Malnutrition and nutrition related problems in adults with mitochondrial diseases

UMC St Radboud

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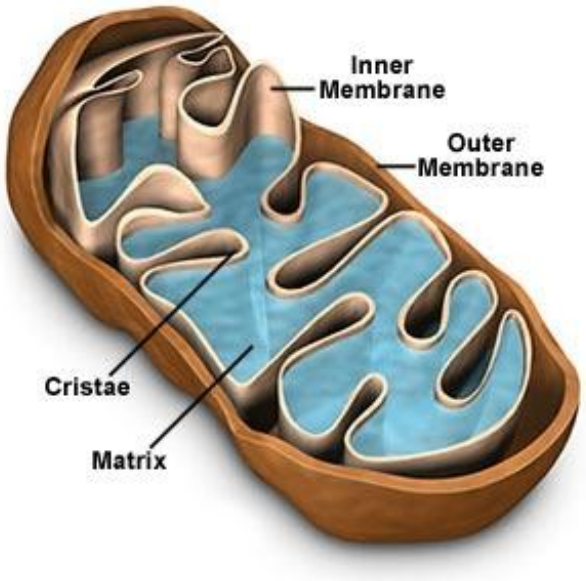
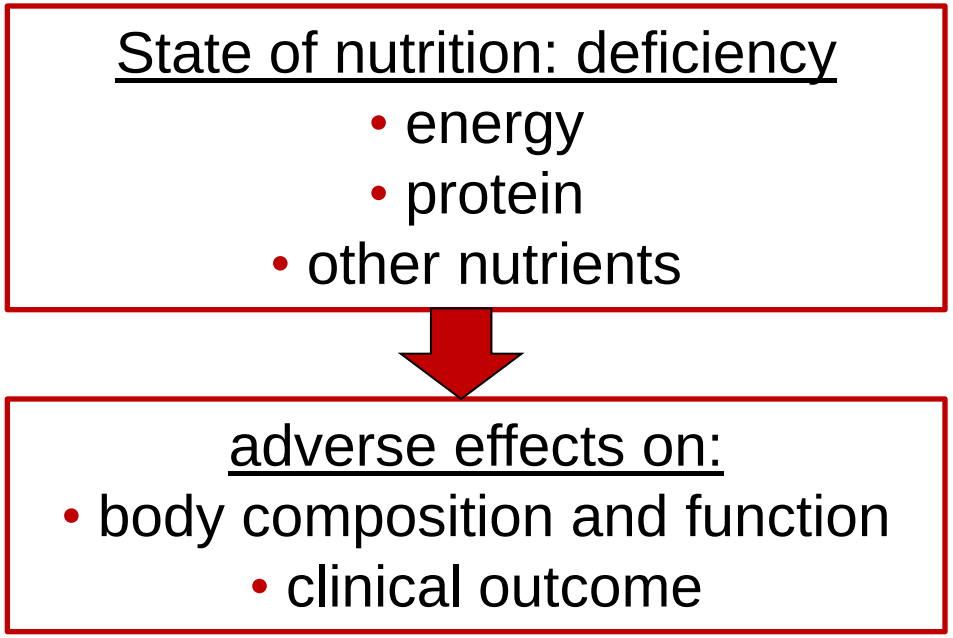
Mitochondrial diseases

prevalence 1:10,000
treatment supportive
Nijmegen Center Mitochondrial Disorders



Cross section mitochondrion
source: www.ncmd.nl

Malnutrition:



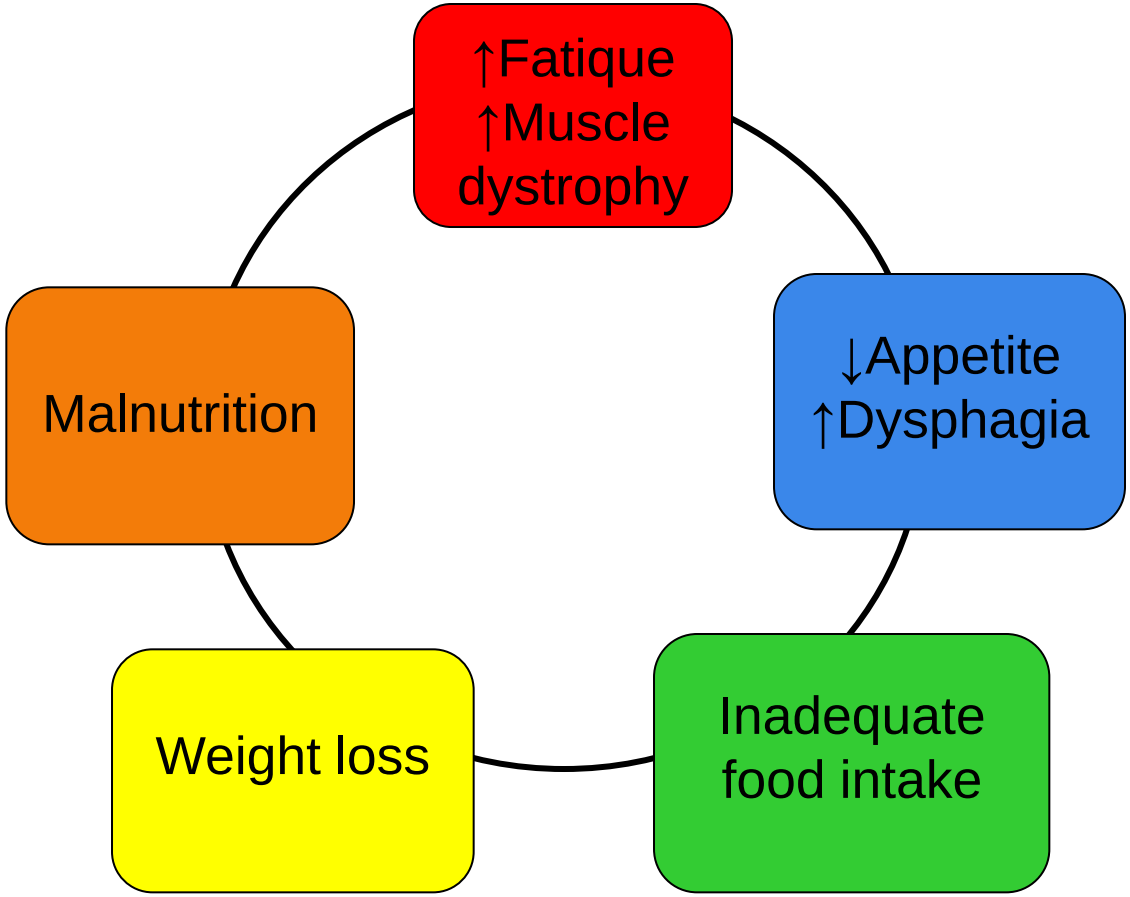
Mitochondrial disorder =

malnutrition on cellular level

Stratton R.J. USA 2003

Malnutrition in Mitochondrial Disorders

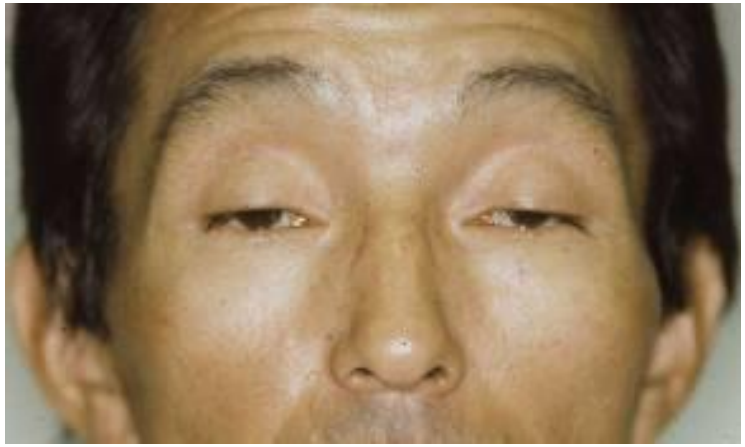
- Secondary mitochondrial dysfunction
- ↑ existing complaints → ↓Quality of Life



Morava E. Nutrition 2006.

