**Autosomal dominant spinocerebellar ataxia type 40 caused by mutations in the CCDC88C gene - report of the first European family**

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Autosomal dominant spinocerebellar ataxias form a group of clinically and genetically diverse disorders. There has been only one report of the spinocerebellar ataxia 40 in a Chinese family. We identify four-generation Polish family with tremor, cerebellar ataxia, Parkinsonism and dementia history. The proband is a 59 years old man with a 10-year history of asymmetric upper limb tremor as a prevailing symptom. Neurological examination also indicates negligible: ataxia, dysdiadochokinesia, bradykinesia, hyperreflexia and mild cognition function impairment. MRI of the proband's brain is normal. The whole exome sequencing analysis was done in four individuals of this family, including three affected and one unaffected. The mutations in genes associated with Parkinson's disease and Wilson's disease were not confirmed. The genes associated with tremor and ataxia were analyzed. The analysis revealed missense mutation in the CCDC88C gene. Bioinformatic analysis shows that the Asp43Asn mutation of CCDC88C is pathogenic. Cosegregation analysis was done and the mutation cosegregates with the phenotype. Referring to OMIM database this mutation is related with SCA 40.