

The neuroscience and phenomenology of sensory loss

Jonathan Cole

Introduction

Historically deafferentation has been seen in the context of a late effect of syphilis, (before the spirochaete's effects were largely killed off by antibiotics, at least in Western societies). One of the consequences of the chronic late stage of the disease is atrophy of the dorsal columns of the spinal cord, leading to loss of the tracts carrying touch and proprioception. This in turn led to the characteristic broad based staggering gait and uncontrolled ataxic movements. Dispassionate accounts of the consequences of this may have been made more difficult by the fact that with ataxia comes intractable lightning pains.¹ That such deafferentation could arise from other peripheral and central causes of loss of proprioception was less clear.²

This chapter will consider neurological causes of haptic sensory loss, from peripheral diseases leading to losses of large myelinated sensory nerve fibres alone to more central conditions including dorsal column loss and cortical damage. Perhaps the purest form of loss of movement and position sensation and cutaneous touch is seen in large fibre peripheral neuropathy. This syndrome was first described by Schaumburg and colleagues in 1980 [1].³ Since then a number of different causes has been found, though their original post infective one remains the paradigm and, being non-progressive, has allowed the most long-term study and an understanding of the importance of afferents in haptic function. The chapter will therefore end with an analysis of function in subjects with this syndrome.

Before this, however, will be given a short précis of how sensation is now tested, a century after Head's techniques (see Chapter 3).

The testing of somatic sensation

When sensation was tested by Henry Head at the turn of the 19th and 20th century it took so long and was so detailed that such a schedule would have been impossible clinically. Faster techniques became accepted. Light touch was tested with cotton wool gently stroked across the skin, pain using a pin or hypodermic syringe needle, temperature with a cold metal object and movement and position sense by small movements of the digits, either fingers or toes. Two point discrimination did require special retractors and higher cortical sensation was sometime tested *via* the drawing of numbers on the outstretched palm and rarely by placing an object in the hand for manipulation in a manner Head would have recognised. These methods were adequate for many clinical situations, when related to clinical presentation and further investigations, whether of nerve conduction, blood tests for diabetes, B12 deficiency, etc., and radiology. But they could not be considered quantitative and there was a clear need for better testing. Fortunately methods of quantitative sensory testing (QST) have been developed and are becoming increasingly accepted and developed to be rapid and reproducible for widespread clinical use (ideally when part of assessment of patients by clinical history and examination, neurophysiology (nerve conduction studies and EMG), laser evoked potentials, neuroimaging and even skin biopsy).

QST has suffered because there has been little agreement about their standard use and because of the length of time needed to perform them as well as because of problems with reproducibility. Rolke et al. [2] have tried to address this by

using a battery of tests of all aspects of sensation within approximately 30 min.

They tested thermal sensation using a small probe (7.84 cm²) placed on the skin of the hand and foot which can be cooled or warmed rapidly. Cold and warm detection thresholds are measured using stimuli which changed in a ramped manner (1° per second). In addition the thresholds for heat pain and cold pain were also determined. All tests were done three times.

Mechanical detection thresholds were determined with von Frey filaments of 0.25–512 mN made without a sharp edge which might activate nociceptors. The threshold was determined as the geometric mean of five series of ascending and descending intensities. Mechanical pain thresholds were measured with a set of seven pinprick stimulators exerting 8 to 512 mN again using the mean of a series of five ascending and descending intensities.

They also tested psychophysical ratings of some stimuli, with mechanical pain sensitivity being determined using the same weighted pinprick stimuli as for the mechanical pain threshold but this time applied in an order five times each and the subject asked to give a pain rating for each. Three light tactile stimulators (cotton wisp, cotton wool and a brush) were used to assess dynamic mechanical allodynia by stroking them across the skin in a single stroke over 1–2 cm and subjects asked to rate the pain as for pinprick.

Temporal summation or wind up of a pinprick was also tested using 10 stimuli at 1 per second and the subjects asked to rate the pain at the end numerically and this compared with single pricks at the same site. Vibration detection thresholds were tested using a tuning fork (64 Hz) placed on a bony prominence. Pressure pain thresholds were determined with a pressure gauge device with an area of 1 cm² and a pressure of up to 20 kg/cm² with an increasing stimulus repeated three times. Rolke et al. then employed statistical analyses of their data, before using Z scores to compare each subject's results with control data. This group accepts that QST remains a psychophysical test with some of the problems inherent

in this but hopes to bring it in to more standard and standardised clinical practice by reducing the time for comprehensive testing.

In an earlier analysis of QST by the American Academy of Neurology [3], its clinical use, efficacy and safety was assessed. Because of differences in technique, in normal ranges and in reproducibility, they were concerned that it should not be the sole criteria for diagnosing pathology and also recommended that it was not used in medico-legal cases. On the more positive side they did suggest that QST is probably or possibly useful in identifying small and large fibre sensory neuropathies associated with diabetic neuropathy, small fibre neuropathies, uraemic neuropathies and demyelinating neuropathy. As the techniques become more refined and widely agreed upon this list may increase.

Causes of large fibre neuropathy

Diseases of the peripheral nerves usually involve all fibre types with both large and small fibres being affected. It is rare therefore to have neuropathies of the large fibres alone, affecting touch and proprioception selectively. In large series it occurs in less than 5% or so. It is seen, however, in several conditions which fall into two broad areas. The first group involves a toxic agent and/or selective vulnerability to that agent (or its lack) in the axon, in cisplatin and vitamin B6 neuropathies, in Freidreich's ataxia, and in vitamin E deficiency (for review see [4]). In the second group an immune reaction appears to target the large fibre cell bodies in the dorsal root ganglia. This appears to be the case in carcinomatous sensory neuropathy, IgM neuropathy, Sjogren's Syndrome and acute sensory neuronopathy. The onset can be insidious and progressive, as in some carcinomatous and Sjogren's Syndrome related neuronopathy, or can be acute and severe, with all large sensory nerve cells being destroyed in days, as in some acute sensory neuronopathy syndrome. In most large fibre axonopathies sensory loss predominates, but in some neuronopathy syndromes