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Nutrients intake in patients with Huntington disease and its relevance to their nutritional status and disease's severity

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Abstract

Introduction and objective

Huntington Disease (HD) is a rare neurodegenerative disorder which is characterized by psychiatric, cognitive and motor progressive impairment. The aim of our study was to compare the nutritional status with calorie and nutrients intake between HD patients and healthy individuals and determine the correlations between nutritional status as well as macro- and micronutrient intake and severity of HD.

Materials and methods

Research encompassed 26 genetically tested HD patients as a Huntington's disease group (HDG) and 26 healthy individuals as a control group (CG). To assess patients clinically UHDRS motor, functional, independence, behavior and cognitive tests were performed.

Results

Higher intake of vitamin D in HDG correlated with higher fat content, higher BMI and lower annual BMI decrement. Higher intake of fat improved TFC, cognitive general and Stroop's test in HDG. Linear Regression in HDG revealed that predictor of BMI was vitamin D intake and TMS.

Conclusions

Nutritional value of daily diet of patients with HD needs to be improved. Of great importance is an adequate fat, carbohydrates and vitamin D intake which affect - body fat content and comply with physical and mental ability in patients with HD.

Key words: Huntington disease, nutrients intake, weight loss, body mass index, fat content

1. Introduction

Huntington Disease (HD) is an autosomal, dominant neurodegenerative disorder which is characterized by psychiatric, cognitive and motor progressive impairment. In the course of the disease patients' develop choreatic movements often accompanied or preceded by depression, irritability, anxiety, aggression or any other psychiatric symptoms and cognitive decline usually displayed as executive function impairment, lose of flexibility of mind and psychomotor processes retardation [1-7]. Disease is caused by mutation in *HTT* gene that results in expansion of a CAG repeat [2]. A disease-modifying treatment for patients with HD is not available [1-4].

One feature of the HD is weight loss which is not associated with anorexia but rather with increased energy expenditure, particularly with spontaneous physical activity caused by chorea [1]. Surprisingly it was reported that patients with HD despite weight loss, consume more calories than unaffected individuals [10, 11]. Some observations explained that unintended weight lose might be caused by central degeneration in the cortex, striatum and hypothalamus which in turn may affect food intake or energy expenditure [9]. It is believed that patients with HD present increased basal metabolic rate which may be also a cause of weight loss [3]. Body weight is reported to be a strong predictor of disease progression [10, 12] and with the course of the disease body weight is slightly decreasing in early and mild stages leading to dramatic decrease in advanced stages [13]. Furthermore, it was also reported that lower BMI (Body Mass Index) value is associated with higher risk of dementia and mild cognitive impairment in elderly people [14].

2. Objectives

The aim of our study was to compare the nutritional status with calorie and nutrients intake between two groups, patients with HD and healthy individuals. We intended also to determine the correlations between nutritional status as well as macro- and micronutrient intake and severity of HD.

3. Material and methods

3.1 Participants

All participants of the study were enrolled between January 2011 and April 2012. Research encompassed 26 genetically tested and confirmed patients with HD and 26 healthy individuals as a control group (CG). Subjects in CG were caregivers of HD patients, so the dietary patterns were expected to be similar due to the same social environment. Due to this fact any discrepancies between Huntington's disease group (HDG) and CG in food intake could result rather from individual preferences modified by course of the disease than social environment.

Patients with HD were participants of REGISTRY study by European Huntington Disease Network (EHDN). REGISTRY is an international, multicenter observational study on clinical features and progression of HD [15]. For purpose of this study UHDRS (The Unified Huntington's Disease Rating Scale) motor, chorea scale, functional, independence, behavior, cognitive assessments were performed in HD group. TFC (Total Functional Capacity), GCI (Global Clinical Impression) scales (appendix 1) were done in each patient and disease duration, disease burden [15], based on CAG repeats number in larger *HTT* allele in all affected participants were calculated.

At the time of the visit each patient was weighed and body fat content was measured using TANITA MC 980 MA. The research tool was 24-h diet recall, which was performed in each subject in the presence of caregiver and HD patient. Nutritional data were analyzed using Dietetyk2006 software. BMI value was calculated in each subject twice, at interval of one year and the difference between these two calculations was called BMI shift.

All participants gave informed written consent according to the International Conference on Harmonisation – Good Clinical Practice (ICH-GCP) guidelines (http://www.ich.org/LOB/media/MEDIA482.pdf). Ethical approval was obtained from the local ethics committee of Poznan University of Medical Sciences, Poland. The REGISTRY protocol was approved by the EHDN Scientific and Bioethics Advisory Committee and by local ethical committee of Poznan University of Medical Sciences, Poland.

