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ORIGINAL ARTICLE

# Hirayama disease: analysis of cases in Russia

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## Abstract

The fourteen cases of Hirayama disease (HD) are presented in this article. HD is seldom disease characterized by juvenile muscular atrophy of upper extremities and benign course. All cases were diagnosed in the Research Center of Neurology (Moscow, Russia) during the year 2015–2017. Such methods as MRI (magnetic resonance imaging), EMG (electromyography), and NCS (nerve conduction studies) have been used to confirm diagnosis of HD. Transcranial magnetic stimulation was used to exclude upper motor neuron involvement in two cases. The original scale of neurological disturbances in HD has been proposed by authors to reveal correlations of HD severity with age of patients and duration of disease.

Most of patients with HD are young males with common clinical signs. Detected MRI and EMG data were also comparable with previous publications. Independence of HD severity from age and duration of the disease may be the result of individual physical characteristics of dura mater and other structures of the cervical vertebra. In some our cases, amyotrophic lateral sclerosis and other neurological disorders were misdiagnosed before. In view of different prognosis in these pathologies and possible correction of HD, early diagnosis is very important.

**Keywords** Hirayama disease · Monomyelic amyotrophy · Cervical myelopathy · Motor focal amyotrophy · Motor neuron disease · Arm flaccid paresis

## Introduction

Juvenile muscular atrophy of unilateral upper extremity with benign course was first described by Keizo Hirayama in 1959 [1]. Decreased anteroposterior diameter of the lower cervical spinal cord for these patients was demonstrated by computed tomography with intrathecal contrast, but the underlying mechanisms of this condition were unclear. In 1987, the results of the first spinal cord Hirayama disease (HD) patient biopsy were published. Authors reported the decreased number of both large and small nerve cells and degenerative changes, and suggested circulatory insufficiency in the lower cervical cord as the leading cause [2]. In-vivo diagnosis of HD could be verified only after a long dynamic observation. In 1987, MRI in cervical flexion position was performed, and anterior shift of posterior dura mater was discovered [3, 4]. Nowadays, a nerve conduction study, needle electromyography, and cervical spine MRI in neutral and flexion positions permit HD confirmation in clinically supposed cases during the first specialist visit. Clinical characteristics of Hirayama disorder include a greater prevalence in Far East and Asia regions (Japan, China, India) with a number of published cases in Europe, Australia, and America; prevalence of male gender; age of 15–25 years; and period of 1–5 years of symptom progression with further stabilization of neurological deficit [5–12]. Predominantly, the pathological process involves one upper extremity; however, involvement of both upper extremities and pyramidal signs can also occur [13, 14].

There are a lot of case reports of HD published in different countries and some case series reviews. This article presents the analysis of 14 cases of HD in Russia that were examined in The Research Center of Neurology from 2015 till 2017.

## Materials and methods

Fourteen patients (12 males, 2 females; age range 18–30 years) with clinical suspicion of Hirayama disease underwent clinical, electrophysiological, and MRI diagnostic study between 2015 and 2017 at Research Center of Neurology, Moscow,

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Russia. The study was approved by the local ethical committee. An informed consent was obtained from all patients prior to study-related procedures.

Diagnostic inclusion criteria for HD were the following:

- weakness and atrophy of the upper limbs, predominantly forearms and hands (upper flaccid paraparesis);
- insidious onset among the teens or the patients in their early 20s;
- absence of constant sensory deficit, signs of pyramidal pathology at cervical level, signs of lower motoneuron involvement at lumbar level, cranial nerves deficits, sphincters malfunction, coordination disturbances;
- needle electromyography evidence of the chronic denervation at C7-T1 myotomes level and fasciculation potentials.
- standard nerve conduction study evidence of motor axonopathy in upper limbs only (decrease in the compound muscle action potentials (CMAPs), a preserved nerve conduction velocity, and duration of the motor response at the early stage of the disease with signs of more severe damage with disease progression);
- exclusion of other diseases.

Instrumental examination included the following:

1) Nerve conduction study (NCS).
2) Needle electromyography (EMG).
3) Transcranial magnetic stimulation (TMS).
4) MRI imaging.

