Review

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Do patients with Hirayama disease require surgical treatment? A review of the literature

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SUMMARY The main clinic characteristic of Hirayama disease (HD) is atrophy of the distal muscles in the upper limbs. Recently, an increasing number of HD cases have been reported. Many HD patients have persistently progressive symptoms and conservative treatments failed. This article aims to review the current status of the field and summarizes the main surgical treatment options for patients with HD. A comprehensive search of the PubMed and the Web of Science databases was conducted from their inception to September 15th, 2022. Search terms included "juvenile muscular atrophy of upper extremity", "Hirayama disease" and "surgery". A total of 169 relevant publications were identified and 29 articles were finally reviewed. Current surgical treatments for HD are either anterior cervical surgery or posterior cervical surgery. The two approaches can effectively stop the disease. However, no studies have compared the advantages and limitations of the two surgical methods. The previous view that HD can be improved with conservative treatment has been challenged. In many studies, surgical treatment has been shown to improve the hand function in patients with HD. However, there is still controversy about the methods of anterior and posterior cervical surgery. Future research could focus on exploring the advantages and limitations of different surgeries.

Keywords Hirayama disease, posterior cervical surgery, anterior cervical surgery

1. Introduction

Hirayama disease (HD), also referred to as juvenile muscular atrophy of the distal upper extremity, was first reported by Keizo Hirayama (*1*) in 1959. Previously, most reports of HD were in Asian countries such as Japan, China and India (*2-4*). Recently, an increasing number of HD cases have been reported in regions outside Asia. Table 1 summarizes cases of HD reported in PubMed in regions outside Asia in the past five years (*5-32*). Developments in imaging and electromyography (EMG) examination have improved the understanding of HD. Although the pathogenesis of HD is not fully understood, two main hypotheses have been proposed. The first is the hypothesis of dynamic cervical flexion compression. Repeated or continuous flexion of the cervical spine causes the posterior wall of the dural sac to move forward causing compression of the spinal cord that results in microcirculation disorders and chronic injury in the anterior horn of the spinal cord (*33,34*). The study of Sun *et al.* used diffusion tensor imaging (DTI) to scan patients with HD also supported the hypothesis (*35*). The other hypothesis is based on growth and development. The spinal cord and the dura mater are unbalanced during rapid growth and development in adolescence. During this time, the posterior roots become shortened. In the neutral position, the posterior root is in a relaxed state. When the neck is flexed, the shortened posterior root pulls the cervical spinal cord forwards resulting in compression of the cervical spinal cord and HD (*2,36*). Both hypotheses suggest that repeated and persistent cervical flexion is an important causative factor in HD.

Based on the above hypotheses, many physicians began to use cervical collar therapy to treat HD patients. In 1992, Tokumaru *et al.* compared 14 HD patients who had worn a cervical collar with 18 untreated patients (*37*). The study showed that cervical collar therapy can stop the progression of HD in a short period. Another study by their team also supported the reliability of this conclusion (*38*). However, conservative treatment has limitations. First, some reports showed that many patients still have continuous progress of symptoms 10 years after the onset of the disease. A small number of patients experience rapid progression months to years after symptoms have developed (*39,40*). Second, the efficacy of cervical collar

Table 1. Summary of the cases of HD published in PubMed in regions outside Asia in the past five years (2016-2022)

References	Country	Cases	Treatment
Kieser DC, *et al.* (*6*)	New Zealand	1	Collar therapy
Kumar M, *et al.* (*8*)	United States	1	Collar therapy
Baumann M, *et al.* (*9*)	Austria	1	Collar therapy
Lewis D, *et al.* (*5*)	United Kingdom	1	Collar therapy
Vachon C, *et al.* (*11*)	United States	1	Conservative treatment
Brems M, *et al.* (*12*)	Belgium	1	No treatment
Filiz MB, *et al.* (*13*)	Turkey	1	Physical therapy
McGregor S, *et al.* (*14*)	Canada	2	ACDF
Lolli VE, *et al.* (*15*)	Belgium	1	Not reported
Antonioni A, *et al.* (*16*)	Italy	1	Conservative treatment
Galletta K, *et al.* (*17*)	Italy	1	ACDF
Salome M, *et al.* (*18*)	The Netherlands	1	ACDF
Ayas ZO, *et al.* (*19*)	Turkey	1	Conservative treatment
Kapetanakis S, *et al.* (*20*)	Greece	1	Conservative treatment
Alpaydin Baslo S, *et al.* (*21*)	Turkey	1	Conservative treatment
Chanson JB, *et al.* (*22*)	France	1	Conservative treatment
Ay H, *et al.* (*23*)	Turkey	1	Conservative treatment
Tolu S, *et al.* (*24*)	Turkey	1	Collar therapy
Kandukuri GR, *et al.* (*25*)	United States	1	Conservative treatment
Witiw CD, *et al.* (*26*)	United States	1	ACDF
Macey MB, *et al.* (*27*)	United States	1	Not reported
Cabona C, *et al.* (*7*)	Italy	3	Conservative treatment
Wang H, *et al.* (*10*)	United States	1	Rehabilitation treatment
Koutsis G, *et al.* (*28*)	Greece	1	Not reported
Ashour M, *et al.* (*29*)	Canada	1	Posterior surgery
Abreu Tanure A, *et al.* (*30*)	Brazil	1	Nerve transfer
Hayden ME, *et al.* (*32*)	United States	1	Tendon transfer

