

CASE REPORT

# Hirayama Disease—Dynamic Cervical Compressive Myelopathy: An Indian Perspective

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**ABSTRACT**

**Introduction:** Hirayama disease is a rare form of dynamic cervical flexion myelopathy. The incidence is more in Asian countries, like Japan, India, etc. Early diagnosis and treatment is the mainstay for the prognosis for this disease.

**Materials and methods:** Our aim is to assess the outcome of anterior cervical stabilization in this disease. We had six patients who was diagnosed to have Hirayama disease in our center and underwent anterior cervical stabilization.

**Results:** All six patients had fair to good outcome according to Odom’s criteria.

**Conclusion:** Early diagnosis and prompt treatment is the mainstay for better outcome. Mode of treatment depends on surgeons preference.

**Keywords:** Cervical compressive myelopathy, Dynamic myelopathy, Hirayama.

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**INTRODUCTION**

Hirayama disease is first reported by Hirayama in 1959.<sup>1</sup> This is also called as Juvenile muscular atrophy of upper extremity. The most common clinical features include predominance in young males with asymmetrical hand and forearm muscles atrophy and preserved brachioradialis and radial group of muscles, minimal or no sensory disturbances without no involvement of lower limbs. On imaging, there will be forward migration of posterior dura in flexion at lower cervical levels. This is seen better with myelography. The symptoms worsen in cold weather and, after few years, there will be cessation of progression. There is no family inheritance found so far. The same condition is also reported in thoracic levels too.<sup>2</sup>

There are various treatment options advocated for this disease. Here, we report this case series where

anterior stabilization helps in arresting the progression and improvement of the disease. In India, studies of Hirayama disease is scarcely available.

**CASE REPORT**

In our series, we had six patients over a period of 2008 to 2011 in our center. All the patients were adolescent males with age ranging from 16 to 27 years and a median of 18 years. Clinically, most of these patients had predominant right hand muscle wasting. They were evaluated with dynamic magnetic resonance imaging (MRI) showing migration of posterior dura and thinning of the cord at the corresponding levels in flexion (Fig. 1). Electromyography (EMG) monitoring revealed high-amplitude action potentials of long duration in association with a reduction of the total motor-unit voltage during the maximum voluntary contraction of the atrophic muscles suggesting neurogenic EMG pattern. In all these patients, nerve conduction velocities showed normal motor and sensory conduction velocities in both ulnar and median nerves. All these patients underwent anterior cervical stabilization with or without decompression. They were assessed post-operatively and followed up for a minimum period of 2 years. Odom’s outcome criteria (Table 1) were used to assess the improvement in the preoperative features. Two of these patients had good outcome with significant symptom relief. Table 2 gives the details of the patients and the observations.

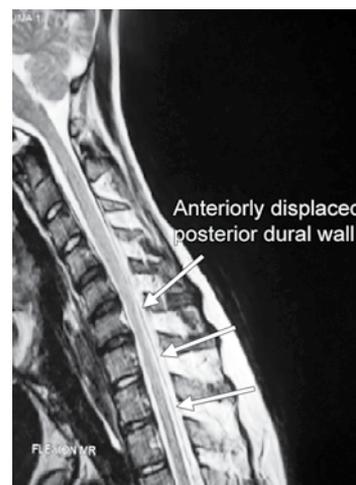


Fig. 1: Posterior dura displaced anteriorly

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**Table 1:** Odom's criteria

Excellent	All preoperative symptoms relieved; abnormal findings improved
Good	Minimal persistence of preoperative symptoms; abnormal findings unchanged or improved
Fair	Definite relief of some preoperative symptoms; other symptoms unchanged or slightly improved
Poor	Symptoms and signs unchanged or exacerbated

