

## MRI<sup>1</sup>

· · · · ·<sup>2</sup> · ·<sup>3</sup>

: (Hirayama )

: 2001 1 2002 6 Hirayama 7  
(17 - 43 , 23.7 ) 5 (25 - 32 ) MRI  
MRI 1.5T (Sonata, Siemens, Germany)

가

가  
: 5 MR

MR Hirayama 7

C4 T1

C5 - 6

가 가

: Hirayama MRI  
가

(Hirayama 가 Hirayama  
) 7, 8 1 7 (neutral position)  
(1 - 3). (neck flexion)

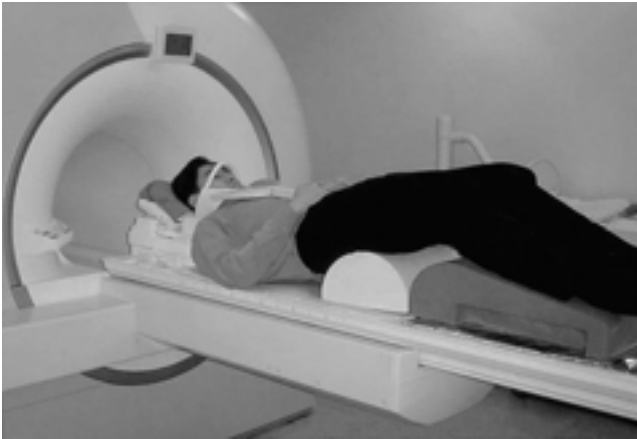
amyotrophy) (1, 4 - 6). 15 - 25 (oblique  
가  
(1, 4, 7).

(7). 2001 1 2002 6 Hirayama  
7 (17 - 43 , 23.7 ) 18.2  
(13 - 18 )  
가 3 . 7  
가 가 300 가  
(5, 8 - 10).

<sup>1</sup>  
<sup>2</sup>  
<sup>3</sup>

가 (fasciculation) 가 (Fig. 4). 가 (tremor) 1 가 (Table 1). MRI 1.5 T (Sonanta, Siemens, Germany) Hirayama (3, 6, 11, 12). (Fig. 1). T2 (TR/TE 5160/122), T1 (TR/TE 410/11), T2 (TR/TE 4000/131) T1 (700/11) T2, (Fig. 2). (fascic- ulation) 가 가 (7). 가 (cold paresis) (6, 14). 1959 Hirayama 가 12 1963 20 , 1972 38