3.2 Statistical analysis

To calculate disease burden following calculation was used: $(CAGn_{larger\ allele} - 35.5) *$ age at examination [16]. Statistical analysis was performed using SPSS. To calculate differences between groups t-student test was used and to calculate correlations r-Pearson test was used. Linear regression was used to verify potential predictors of BMI, annual BMI shift, fat content and body mass, introducing to regression equation variables which statistically correlated respectively with BMI, annual BMI shift fat content, body mass (table 5). Lacking data were not implemented. P-values ≤ 0.05 were considered significant.

4. Results

No significant differences between HDG and CG in caloric, nutrients, vitamins and minerals intake were observed (table 1). One exception was a finding that patients with HD consumed twice as less of vitamin C as healthy individuals (55.59 vs. 135.41 mg).

Parameter	*Patients with HD*	*Control Group*	*P*
General groups characteristics			
Age (mean ± SD)	50.73 ± 11.30	38.04 ± 18.54	0.004
Body mass (kg, mean ± SD)	63.21 ± 21.22	68.47 ± 14.57	0.556
Height (cm, mean ± SD)	170.38 ± 8.92	172.54 ± 10.38	0.426
Fat content (%, mean ± SD)	19.55 ± 12.76	23.99 ± 7.94	0.212
BMI (kg/m^2, mean ± SD)	23.35 ± 3.44	22.81 ± 3.03	0.856
Annual BMI shift (kg/m^2 ± SD)	- 0.46 ± 2.01	-	-
Calorie intake (kcal, mean ± SD)	2504.34 ± 641,16	2427.76 ± 1322.96	0.986
HD group characteristics			
CAG repeats large allele (mean ± SD)	42.58 ± 9.26	-	-
TMS (mean ± SD)	47.92 ± 27.19	-	-
GCI (mean ± SD)	2.46 ± 1.24	-	-
Independence (%, mean ± SD)	64.23 ± 27.26	-	-
Disease burden (mean ± SD)	415.48 ± 130.72	-	-
Chorea (mean ± SD)	12.08 ± 6.57	-	-
Function (mean ± SD)	18.31 ± 6.40	-	-
TFC (mean ± SD)	8.50 ± 3.57	-	-
Behaviour (mean ± SD)	1131 ± 10.66	-	-
Cognitive (mean ± SD)	148.85 ± 105.55	-	-
Verbal fluency test (mean ± SD)	41.11 ± 51.17	-	-
SDMT **(mean ± SD)**	16.92 ± 11.16	-	-
Stroop's test (mean ± SD)	90.81 ± 54.61	-	-
Duration of disease (years, (mean ± SD)	7.46 ± 4.83	-	-
Nutrients intake characteristics			
Protein (g, mean ± SD)	93.68 ± 29.12	99.96 ± 75.46	0.568
Protein (%,mean ± SD)	14.85 ± 2.80	16.23 ± 4.65	0.425
Plant protein (g, mean ± SD)	29.96 ± 9.66	33.57 ± 21.93	0.357

