PAIN THRESHOLD IMPAIRMENT IN PRADER WILLI SYNDROME:
A NEUROPHYSIOLOGICAL STUDY

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RATIONALE: The neurophysiology of Prader-Willi syndrome (PWS) has been poorly investigated,
although the central nervous system is one of the main targets of the underlying genetic
defect. A hypothalamic involvement has been proposed to explain altered pain perception,
but no neurophysiological demonstration has been given yet. The present study was
undertaken to analyse and objectively investigate the sensory pathway functioning, with the
aim of identifying a possible site responsible for the altered pain perception.

PATIENTS AND METHODS: 14 PWS patients, 10 obese non-diabetic people and 19 age-matched
controls, underwent: a) MNCS (median, ulnar, peroneal, tibial) and SNCS (median, ulnar,
sural); b) somatosensory evoked potentials (SSEP) from upper and lower limbs; c)
Quantitative sensory testing to measure sensory threshold for vibration, warm and cold
sensation (WS-CS), heat and cold-induced pain (HP-CP); d) blood sample analysis to evaluate
glucose and insulin levels and calculate the quantitative insulin-sensitivity check index
(QUICKI). All the PWS patients had a deletion at chromosome 15.

RESULTS: Electroneurography was in the normal range in PWS, although PWS patients like obese
people showed significantly decreased C-MAP amplitude of the tibial and peroneal nerves,
and decreased SAP amplitude of the sural nerve. The SSEP wave latencies were all within
normal limits. In the whole PWS group, thermal and pain thresholds but not vibratory were
significantly higher than in healthy and obese people. Most of the sensory thresholds were
altered in PWS people. Insulin serum levels were significantly increased and QUICKI
decreased but less than in obese people. The sensory threshold did not correlate either with
BMI or with insulinemia levels or with QUICKI index.

CONCLUSIONS: Our preliminary data suggest that a significantly impaired thermal and pain
stimulus perception, much more evident than in our obese group, is present in PWS, but it
does not seem attributable to peripheral nerve derangement and is not related to
hyperinsulinemia and insulin sensitivity. In particular, the high pain threshold might suggest a
hypothalamic dysfunction or complex derangement of the neurotransmitter balance, as
described in PWS.