HD, Hirayama disease; ACDF, anterior cervical discectomy and fusion.

therapy is greatly affected by patient compliance. A study showed that most patients wear cervical collars for less than half a year due to factors including appearance, inconvenience and progression of the disease (*41*). Finally, a study published in 2021 suggested that surgery may be better at helping patients with HD achieving symptomatic improvement than conservative treatment (*42*).Therefore, in patients with progressive disease, surgery should be considered for treatment. At the same time, a large number of studies including guidelines established by clinical multidisciplinary teams have highlighted that surgical treatment can effectively limit abnormal flexion of the cervical spine and expand the volume of the dural sac (*43-46*). Based on these studies, many HD patients who have failed conservative treatment have benefited from surgery. HD has become an area of intense research for spinal surgeons. In this article, we review the current knowledge of HD and provide a review of surgical treatments.

We conducted a structured search of PubMed and Web of Science. Publication dates were included from the inception of each database to September 15th, 2022. Search terms included "juvenile muscular atrophy of upper extremity", "Hirayama disease" and "surgery". A total of 169 relevant publications were identified. After the exclusion of duplicate publications and a further detailed review of the articles, 29 articles were finally included in the review. The inclusion criteria were case reports focusing on patient surgical treatment and prognosis, observational studies and randomized controlled trials (RCTs). Reviews, systematic reviews and meta-analyses were excluded.

2. Indications for surgical treatment

At present, the surgical treatment of HD aims to relieve the compression of the cervical spine and reconstruct normal cervical spine alignments. The guidelines established by the clinical multidisciplinary team showed that surgical treatment can be performed when the disease continues to progress after long-term wearing of a cervical collar, in patients who cannot tolerate the long-term wearing of a cervical collar and when symptoms progress (*43*). In addition, the Huashan clinical classification system for patients with HD was established in 2021 and its internal consistency was primarily verified (*47*). According to this system, HD patients can be divided into types I, II and III. Patients with type I HD have typical symptoms such as unilateral muscular atrophy without pyramidal tract signs or sensory disturbances. If the symptoms do not progress within 6 months, the patient is classified as subtype Ia; however, if the patient progresses, then the classification is subtype Ib. Patients with type II HD have typical symptoms accompanied by sensory disturbances or pyramidal tract signs. Patients with type III HD have an atypical form of HD involving the proximal muscles of the upper limbs or bilateral symptoms. Some physicians believe patients with HD of type Ib or above need surgical treatment (*47-49*).

3. Posterior cervical surgery

The literature and key conclusion on posterior cervical surgery are summarized in Table 2. Masaki *et al.* began to treat HD patients with surgery as early as 1990 (*50*). They believed that the main cause of the disease was increased imbalance of the cervical spine alignment. A posterior C2-C6 segmental fusion was performed on a HD patient. After surgery, the patient showed signs of improved muscular strength. In 1999, Kohno *et al.* performed posterior C4-C5 decompression and fusion on 3 HD patients (*51*). The surgery was successful and stopped the progression of the disease. Xu *et al.* performed posterior cervical fixation from the C3-C7 segment on a patient who had not benefited from 2-years of cervical collar therapy (*52*). Follow-up at three months and four years after surgery showed improvements in muscle atrophy. Goel *et al.* postulated that HD patients have multiple levels of cervical spine alignment imbalance that are not limited to the diseased segment. Therefore, they performed C1-C6/C7 posterior fixation on 4 patients. The patients were followed after surgery and showed no progression of hand symptoms (*53*).