Dysphagia in Mitochondrial Disorders

Cause:



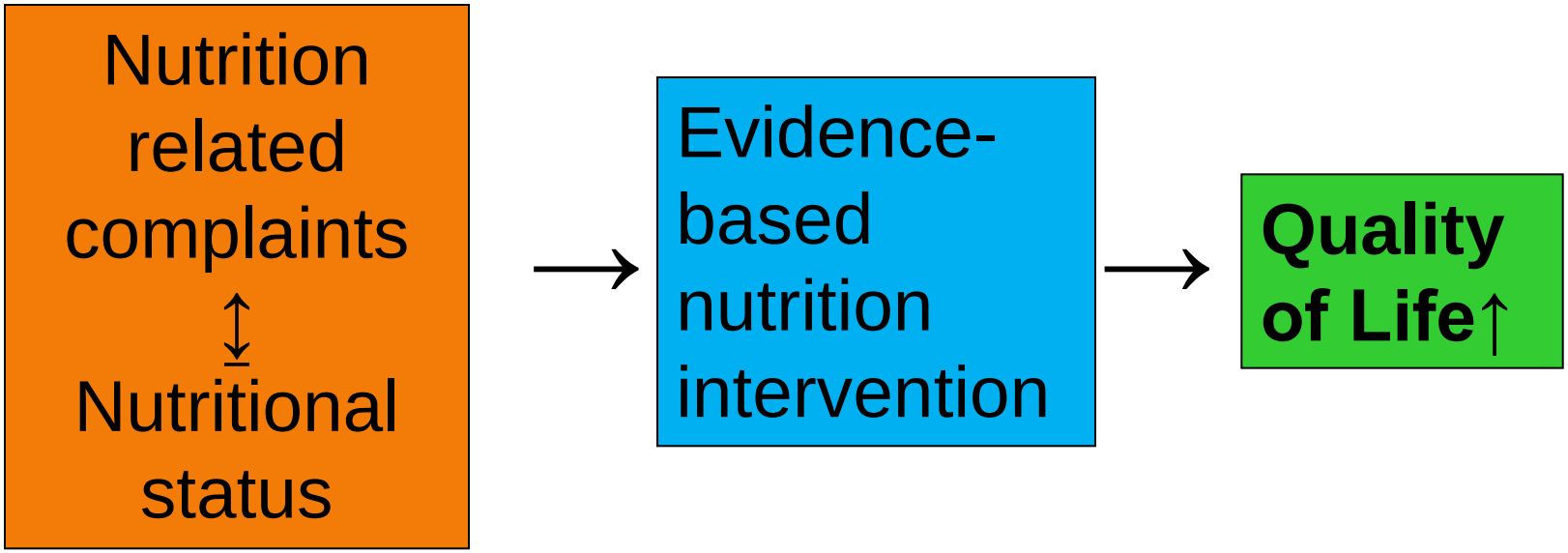
- muscle weakness
- fatigue
- ptosis
- slow
- accelerated aging
- centrally controlled: strokes and epilepsy
- coordination problem

Result:

↓nutritional state

Aspiration →pneumonia →↑ mortality

Research aim



Method:

Pilot:

- 23 adults with mitochondrial disease
- June 2010-June 2011 evaluation dietician
- food diary:

Kcal Protein Carbohydrates Fat Water Fiber	use special diet products use vitamin or mineral supplements food groups

- anthropological data
- diet-related complaints
- nutritional assessment (n=10)

Method

Nutritional Assessment (NA):

- IC with Delta Track
  (REE (kcal) and RQ)
- BIA (FFM and % fat)
- Anthropometry : height / weight