Hirayama 5 MR Hirayama 7 MR (Fig. 2) C4 T1 C5 - 6 가 가 (Fig. 2, 3). 5 2

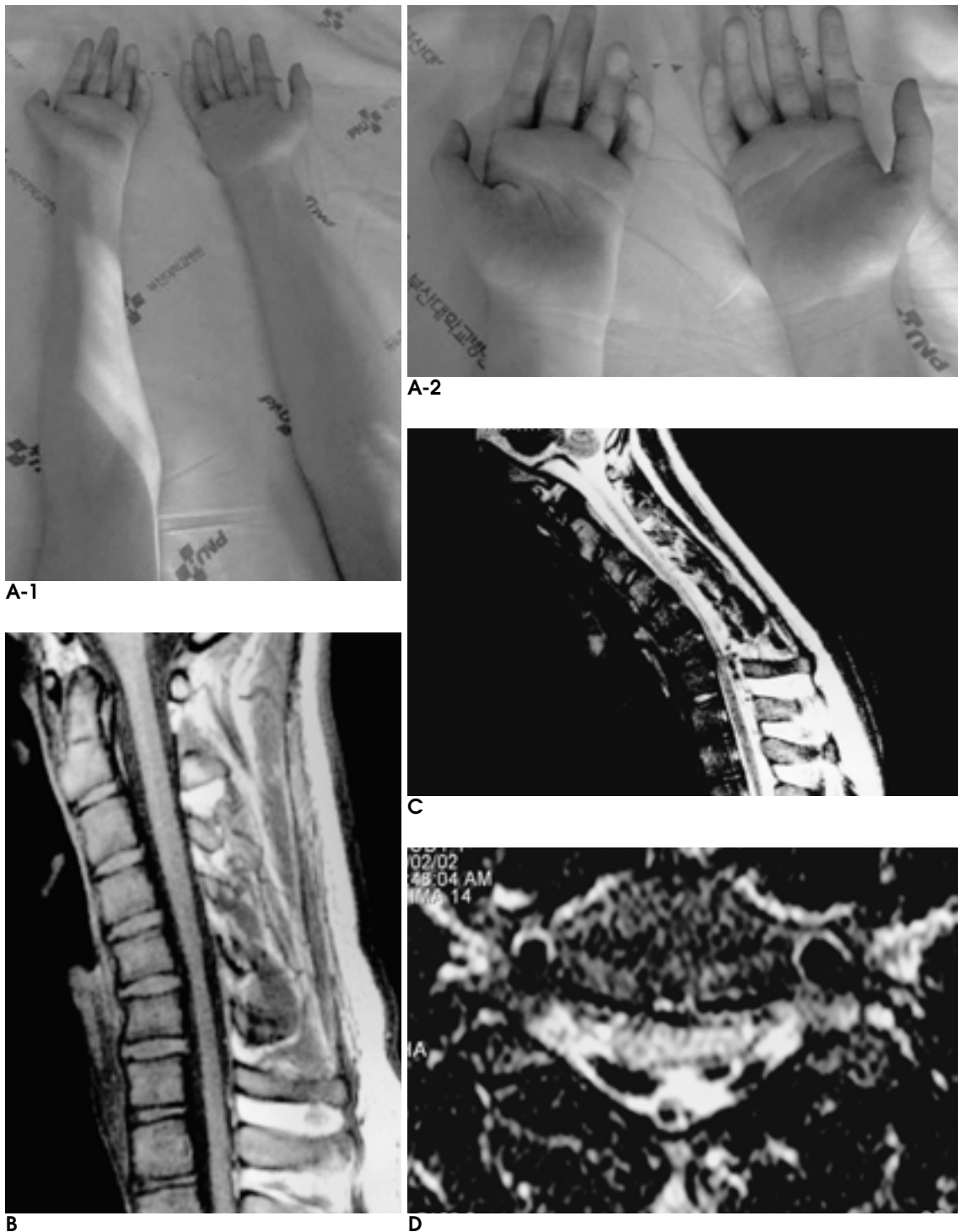


**Fig. 1.** Positioning on the MR table to obtain full flexion of the neck. Maximal elevation of the head against the trunk is possible when the trunk is tilted down rostrally using a pelvic wedge.

**Table 1.** Clinical Data on Seven Patients with Hirayama's Disease

Patient no.	Age(years)	Onset age	Approx. * period for stabilization (yr)	Involved side	Fasc <sup>†</sup>	Tremor
1	19	16	3	L	+	-
2	43	13	2	L > R	+	+
3	18	17	1	R > L	+	-
4	21	15	2	R > L	+	-
5	17	16	1	L	-	-
6	25	17	3	L	-	-
7	23	18	2	L	+	-

\* Approximately, <sup>†</sup> Fasciculation



**Fig. 2.** A 19-year-old man presented with progressive weakness and atrophy in left forearm and hand since 16-years-old.  
**A.** The muscular atrophy of the hand and forearm with relative sparing of the brachioradialis muscle leads to the characteristic clinical picture of a so-called oblique amyotrophy.  
**B.** Nonflexion sagittal T1-weighted MR image (TR/TE 410/11) shows cord atrophy at the C5-6 vertebral level.  
**C, D.** Sagittal T2-weighted MR image (TR/TE 4000/131) (**C**) and axial T2-weighted image (TR/TE 5160/122) (**D**) at maximum flexion of the cervical spine show anterior shifting of the posterior wall of the dural canal below C-3, which causes marked flattening of the lower cervical cord. An epidural mass posterior to the shifting dura matter is noted, with small flow void signals inside it.

(5).

1997 Robberecht

375

94%

Hirayama

가

가 15 - 25

10.7%

, 37%

(4).

(13, 14).