Based on the clinical and imaging characteristics of HD patients, Konno *et al.* proposed that the dura mater lacks elasticity causing the spinal cord to become compressed and flattened and the spinal cord moves forward when patients flex their necks. They performed a duraplasty and fusion in patients and showed improvement of symptoms after surgery (*54*). In 2014, Ito *et al*. performed cervical duraplasty with tenting sutures *via* laminoplasty on HD patients. Their surgery achieved positive results (*44*). In 2009, Patel *et al.* performed C4-T1 laminectomy and duraplasty to treat HD patients. The muscle strength of the patients improved significantly after the surgery (*55*). Brandicourt *et al.* performed a posterior cervical laminectomy and posterior venous plexus micro-resection on HD patients. All of the patients' symptoms stopped progressing after the surgery (*56*).

In general, most of the studies on posterior cervical surgery are case reports or case series. Compared to anterior cervical surgery, there is still a lack of advanced evidence-based medical studies to support its effectiveness and reliability.

4. Anterior cervical surgery

The literature and key conclusion on anterior cervical surgery are summarized in Table 3.

4.1. Is anterior cervical surgery effective?

Paredes *et al.* (*57*) admitted a Caucasian male with HD, and cervical collar therapy was ineffective. They chose C5-C6 anterior cervical discectomy and fusion (ACDF). One year after the surgery, the lower limb muscle strength in the patient had completely recovered and the symptoms of upper limbs stopped progressing. Imamura *et al.* (*58*) reported a case in a 16-year-old HD patient who underwent anterior cervical corpectomy decompression and fusion (ACCF). The patient was in good condition after surgery without complications and follow-up at six months showed the symptoms had stopped progressing. Wu *et al.* (*59*) reported a 34-year-old patient with advanced HD who underwent ACCF surgery and recovered well after surgery. Kohno *et al.* (*51*) performed ACCF on 4 patients with HD which stopped the progression of the disease. Their case illustrates the effectiveness of anterior cervical surgery.

Zhang *et al.* (*60*) performed anterior cervical internal fixation with or without fusion on 19 HD patients. For the internal fixation group, titanium plates and screws were used for fixtures during the operation, while the fusion group used intervertebral fusion cages or autologous iliac bone graft. After an average of 70 months of follow-up, the patient had a reduced range of flexion in the cervical spine, the lower cervical spine mobility angle was reduced, the cervical lordosis was restored and the cross-sectional area of the spinal cord increased. Liu *et al.* (*61*) investigated short-term to midterm clinical outcomes after ACDF surgery among 115 patients with HD. Their study found that most patients showed improvement in postoperative cervical sagittal alignments, hand function rating scales, and EMG data. Therefore, they concluded HD patients can benefit from ACDF surgery. These data indicated that anterior cervical surgery has significant clinical efficacy and is associated with improvements detected on imaging.

In addition to case control and cohort studies, a RCT showed that anterior cervical surgery is effective in treating HD. In 2013, Lu *et al.* carried a prospective RCT (*45*) involving anterior cervical decompression and fusion in 48 male HD patients. 24 patients underwent ACDF and 24 underwent ACCF. Symptoms were assessed and EMG was performed before and after surgery. Both groups showed improvements of > 60% demonstrating that anterior cervical surgery is effective in treating HD.

4.2. What factors affect the effectiveness of anterior cervical surgery?

As mentioned above, many physicians tend to perform posterior cervical surgery on HD patients based on the fact that posterior surgery can reduce cervical spine alignments imbalance. Anterior surgery is also an active area of research. A retrospective study by Song *et al.* (*62*) included 23 HD patients. ACDF were performed and the alignment of the cervical spine in sagittal position was compared to 21 healthy participants before and after the surgery. Specifically, the alignments included the C2-C7 Cobb angle, the thoracic inlet angle, the C2-C7 sagittal vertical axis and the neck tilt angle. The study showed an

Table 2. Summary of the studies about posterior cervical surgery

References	Type of study	Key conclusion
Ashour M, *et al.* (*29*)	Case report	Patient got symptom improvement with Posterior cervical surgery.
Masak T, *et al*. (*50*)	Case report	Patient got symptom improvement with posterior C2-C6 segmental fusion.
Kohno M, *et al.* (*51*)	Case report	Patient got symptom stabilization with posterior C4-C5 decompression and fusion.
Xu Q, *et al.* (*52*)	Case report	Patient got symptom improvement with posterior cervical fixation from the C3-C7 segment.
Goel A, *et al.* (*53*)	Case report	Patient got symptom improvement with posterior C1-C6/C7 posterior fixation.
Konno S, *et al.* (*54*)	Case report	Patient got symptom improvement with posterior duraplasty and fusion surgery.
Ito H, *et al.* (*44*)	Case report	Patient got symptom improvement with posterior duraplasty surgery.
Patel TR, *et al.* (*55*)	Case report	Patient got symptom improvement with C4-T1 laminectomy and duraplasty.
Brandicourt P, *et al.* (*56*)	Case report	Patient got symptom improvement with posterior cervical laminectomy and posterior venous plexus micro-resection.
Thakar S, *et al.* (*42*)	Case control study	Compared with conservative treatment, cervical duraplasty can help HD patients get better clinical and electrophysiological improvement.