The MRI imaging was performed on a 3 Tesla Siemens Verio system (Erlangen, Germany). The neutral position MRI protocol included a sagittal T1W sequence (turbo-spin echo, repetition time/echo time in ms of 700/10, 3-mm thickness), sagittal T2tirm sequence (turbo inversion recovery magnitude, TR/TE 4000/57 ms, 3-mm thickness), sagittal and axial T2W sequence (turbo spin echo, TR/TE 3000–4000/101–123 ms, 3-mm thickness). The flexion imaging protocol consisted of sagittal and axial T2W sequence (turbo spin echo, TR/TE 3000–4000/101–123 ms, 3-mm thickness), with maximum flexion obtained using a positioning sponge. Cervical flexion angle was not less than 35° in all cases as it is recommended [15]. MRI assessment included the presence or absence of the following signs:

In neutral position:

1. abnormal cervical lordosis;
2. loss of attachment of the posterior dural sac and subjacent lamina;
3. localized spinal cord atrophy and flattening (either symmetric or asymmetric);
4. T2W intramedullary hyperintensity at the C5-T1 levels;
5. evidence of other causes of cord compression.

In flexion position:

1. forward migration of the wall of the dura mater with an enlarged posterior epidural space;
2. crescentic epidural mass appearance showing curvilinear flow voids in the posterior epidural space.

Cervical lordosis was considered normal when the vertebral bodies C3 through C6 were anterior to the line drawn from C2 through C7. An abnormal, a curvature was defined when a part or all of the vertebral bodies from C3 to C6 met or crossed the line from C2 to C7 (Fig. 1).

Localized spinal cord flattening was defined as decrease in cord size in comparison to the normal cord above and below the affected level on sagittal and/or axial images.

To appreciate severity of HD, we used our new self-developed clinical and physiological scale (Table 1).

This kind of scaling is close to suspected pathophysiology of the disease and represents the spreading of the lesion in spinal cord from gray matter of one side to anterior horn of the other side and then to white matter. White matter involvement results in pyramidal syndrome due to damage of corticospinal tracts at cervical level.

The Kruskal–Wallis test and Spearman correlation analysis were used as the methods of statistics.

## Results

### Clinical findings

Fourteen patients fulfilled the study inclusion criteria. All the patients were Caucasians, 2 females and 12 males. The group could be characterized by the following median and quarters M (Q1, Q3): age – 20.5 years (19–21.8), age of first disorder sign – 17 years (16–17.8), period from onset of disease to HD diagnosis– 4 years (2.3–6.3). Ten from 14 patients had bilateral upper limb involvement and 4 patients had pyramidal signs in lower limb (most likely due to the cervical myelopathy). More detailed epidemiological, clinical data, and anamnesis are summarized in Table 2.

The presented cases have a lot of traits that are common among patients with HD: most of the patients were young males (mean age of disease onset is 16.3 years) and had specific neck flexion activity in anamnesis (exercise with a high weight for trapezius muscle). One of HD patients had somatotropin deficiency. There are no details worth discussing in other cases.

All patients complained about weakness and atrophy of the upper limbs, predominantly forearms and hands. Eight patients had wrist muscle cramps caused by a cold temperature.

