COGNITIVE-BEHAVIOURAL PROFILE IN WILLIAMS SYNDROME

Carrubba Angelica[1], Zingale Marinella[1], Greco Donatella[2], Di Blasi Francesco[3], Buono Serafino[1]

Williams Syndrome (WS) is a genetic disorder, caused by the deletion of about 25 genes from the region q11.23 of chromosome 7. The clinical phenotype is characterized by: cardiovascular anomalies, psychomotor delay, dysmorphic facial features, connective-tissue abnormalities, hyperacusis etc. About 75% of people with WS have intellectual disability, usually mild-to-moderate, learning disabilities and unique personality features (e.g. anxiety, attention deficit disorder, social disinhibition, etc). Studies on neuropsychological profile of people with WS showed strength in some language abilities, short-term auditory rote memory, facial recognition, social drive and music skills; while weakness were found in visuospatial processing abilities, planning activities, problem-solving, number processing and implicit learning.
As far as adaptive functioning profile is concerned, people with WS show good communication and social skills, while daily living skills are relatively impaired. The cognitive-behavioral phenotype (visuospatial deficits, behavioral features, motor coordination impairment and fine/gross motor control problems) influences considerably the independence level of people with WS in the execution of activities related to self-sufficiency and personal care.

1 Unità Operativa di Psicologia, IRCCS Oasi Maria SS., Troina (EN). E-mail: fbuono@oasi.en.it
2 Unità Operativa di Pediatria, IRCCS Oasi Maria SS., Troina (EN).
3 Modulo Organizzativo di Pedagogia, IRCCS Oasi Maria SS., Troina (EN).