## FP86

### Juvenile amyotrophy of the distal upper extremity (Hirayama disease): case series and surgical management

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Hirayama firstly report (1959) a disorder of the juvenile age (11-25 years), characterized by upper extremity, slowly progressive asymmetric amyotrophy occurring in late childhood-adolescence. This disease was named also Monomyelic Amyotrophy or Cervical Flexion Myelopathy (CFM); this definition, proposed by Kikuchi is based on the possible etiology: a compression of the cervical spinal cord occurring during neck flexion, but the exact etiology is still unknown. The value of conservative and surgical treatments has not been established: conservative prolonged collar therapy, cervical arthrodesis, cervical duroplasty – laminoplasty and muscolotendineous transfer have been proposed in the few published series Series: The clinical and MRI characteristics of 16 patients affected by CFM were examined. Age at onset ranged between 14 and 25 years, with a pick at 16. We observed a male prevalence (14/17), as already reported. Nine patients with progressive deterioration were treated surgically with expansive duraplasty and suspension in combination with laminotomy. Dynamic MRI and neurophysiology were performed before and after surgery. The remaining 7 cases were stabilized at diagnosis or mildly deteriorating and were managed conservatively.Results: Dynamic MRI taken with the neck in a neutral position showed that the spinal cord was flattened and when the neck was flexed, the dura and the spinal cord were compressed further. 3 out of 9 operated cases experienced transient worsening, one progressed further despite surgery, while the remaining stabilized at the preoperative neurological level. Venous engorgement, presumed by MRI, was documented at surgery in all the cases, as well as spinal cord tendency to herniate trough the dural opening; in 1 case arachnoid adhesion was documented at surgery.Conclusions: The early diagnosis of the rare Hirayama Disease by dynamic MRI is advisable, because clinical outcome of the progressively deteriorating cases may be improved by duraplasty-laminotomy if performed before the neurological deterioration stabilized.