Method

Status research :
anthropological data of 77 MELAS, CPEO and MIDDS patients

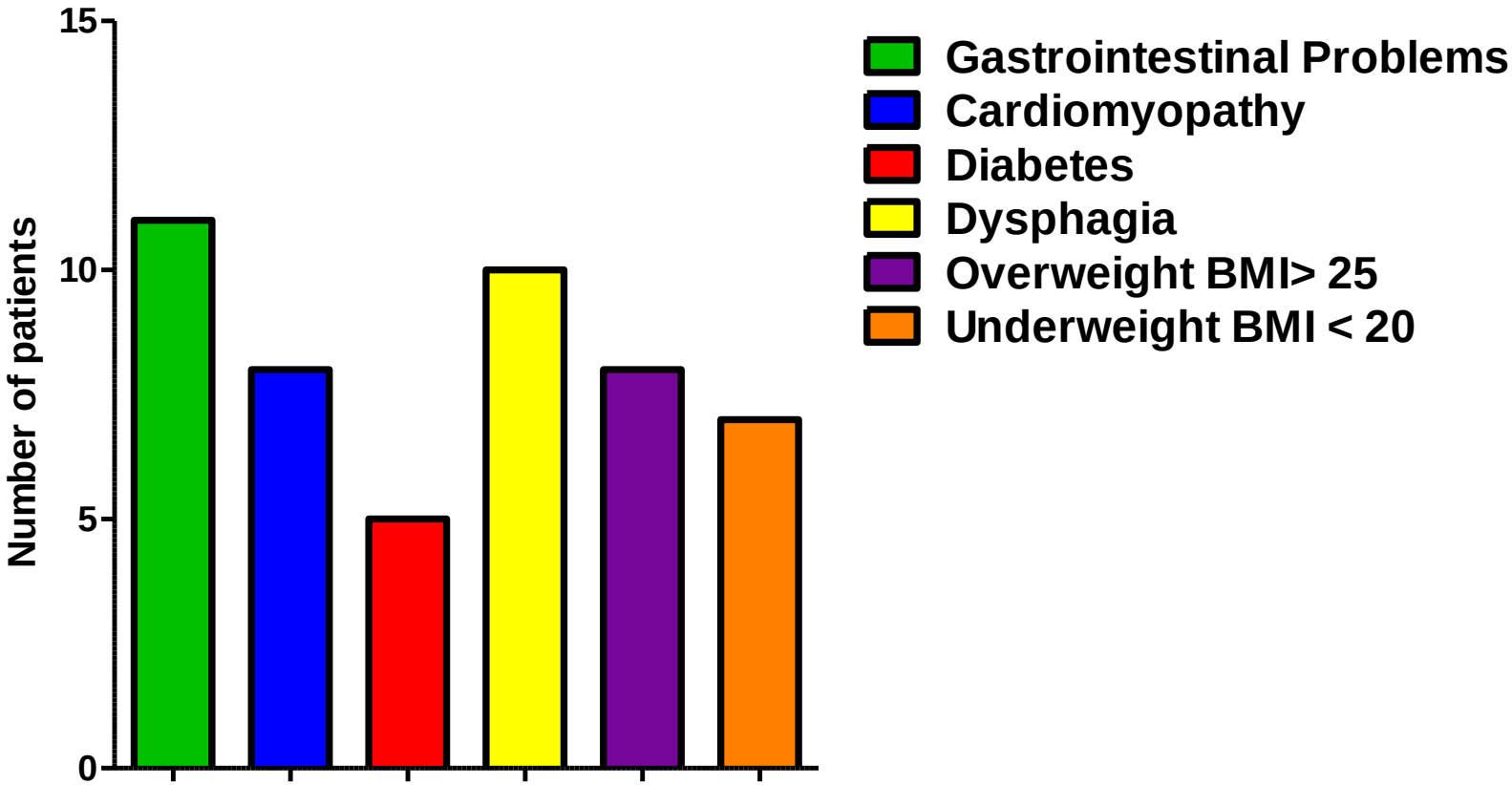
23 CPEO patients:
Swallow Speed Test
(Hughes and Wiles)

