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<u>The Need to Accurately Measure Energy Intake and Expenditure in</u> <u>Patients with Systemic Sclerosis</u>

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ABSTRACT

Background

Malnutrition is common in systemic sclerosis (SSc) and patients are frequently underweight. However, the balance between assessed dietary energy intake vs. expenditure has been neglected to date. This study aimed to assess energy (dietary) intakes and expenditures and to compare discrepancies in SSc.

Methods

36 outpatients with SSc completed the study. Demographics and clinical data were recorded. Functional questionnaires were completed. Predicted energy requirements were calculated. Over a consecutive 3-day period, patients completed an estimated food diary and wore a specialist energy expenditure monitor (SenseWear® Armband). Assessments of intake and expenditure were compared for individual patients and the impact according to patient demographics, clinical manifestations, and disease severity evaluated.

Results

Energy intake did not correlate with predicted (s= 0.117, P=0.511) or measured (s=-0.039; P=0.825) expenditures. Predicted and measured energy expenditures correlated, but actual values differed for individuals (Intraclass correlation = 0.62; 95% limits of agreement -459 to 751kcal). Respiratory involvement was negatively correlated with number of steps (s=-0.350, P=0.04) and time spent lying (s=0.333, P=0.05). There was a significant correlation between BMI and predicted vs. measured energy discrepancy (s=0.41; P=0.02), and this discrepancy was greater with higher BMIs.

Conclusions

There was no correlation between intake and either predicted or measured energy expenditure. Predicted and measured energy were strongly correlated yet differed for the individual patient. In patients with SSc, where energy expenditure must be accurately assessed, it should be directly measured. Keywords: Systemic Sclerosis; Energy Intake; Energy Expenditure; Physical Activity; SenseWear® Armband

INTRODUCTION

Systemic Sclerosis (SSc) is characterised by widespread tissue fibrosis of the skin and internal organs and its complex pathobiology also includes vascular and neural abnormalities (1). Malnutrition is common in patients with SSc and often multi-factorial (2,3). The entire length of the gastrointestinal (GI) tract can be involved and other factors (e.g., musculoskeletal involvement and low mood) can also result in nutritional impairment (1,4). Although malnutrition is an important cause of disease-related mortality in SSc (2,5), this has been little studied to date. In particular, few studies have specifically investigated the relative effects of reduced oral intake (e.g., as a consequence of GI manifestations) compared to increased active and/or sedentary energy expenditures in patients with SSc.

To date, investigators have compared the energy intakes between patients with SSc and healthy controls. For example, no difference was identified in the energy intakes of 61 patients with SSc compared to 67 matched healthy participants with statistically different body mass indices (BMIs) (6). Using an activity questionnaire, the latter study also found similar proportions of patients and healthy participants to be physically active for at least 150 minutes per week. However, this approach did not account for differences in sedentary activity, and also did not compare outcomes between patients. This lack of any detectable difference in energy intake between patients and healthy volunteers with similar BMIs has also been reported by another study (7). Likewise, Caporali et al (8) measured energy intake via 3-day dietary records and also found that there was no difference in the percentage of SSc patients with and without disease-related malnutrition who were not consuming an 'adequate' intake. An earlier study using 7 day weighed records reported a higher dietary energy intake in 12 patients with SSc compared to healthy controls (9).

To-date, only one study has measured energy expenditure in patients with SSc (10). In this study, the SenseWear® Armband was utilised and compared the physical activity of 27 patients with SSc and preserved nutritional status to 11 matched healthy participants over at least 6 days (10). However, dietary intake was not assessed. Patients with SSc performed less daily physical activity than healthy participants, and this activity reduction (duration and level) occurred in patients with very early respiratory involvement affecting gaseous diffusion.

Against this background, we therefore sought to investigate the relationship between energy intake and expenditures, including differences between predicted and measured expenditures. We also compared discrepancies between 1) intake and expenditure, and 2) predicted vs. measured expenditures, with patient demographics, functional impact and disease severity.

MATERIALS

Patients

A semi-selective approach was used to recruit consecutive outpatients with SSc attending routine clinic appointments between June 2012 and May 2014 at a tertiary referral centre for SSc. Patients had a clinical diagnosis of SSc and were classified as either the diffuse or limited subset of the disease according to LeRoy et al (11). Targeted patient identification ensured the inclusion of patients with a representative spectrum of clinical manifestations from SSc. Patients were ineligible if they had an eating disorder, severe psychiatric illness or other GI disease leading to weight loss, were acutely unwell, pregnant, or had an implanted electrical device. Ethical approval was granted by the North West Ethics Committee (12/NW/0247).

