Classic foramen magnum syndrome is defined by development of unilateral arm sensory and motor deficits, which progress to the ipsilateral leg, then the contralateral leg, and finally contralateral upper extremity. Long tract findings characteristic of upper motor lesions are found paradoxically in the presence of atrophy in the intrinsic muscles of the hands. Later findings include spastic quadriparesis and lower cranial nerve palsies. Slowly progressive lesions such as these allow the development of accessory muscles to replace trapezius and sternocleidomastoid function. We therefore highly recommend that the patient be undressed and the sternocleidomastoid and trapezius muscles be closely inspected for atrophy. Likewise, the tongue should be inspected at rest for atrophy and fasciculation. Close attention to sensory testing of the C-2 dermatome will help establish the diagnosis. Patients attest to initial sensory disturbances such as cold or burning dysesthesias, astereognosis, and anesthesia but often do not seek medical attention until intractable pain, motor deficits, or ataxia ensue. Terminal progression includes quadriplegia, an inability to maintain airway protection with secondary pneumonitis, and ultimately respiratory arrest.

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