B. Smits, J Neurology (2011)

Nutrition related complaints

n= 23 (6 ♂ 17 ♀)

mean(range): 38 (18-58) years



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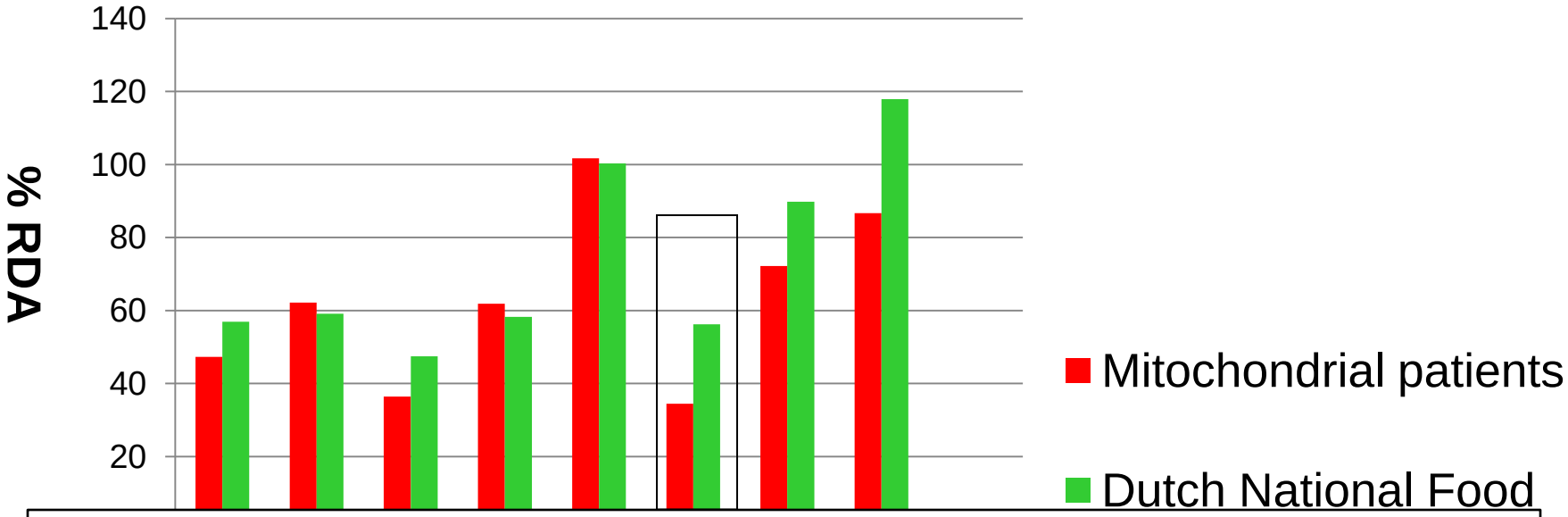
Food diary

n= 20 (6 ♂ 14♀)
mean(range): 38(18-58) years

- No adequate intake
- Special diet products (n=5)
- Low protein intake (n=4)
- High sugar intake (n=12)

Food diary compaired to the Recommended Dietary Allowance (RDA)



Milkproducts:

- Mitochondrial patients — mean 34%RDA range 0-81%
- Dutch healty adults — mean 56%RDA range 0-230%

Body composition

n=12 3 ♂ 9 ♀
mean(range):40(20-58) years

Sex	Fat Free Mass (%) healthy	Fat Free Mass (%) patients	Fat Mass (%) Healthy	Fat Mass (%) Patients
M	80.8	76.1 (SD 11.6)	19.2	23.9 (SD 11.6)
V	71.9	63.1 (SD 10.4)	28.1	36.9 (SD 10,5)