HD, Hirayama disease.

Table 3. Summary of the studies about anterior cervical surgery

References	Type of study	Key conclusion
McGregor S, *et al.* (*14*)	Case report	Patient got symptom stabilization with ACDF.
Galletta K, *et al.* (*17*)	Case report	Patient got symptom stabilization with ACDF.
Salome M, *et al.* (*18*)	Case report	Patient got symptom stabilization with ACDF.
Witiw CD, *et al.* (*26*)	Case report	Patient got symptom improvement with ACDF.
Paredes I, *et al.* (*57*)	Case report	Patient got symptom improvement with C5-C6 ACDF.
Imamura H, *et al.* (*58*)	Case report	Patient got symptom improvement with ACCF.
Wu W, *et al.* (*59*)	Case report	Patient got symptom improvement with ACCF.
Kohno M, *et al.* (*51*)	Case report	Patient got symptom improvement with ACCF.
Zhang H, *et al.* (*60*)	Cohort study	Patients can benefit from anterior cervical surgery; fusion and fixation have similar efficacy.
Liu S, *et al.* (*61*)	Case control study	Patients showed improvement in cervical sagittal alignments, hand function rating scales, and electromyographic data after ACDF surgery.
Lu F, *et al.* (*45*)	RCT	Patients can benefit from anterior cervical surgery; ACDF and ACCF have similar efficacy.
Song J, *et al.* (*62*)	Cohort study	HD patient have an imbalance of the cervical spine alignments compared to healthy individuals and the imbalance can be corrected by ACDF.
Lu X, *et al.* (*49*)	Case control study	Preoperative sagittal alignments of the cervical spine in HD patients may be a predictor of postoperative outcomes.
Zheng C, *et al.* (*46*)	Cohort study	ACDF for HD patients can restore nerve conduction in the cervical spinal cord and restore muscle strength.
Wang HL, *et al.* (*64*)	Cohort study	ACDF for HD patients can promote the functional reconstruction of upper motor neurons.
Zou F, *et al.* (*65*)	Case control study	The degree of spinal cord flatness, spinal atrophy, and postoperative spinal cord recovery is related to the outcome of ACDF for HD patients.
Song J, *et al.* (*62*)	Case control study	Age of onset, course of the disease, physiological reflexes and pathological reflexes were risk factors that affect the outcome of ACDF for HD patients.
Yu Q, *et al.* (*66*)	Case control study	Loss of attachment is an important factor affecting surgical outcomes in patients with HD.
Wang H, *et al.* (*67*)	Case control study	In addition to neck flexion MRI, dynamic X-ray is also an important reference index for selecting surgical segments in patients with HD.
Lu X, *et al.* (*63*)	Cohort study	HD patient have increased stress in the entire cervical spine and can be relieved by ACDF.

HD, Hirayama disease; ACDF, anterior cervical discectomy and fusion; ACCF, anterior cervical corpectomy decompression and fusion.

imbalance of the cervical spine alignments in HD patients compared to healthy individuals, and the imbalance can be corrected by ACDF. Another study carried out by the team concluded that preoperative sagittal alignment of the cervical spine in HD patients may be a predictor of postoperative outcomes (*49*). In addition, the study of Lu *et al.* pointed out that HD patients have increased stress in the entire cervical spine and it can be relieved by ACDF (*63*).

A study by Zheng *et al.* (*46*) showed that ACDF for HD patients can immediately eliminate the abnormality of the F wave in the flexion position of the neck. The results showed ACDF can help to restore not only the nerve conduction in the cervical spinal cord but also the muscle strength of the upper limbs. Also, Wang *et al*. (*64*) showed that ACDF can promote the functional reconstruction of upper motor neurons. By measuring brain functional magnetic resonance imaging (fMRI) and handgrip strength before and after surgery, they found that the contralateral and ipsilateral cortex were simultaneously activated before surgery. After surgery, the observed pathological activation decreased and the handgrip strength increased.