Animal protein (g, mean ± SD)	63.18 ± 24.13	62.47 ± 58.47	0.957
Isoleucine (mg, mean ± SD)	4458.11 ± 1496.61	4599.65 ± 3792.63	0.848
Leucine (mg, mean ± SD)	6983.97 ± 2221.17	7068.27 ± 5839.38	0.891
Lysine (mg, mean ± SD)	6250.27 ± 2179.48	6278.94 ± 5467.03	0.984
Methionine (mg, mean ± SD)	2239.36 ± 780.15	2271.95 ± 1926.90	0.892
Cysteine (mg, mean ± SD)	1383.29 ± 472.16	1439.83 ± 1166.61	0.816
Phenylalanine (mg, mean ± SD)	4114.73 ± 1263.90	4146.77 ± 3347.64	0.877
Tyrosine (mg, mean ± SD)	3209.89 ± 1039.22	3283.78 ± 2672.79	0.849
Threonine (mg, mean ± SD)	3863.20 ± 1228.12	3815.26 ± 3163.93	0.997
Tryptophan (mg, mean ± SD)	1188.31 ± 394.34	1198.34 ± 958.52	0.993
Valine (mg, mean ± SD)	5191.08 ± 1665.12	5298.30 ± 4106.18	0.892
Arginine (mg, mean ± SD)	4888.12 ± 1758.15	4576.94 ± 4206.69	0.978
Histidine (mg, mean ± SD)	2707.23 ± 1143.10	2981.33 ± 3214,32	0.625
Alanine (mg, mean ± SD)	4634.32 ± 1659.24	4737.37 ± 4141.61	0.923
Asparagine (mg, mean ± SD)	8490.52 ± 2496.43	8255.48 ± 6311.51	0.885
Glutamine (mg, mean ± SD)	17815.08 ± 5297.23	18104.36 ± 14467.93	0.883
Glycine (mg, mean ± SD)	4186.71 ± 1564.71	4248.31 ± 3617.25	0.987
Proline (mg, mean ± SD)	6309.66 ± 1984.76	6324.83 ± 4458.99	0.992
Serine (mg, mean ± SD)	4422.48 ± 1412.28	4488.42 ± 3350.02	0.859
Carbohydrates (g, mean ± SD)	306.20 ± 84.21	315.37 ± 155.45	0.663
Carbohydrates (%, mean ± SD)	46.85 ± 8.74	49.85 ± 11.04	0.534
Saccharose (g, mean ± SD)	76.34 ± 48.53	73.09 ± 67.50	0.964
Saccharose (%, mean ± SD)	11.96 ± 6.64	12.57 ± 8.32	0.658
Dietary fibre (g, mean ± SD)	19.45 ± 6.59	24.12 ± 14.54	0.122
Saturated fatty acids (g, mean ± SD)	39.88 ± 17.41	31.53 ± 21.81	0.245
Monounsaturated fatty acids (g, mean ± SD)	44.34 ± 19.49	37.55 ± 30.79	0.641
Polyunsaturated fatty acids (g, mean ± SD)	15.36 ± 6.94	13.92 ± 10.97	0.988
Omega-3 fatty acids (g, mean ± SD)	1.85 ± 1.00	2.63 ± 5.49	0.401
Omega-6 fatty acids (g, mean ± SD)	13.50 ± 6.37	11.27 ± 9.15	0.651
Fat (g, mean ± SD)	107.85 ± 40.18	94.25 ± 65.47	0.670

Fat (%, mean ± SD)	38.28 ± 7.79	33.90 ± 10.64	0.323
Cholesterol (mg, mean ± SD)	407.55 ± 250.38	389.05 ± 340.56	0.890
Vitamin A (µg, mean ± SD)	1409.59 ± 2192,74	1124.85 ± 893.31	0.609
β-caroten (µg, mean ± SD)	2749.71 ± 2578.09	4602.94 ± 4740.09	0.062
Vitamin D (µg, mean ± SD)	4.32 ± 3.82	3.35 ± 4.29	0.395
Vitamin E (mg, mean ± SD)	10.19 ± 4.84	11.54 ± 8.64	0.492
Vitamin C (mg, mean ± SD)	55.59 ± 41.26	135.41 ± 132.17	0.005
Folacin (µg, mean ± SD)	276.77 ± 79.16	354.92 ± 225.58	0.102
Vitamin B_1 (mg, mean ± SD)	1.37 ± 0.53	1.44 ± 1.30	0.814
Vitamin B_2 (mg, mean ± SD)	1.65 ± 0.80	1.76 ± 0.93	0.665
Vitamin B_6 (mg, mean ± SD)	1.86 ± 0.57	1.99 ± 1.44	0.667
Vitamin B_{12} (mg, mean ± SD)	4.43 ± 5.16	3.92 ± 3.62	0.679
Niacin (mg, mean ± SD)	21.02 ± 8.46	24.41 ± 24.59	0.512
Natrium (mg, mean ± SD)	2732.94 ± 1281.00	2573.43 ± 2548.40	0.777
Potassium (mg, mean ± SD)	3781.44 ± 866.57	3835.52 ± 1858.89	0.894
Calcium (mg, mean ± SD)	653.09 ± 333.52	733.21 ± 506.29	0.388
Phosphorus (mg, mean ± SD)	1416.42 ± 409.58	1508.32 ± 999.62	0.667
Magnesium (mg, mean ± SD)	336.27 ± 118.98	360.77 ± 232.01	0.635
Iron (mg, mean ± SD)	12.79 ± 5.14	13.31 ± 9.07	0.800
Manganese (mg, mean ± SD)	4.33 ± 2.09	5.30 ± 3.69	0.248
Copper (mg, mean ± SD)	1.28 ± 0.36	1.33 ± 0.69	0.736
Zinc (mg, mean ± SD)	11.82 ± 3.92	11.91 ± 9.42	0.964
Ca/P ratio (mean ± SD)	0.46 ± 0.19	0.50 ± 0.22	0.344
Na/K ratio (mean ± SD)	0.73 ± 0.33	0.72 ± 0.61	0.910

Table 1. Descriptive statistics.