Status research

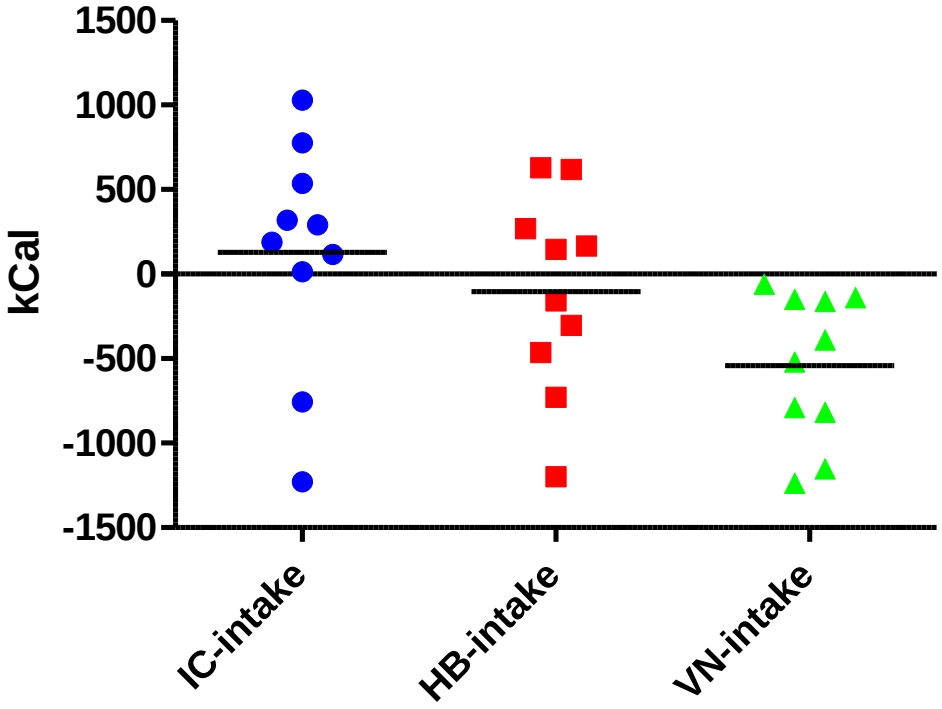
Stature

Sex		N	Min.	Max.	Mean (SD)	Dutch healthy (CBS)
M	Height (m)	32	1,62	1,96	1,78 (0,8)	1,81
V	Height (m)	45	1,45	1,82	1,65 (0,7)	1,68

Energy requirements and energy intake

n=10 1 ♂ 9 ♀

mean(range): 38(18-58) years



REE Harris and Benedict
mean 158 kCal =11%
overestimation of
REE Indirecte calorimetrie
(-25 - +386 kCal SD 143 kCal)

- **Intake - REE Indirecte calorimetrie + activity allowance (kCal)**
- **Intake – Harris en Benedict + activity allowance (kCal)**
- **Intake – Dutch energy reccomendations (kCal)**

Underweight / overweight

n= 77 32 ♂ 45 ♀

mean(range):46(19-69) years .

51 patients with m3243AG mutation: mean age 44 years

26 CPEO patients: mean age 50 Years

	CPEO (%)	m3243AG mutation (%)	Total (%)	Healthy (CBS) (%)
Underweight BMI<20	27	26	26	1.5
Normal weight BMI 20-25	31	53	46	51
Overweight BMI > 25	42	22	29	47.5
Obesity BMI>30	12	4	6,5	12

Dysphagia and BMI

n= 39 17 ♂ 22 ♀

16 patients with m3243AG mutation and 23 CPEO patients

mean(range): 47(28-69) years

		N	Minimum	Maximum	Mean	SD
dysphagia	BMI	16 (41%)	16.8	27.8	20.4	3.4
no dysphagia	BMI	23 (59%)	16.9	34.7	24.3	4.7

Conclusion

Adults with mitochondrial disease:

- low energy requirement
- need for high nutrient density
- adequate intake not achieved

Cause:

- dysphagia,
- bowel problems
- fatigue

Increased risk of malnutrition.