Studies have analyzed the factors that affect surgery outcomes. Zou *et al*. (*65*) retrospectively analyzed 40 HD patients and performed ACDF on these patients. The patients were followed for an average of 18 months and Odom scores were recorded after surgery. Based on the scores, the patients were divided into effective and ineffective groups and the imaging findings were

compared across the two groups. The data indicated that the degree of spinal cord flatness, spinal atrophy, and postoperative spinal cord recovery is related to the outcome of the surgery. Another retrospective study by Song *et al.* (*62*) included 210 patients who underwent ACDF. The Odom score was recorded after surgery, and the patients were divided into effective and ineffective groups. The age, gender, age of onset, course of the disease, cervical alignment, physiological and pathological reflexes, and involvement of the bilateral extremities were compared across the groups. The authors concluded that four factors, age of onset, course of the disease, physiological reflexes and pathological reflexes were risk factors that affected the outcome of the surgery.

The above studies have confirmed reliability and effectiveness of anterior surgery for HD. In addition, a preliminary exploration of the factors affecting the surgical effect was also carried out in these studies. However, the choice of surgical segments remains controversial.

4.3. How many segments and which segments should be chosen?

The first question is how many segments should be chosen. Paredes (*57*) chose one segment. In the study of Zhang *et al.* (*60*), there are examples of choosing three segments. Lu *et al.* (*45*) and Zheng *et al.* (*46*) chose two segments and believed that fusion of the two segments could limit the abnormal movement of the cervical spine while retaining sufficient cervical spine mobility. The study of Yu *et al.* (*66*) showed that when the longitudinal separation range is ≥ 5 cervical segments, longer segment fusion may be required to further improve the surgical efficacy in such patients. Wang *et al.* (*67*) proposed that as the cervical spine pressure of HD patients changes when standing and lying down, previous studies only using magnetic resonance imaging (MRI) to determine stability of the cervical spine are not comprehensive. They then conducted a retrospective analysis of 50 HD patients and measured the Cobb angle of the cervical spine and the range of motion of the cervical spine with dynamic X-ray imaging and MRI. The study concluded that two imaging techniques should be used to select the most appropriate surgical segments. The unstable segment of the dynamic position X-ray is also a factor that should be considered. Clinical guidelines (*43*) and update reviews (*48*) indicate that patients with HD always have an imbalance in the cervical spine sagittal alignments. In addition, the patient also has imaging manifestations characterized by loss of attachment between the posterior dural sac and the subjacent lamina, and the clinical symptoms are mainly hand dysfunction. Therefore, the selection of surgical segments needs to combine dynamic X-ray, neck flexion MRI and EMG.

In summary, many studies have confirmed the effectiveness and reliability of anterior surgery. However, there is still a lack of reports comparing anterior and posterior cervical surgery.

5. Conclusions

With an improved understanding of the clinical manifestations and pathogenesis of HD, an increasing number of cases are being diagnosed. The treatment of HD can be divided into conservative and surgical treatments. When conservative treatment fails to stop progression, surgery can be used. However, the indications for surgery remain controversial and more evidence is required to support its use. Surgical treatments can be divided into anterior and posterior cervical surgery. These two surgical methods can effectively stop the disease. Anterior cervical surgery has more detailed studies concerning the choice of surgical methods, surgical segments, and prognosis. However, no study has demonstrated superiority of either approach. Future research should focus on exploring surgical indications and comparing the advantages and limitations of anterior and posterior cervical surgery.

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References

1. Hirayama K, Tsubaki T, Toyokura Y, Okinaka S. Juvenile muscular atrophy of unilateral upper extremity. Neurology. 1963; 13:373-380.
2. Tashiro K, Kikuchi S, Itoyama Y, Tokumaru Y, Sobue G, Mukai E, Akiguchi I, Nakashima K, Kira J, Hirayama K. Nationwide survey of juvenile muscular atrophy of distal upper extremity (Hirayama disease) in Japan. Amyotroph Lateral Scler. 2006; 7:38-45.
3. Zhou B, Chen L, Fan D, Zhou D. Clinical features of Hirayama disease in mainland China. Amyotroph Lateral Scler. 2010; 11:133-139.
4. Nalini A, Gourie-Devi M, Thennarasu K, Ramalingaiah AH. Monomelic amyotrophy: clinical profile and natural