In the CG higher calories, protein, amino acids, saturated fatty acids (SFA), monounsaturated fatty acids (MUFA), vitamin D, B_{12} and natrium intake negatively correlated with a fat content but not with weight and/or BMI (table 2). These correlations with a fat content were not observed in HDG (table 3). In HDG, the higher intake of β-caroten correlated with lower fat content but not weight, BMI and annual BMI shift and in oposite, higher intake of vitamin D positively correlated with higher fat content, higher BMI and lower annual BMI decrement. Intake of calories, amino acids (with the exception of arginine), cholesterol, vitamin A, B_{12} and Ca/P ratio was significantly reduced among the age in HDG when this phenomenon in CG was not observed (table 3). Lower intake of calories and listed nutrients did not correlate with disease severity measured in scales used (table 3).

	Body mass	BMI	Fat content
Calories intake	0.238	- 0.069	- 0.616**
Carbohydrates [%]	0.369	0.290	0.013
Saccharose [%]	0.145	0.111	- 0.006
Fat [%]	- 0.425*	- 0.345	0.035
SFA	0.000	- 0.267	- 0.549*
MUFA	0.033	- 0.197	- 0.556**
Omega-3 fatty acids	0.074	- 0.143	- 0.344
Cholesterol	0.307	0.013	- 0.486*
Protein [g]	0.221	- 0.051	- 0.536**
Plant protein	0.292	0.014	- 0.442*
Animal protein	0.186	- 0.080	- 0.552**
Isoleucine	0.249	- 0.023	- 0.567**
Leucine	0.222	- 0.063	- 0.580**
Lysine	0.230	- 0.037	- 0.557**
Methionine	0.256	- 0.027	- 0.564**
Cysteine	0.310	0.000	- 0.575**
Phenylalanine	0.234	- 0.057	- 0.585**
Tyrosine	0.211	- 0.068	- 0.575**
Threonine	0.232	- 0.040	- 0.572**
Tryptophan	0.280	0.014	- 0.569**
Valine	0.264	- 0.013	- 0.574**
Arginine	0.169	- 0.116	- 0.576**
Asparagine	0.284	0.015	- 0.552**
Glutamine	0.242	- 0.057	- 0.587**
Glycine	0.288	0.045	- 0.531**
Proline	0.211	- 0.068	- 0.588**
Serine	0.254	- 0.034	- 0.587**
Vitamin A	0.277	0.028	- 0.346
Vitamin D	0.123	- 0.132	- 0.445*
β-caroten	0.220	0.034	- 0.233
Vitamin B_{12}	- 0.010	- 0.219	- 0.486*
Natrium	0.030	- 0.194	- 0.501**
Manganese	0.197	0.137	- 0.268
Ca/P ratio	- 0.419*	- 0.382	- 0.079

Table 2. Correlations between anthropometric measurements and nutrients intake – control group.

*** *p*-value ≤ 0.05; ** *p*-value ≤ 0.01**

MUFA – Mono Unsaturated Fatty Acids

SFA – Saturated Fatty Acids

	Body mass	BMI	Annual BMI	Fat content	Age	Disease burden	Disease duration	Motor TMS	Chorea	Functional
Calories intake	-0.013	-0.216	-0.058	-0.051	-0.484*	0.282	0.040	-0.139	-0.129	0.225
Carbohydrates [%]	-0.083	-0.205	-0.356	-0.377	0.174	0.117	0.156	0.347	0.418*	-0.401*
Saccharose [%]	0.056	-0.340	-0.358	-0.337	-0.081	0.226	0.184	0.194	0.239	-0.274
Fat [%]	0.032	0.102	0.299	0.323	-0.159	-0.105	-0.247	-0.372	-0.469*	0.374
SFA	0.067	-0.094	0.033	0.140	-0.435*	0.079	0.055	-0.267	-0.302	0.330
MUFA	-0.015	-0.124	0.192	0.177	-0.327	0.126	-0.152	-0.285	-0.331	0.298
Omega-3 fatty acids	0.041	-0.062	0.062	0.132	-0.174	0.159	-0.409*	-0.127	-0.223	-0.284
Cholesterol	0.283	0.066	-0.014	-0.158	-0.507**	0.044	0.026	-0.242	-0.215	0.176
Protein [g]	0.041	-0.002	0.100	0.086	-0.413*	0.133	0.116	-0.135	-0.126	0.275
Plant protein	-0.193	-0.043	0.061	0.001	-0.256	0.207	-0.197	0.024	0.062	0.185
Animal protein	0.150	0.005	0.101	0.100	-0.399*	0.069	0.225	-0.162	-0.189	0.257
Isoleucine	0.055	-0.032	0.095	0.068	-0.457*	0.127	0.102	-0.128	-0.144	0.251
Leucine	0.064	-0.009	0.055	0.059	-0.418*	0.147	0.120	-0.102	-0.120	0.260
Lysine	0.111	0.026	0.113	0.123	-0.413*	0.092	0.186	-0.143	-0.177	0.268
Methionine	0.113	0.050	0.150	0.095	-0.417*	0.074	0.098	-0.145	-0.150	0.268
Cysteine	0.033	0.027	0.123	0.058	-0.508**	0.177	0.019	-0.081	-0.047	0.216
Phenylalanine	0.066	0.033	0.043	0.032	-0.415*	0.150	0.098	-0.133	-0.147	0.290
Tyrosine	0.048	0.042	0.001	0.054	-0.403*	0.121	0.127	-0.123	-0.153	0.300
Threonine	0.111	0.037	0.145	0.105	-0.412*	0.128	0.146	-0.139	-0.147	0.280
Tryptophan	0.037	-0.101	0.012	0.010	-0.479*	0.112	0.124	-0.144	-0.171	0.257
Valine	0.079	-0.031	0.033	0.036	-0.466*	0.139	0.127	-0.120	-0.132	0.259
Arginine	-0.013	0.139	0.249	0.154	-0.23	0.007	-0.003	-0.169	-0.187	0.341

