presents a individual challenge because of its oddity and stage with other neuromuscular conditions. Dynamic cervical MRI is necessary for attesting the opinion and demonstrating pathognomonic dural shift and cord compression during neck flexion. Beforehand conservative operation of cervical immobilization limits functional loss. Surgical stabilization may be demanded for progressive complaint. Further high- quality studies are demanded to establish clear suggestions and optimal ways.

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