## DISCUSSION

### Pathophysiology

Autopsy of patients with Hirayama disease revealed focal anterior horn cell necrosis with preserved white matter of the cervical cord.<sup>3,4</sup> This is due to circulation disturbances caused by repetitive or persistent compressive force applied to the cervical cord with neck in flexion. Reid et al<sup>5,6</sup> and Beig et al<sup>7,8</sup> hypothesized that, in the flexed-neck position, compression of the cervical spinal cord by anterior structures, such as the vertebral bodies or intervertebral disks, causes cervical myelopathy (contact pressure mechanism). Iwasaki et al<sup>9</sup> and Kikuchi et al<sup>10</sup> hypothesized that the cervical spinal cord was compressed by the migrating posterior wall of the dura mater in the flexed-neck position (tight dural canal in flexion mechanism). Tokumaru and Hirayama<sup>11</sup> proposed that the anterior shift of the posterior dura mater provides a clue to the cause and a possible method for arresting the progression of CFM. The clinical features of this disease include predominance in adolescent males, muscular atrophy of the upper extremities with laterality, slight or no sensory disturbance, and forward migration of the posterior surface of the lower cervical dura mater, which is recognized on myelography when the neck is flexed.<sup>11</sup> The distribution of the muscular atrophy is characteristic and is recognized in the distal upperextremity muscles that are innervated by the lower cervical spinal cord.<sup>12,13</sup> Toma and

Shiozawa<sup>14</sup> have hypothesized that cervical dorsal roots, which had become relatively shortened during the juvenile growth period, caused forward migration of the spinal cord and the dura mater. Motor neuron disease usually has bilateral effects and affects not only the hand and ulnar-side forearm but also the legs. It also shows continuous progression of muscular atrophy, whereas muscle atrophy in cervical flexion myelopathy ceases to worsen after a few years of insidious progression. Myogenic diseases can be excluded by examining EMG findings.<sup>15</sup>

### TREATMENT

There are various treatment modalities being carried out all over. It includes conservative treatment with continuous cervical collar, anterior decompression and stabilization, posterior decompression and stabilization with or without duroplasty and duroplasty alone by posterior approach. The presence of ischemic necrosis in the anterior part of the cervical cord and improvement of the clinical symptoms by short-segment ADF support the hypothesis of the contact pressure mechanism.<sup>16</sup> Masaki et al,<sup>17</sup> Ohnari et al,<sup>18</sup> and Kohno et al<sup>19</sup> have described a posterior fusion procedure and reported good results. However, longer fusion segments were required. Because most of the patients are children and commonly exhibit neurological symptoms specific to C-7 or C-8, shorter-length fusion to prevent kyphotic alignment in flexion and to decrease cervical movement at only the corresponding segments is ideal.

Tokumaru et al,<sup>20</sup> in a study on patients with juvenile muscular atrophy, placed their patients in a cervical collar for 3 to 4 years if the patient had kept the neck flexed for more than 10 to 20 minutes, and they considered this therapy to be effective. In five of the 14 patients in their series, improvement in upperextremity paresis was recognized, and in another eight patients symptom progression was halted. Konno et al<sup>21</sup> and Mochizuki et al<sup>22</sup> reported

**Table 2:** List of cases enrolled in this study

Patients	Age	Duration of symptoms	Wasting of muscles	Sensory involvement	Cervical full flexion MRI	Surgery	Odom's outcome status
1	22	2-3 yrs	Hands R>L	Nil	Atrophy at C4-6	C4-6 anterior stabilization	Fair
2	17	2 yrs	Hands and forearms R>L	Nil	Atrophy from C6-7 to D2-3, C4-5 and D5-6 disk herniation	C4-5 and C5-6 anterior decompression and stabilization	Fair
3	17	1 yr	Hands R>L	Rt UL- numbness	Atrophy at C4-6	C3-6 anterior stabilization	Good
4	27	2 yrs	Hands R>L	Nil	Atrophy at C6-7 C6-7 disk herniation	C6-7 anterior decompression and stabilization	Good
5	19	1 yr	Right thenar and forearm	Rt hand tingling o/e mild involvement	Atrophy at C5-D1	C4-5, C5-6, C6-7 anterior decompression and stabilization	Fair
6	16	5 yrs	Right hand and forearm	Nil	Atrophy at C6-D1	C3-6 anterior stabilization	Fair



the effectiveness of adding duroplasty to posterior cervical fusion, whereas Fujimoto et al<sup>23</sup> have stated that duroplasty alone was effective for treating this disease.

## CONCLUSION

Suspicion of Hirayama disease in adolescent male patients with atrophy of hand and forearm muscles and early diagnosis helps in further progression of the disease. This helps in preventing the debilitating deficits. Clinicians in countries other than Asian countries should also consider this disease as diagnosis of exclusion. Anterior cervical stabilization with or without decompression also can be considered as choice of treatment. However, long-term effects are yet to be proved.

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