Risk Factors for Surgical Results of Hirayama Disease: A Retrospective Analysis of a Large Cohort

Jian Song, Hong-Li Wang, Chao-Jun Zheng, Jian-Yuan Jiang

OBJECTIVE: **To explore risk factors affecting surgical results of Hirayama disease.**

METHODS: **A retrospective analysis of 210 patients was performed to identify risk factors affecting surgical results of Hirayama disease by using univariate and multivariate analyses. A receiver operating characteristic curve and area under the curve were applied to evaluate the significant results of the multivariate analysis and the optimal reference value.**

RESULTS: **The mean follow-up period was 27.3 months (range, 14–45 months), and 194 patients with clinical and radiographic data completed the final follow-up. Multivariate analysis identified age of patients (cutoff value 22.5 years), duration of the disease (cutoff value 33 months), physiologic reflex, and pathologic reflex as independent risk factors for surgical results of Hirayama disease. The receiver operating characteristic curve analysis and area under the curve showed that good reference value was obtained for the risk factors.**

CONCLUSIONS: **Age of patient, duration of the disease, physiologic reflex, and pathologic reflex are the main risk factors affecting surgical results of Hirayama disease. Receiver operating characteristic analysis shows that good reference value was obtained for the risk factors.**

INTRODUCTION

Hirayama disease (HD), first reported in 1959 by Hirayama, is a slowly progressing benign myelopathy characterized by juvenile muscular atrophy of the distal upper extremity, with accompanying symptoms of oblique atrophy, cold paralysis, fasciculation, and tremor.[1] The disease is frequently observed among teenage boys and young men in their 20s of Asian origin who are still growing, and the body parts most often affected parts are unilateral upper extremities.[2,3] The pathologic and physiologic mechanisms are unknown. Current studies show that anterior shifting of the dural sac may be the main cause, which occurs in the case of neck flexion leading to ischemic change in the anterior part of the spinal cord.[4,5] No family history has been recorded for HD, although some research suggests that genes such as *KIAA1377* and *C5orf42* might be relevant.[6]

Since neck flexion has been considered as a possible cause of HD, several therapies have proved useful, shortening the progression period and improving patients' strength. The most frequently used conservative therapy is a cervical collar. Surgical methods, including anterior decompression and fusion and posterior decompression with dural sac augmentation, with and without fusion, have been applied in cases in which the disease progresses despite conservative treatment.[7-9] Many factors have been found to be associated with the severity of HD, including gross hand function score, motion of the cervical vertebrae, and degree of muscular atrophy[10-12]; however, factors affecting the operative prognosis still are unknown. The aim of the present study was to predict the risk factors affecting surgical results of HD to improve the operative outcome.

Key words
- Extremity involved
- Hirayama disease
- Pathologic reflex
- Physiologic reflex
- Risk factors

Abbreviations and Acronyms
AUC: Area under the curve
CI: Confidence interval
HD: Hirayama disease
MRC: Medical Research Council
ROC: Receiver operating characteristic

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MATERIALS AND METHODS

Patients and Diagnostic Criteria

The study protocol was approved by the ethics committee of Huashan Hospital, Fudan University. Informed consent from the enrolled patients had been acquired previously. We retrospectively analyzed 210 patients with HD from October 2007 to December 2016 initially; all the patients had undergone surgery (anterior decompression and fusion) at Huashan Hospital, Fudan University. The diagnostic criteria of HD were as follows: 1) unilateral or bilateral muscular atrophy and weakness of the distal upper extremity with asymmetric signs and symptoms; 2) insidious onset in the teens or early 20s; 3) absence of substantial sensory and reflex abnormalities of pyramidal tracts, lower limbs, or cranial nerves or sphincteric or cerebellar deficits; 4) magnetic resonance imaging acquired in neutral position showing focal areas of cord atrophy in the lower cervical spine and cervicodorsal junction as asymmetric flattening of cord with areas of gliosis appearing hyperintense on T2-weighted images or magnetic resonance imaging acquired in flexion position revealing anterior displacement of the detached posterior dura from the underlying lamina compressing the thecal sac, widened posterior epidural space seen as a crescentic area, or epidural mass of signal alteration appearing with flow voids within[13,14]; and 5) electromyography evidence of chronic denervation at lower cervical spine (**Figures 1–5**). Exclusion criteria of discogenic low back pain included 1) unclear diagnosis of HD, 2) improvement of symptoms after conservative therapy of wearing cervical collar for 3 months, 3) infection of incisional wound of cervical spine, and 4) the presence of other disease.