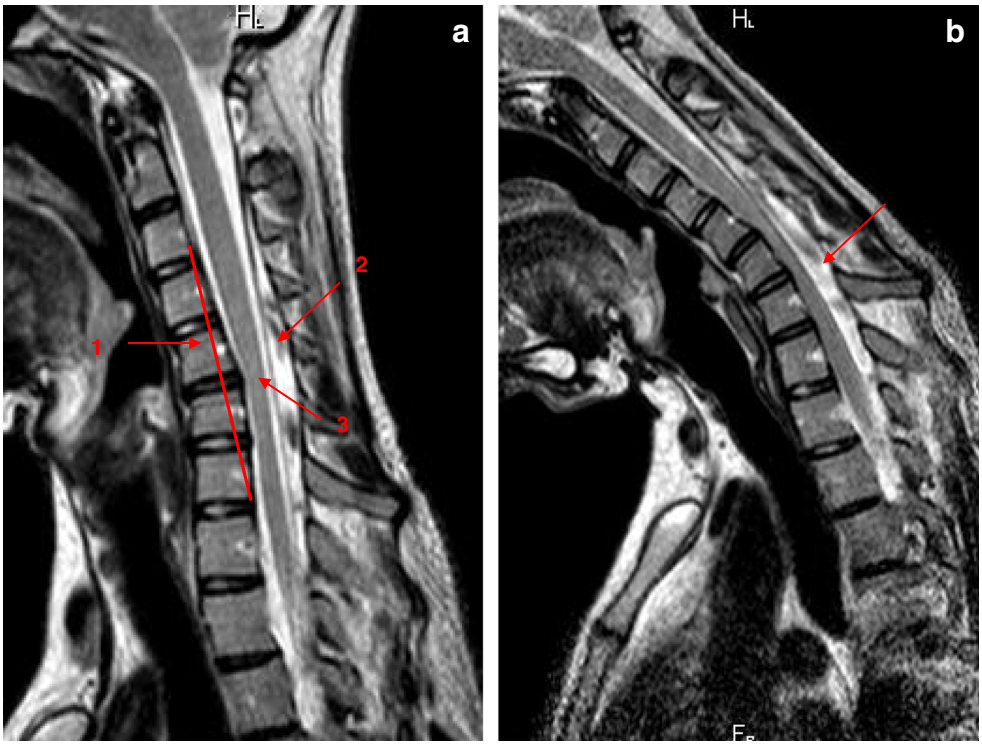


**Fig. 1** Neutral (**a**) and flexion (**b**) position T2W sagittal MRI in HD. (**a**1), abnormal cervical lordosis; (**a**2), prominent dorsal epidural compartment due to anterior displacement of the dorsal dura compressing the thecal sac, at the C5-C7 levels; (**a**3), flattening spinal cord at the same levels. **b** More prominent anterior dura displacement leading to C5-C7 spinal cord compression

No sensory deficits and weakness of lower limbs were reported by any of the patients. Signs of pyramidal tract pathology were detected in four patients (hyperreflexia in legs in four cases and unilateral positive Babinski in one case). The clinical involvement at onset was unilateral in five patients and bilateral asymmetric in nine patients (Fig. 2). These clinical signs were prevalent on the left side in ten patients and on the right - in four patients. The average time from onset of disease to HD diagnosis was 4.4 years. It is also worth noting that in five cases, ALS was preliminary diagnosed, which proves the similarity of ALS at an early stage and HD.

We analyzed correlation of HD severity according to Original Severity HD scale (Table 1) with age of patients, with age of disorders' onset, and period from the onset to the right diagnosis. Significant correlations and differences were not detected. Mean age, age of first disorder's sign, and period from the onset of disease to HD diagnosis corresponding to each severity group are presented in Fig. 3.

## Electrophysiological findings

All patients underwent NCS and EMG. Median and ulnar nerves were stimulated at the wrist and elbow. The initial negative-peak amplitudes were used for all compound muscle action potentials. CMAPs were recorded from the abductor pollicis brevis and abductor digiti minimi muscles. Motor NCS typically show low ulnar CMAP amplitudes, that are more severely reduced than the median CMAP in all patients. Sensory conduction and electrophysiologic features in lower limbs in all patients were normal. In the studied series, chronic denervation at EMG of C6, C7, C8, and T1 myotomes bilaterally was evident for all patients (prevalent on the left side for

**Table 1** Severity HD scale based on clinical and physiological traits

| Grade of HD severity | Clinical and electrophysiological signs of severity |
|---|---|
| 1 | Weakness in one arm without other arm involvement* |
| 2 | Weakness in one arm with signs of other arm involvement |
| 3 | Weakness in both arms without signs of leg involvement |
| 4 | Weakness in both arms with signs of one leg involvement |
| 5 | Weakness in both arms with signs of both legs involvement |

*Signs of arm involvement—typical EMG-signs, hyporeflexia, hypotrophies, without disability (without change of strength)

Signs of leg involvement—pyramidal signs (hyperreflexia, positive Babinski, or other pathological reflexes) without disability (without change of strength)