Patient and disease-related information

Following recruitment, demographic and clinical data (date of birth, gender, handedness, smoking status, SSc disease-related characteristics and GI manifestations) were obtained. GI involvement was based on clinically performed investigations: oesophageal – reflux +/ dysmotility demonstrated on investigations (e.g., OGD, barium swallow, pH manometry) and small intestinal – dilatation or SIBO (based on Barium follow through/CT/MR/breath test or proven need for rotational antibiotics). Disease onset was defined by first non-Raynaud's symptom (11). Respiratory and cardiac disease severity was defined using the Medsger SSc Severity Scale, which defines the effect (reversible and irreversible) of disease on the organ's function (12). This is an ordinal scale of severity: 0 (normal), 1 (mild), 2 (moderate), 3 (severe) and 4 (end-stage). All patients completed the Scleroderma Heath Assessment Questionnaire (SHAQ) which assesses function and incorporates the Health Assessment Questionnaire (HAQ) disability index (HAQ-DI) and five additional visual analogue scales, including patient global assessment and lung involvement. (13-15). Malnutrition Universal Screening Tool (MUST) scores were determined using the BMI and details of any unintentional weight loss

(>5% over 3 to 6 months) (16); patients were graded as being at low, medium, or high risk of malnutrition according to MUST score.

Energy (dietary) intake assessment

Energy and macro-nutrient consumption (intake) were reported using a 3 day estimated food diary, completion of which was supported by a selection of validated food portion photographs (17). Agreed portion size estimates were used to interpret qualitative portion size estimates (18). Microdiet version 2.0 (Downlee Systems Ltd, High Peak, UK), which utilises validated food composition tables, was used to quantify nutrient consumption (19).

Energy expenditure assessment

Predicted energy expenditure

Predicted energy requirements were calculated using Schofield's basal metabolic rate equation and the agreed UK average Physical Activity Level of 1.4 (20).

Measured energy expenditure

Expenditure was measured over 3 consecutive days (two weekdays and one weekend day) using the SenseWear® Armband (BodyMedia Inc, Pittsburgh, PA), which was to be worn continuously unless bathing. This measured 3-axis acceleration, heat flux, galvanic skin response and skin temperature and counted steps. From these recorded data, the SenseWear® Basic version 7.0 software deduced the percentage time worn, time spent lying and sleeping and total energy and time spent at different energy intensities: Metabolic Equivalent of Task (METs). Sedentary activity was defined as ≤ 1.5 METs. Light intensity activity was classed as 1.1 to 2.9METs, moderate intensity as 3.0 to 5.9METs and vigorous intensity as >6.0METs (21).

Statistical analysis

For our statistical analysis we utilised SPSS version 22 and StatsDirect version 3. The discrepancy (expended minus consumed) between dietary and expended energies was calculated. Analysis used non-parametric methods including Spearman's test (s), Mann-Whitney U and Kruskal-Wallis as appropriate. Agreement studies (ICC) were used to compare

energy intakes and expenditures. The cut-off for statistical significance was accepted as a p value of ≤ 0.05 .

RESULTS

Patients

Forty-two patients were consented. However, due to intercurrent medical problems, 5 withdrew prior to commencement. Thus, 37 patients commenced the study, but 2 failed to complete the diary component, and one was later excluded as they deviated from protocol. The results are reported for the 36 patients who completed the whole or part (no dietary component) of the study. Demographics and clinical data are detailed in Table 1.

Daily energy intake

Mean daily intakes are depicted in Table 2. Energy intakes did not correlate with BMI (s=-0.162; p=0.36). Food substances had differing energy densities. Carbohydrates formed the bulk of most diets (mean 45%; range 31% to 61%). However, patients also acquired a significant proportion of their energy from fat (mean 37%; range 18 to 51%). There were no differences in the percentage energy intakes from these different food groups in patients with and without oesophageal or small intestinal involvement.

Predicted and measured expenditures

Predicted and measured expenditures are shown in Table 2. Patients were mostly resting or undertaking light exertion. Mean daily time at >1.5METS was 20% (299/1440 min) and ranged from 5% (70/1440 min) to 40% (575/1440 min). Few patients performed high intensity activity and those who did, only did so for short periods.

Comparison between energy intake and expenditures

There was no correlation between energy intake and either predicted (s=0.117, P=0.511; n=34) or measured (s=-0.039; P=0.825) expenditure. For all patients (n=36), the mean difference between intake and expenditure was 241±709kcal/day (range -1654 to 1784). Fourteen patients reported having consumed more energy than they expended (mean 400±416kcal/day; range 44 to 1654). Twenty patients expended more energy that they reported consuming (mean

 689 ± 491 kcal/day; range 68 to 1784). Of the 5 patients with weight loss, only 2 had expenditures greater than their intakes (excess expenditures = 116kcal and 1438kcal).

