

Resting Energy Expenditure in the Kennedy's disease

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Aims

Spinal and bulbar muscular atrophy (SBMA), known as Kennedy's disease, is a rare and incurable X-linked recessive genetic, progressive, adult-onset neuro-muscular disease. SBMA has a wide non-neural clinical phenotype, including metabolic disorders such as insulin-resistance and dyslipidemia. While in other neuro-muscular diseases an increase of energy expenditure has been described, in SBMA patients this aspect have not yet been assessed.

Methods.

Eighteen SBMA patients ranged in age from 42 to 67 years and 18 age matched healthy men were evaluated. Resting energy expenditure (REE) was measured by an open-circuit indirect calorimeter after an overnight fast. Harris-Benedict equation was used to calculate the predicted REE. Body composition was assessed by bioimpedance analysis (BIA 101 RJL, Akern, Bioreserch, Florence, Italy). Free fat mass (FFM) was obtained by subtracting fat mass from total body weight.

Results

BMI and age did not differ significantly between the two groups. In SBMA subjects FFM was $46.8 \pm 5,8$ kg ($64.7 \pm 4.9\%$), REE was 1550 ± 170 kilocal/24h, predicted value 1605 ± 118 kilocal/24h ($+15 \pm 12,4\%$; $p < 0,01$), REE/body weight was $22.2 \pm 2,5$ kilocal/kg, REE/FFM was $34,8 \pm 3,5$ kilocal/kg. In control subjects REE was $1638 \pm 148,0$ kilocal/24h, REE/body weight was $21,7 \pm 1,3$ kilocal/kg and REE/FFM was $31,6 \pm 2,1$ kilocal/kg ($p < 0,05$).

Conclusions

Our results indicate that SBMA patients expend more energy respect to normal subjects when the REE is normalized for the FFM. An altered protein turnover and respiratory chain defects were observed in SBMA animal model (1,2). These abnormalities can explain the hypermetabolism observed in SBMA patients.

References

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Body composition

(mean \pm DS)

	SBMA subjects	Controls
BMI kg/m ²	24,4 \pm 3,4	24,5 \pm 3,5
FM Kg	25,5 \pm 6.4	24,3 \pm 7,3
FM %	35.3 \pm 6,3	31,7 \pm 7.4
FFM Kg	46.8 \pm 5,8	51,9 \pm 5.4
FFM %	64.7 \pm 4.9	58.9 \pm 4.4

Tab. 1- BMI and body composition in SMA and in control subjects.

Resting Energy Expenditure

	SBMA subjects	Controls
Kilocal/ Kg body weight/24h	22.2 \pm 2,5	21,7 \pm 1,3
Kilocal /kg FFM/24h	34,8 \pm 3,5	31,6 \pm 2,1*

* $p < 0,005$

Tab.2 - REE in SBMA and in control subjects.