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PREPULSE INHIBITION IN PATIENTS WITH FXTAS

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Fragile X associated tremor/ataxia syndrome (FXTAS) is a late-onset neurodegenerative disorder that affects carriers of the fragile X premutation, typically after age 50. White matter disease, neuropathy, tremor, ataxia, cognitive decline, and dementia have been described extensively. Prepulse inhibition (PPI) is used as a measure for sensorimotor gating. Studies in animals have indicated that hippocampus and entorhinal cortex, structures which are affected in FXTAS, are involved in the regulation of PPI. The objectives of this study were to determine if patients with FXTAS have altered PPI, and to study possible correlations between PPI and cognitive performance. A passive acoustic PPI paradigm was applied in 57 subjects, 35 carriers of the fragile X premutation (Mean_{age}= 60.92 years), and 22 healthy controls (Mean_{age}= 64.68 years). Of the 35 premutation carriers, 22 subjects showed usable PPI data (13 male, 9 female), and 13 of the controls (7 male, 6 female). 9 premutation carriers had no signs of FXTAS, 13 patients had a FXTAS stage of 3, 4 or 5 (severely impaired). There were no significant differences in the PPI in the female subjects. The male premutation carriers had a tendency towards a reduced PPI at 120ms stimulus interval. The mean latency of the startle response at 105dB was longer in the male premutation carriers (p=.008) compared to the controls. The results show a subtle change in sensorimotor gating processes in older male carriers of the fragile X premutation.