Comparison between predicted and measured energy expenditures

Predicted and measured energy expenditures were strongly correlated (s=0.706, P=<0.01). However, actual values differed for individual patients (Intraclass correlation= 0.62; 95% limits of agreement -459 to 751kcal; Figure 1). For those patients with higher mean energies per day, the expenditures often exceeded predicted requirements. In contrast, for those patients with lower mean energies, predicted energies often exceeded measured expenditures.

Energy discrepancy comparisons

Active and sedentary activities

Active (mean daily steps and mean time at >1.5 METS) and sedentary (mean time lying and sleeping) activities were compared to patient demographics, functional impairment, and SSc disease severity (Table 3). Age was negatively correlated with number of steps (s=-0.667, P=<0.01) and time >1.5 METS (s=-0.390, P=0.02). The severity of respiratory involvement based on Medsger scoring was negatively correlated with number of steps (s=-0.350, P=0.04) and time spent lying (s=0.333, P=0.05). There was a trend that the severity of cardiac involvement was negatively correlated with number of steps (s=-0.313, P=0.07).

Measured vs predicted energy expenditures

Patient demographics

All patients (n=34) had a discrepancy between energy expended and consumed, but the magnitude differed between individuals. This discrepancy correlated (s=-0.463; P=<0.01) with age. Younger patients expended more energy than they consumed (Figure 2). In comparison, discrepancies were smaller in older patients, and a few patients appeared to consume more energy than they expended. There were no correlations (n=34) between the energy discrepancy and gender (s=0.11; P=0.54) or disease subtype (s=-0.23; P=0.19). However, there was a significant correlation (n=34) between BMI and energy discrepancy (s=0.41; P=0.02). The discrepancy was greater with higher BMIs (Figure 3).

Functional impairment

There was no correlation (n=34) between energy discrepancy and total HAQ-DI (s=-0.027; P=0.88) or SHAQ global disability (s=0.100; P=0.57). There was no difference in the mean energy discrepancies of patients with $(260\pm795\text{kcal/day})$ and without $(214\pm594\text{kcal/day})$ oesophageal (p=0.99) or with (-73±766kcal/day) and without (354±66kcal/day) small intestinal (p=0.19) involvement.

SSc disease severity: lung and cardiac involvement

Based on the Medsger severity scale, the majority of patients (69%) had normal cardiac function. The remaining patients (28%) had mild disease. Data was unavailable for 1 patient. None of the patients with moderate or severe cardiac disease were included in this study. In contrast, based on the Medsger severity scale, most of the patients studied had respiratory involvement (39% = normal respiratory function; 22% = mild; 25% = moderate; 11% = severe; 3% = end-stage involvement). There were no correlations between energy discrepancies and the Medsger cardiac (s=-0.164; P=0.36; n=33) or lung (s=-0.254; P=0.15; n=34) severity scores.

There were no correlations between energy discrepancies and the Medsger cardiac (s=-0.164; P=0.36; n=33) or lung (s=-0.254; P=0.15; n=34) severity scores.

DISCUSSION

To our knowledge, this is the first study to directly compare measured energy intakes and expenditures in patients with SSc. A key finding of our study is that there was no correlation between intake and either predicted or measured energy expenditure. Furthermore, although predicted and measured energy were strongly correlated, the actual values differed for individual patients. Therefore, where energy expenditure must be accurately measured in clinical practice (e.g., in patients with or at risk of malnutrition) or research, energy expenditure should be directly measured.

Our study highlights some important practical considerations about the energy assessment in patients with SSc. Older patients were more likely to report consuming more energy than they

expended (and vice versa). Previous research has suggested that this discrepancy could be related to differences in ability to estimate portion and food group size with increasing age (22,23). There was also a significant correlation between BMI and the discrepancy between predicted and measured energy expenditure and this was greater with higher BMIs. This is supported by a well-established link between BMI and under-reporting, with frequent under-reporters tending to have greater BMIs (24). Furthermore, as with other studies we did not find any significant correlation between energy intake and BMI (6).

Considering the systemic nature of the disease, patients with worse lung involvement took significantly fewer steps and spent more time sleeping. They also spent less time at >1.5METS, although this just approached statistical significance. This concurs with the finding of the previous study of reduced daily activity even with early lung involvement (10). Patients with cardiac involvement also took fewer steps, although this did not meet the predetermined level of statistical significance. However, it should be noted that none of the patients in our study were considered to have significant cardiac involvement. Patient-reported outcome measures are widely used in clinical practice and trials to assess the impact and severity of disease (25). Of interest, in our study, no association was seen between energy expenditure (activity) and patient-reported measures of function.

