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PHYSICAL THERAPY FOR HIRAYAMA DISEASE: IS THE EVIDENCE BENDING TOWARDS OR AWAY FROM FLEXION?

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ABSTRACT Hirayama disease (HD) or juvenile muscular atrophy of distal upper extremity is a cervical myelopathy characterized by progressive muscular weakness and atrophy of distal upper limbs, secondary to forward displacement of the posterior cervical dural sac during neck flexion, causing compression of the cervical cord, resulting in atrophic and ischemic changes in the anterior horn.The objective of this study was to perform a scoping review of existing evidence through studies from Pubmed to answer two questions: whether HD has a mechanical pathogenetic component? And if so, what could be the role of Physical therapy in such patients?Evidence demonstrated that excessive flexion range of motion was present in patients with HD, and that flexion was equivocally associated with abnormal spinal cord compression and canal dynamics, cervical spine instability (CSI) as an entity should thus encompass HD at one end of its clinical spectrum. Two major issues arose from this review: one, literature on physical therapy examination and treatment for HD is albeit absent if scarcely available, and two, proven methods of physical therapy for clinical cervical spine instability had not been attempted for HD despite their low risk for complications.

KEYWORDS : Juvenile muscular atrophy, Hirayama disease, clinico-radiological correlation, neuroorthopaedic rehabilitation.

INTRODUCTION:

Hirayama disease (HD) or juvenile muscular atrophy of distal upper extremity is a cervical myelopathy characterized by progressive muscular weakness and atrophy of distal upper limbs, secondary to forward displacement of the posterior cervical dural sac during neck flexion, causing compression of the cervical cord, resulting in atrophic and ischemic changes in the anterior horn.[1]

The objective of this study was to perform a scoping review of existing evidence through studies from Pubmed to answer two questions: whether HD has a mechanical pathogenetic component? And if so, what could be the role of Physical therapy in such patients?

NECK FLEXION AND PLAIN RADIOGRAPHIC FINDINGS:

Towards:

Xu et al (2011) compared cervical range of flexed motion in 31 HD patients and 40 healthy controls using conventional flexion radiographs of cervical spine and flexion cervical MRI. The authors demonstrated greater segmental and overall range of cervical flexed motion of patients, which correlated well with flexion MRI. The authors concluded that HD patients had an increased flexed motion range of cervical spine which had contributed to the pathophysiological change and would thus determine its treatment.

NECK FLEXION AND MAGNETIC RESONANCE IMAGING (MRI) FINDINGS:

Towards:

Yin et al (2011) studied fully flexed cervical MRI findings in 13 male HD patients and 12 normal males, and found significant differences between patients and controls in localized lower cervical-cord atrophy and cord flattening, with patients having a crescent-shaped high-intensity mass on T2WI MRI and a low-intensity mass on T1WI MRI in the posterior epidural space, and characteristic findings of HD on fully flexed MRI.

Hou et al (2012) performed cervical MRI in neutral and different flexion positions (cervical flexion angle 20°, 25°, 30°, 35°, and 40°) in 45 HD patients and found that anterior shifting of the posterior wall of the cervical dural canal (ASD) through a ratio of the widest cervical epidural space with the maximum sagittal diameters (d) and cervical canal sagittal diameter (D) at the same level, and epidural flow voids (EFV) were influenced significantly by flexion angles especially more so at 35^0. The authors recommended 25^0 flexion for the least effective diagnostic flexion angle for MRI diagnosis of HD and 35^0 flexion the best one of choice.

Hassan et al (2012) performed cervical MRI in neutral position in 11 patients and flexion contrast imaging in 10 patients with HD, and found that neutral position MRI showed loss of cervical lordosis in 10/11 (91%), localized lower cervical cord atrophy in 9/11 (82%), asymmetric cord flattening in 11/11 (100%) and intramedullary hyperintensity in 2/11 (18%); while flexion study showed loss of dural attachment, anterior displacement of dorsal dura, epidural flow voids in 9/10 (90%) and enhancing epidural crescent in 10/10 (100%). Interestingly, collar therapy slowed pathogenetic progression in most patients with HD.

AWAY:

Lai et al (2011) measured the degree of forward shift of posterior dural sac during flexion in 50 healthy volunteers and 3 HD patients and found that in normal subjects, the forward shifting of the posterior cervical dural sac occurred during flexion, but without associated cord compression due to intrinsic expansion of the spinal canal volume. In HD patients, inadequacy of this compensatory mechanism lead to significant increment in ratio of anteroposterior diameter of forward displacement of posterior dural wall/anteroposterior diameter of spinal canal ("x/y"), and decrement in ratio of anteroposterior diameter of spinal cord/perpendicular transverse diameter of spinal cord ("a/b"). The authors suggested that depiction of forward shift alone would not be reliable, and it should be correlated with increased x/y and decreased a/b on cervical flexion.