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Asparagine	0.193	0.053	0.132	0.075	-0.443*	0.133	0.139	-0.200	-0.193	0.322
Glutamine	-0.017	-0.083	0.045	0.049	-0.412*	0.191	0.118	-0.054	-0.063	0.225
Glycine	0.046	0.024	0.217	0.146	-0.421*	0.049	0.084	-0.164	-0.183	0.262
Proline	-0.047	-0.187	-0.115	-0.020	-0.420*	0.192	0.222	-0.044	-0.062	0.200
Serine	0.071	-0.027	-0.018	-0.002	-0.480*	0.155	0.124	-0.104	-0.130	0.255
Vitamin A	0.202	0.058	-0.207	-0.281	-0.085	0.294	0.235	0.059	0.177	-0.024
Vitamin D	0.338	0.643**	0.553**	0.399*	-0.038	-0.014	-0.099	-0.131	0.010	0.261
β-caroten	0.005	-0.170	-0.163	-0.431*	0.260	0.038	0.056	-0.100	0.154	-0.161
Vitamin B_{12}	0.158	-0.034	-0.221	-0.257	-0.172	0.277	0.214	0.035	0.074	0.036
Natrium	-0.031	-0.067	0.118	0.117	-0.315	-0.203	0.043	-0.318	-0.390*	-0.397*
Manganese	0.088	0.322	0.252	0.233	0.235	-0.079	-0.092	-0.105	0.063	0.259
Ca/P ratio	-0.256	-0.255	-0.434*	-0.147	0.175	-0.099	0.128	0.161	0.056	-0.127

Table 3a. Correlations between anthropometric measurements/clinimetric tests' results and nutrients intake – Huntington's disease group.