history of 279 cases seen over 35 years (1976-2010). Amyotroph Lateral Scler Frontotemporal Degener. 2014; 15:457-465.
5. Lewis D, Saxena A, Herwadkar A, Leach J. A confirmed case in the United Kingdom of Hirayama disease in a young white male presenting with hand weakness. World Neurosurg. 2017; 105:1039.e7-1039.e12.
6. Kieser DC, Cox PJ, Kieser SCJ. Hirayama disease. Eur Spine J. 2018; 27:1201-1206.
7. Cabona C, Beronio A, Martinelli I, Briani C, Lapucci C, Serrati C, Delucchi S, Mannironi A, Benedetti L. Are basketball players more likely to develop Hirayama disease? J Neurol Sci. 2019; 400:142-144.
8. Kumar M, Athwal PSS, Rhandhawa S, Kahlon S, Shiv Kumar J. Hirayama's disease in a young male: a rare case report. Cureus. 2019; 11:e6204.
9. Baumann M, Finsterer J, Gizewski ER, Loscher WN. Early-onset Hirayama disease in a female. SAGE Open Med Case Rep. 2017; 5:2050313X16686710.
10. Wang H, Chen Y. Poster 469 A variant of motor neuron disorder: Hirayama disease: a case report. PM R. 2016; 8:S313.
11. Vachon C, Abu Libdeh A. Clinical reasoning: A 17-year-old baseball player with right hand weakness. Neurology. 2019; 92:e76-e80.
12. Brems M, Van Goethem J, De Praeter M, Willekens B. Subacute onset Hirayama disease with extensive myelopathy on MRI in flexed posture. Acta Neurol Belg. 2017; 117:795-797.
13. Filiz MB, Cakir T, Toraman NF, DoGan SK, Toslak IE, Uysal H. Late-onset Hirayama disease presenting with ulnar neuropathy: a case report. J Pak Med Assoc. 2017; 67:1280-1282.
14. McGregor S, Joswig H, Duggal N, Miller TA. Hirayama disease: a diagnostic and therapeutic challenge. Can J Neurol Sci. 2017; 44:754-756.
15. Lolli VE, Sarbu N, Pezzullo M, Mavroudakis N. Flexion MRI in a case of Hirayama disease. Radiol Case Rep. 2020; 15:1701-1704.
16. Antonioni A, Fonderico M, Granieri E. Hirayama disease: a case of an Albanian woman clinically stabilized without surgery. Front Neurol. 2020; 11:183.
17. Galletta K, Gaeta M, Alafaci C, Vinci S, Longo M, Grasso G, Granata F. Hirayama disease - early MRI diagnosis of subacute medullary ischemia: a case report. Surg Neurol Int. 2020; 11:115.
18. Salome M, Barkhof F, Visser L. Hirayama disease; an atypical clinical manifestation of a cervical myelopathy with typical MRI features. BMJ Case Rep. 2017; 2017:bcr2016217780.
19. Ayas ZO, Asil K. Atypical type of Hirayama disease: onset of proximal upper extremity. Ideggyogy Sz. 2018; 71:284-287.
20. Kapetanakis S, Chourmouzi D, Terzoudi A, Georgiou N, Giovannopoulou E. Hirayama disease: diagnostic essentials in neuroimaging. Clin Case Rep. 2017; 5:2151-2152.
21. Alpaydin Baslo S, Erdogan M, Balcik ZE, Ozturk O, Atakli D. Is Hirayama a Gq1b disease? Neurol Sci. 2019; 40:1743-1747.
22. Chanson JB, Renaud M, Echaniz-Laguna A, Koob M. Dilation of epidural space and posterior soft tissue veins in Hirayama disease. BMJ Case Rep. 2017; 2017:bcr2017220787.
23. Ay H. Hirayama disease (monomelic amyotrophy) clinically confused for carpal tunnel syndrome. Neuropsychiatr Dis Treat. 2017; 13:1385-1388.
