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# BEHAVIOURAL PHENOTYPE OF NOONAN SYNDROME IN ADULTHOOD

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Introduction: Noonan syndrome (NS) is an autosomal dominant genetic disorder with an estimated incidence of 1:1,500 live births and is characterized by short stature, facial dysmorphisms and congenital heart defects. At present, mutations in seven different genes have been identified. NS is associated with impaired affective processing and subsequently increased levels of anxiety.
Objectives: Neuropsychological investigation of social cognition.
Aims: The use of neuropsychological assessment as a tool for studying the contribution of cognition and behaviour to the expression of the Noonan phenotype.
Methods: Forty adult NS-patients and a matched group of healthy controls underwent extensive neuropsychological assessment. Next to the standard cognitive domains (i.e. intelligence, attention, memory, executive functioning) several tests for social cognition were included to explore affective information processing. Correlation analysis and repeated measures MANCOVA were used.
Results: Marked problems were found in the recognition of own and other's emotions, as well as in the ability to verbally express feelings. Alexithymia was significantly more prevalent in the NS-group. In addition, NS-patients displayed more mood and anxiety complaints than controls. A tendency was found to social desirability and agreeableness.
Conclusions: Impairments in social cognition are common elements of NS behavioural phenotype in adults. With neuropsychological assessment, psychosocial immaturity, amenable traits and alexithymia could be identified. The latter increases the vulnerability for the development of mood and anxiety disorders.

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