	TFC	Independence	GCI	Cognitive	Verbal fluency test	SDMT	Stroop's test
Calories intake	0.177	0.195	-0.011	0.020	-0.105	0.079	0.121
Carbohydrates [%]	-0.454*	0.074	0.300	-0.464*	-0.253	-0.487*	-0.561**
Saccharose [%]	-0.321	-0.146	0.251	-0.375	-0.200	-0.309	-0.475*
Fat [%]	0.456*	-0.198	-0.335	0.472*	0.287	-0.487*	0.545**
SFA	0.318	0.013	-0.173	0.226	0.023	0.289	0.356
MUFA	0.373	-0.028	-0.192	0.287	0.102	0.329	0.393*
Omega-3 fatty acids	0.395*	-0.059	-0.206	0.262	0.151	0.308	0.303
Cholesterol	0.134	0.180	-0.072	0.066	0.003	0.058	0.114
Protein [g]	0.208	0.307	-0.024	0.091	-0.065	0.143	0.207
Plant protein	0.066	0.441*	0.094	-0.086	-0.202	-0.007	0.024
Animal protein	0.231	0.186	-0.074	0.142	0.000	0.177	0.239
Isoleucine	0.203	0.253	-0.028	0.106	-0.037	0.154	0.208
Leucine	0.200	0.277	-0.008	0.076	-0.082	0.130	0.197
Lysine	0.219	0.213	-0.072	0.138	0.002	0.164	0.230
Methionine	0.229	0.296	-0.039	0.116	-0.042	0.177	0.228
Cysteine	0.150	0.390*	0.019	0.025	-0.092	-0.050	0.124
Phenylalanine	0.224	0.319	-0.035	0.071	-0.100	0.134	0.204
Tyrosine	0.236	0.257	-0.049	0.107	-0.073	0.171	0.241
Threonine	0.216	0.309	-0.030	0.115	-0.054	0.167	0.238
Tryptophan	0.206	0.250	-0.056	0.105	-0.025	0.137	0.198
Valine	0.194	0.258	-0.023	0.084	-0.058	0.133	0.189
Arginine	0.253	0.434*	-0.099	0.171	-0.011	0.211	0.298
Asparagine	0.247	0.296	-0.064	0.115	-0.033	0.156	0.222
Glutamine	0.159	0.302	0.023	0.010	-0.127	0.074	0.124
Glycine	0.239	0.276	-0.078	0.154	0.025	0.163	0.240
Proline	0.144	0.236	0.009	-0.034	-0.141	-0.001	0.067
Serine	0.180	0.287	-0.019	0.055	-0.084	0.085	0.168
Vitamin A	-0.247	0.072	0.288	-0.145	-0.134	-0.115	-0.132
Vitamin D	0.159	0.242	0.041	0.198	0.042	0.331	0.275
β-caroten	-0.117	0.152	0.204	-0.299	-0.234	-0.320	-0.293
Vitamin B_{12}	-0.172	0.045	0.217	-0.063	-0.092	-0.019	-0.033
Natrium	0.406*	0.169	-0.284	0.264	0.081	0.328	0.367
Manganese	0.130	0.496**	0.007	0.092	-0.167	0.253	0.282
Ca/P ratio	-0.047	-0.323	-0.049	-0.084	-0.014	-0.109	-0.128

Table 3b. Correlations between anthropometric measurements/clinimetric tests' results and nutrients intake – Huntington's disease group.

*** *p*-value ≤ 0.05; ** *p*-value ≤ 0.01**

BMI – Body Mass Index; SDMT - Symbol-Digit Modality Test; TMS – Total Motor Score; MUFA – Mono Unsaturated Fatty Acids**;** GCI - Global Clinical Impression; TFC - Total Functional Capacity; SFA – Saturated Fatty Acids

In the HDG, BMI positively correlated with some clinimetric tests' results like UHDRS functional, Symbol-Digit Modality Test (SDMT), Stroop's test and negatively with TMS (Total Motor Score), indicated better clinical status of these patients with higher BMI (table 4).

	Body mass	**BMI**	**Annual BMI**	**Fat content**	**Age**	**Disease burden**	**Disease duration**
Motor TMS	-0.405*	-0.439*	-0.193	-0.332	0.123	0.373	0.312
Chorea	-0.154	-0.191	-0.074	-0.189	0.210	0.310	0.367
Functional	0.427*	0.485*	0.112	0.417*	-0.186	-0.239	-0.398*
TFC	0.390*	0.361	0.070	0.375	-0.144	-0.387	-0.455*
GCI	-0.175	-0.276	-0.088	-0.360	0.163	0.401*	0.371
Cognitive	0.173	0.380	0.176	0.557**	-0.266	-0.401*	-0.287
Verbal fluency test	-0.009	0.194	0.091	0.424*	-0.402*	-0.320	-0.253
SDMT	0.346	0.484*	0.214	0.570**	-0.092	-0.396*	-0.302
Stroop's test	0.272	0.453*	0.210	0.563**	-0.119	-0.395*	-0.257
Behaviour	-0.178	-0.370	-0.206	-0.554**	0.319	0.073	0.146

Table 4. Correlations between anthropometric measurements and clinimetric tests' results – Huntington's disease group.

*** *p*-value ≤ 0.05; ** *p*-value ≤ 0.01**

BMI – Body Mass Index

GCI - Global Clinical Impression

SDMT - Symbol-Digit Modality Test

TFC - Total Functional Capacity

TMS – Total Motor Score

In this study fat intake seemed to improve physical and mental functioning of patients with HD. Higher intake of fat improved TFC outcome, cognitive and Stroop's test. On the other hand, intake of carbohydrates seemed to worsen physical as well as mental skills. Carbohydrates aggravated outcome in UHDRS functional scores, TFC, cognitive, SDMT and Stroop's test. Furthermore, higher intake of saccharose also correlated with worse outcome in Stroop's test. Higher intake of plant proteins, cysteine, arginine and manganese correlated with better outcome in Independence scale.