Robberecht

가

3

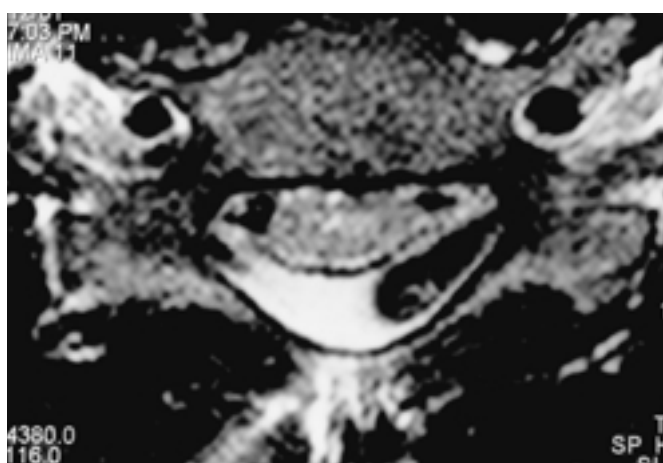
Hirayama



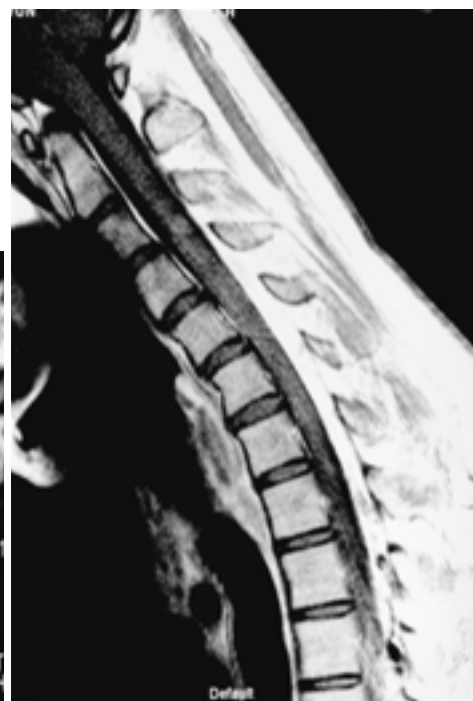
A



B



C



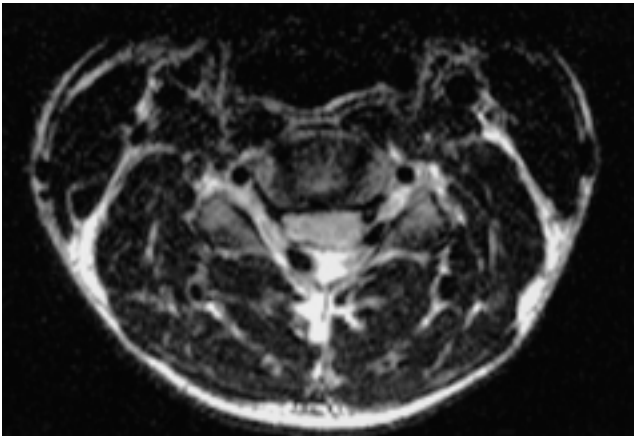
D

**Fig. 3.** A 23-year-old man with slowly progressive weakness and atrophy of the left forearm and hand.

**A.** Nonflexion sagittal T1-weighted MR image (TR/TE 410/11) did not show any abnormalities.

**B, C.** Flexion MR studies. Sagittal T2-weighted MR image (TR/TE 4000/131) (**B**) and axial T2-weighted MR image (TR/TE 5160/122) (**C**) show anterior shifting of the posterior wall of the dural canal below C-3, which causes marked flattening of the lower cervical cord. An epidural mass posterior to the shifting dura matter is noted, with small flow void signals inside it.

**D.** Contrast-enhanced flexion sagittal T1-weighted MR image (TR/TE 416/11) shows strong enhancement of the epidural mass.



**Fig. 4.** A 21-year-old male patient complained with bilateral muscular atrophy of the hand and forearm predominantly on the right side. On neck flexed axial T2-weighted MR image, right-sided spinal cord atrophy is noted. This finding presents that the cord flattening is asymmetrical, with the more flattened side corresponding to the more atrophied limb.

(6, 14).  
(dural canal)  
(dural canal) 가  
,  
(1, 6).  
가  
가  
(1, 4, 5, 7). Hirayama  
가 가  
Hirayama  
(1, 4, 7).  
Hirayama  
가가 ,  
가  
(1). Hirayama  
(4).  
(5).  
5  
가  
가  
(7). 가 (6, 17). MR  
가  
MR  
(5). 가  
Hirayama  
Willeit  
가  
3 Hirayama  
Hirayama  
(9).  
(4). 가 1982  
C5 - T1, C - 7,8  
(5). Hirayama  
(cervical color)  
Hirayama  
(5).  
Hirayama  
(6).  
Hirayama 26  
(7). Chen  
MR  
MRI  
(6, 17).  
(6).  
Hirayama  
(7). Chen  
(6).  
Hirayama 26  
(7). Chen  
(6).  
Hirayama  
(cervical color)  
Hirayama  
(5).  
Hirayama

MR  
가  
가  
MRI  
Hirayama  
가  
MRI  
Hirayama  
7 MRI,  
가  
MRI  
Hirayama  
가

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## MRI Findings of Nonprogressive Juvenile Spinal Muscular Atrophy of the Distal Upper Limbs (Hirayama's Disease)<sup>1</sup>

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**Purpose:** The aim of this study was to describe the dynamic changes of the cervical dural sac and the spinal cord during neck flexion in patients suffering from Hirayama's disease and to present the usefulness of flexion MR study for the diagnosis.

**Materials and Methods:** Seven consecutive male patients (age ranging 17 - 43 years, mean age 23.7 years) with the clinical diagnosis of Hirayama's disease and 5 healthy subjects (aged 25 - 32 years) for controls had done cervical MRI from January 2001 through June 2002. Cervical MRI was done in neutral and neck flexed positions using 1.5 T system (Sonata, Siemens, Germany) and obtained images were reviewed by two radiologists. We compared the cervical MRI findings of 7 patients with those of 5 healthy controls regarding neck flexion induced changes in the lower cervical segments.

**Results:** Neutral positioned cervical sagittal MR images revealed subtle or mild cord atrophy in only 2 patients. On maximal neck flexion, AP diameter of the crescent posterior epidural space was increased and also cord flattening with anterior shifting of posterior wall of the lower cervical dural canal was noted in all 7 patients. In all 7 cases, the level and side of spinal cord changes corresponded to the clinical phenotype. All control subjects showed neither cord flattening nor widening of posterior epidural space on neck flexion.

**Conclusion:** In patients with the clinical diagnosis of Hirayama's disease, MRI scans obtained on maximal neck flexion showed characteristically dynamic flattening of lower cervical cord and widening of posterior epidural space. Therefore, a flexion MR study is needed to prove the diagnosis.

**Index words :** Muscular atrophy, spinal  
Muscular atrophy  
Spinal muscular atrophies of childhood

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