24. Tolu S, Aysal F, Ormeci T, Kirez IE, Hindioglu N. Flexion-induced cervical cord compression: Hirayama Disease. Balkan Med J. 2019; 36:190-191.
25. Kandukuri GR, Acosta NR. Significance of dynamic imaging in diagnosis of Hirayama disease: a rare case report and literature review. Kans J Med. 2020; 13:58-60.
26. Witiw CD, O'Toole JE. Electric shocks and weakness of the right hand in a young man: Hirayama disease. Lancet. 2019; 394:684.
27. Macey MB, Ho DT, Parres CM, Small JE. Spinal epidural venous plexus pathology in Hirayama disease. J Clin Neuromuscul Dis. 2019; 21:47-51.
28. Koutsis G, Velonakis G, Breza M, Karavasilis E, Zambelis T, Panas M. Neck flexion-induced finger tremor: A novel observation in Hirayama disease. J Neurol Sci. 2016; 364:42-44.
29. Ashour M, Fortin O, Parent L, Sébire G, Saint-Martin C, Poulin C, Dudley R. Hirayama disease in a patient with a history of late-onset symptomatic vein of galen aneurysmal malformation. Pediatr Neurosurg. 2021; 56:555-562.
30. Abreu Tanure A, Rosifini Alves Rezende LG, Pazim AC, Leal Ribeiro M. Supercharged end-to-side anterior interosseous to ulnar motor nerve transfer for Hirayama disease: a case report. Hand (N Y). 2022; 17: NP12-NP16.
31. Tadokoro N, Hashimoto K, Kiyasu K, Kasai Y, Aoyama N, Takemasa R, Ikeuchi M. Finger trembling improvement after surgery in Hirayama disease: a case report. Spinal Cord Ser Cases. 2022; 8:44.
32. Hayden ME, Kim J, Arányi Z, Wolfe SW. Outcome of tendon transfer for monomelic amyotrophy (Hirayama disease). J Hand Surg Am. 2022. S0363-5023(21)00754-1.
33. Hirayama K. Juvenile muscular atrophy of distal upper extremity (Hirayama disease). Intern Med. 2000; 39:283-290.
34. Fujimoto Y, Oka S, Tanaka N, Nishikawa K, Kawagoe H, Baba I. Pathophysiology and treatment for cervical flexion myelopathy. Eur Spine J. 2002; 11:276-285.
35. Sun C, Zhou S, Cui Z, Zhang Y, Wang H, Jiang J, Lu F, Ma X. The evaluation on neural status of cervical spinal cord in normal and Hirayama disease using diffusion tensor imaging. Eur Spine J. 2019; 28:1872-1878.
36. Toma S, Shiozawa Z. Amyotrophic cervical myelopathy in adolescence. J Neurol Neurosurg Psychiatry. 1995; 58:56-64.
37. Tokumaru Y, Hirayama K. A cervical collar therapy for non-progressive juvenile spinal muscular atrophy of the distal upper limb (Hirayama's disease). Rinsho Shinkeigaku. 1992; 32:1102-1106. (in Japanese)
38. Tokumaru Y, Hirayama K. Cervical collar therapy for juvenile muscular atrophy of distal upper extremity (Hirayama disease): results from 38 cases. Rinsho Shinkeigaku. 2001; 41:173-178. (in Japanese)
39. Huang YC, Ro LS, Chang HS, Chen CM, Wu YR, Lee JD, Lyu RK. A clinical study of Hirayama disease in Taiwan. Muscle Nerve. 2008; 37:576-582.
40. Ciceri EF, Chiapparini L, Erbetta A, Longhi L, Cicardi B, Milani N, Solero CL, Savoiardo M. Angiographically proven cervical venous engorgement: a possible concurrent cause in the pathophysiology of Hirayama's myelopathy. Neurol Sci. 2010; 31:845-848.
41. Fu Y, Qin W, Sun QL, Fan DS. Investigation of the