**Table 2** Epidemiological, clinical data, and anamnesis of 14 HD cases

| Case<br>Sex | 1<br>Male | 2<br>Male | 3<br>Male | 4<br>Male | 5<br>Male | 6<br>Male | 7<br>Female | 8<br>Male | 9<br>Male | 10<br>Male | 11<br>Female | 12<br>Male | 13<br>Male | 14<br>Male |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| Age | 21 | 19 | 21 | 30 | 20 | 18 | 21 | 27 | 19 | 19 | 25 | 19 | 22 | 18 |
| Age of first disorder sign | 17 | 15 | 18 | 23 | 18 | 12 | 17 | 16 | 17 | 16 | 17 | 18 | 15 | 16 |
| Period from onset of disease to HD diagnosis, years | 4 | 4 | 3 | 7 | 2 | 4 | 4 | 11 | 2 | 3 | 8 | 1 | 7 | 1,5 |
| Period from onset of disease to stabilization stage (in case of stabilization), years | 1 | NS* | NS | 2 | NS | 4 | NS | 5 | NS | NS | NS | NS | NS | NS |
| Previous dignosis | ALS | Spinal tu-mor | Cubital Tunnel Syndrome | Parsonage-Turner Syndrome | No | No | No | Myelopathy | Myelopathy | CIDP | ALS | SMA | Cubital Tunnel Syndrome | ALS |
| Severity of the disease at the moment of final diagnosis** | 5 | 5 | 3 | 2 | 3 | 2 | 5 | 5 | 3 | 3 | 2 | 2 | 2 | 2 |
| Physical load of neck muscles in anamnesis | Yes | Yes | Yes | Yes | Yes | Yes | Yes | No | Yes | Yes | No | No | No | Yes |
| EMG-signs (fibrillation potentials and moderate active denervation only in the muscles innervated by the cervical level) | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes |
| NCS-signs (ulnar CMAP amplitudes more severely reduced than the median CMAP) | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes |
| Cervical MRI findings: | | | | | | | | | | | | | | |
| - Normal cervical lordosis loss | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes |
| - Dural sac displacement during flexion | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes |
| - Spinal cord atrophy at C5-C7 level | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes | Yes |
| - C5-C7 myelopathy signs | No | Yes | Yes | Yes | Yes | No | No | Yes | Yes | Yes | Yes | No | No | No |

*NS - no stabilization

**According to Original Severity HD scale (Table 1)

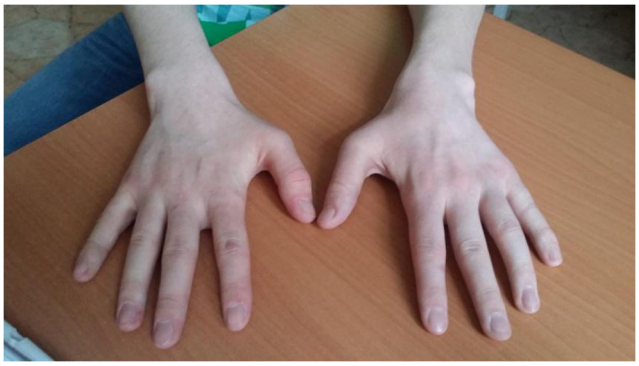

**Fig. 2** Both hands changes in HD case No. 1

ten patients and on the right - for four). Proximal muscles (biceps and deltoid) controlled by C5 and C6 nerves, generally appeared normal in all patients. It should be, however, noted that EMG exams revealed both distal and proximal muscles bilateral chronic denervation in two patients.

### Transcranial magnetic stimulation

TMS was performed in two patients with HD to exclude subclinical upper motor neuron dysfunction. In these cases, neurological and EMG exams revealed bilateral weakness and hypotrophy in both distal and proximal upper limb muscles. This caused difficulties in differential diagnosis with motor neuron disease, in particular, with flail arm syndrome. In both cases, all TMS parameters (resting motor threshold, motor evoked potentials latency and amplitude, cortical silent period duration, and central motor conduction time) were normal.

### Cervical MRI findings

All patients had loss of the normal cervical lordosis and anterior displacement of the dural sac on flexion MRI (Fig. 1). Also all patients showed localized cord atrophy at C5-C7 level. Intramedullary bilateral anterior horns T2W-hyperintensity lesions were revealed at the same level asymmetrical in six patients (more prominent on left side in five patients and on the right - in two patients) and symmetrical in two patients (Fig. 4) – the so-called snake eyes sign. The sign itself is not specific to Hirayama disease and is supposed to be the outcome of anterior spinal cord circulation insufficiency, that can be present in flail arm syndrome, spinal stroke, and, etc. [16–18].

## Discussion

Literature analysis of the recently published studies compared to our dates are showed in Table 3.

As in previous reports, most of the patients with HD are young males. Most of them had anamnesis about prolonged (from months to years) period with repeated neck flexion activity. Authors noted about different kinds of activities (arm sport exercises, writing at a desk, and others) [4, 5, 29]. Ten of our patients reported about intense exercises with a high weight for trapezius muscle. Clinical course and HD signs of all our patients (including with sport anamnesis) are comparable to the described in previous articles, but legs involvement (pyramidal signs) are largely exceeding. We must to emphasize that in all described cases, neutral MRI of the cervical spine was performed before consultation in The Research Center of Neurology but signs of HD in neutral position were not noted. According to V.T. Lehman and coauthors, sensitivities of neutral and flexion MRI are about the same and investigation in flexion position just augments diagnostic confidence [11].

