



6th International IPWSO Conference  
Cluj-Napoca 21.06.07 - 24.06.07  
"Coming here, you will discover yourself, discovering others"  
Romanian Prader-Willi Association

ASOCIACIÓN MADRILEÑA  
PARA EL SÍNDROME DE  
PRADER-WILLI



## PAIN THRESHOLD IMPAIRMENT IN PRADER WILLI SYNDROME: A NEUROPHYSIOLOGICAL STUDY

Lorenzo Priano<sup>1</sup>, Giacinta Miscio<sup>1</sup>, Graziano Grugni<sup>2</sup>, Silvia Baudo<sup>1</sup>, Alessandro Mauro<sup>1, 3</sup>

<sup>1</sup> Department of Neurology, Istituto Auxologico Italiano, IRCCS, Piancavallo (VB), Italy; <sup>2</sup> Department of Auxology, Istituto Auxologico Italiano, IRCCS, Piancavallo (VB), Italy; <sup>3</sup> Department of Neurosciences, University of Turin, Italy.

**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.