Discrepancy between need and intake → nutritional intervention useful

Future

More research

- Actometre
- Validated Fatique questionaire
- Gastrointestinal questionaire

Before and after diet intervention

- body composition,
- muscle strength
- Quality of Live

Questions?

Dysphagia and BMI

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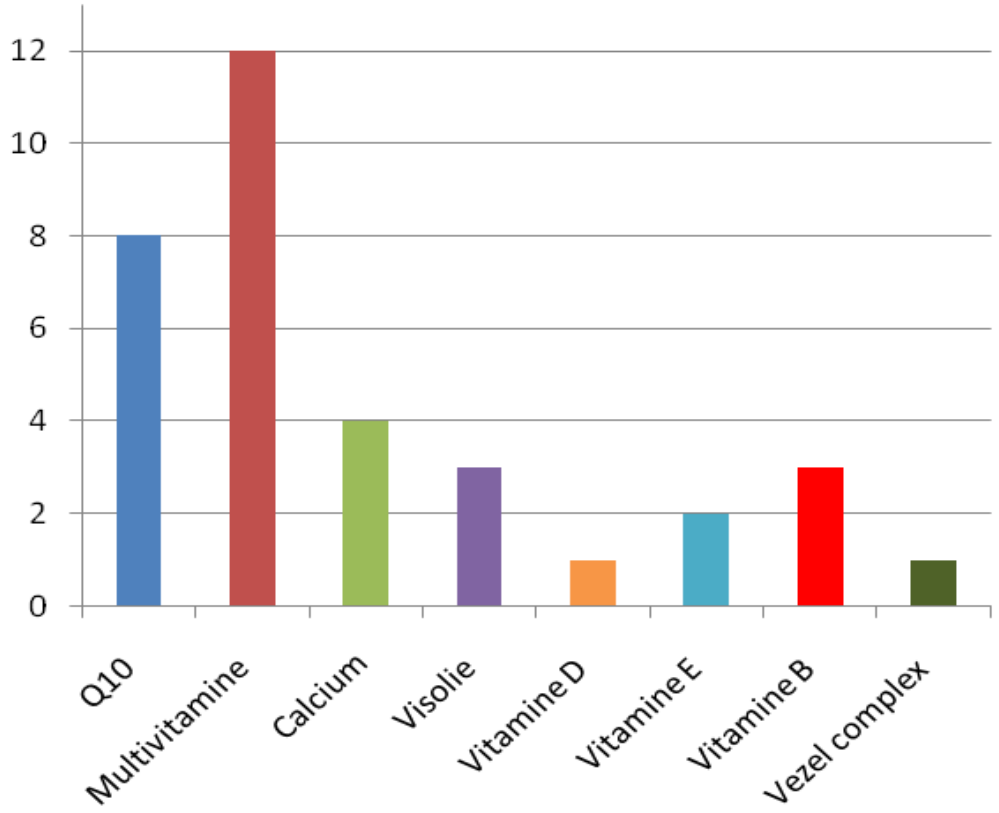
Mann-Whitney test p = 0.07

Supplementation vitamins and minerals

- 12 of 23 patients takes multivitamins
- 8 of 23 patients takes Q10



Underweight / overweight

n= 77 32 men 45 women 19-69 mean 46 years
mean age m3243AG mutation: 44 Years
mean age CPEO: 50 Years

	CPEO			m3243AG mutation			total	Healthy (CBS)	
SEXE	M (%)	V (%)	%	M (%)	V (%)	%	%	M %	V %
Underweight BMI<20	4 (31)	3 (23)	27	7 (37)	6 (19)	26	26	1	2
Normal weight BMI 20-25	2 (15)	6 (46)	31	7 (37)	20 (63)	53	46	46	56
Overweight BMI > 25	7 (53)	4 (31)	42	5 (26)	6 (19)	22	29	53	42
Obesity BMI>30	0 (0)	3 (23)	12	0 (0)	2 (6)	4	6,5	11	12