Linear Regression in HDG revealed that predictor of BMI was vitamin D intake and TMS, predictor of annual BMI shift was vitamin D intake, predictor of body mass was UHDRS functional scores, and predictor of fat content was Stroop's test result (table 5).

Dependent variable	**Independent variables**	**β coefficient**	**F value**	**p**
BMI shift	**Vitamin D**	0,553	10,59	0,003
	BMI	0,083		0,718
	Ca/P	-0,199		0,328
Body mass	**Functional**	0,427	5,36	0,029
	Motor TMS	-0,157		0,652
	TFC	-0,003		0,994
Fat content	**Stroop's Test**	0,563	11,14	0,003
	Behavioural	-0,364		0,060
	Cognitive	0,262		0,583
	Verbal Fluency Test	0,108		0,634

Table 5. Results of linear regression with annual BMI shift, body mass and fat content as a dependent variable among HDG (n=26)

BMI – Body Mass Index

TMS – Total Motor Score

TFC – Total Functional Capacity

5. Discussion

Weight loss, which leads to deterioration in overall state of health, was often reported in individuals with HD [11, 17, 18]. Moreover, patients with a larger CAG expansion presented faster weight loss [16]. Clinically weight loss of ≥ 1 kg/year correlated with more severe motor impairment and chorea in patients with HD [19]. The weight loss occurred even when caloric intake is adequate or higher than energy needs [19]. In this study no significant differences between groups in respect to weight, height, BMI and fat content were observed. Higher caloric intake reported earlier in HD patients was not confirmed in this study [11, 13].

Patients with HD consumed significantly less than controls and inadequate amount of vitamin C. Recommended Dietary Allowances (RDA) for vitamin C equals 90 mg for men and 75 mg for women [20], therefore HDG seems to be slightly undernourished with vitamin C. The reason of decreased vitamin C consumption in HDG may be mastication difficulties and choking episodes. Rich sources of vitamin C are vegetables and fruits and those food products are often tough and difficult to chew causing problems in HD patients who display tongue and cheeks involuntary movements. Furthermore, the mean value of consumed iron in HDG was low (± 12.79 mg) and in conjunction with low intake of vitamin C, which significantly increases Fe absorption, risk of iron deficiency development and consecutively anaemia is higher.

It was previously reported that patients with HD have lower plasma amino acids levels, especially branched chain amino acids (BCAA) levels [11]. We did not found statistically significant differences in amino acids intake between both groups but the intake reduction with age could be possible explanation for lower plasma BCAA concentration especially in older HD patients. Higher amount of amino acids especially exogenous leucine, isoleucine and valine in a diet of HD patients could therefore help to correct deficits in serum [21]. BCAA are crucial for proper muscle protein metabolism and they increase protein synthesis [22].

It was reported that higher BMI at HD onset was associated with slower HD progression. Those patients with higher BMI exhibit better outcome in motor performance measured using UHDRS, milder chorea and less incidences of depression

[19]. Study conducted on elderly demented patients revealed that higher BMI value correlated with better cognitive status measured with MMSE (Mini Mental State Examination) [14]. We confirmed previously reported phenomenon that HD patients with higher BMI are cognitively and physically better.

Here we reported also that patients with higher fat content function better and their behavior is less affected and this observation reflect previous reports from patients with dementia [14]. This confirms the hypothesis that HD affected individuals need more energy to maintain body weight and support metabolic processes with a special pressure on these metabolic processes in brain. Patients with HD exhibit mitochondrial dysfunction and calcium dyshomeostasis. Increased mitochondrial pores permeability and toxic Ca^{2+} influx may also affect energy balance of patients with HD [23].

It is believed that high intake of calcium causes decrease in PTH and consecutively decrease in cytosolic [Ca^{2+}] in human adipocytes switching their metabolism from lipogenesis to lipolysis [24, 25]. As we observed that annual BMI decrement is higher in HD patients with higher Ca compared to P intake then is important to avoid high amount of calcium in HD diet. Recommended daily allowance (RDA) for calcium equals 1000 mg per day for individuals between 31 and 50 year of age [26]. Higher vitamin D intake in patients with HD results in higher BMI value. Calcium is important for appropriate action of vitamin D which is also known as a hormonal vitamin. Vitamin D affects bone metabolism, so sun exposition is advisable.