NECK FLEXION AND ELECTROPHYSIOLOGICAL RESPONSES:

Towards:

Abraham et al (2013) demonstrated alterations in motor evoked potentials recording in 15 men with HD and 7 age-matched controls, with lower amplitudes, and slightly prolonged latencies in HD patients compared to control subjects. Neck flexion was associated with a significant drop in mean upper limb amplitude more in HD patients than healthy subjects. The somatosensory evoked potentials did not show such differences of neck flexion-induced changes.

AWAY:

Misra et al (2006) studied SSEPs (N9, N13, and N20), median and ulnar F-waves (minimal latency, FM ratio, persistence, and chronodispersion) and MRI findings in 8 HD patients and 7 matched controls, in neutral and neck flexion positions. The authors neither found electrophysiological differences between neutral and neck flexed positions nor those responses between patients and normals, which suggested other probable mechanisms for HD.

Ammendola et al (2008) reported three male HD patients who did not have significant alterations in Median and ulnar F waves, median, ulnar and posterior tibial SEP, and upper and lower limb MEP. Of the three patients, the flexion MRI in third patient showed that the spinal cord was normally located, was not compressed within the cervical canal and that there were no abnormalities of the dural sac, which

suggested that HD may have a different pathogenetic mechanism other than just "flexion myelopathy".

NECK FLEXION AND EPIDURAL PRESSURE: AWAY:

Patel et al (2009) reported a case of 18-year old man with electromyographic, MRI and angiographic findings of HD, on whom the authors used a microcatheter to measure the pressure in the epidural space (through changes with and without Valsalva maneuver in the inferior vena cava, vertebral veins, and epidural space at C4 and C6) in both the flexion and neutral positions. The authors did not find significant change in epidural pressure which suggested that dural venous engorgement was passive and was not the direct cause for spinal cord injury.

Now, it is very much evident that excessive flexion range of motion was present in patients with HD, and that flexion was equivocally associated with abnormal spinal cord compression and canal dynamics, cervical spine instability (CSI) as an entity should thus encompass HD at one end of its clinical spectrum (Gui et al, 1982). Physical therapists and manual therapists are in a clinical dilemma of fact or fiction about CSI (Swinkels and Oostendorp, 1996) and associated clinical instability (potential involvement of passive fibroligamentous, active musculotendinous and neural control subsystems) co-existing with mechanical instability (Swinkels et al, 1996).

CSI was perceived to clinically present with a history of major trauma; reports of the neck catching or locking or giving way; poor muscular control; signs of hypermobility on X-ray; excessively free end-feel on passive motion testing and unpredictability of symptoms (Niere et al and Torney, 2004). The above mentioned findings together with observable or palpable abnormalities of motion during movement assessment were associated with increased physical therapists' confidence in diagnosis of CSI (Cook et al, 2005).

Cook et al (2005b) also described the list of subjective descriptors and identifiers which were suggestive of clinical CSI as follows; "intolerance to prolonged static postures," "fatigue and inability to hold head up," "better with external support, including hands or collar," "frequent need for self-manipulation," "feeling of instability, shaking, or lack of control," "frequent episodes of acute attacks," and "sharp pain, possibly with sudden movements." The physical examination findings related to cervical instability that reached the highest consensus among respondents included "poor coordination/neuromuscular control, including poor recruitment and dissociation of cervical segments with movement," "abnormal joint play," "motion that is not smooth throughout range (of motion), including segmental hinging, pivoting, or fulcruming," and "aberrant movement."

Current management of mechanical CSI is mainly operative fusion (Garfin and Ahlgren, 1995) orcollar therapy, while clinical CSI has multiple therapeutic options ranging from cervical deep flexor endurance training, motor control or movement retraining, proprioceptive training, cervical extensor strengthening and postural re-education. It is also well understood that patients with mechanical instability do derive a large benefit from use of therapies for clinical instability as witnessed in physical therapy management of spondylolisthesis, hypermobility syndromes and post-traumatic ligament injuries.

We have two major issues: one, literature on physical therapy examination and treatment for HD is albeit absent if scarcely available, and two, proven methods of therapy for clinical cervical spine instability had not been attempted for HD despite their low risk for complications.

To all expert clinicians in Physical therapy, we ask:
Physical therapy for Hirayama disease: Is it useless or was it used less?

And to all expert researchers in Physical therapy, we ask:
Research unreported is research undone: Does absence of evidence mean evidence of absence?

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