compliance of cervical collar therapy in 73 patients with Hirayama disease. Zhonghua Yi Xue Za Zhi. 2016; 96:3485-3488. (in Chinese)
42. Thakar S, Arun AA, Rajagopal N, Aryan S, Mohan D, Vijayan JE, Hegde AS. Outcomes after cervical duraplasty for monomelic amyotrophy (Hirayama disease): results of a case-control study of 60 patients. J Neurosci Rural Pract. 2021; 12:642-651.
43. Lyu F, Zheng C, Wang H, *et al*. Establishment of a clinician-led guideline on the diagnosis and treatment of Hirayama disease using a modified Delphi technique. Clin Neurophysiol. 2020; 131:1311-1319.
44. Ito H, Takai K, Taniguchi M. Cervical duraplasty with tenting sutures *via* laminoplasty for cervical flexion myelopathy in patients with Hirayama disease: successful decompression of a "tight dural canal in flexion" without spinal fusion. J Neurosurg Spine. 2014; 21:743-752.
45. Lu F, Wang H, Jiang J, Chen W, Ma X, Ma X, Xia X, Wang L. Efficacy of anterior cervical decompression and fusion procedures for monomelic amyotrophy treatment: a prospective randomized controlled trial: clinical article. J Neurosurg Spine. 2013; 19:412-419.
46. Zheng C, Nie C, Lei W, Zhu Y, Zhu D, Wang H, Lu F, Weber R, Jiang J. Can anterior cervical fusion procedures prevent the progression of the natural course of Hirayama disease? An ambispective cohort analysis. Clin Neurophysiol. 2018; 129:2341-2349.
47. Sun C, Xu G, Zhang Y, Cui Z, Liu D, Yang Y, Wang X, Ma X, Lu F, Jiang J, Wang H. Interobserver and intraobserver reproducibility and reliability of the Huashan clinical classification system for Hirayama disease. Front Neurol. 2021; 12:779438.
48. Wang H, Tian Y, Wu J, Luo S, Zheng C, Sun C, Nie C, Xia X, Ma X, Lyu F, Jiang J, Wang H. Update on the pathogenesis, clinical diagnosis, and treatment of Hirayama disease. Front Neurol. 2021; 12:811943.
49. Lu X, Xu GY, Nie C, Zhang YX, Song J, Jiang JY. The relationship between preoperative cervical sagittal balance and clinical outcome of patients with Hirayama disease treated with anterior cervical discectomy and fusion. Neurospine. 2021; 18:618-627.
50. Masaki T, Hashida H, Sakuta M, Kunogi J. A case of flexion myelopathy presenting juvenile segmental muscular atrophy of upper extremities--a successful treatment by cervical spine immobilization. Rinsho Shinkeigaku. 1990; 30:625-629. (in Japanese)
51. Kohno M, Takahashi H, Ide K, Yamakawa K, Saitoh T, Inoue K. Surgical treatment for patients with cervical flexion myelopathy. J Neurosurg. 1999; 91(1 Suppl):33-42.
52. Xu Q, Gu R, Zhu Q, Suya D. A severe case of Hirayama disease successfully treated by posterior cervical fixation without decompression and fusion. World Neurosurg. 2019; 122:326-330.
53. Goel A, Dhar A, Shah A. Multilevel spinal stabilization as a treatment for Hirayama disease: report of an experience with five cases. World Neurosurg. 2017; 99:186-191.
54. Konno S, Goto S, Murakami M, Mochizuki M, Motegi H, Moriya H. Juvenile amyotrophy of the distal upper extremity: pathologic findings of the dura mater and surgical management. Spine (Phila Pa 1976). 1997; 22:486-492.
55. Patel TR, Chiocca EA, Freimer ML, Christoforidis GA. Lack of epidural pressure change with neck flexion in a patient with Hirayama disease: case report. Neurosurgery. 2009; 64:E1196-1197; discussion E1197.
56. Brandicourt P, Sol JC, Aldea S, Bonneville F, Cintas P, Brauge D. Cervical laminectomy and micro resection of the posterior venous plexus in Hirayama disease. Neurochirurgie. 2018; 64:303-309.
57. Paredes I, Esteban J, Ramos A, Gonzalez P, Rivas JJ. A severe case of Hirayama disease successfully treated by anterior cervical fusion. J Neurosurg Spine. 2014; 20:191-195.
58. Imamura H, Matsumoto S, Hayase M, Oda Y, Kikuchi H, Takano M. A case of Hirayama's disease successfully treated by anterior cervical decompression and fusion. No To Shinkei. 2001; 53:1033-1038. (in Japanese)
59. Wu W, Wang S, Lin J. A 34-year-old female patient with Hirayama disease complicated by severe spinal cord injury. World Neurosurg. 2019; 130:84-88.
60. Zhang H, Wang S, Li Z, Shen R, Lin R, Wu W, Lin J. Anterior cervical surgery for the treatment of Hirayama disease. World Neurosurg. 2019; 127:e910-e918.
61. Liu S, Zou F, Lu F, Xia X, Wang H, Zheng C, Gong Z, Ma X, Jiang J. The short-term to midterm follow-up of patients with Hirayama disease after anterior cervical discectomy and fusion. World Neurosurg. 2021; 150:e705-e713.
62. Song J, Cui ZY, Chen ZH, Jiang JY. Analysis of the effect of surgical treatment for the patients with Hirayama disease from the perspective of cervical spine sagittal alignment. World Neurosurg. 2020; 133:e342-e347.
63. Lu X, Zou F, Lu F, Ma X, Xia X, Jiang J. How to reconstruct the lordosis of cervical spine in patients with Hirayama disease? A finite element analysis of biomechanical changes focusing on adjacent segments after anterior cervical discectomy and fusion. J Orthop Surg Res. 2022; 17:101.
64. Wang HL, Wu YW, Song J, Jiang JY, Lu FZ, Ma XS, Xia XL. Cortical activation changes in Hirayama disease after anterior cervical decompression and fusion. World Neurosurg. 2018; 116:e588-e594.
65. Zou F, Yang S, Lu F, Ma X, Xia X, Jiang J. Factors affecting the surgical outcomes of Hirayama disease: a retrospective analysis of preoperative magnetic resonance imaging features of the cervical spine. World Neurosurg. 2019; 122:e296-e301.
66. Yu Q, Tang S, Jiang J, Jin X, Lyu F, Ma X, Xia X. The influence of "loss of attachment" on the outcome of anterior cervical fusion procedures in patients with Hirayama disease. Orthopedics. 2021; 44:30-37.
67. Wang H, Sun C, Yang S, Jiang J, Lu F, Ma X, Xia X. Dynamic cervical radiographs in patients with Hirayama disease: an unneglectable factor on the choice of surgery options. World Neurosurg. 2018; 114:e433-e440.

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