Preoperative Evaluation and Operative Treatment

Preoperative evaluation of HD included the following points:

1. Age of the patient: the current age of enrolled patients

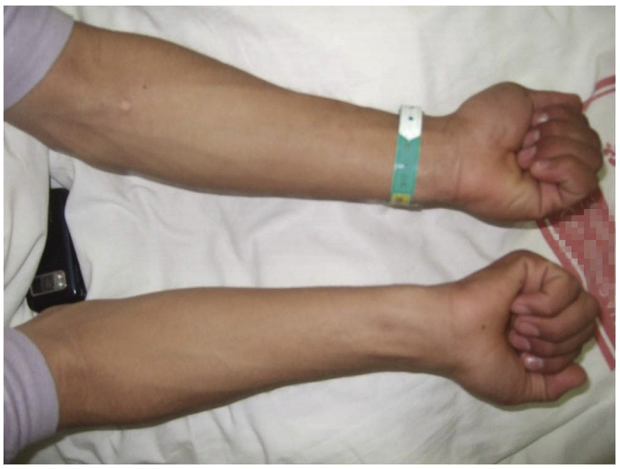

Figure 1. A 24-year-old man had Hirayama disease for >2 years before presentation. Atrophy of his right hand is noticeable compared with his right hand.

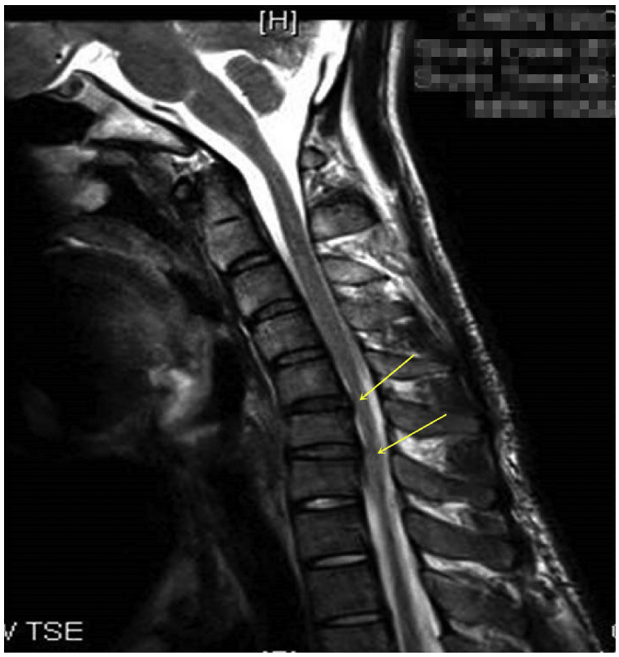

Figure 2. Nonflexion sagittal T2-weighted magnetic resonance imaging shows atrophy of the spinal cord at C5-C7 level (*arrows*).

2. Curvature of the cervical spine: cervical curvature was considered normal when vertebral bodies C3-C6 were anterior to a line drawn from C2 to C7; an abnormal, straight, or kyphotic

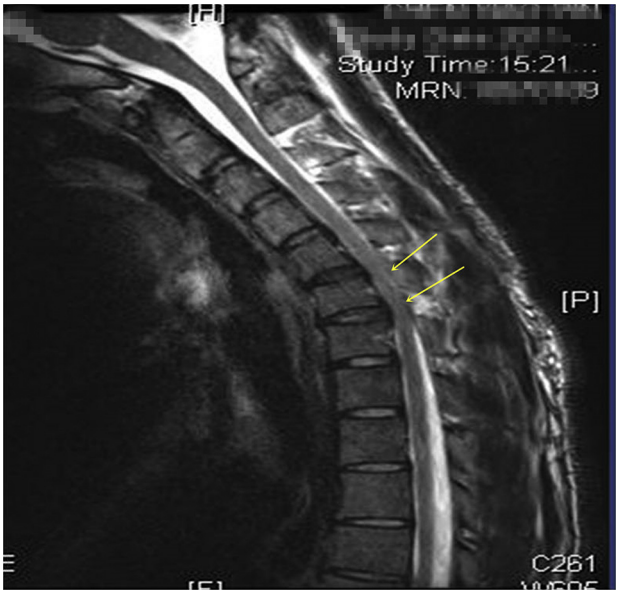

Figure 3. Flexion sagittal T2-weighted magnetic resonance imaging shows anterior shifting of the posterior wall of the dura mater (*arrows*).