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**Title:** Cognitive functioning in adults with Noonan syndrome**Author(s):** Wingbermuhle, Ellen (217863817)  
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Egger, J.I.M. (255791976)**Publication year:** 2013**Document type:** Article in monograph or in proceedings**Book title:** Final Program. The 2013 International Neuropsychological Society Mid-Year Meeting**Start page:** p. 5**Publisher:** [S.l. : s.n.]**Related link(s):** [http://www.ins%2Damsterdam2013.nl/abstracts.php?instance=i88kt7z2ul6%26file\\_name=abstracts%26open\\_in\\_browser=1](http://www.ins%2Damsterdam2013.nl/abstracts.php?instance=i88kt7z2ul6%26file_name=abstracts%26open_in_browser=1)**Abstract:** Objective: Noonan syndrome (NS) is a common genetic disorder characterized by short stature, facial dysmorphism, congenital heart defects and a slightly lowered mean IQ. Genetic research has revealed mutations in nine genes in the RAS-MAPK pathway. Although research on cognitive functioning in NS is scarce, previous studies indicate generalised deficits in children with NS. In this study, functioning within all major cognitive domains was evaluated in a large group of adults with NS, and compared with a control group. Participants and Methods: A group of 42 adult patients with NS (16- 61y), as well as 42 healthy controls matched for age, sex and education level underwent extensive neuropsychological assessment, including the multiple tested domains intelligence, speed of information processing, memory, executive functioning, and visuoconstruction. Domain scores were compared by GLM multivariate analyses of variance and posthoc independent t-tests were performed for between-group comparisons within significant domains. Results: Patients with NS showed a significantly worse performance in the domain speed of information processing ( $p < 0.05$ ,  $\hat{p}2 = 0.059$ ), while their performance on delayed recall was better than that of the control group ( $p < 0.05$ ,  $\hat{p}2 = 0.055$ ). No between-group differences were found on any of the other cognitive domains. Conclusions: While diffuse cognitive problems seem to be present in children with NS, cognitive functioning of adults with this syndrome is characterized by mental slowness, but no other cognitive impairments, taken education level into account. Despite this relatively intact profile, patients frequently report cognitive complaints, indicating that individual neurocognitive and psychological assessment is important in clinical management of NS. Longitudinal research is needed to study cognitive development in NS.**Subject:** Experimental Psychopathology and Treatment  
Neuropsychology and rehabilitation psychology**Subject:** Neuro- en revalidatiepsychologie**Organization:** SW OZ DCC NRP  
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