According to Table 3, there are no obvious difference between onset-diagnosis periods in Russia and other countries. Independence of severity according to Original Severity HD scale from age and duration of HD is very important. These data probably pointed to individual physical characteristics of dura mater and other structures of the cervical spine. Also, small number of cases limits statistical investigation and more dates are required.

In some cases, it was difficult to distinguish HD from early stages of upper-limb onset of ALS. In some HD patients (18% in the study by A. Nalini and coauthors [14] and 2/14 in our study), the subtle signs of upper motor neuron involvement may occur in legs, possibly due to the damage of corticospinal tract at the cervical cord level. The key findings that may differentiate HD from ALS include an earlier age of disease onset, plateau after an initially progressive course, absence of EMG-signs of low motoneuron damage at other levels (bulbar, thoracic, and lumbar), and imaging findings at the cervical

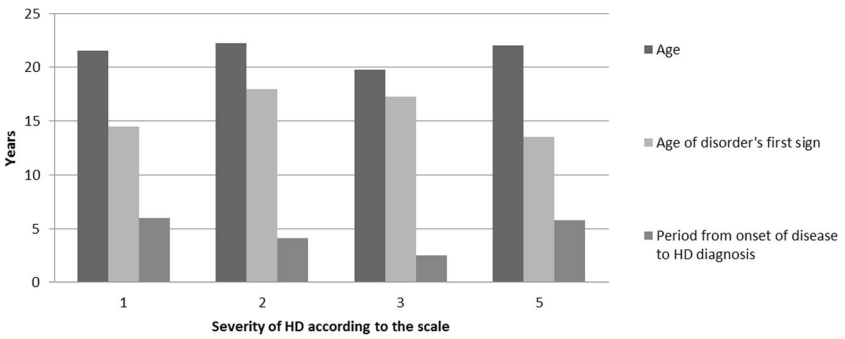


**Fig. 3** Age of patient at the moment of diagnosis, age of first disorder sign, and period from onset of disease to HD diagnosis in patients with different grades of severity

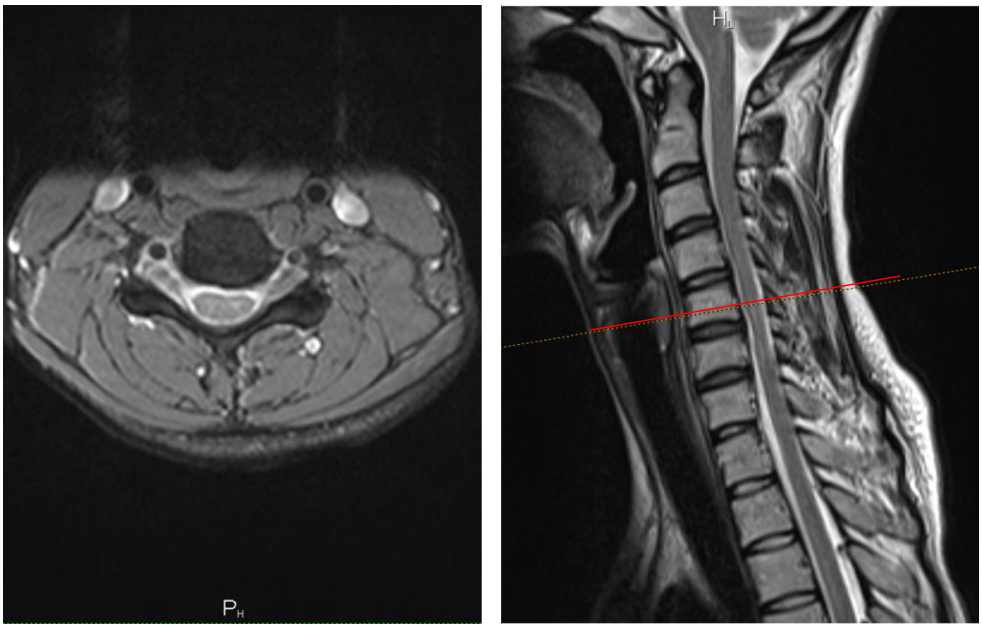

**Fig. 4** Cervical anterior horns snake eyes appearances on axial T2-W at C5-C6 level in HD

level, especially in flexion position. In difficult cases, TMS can be used to detect upper motor neuron involvement, but this method is not included in current ALS diagnostic criteria [30, 31]. Furthermore, to date, there are only a few studies that applied TMS in HD [29, 32]. Although, these studies confirm the intactness of motor cortex and supraspinal pathways of the