The total intake of fat was higher than dietary recommendations in both groups (HDG: 38.28% and CG: 33.90% of total consumed calories). This finding is in conjunction with results obtained in research conducted on healthy Polish population [27, 28]. Intake of fat is not statistically significant but higher in HDG than CG. Moreover, this study revealed that also intake of saturated fatty acids (HDG: 39.88 g vs CG: 31.53 g) is also higher in HDG but this difference is not statistically significant. In the general population higher intake of fat, especially in the form of saturated fatty acids is not advisable due to the deteriorative effect on lipid profile. Excessive amount of saturated fatty acids in a daily diet may lead to the development of hypercholesterolaemia and therefore cardiovascular diseases. Fat in a diet is associated with high palatability and this may be the reason of its increased

consumption. Ingestion of fat causes release of endocannabinoids and consequently fat storage. Here we proved that total fat intake seemed to improve physical and mental functioning of patients with HD. Higher intake of fat improved TFC outcome, cognitive and Stroop's test. So it is important to ensure adequate amounts of high quality fat in daily diet patients with HD in order to prevent from decline in physical and mental ability.

Average carbohydrates intake was reported to be higher in HD patients than in controls [17], but it was not observed here. Some authors indicated that increased consumption of carbohydrates triggers mild cognitive impairment in elderly people. Above finding was confirmed in this research. Patients with HD who consumed more carbohydrates were cognitively worse and obtained worse outcomes in motor tests. Furthermore, higher intake of saccharose contributed to deterioration in intellectual ability. It was already demonstrated that a surplus of simple carbohydrates in opposite to complex carbohydrates causes decline in memory performance. It is also worth to mention that patients with HD exhibit impaired glucose regulation and it may directly affect their cognitive function [17].

In HDG TMS and vitamin D level significantly contributed to BMI namely higher vitamin D intake increased BMI when higher TMS reduced BMI. Longitudinally annual BMI downward shift is reduced by higher vitamin D intake. So adequate intake of vitamin D allow to maintain proper body weight also in a long-term perspective. Therefore, it is important to ensure adequate amount of vitamin D in a diet of HD patients in order to prevent from weight loss. Adequate intake of vitamin D in a daily diet equals 15 μg of cholecalciferol. In this research mean amount of consumed vitamin D was below recommendation (mean value 4.32 μg/d).

Lower value of BMI is associated with worse outcome in TMS. Increased body weight contributed to better physical performance. In many studies scientists revealed that weight loss often precedes motor abnormalities [13, 16]. Subsequently, regression analysis revealed that higher fat content is associated with better outcome in Stroop's test. Body fat influences intellectual skills and in this context of great importance is balanced nutritive dietary plan to preserve proper body fat content.

In CG, as it was expected lower intake of fat in a daily diet contributed to lower body mass. This finding was not confirmed in HDG. Furthermore, lower

calories intake was associated with lower fat content and interestingly this relation was also not confirmed in HDG. It seems that in patients with HD factors other than calories and fat intake make a contribution to the extent of body fat content.

6. Conclusions

This is the first paper considering the composition of nutritious dietary plans for patients with HD in order to prevent weight loss which occur in advanced stages of HD. Balanced diet for people with HD should cover their increased caloric requirements and be tailored to personal disabilities like choking episodes or mastication difficulties. Of great importance is an adequate fat intake which in turn triggers to preservation of body fat content and improves physical and mental ability in patients with HD. There is a need to find some modern strategies helping affected people to improve nutritional status and therefore prevent from cognitive as well as physical decline. Dietary counseling and monitoring nutrition risk indicators in times when casual treatment for HD patients is not yet available should be an integral part of treatment of patients with HD in order to provide better overall assistance.

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Appendix 1

The UHDRS'99 motor assessment includes 15 items with five response options (0-4), for a maximum total score of 124, with higher scores reflecting more severe disease, progression in this scale therefore is defined as scores' increase between subsequent visits; the functional assessment includes 25 items describing patients potential independence in particular functions with response options yes or no, score range 0-25, where lower scores reflect lower functional capacity, progression in this scale therefore is defined as scores' decrease between subsequent visits; the cognitive assessment consists of 5 items: verbal fluency test, symbol-digit modality test (called symbol test), and 3 Stroop tests: word naming, word reading and interference, with lower scores reflecting poorer cognitive function, progression in this scale therefore is defined as scores' decrease between subsequent visits; the behavioural assessment includes 11 items with 5 response options (0-4) in 2 sections ('frequency' and

'severity') and 5 rater marks with answers yes or no, higher scores express disease severity, progression in this scale therefore is defined as scores' increase between subsequent visits; independence scale, where the subject's independence is measured in percents with 100% reflecting complete independence, progression in this scale therefore is defined as scores' decrease between subsequent visits. The TFC scale, a standardized measure to assess capacity to work, handle finances, perform domestic chores and self-care tasks, and live independently was also administered; the scale ranges from 13 (normal) to 0 (severe disability) , progression in this scale therefore is defined as scores' decrease between subsequent visits.

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