**Table 3** Published articles about series of HD cases for the last 5 years

| Investigation | Nationality | Period of investigation | Number of patients | Sex (m/f) | Age, years Mean ± δ (min – max) | Age of disorder's first sign | Period from onset of disease to HD diagnosis, months/ years | Period from onset of disease to stabilization stage, years |
|---|---|---|---|---|---|---|---|---|
| Zheng C et al., 2017 [19] | China | March 2011–December 2015 | 33 | 32/1 | 19.2 ± 3.2 | NA* | 28.3 + 17.5 months | NA |
| Goel A. et al., 2016 [20] | India | June 2014–January 2016 | 5 | 5/0 | 20 (16–28) | NA | 1–2 years | NA |
| Singh RJ et al., 2016 [21] | India | January 2014–April 2015 | 10 | NA | 25.8 ± 3.8 | NA | 8.1 ± 5.7 years | NA |
| Liao MF et al., 2016 [22] | Taiwan | 1995–2010 | 44 | 40/4 | NA | 16.4 ± 2.5, 17.2 ± 3.3 | 4.3 ± 4.2, 5.3 ± 4.1 years | 3.4 + 2.4,3.1 + 2.3 |
| Agundez M et al., 2015 [23] | Spain | NA | 4 | 4/0 | 26.3 (19–40) | 18.5 (18–20) | 7.9 (1.5–20) years | NA |
| Ding Y et al., 2015 [24] | China | 2009–2011 | 60 | 57/3 | NA | 12–25 (17.0 ± 2.4) | NA | NA |
| Khadilkar S et al., 2015 [25] | India | February 2014–April 2015 | 26 | NA | NA | 17.9 | NA | NA |
| Shao M et al., 2015 [26] | China | NA | 64 | 63/1 | 16–26 | NA | 23.5 (3–86) months | NA |
| Foster E et al., 2014 [27] | Italy(Italian, Spanish, Jewish) | NA | 3 | 3/0 | 22–29 | 16 | 4–10 years | NA |
| Abraham A et al., 2013 [28] | Israel | NA | 15 | 15/0 | 22 ± 3.8 | 18 ± 1.9 | NA | NA |
| Our investigation | Russian | 2013–2017 | 14 | 12/2 | 21.4 (18–30) | 16 (4–23) | 4.4 (1–11) years | 1–5 |

*NA - not available

corticospinal tract in HD patients. Future studies are needed to confirm these findings and establish the diagnostic value of TMS in differential diagnosis of ALS and HD.

One of our HD patients used to have somatotropin deficiency in childhood and was treated with hormone therapy until reaching normal height. K. Hirayama and coauthors suggested that a disproportional growth between the vertebral column and the contents of the spinal canal, especially the dural sac, during the juvenile growth spurt is a reason of the disease [33]. That is why possible link between these two disorders in the case should be discussed. Growth rate usually increases most during the first year of treatment with somatotropin. The patient took hormone therapy since 4 to 13 years old. At 17 years old, he noted weakness of the left hand. Weakness and wasting of the hand were progressing during the year and then plateauing over 4 years (Fig. 2). We have to mark that beginning of HD was not against the background or soon after the start of hormone therapy. Also, we did not find any descriptions of HD in somatotropine deficient patients. So, we conclude that more likely, these two conditions of the patient are concomitant diseases.

## Conclusion

HD is a very rare disease in Russia and has common signs described by K. Hirayama. Years ago, the diagnosis could be made only after long period of clinical supervision. These days' instrumental methods help to determine HD during the first patient's request of medical assistance regarding upper flaccid paresis. NCS and EMG allowed us to prove the only cervical level segments involvement. MRI confirms the change of cervical thecal sac placement. The gradual onset and progressive course of the disease and isolated motor neuron involvement may mimic early stage of the amyotrophic lateral sclerosis and other neurological disorders. HD early diagnosis is very important in terms of recovery prognosis [34]. Neurologists, MRI-specialists, and clinical neurophysiologists should be more informed about HD, considering the importance of early-stage diagnosis.

### Compliance with ethical standards

**Conflict of interest** The authors declare that they have no conflicts of interest.

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