Unlike the previous study to utilise the SenseWear® Armband in 27 patients with SSc (10), we recorded a wide variation in active expenditures. Some patients performed very little activity (steps, time at >3METs). Whereas compared to the previous study, patients had a lower mean active expenditure (steps), but similar overall mean expenditure. However, patients in this study were older and age inversely correlated with activity. There were no associations between physical activity and BMI. However, in the previous study, negative correlations were noted physical activity and BMI, despite both studies having patient cohorts with comparable mean BMIs (10).

To-date, few nutritional studies involving patients with SSc have assessed or estimated energy expenditure. Previous studies comparing intakes to predictions have failed to detect differences between patients with and without disease–related malnutrition (8). Our study was underpowered to assess differences in expenditure between patients with and without recent

weight loss. However, recent research has examined predictors of weight loss in patients with SSc (26,27), and future research on this topic would benefit from measuring expenditures, rather than using predictions. Mean daily energies from total and saturated fat were 37% and 14% respectively, which is similar to the 37 to 39% total fat energy reported in a previous study of unselected patients with SSc (7).

There are a number of aspects to highlight about our study. Intakes were assessed by 3-day estimated dietary records and errors could have occurred. For example, reporting biases may occur with dietary documentation due to an increased awareness of one's own diet and thus, in order to improve the appearance of their diet, participants may over or underreport. A potential limitation, which should be considered when interpreting this study's results, was the patient cohort recruited. Participating patients may not have been representative of all patients due to unforeseen recruitment biases. There could have also been an element of patient self-selection as patients with less disease burden and/or greater dietary interest may have been more likely to enrol. However, the patient demographics and clinical phenotype including diseasesubsetting does suggest that our studied cohort was representative of SSc based on previous registry analyses (28). In addition, following recruitment, some of the more symptomatic patients were withdrawn due to the detection of clinical problems necessitating admission. Future research could assess energy expenditure comparisons with other features of SSc disease severity (e.g., vascular disease). The addition of a healthy control group could also yield important disease-related information. The impact of disease duration should also be considered in the design of future studies. Furthermore, economic evaluations (e.g., the cost of food) could be explored, including the impact of the loss of work productivity and financial income on food choices. Another practical limitation is that we were not able to determine whether oral intake translated into equal absorption of high energy substrates.

In summary, our study highlights the importance of measuring energy intake and expenditure, including measured expenditure rather than relying on predictive estimation, when accurate assessment is required. Energy assessment can provide novel insights into systemic involvement in SSc, including to aid the dietetic management of patients.

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DECLARATION OF COMPETING INTERESTS

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REFRENCES

- 1. Denton CP, Khanna DK. Systemic sclerosis. *Lancet*, 2017; 390: 1685–99.
- Harrison E, Herrick AL, McLaughlin JT, Lal S. Malnutrition in systemic sclerosis. *Rheumatology (Oxford)*, 2012; 51 :1747–56.
- 3. Shreiner AB, Murray C, Denton C, Khanna D. Gastrointestinal Manifestations of Systemic Sclerosis. *J scleroderma Relat Disord*, 2016; 1 :247–56.
- 4. Hughes M, Herrick AL. Systemic sclerosis. *Br J Hosp Med*, 2019; 80: 530–6.
- Steen VD, Medsger TA. Changes in causes of death in systemic sclerosis, 1972-2002. Ann Rheum Dis, 2007; 66 :940–4.
- 6. Marighela TF, Genaro P de S, Pinheiro MM, et al. Risk factors for body composition abnormalities in systemic sclerosis. *Clin Rheumatol*, 2013 ;32 : 1037–44.
- Lundberg AC, Akesson A, Akesson B. Dietary intake and nutritional status in patients with systemic sclerosis. *Ann Rheum Dis*, 1992 ;51 :1143–8.
- 8. Caporali R, Caccialanza R, Bonino C, et al. Disease-related malnutrition in outpatients with systemic sclerosis. *Clin Nutr*, 2012; 31(5):666–71.
- Herrick AL, Worthington H, Rieley F, et al. Dietary intake of micronutrient antioxidants in relation to blood levels in patients with systemic sclerosis. *J Rheumatol*, 1996 ; 23 :650–3.
- Battaglia S, Bellia M, Serafino-Agrusa L, et al. Physical capacity in performing daily activities is reduced in scleroderma patients with early lung involvement. *Clin Respir J*, 2017; 11: 36–42.
- 11. LeRoy EC, Black C, Fleischmajer R, et al. Scleroderma (systemic sclerosis): classification, subsets and pathogenesis. *J Rheumatol*, 1988; 15: 202–5.
- 12. Medsger TAJ, Silman AJ, Steen VD, et al. A disease severity scale for systemic sclerosis: development and testing. *J Rheumatol*, 1999; 26: 2159–67.
- 13. Fries JF, Spitz PW, Young DY. The dimensions of health outcomes: the health assessment questionnaire, disability and pain scales. *J Rheumatol*, 1982; 9: 789–93.
- Steen VD, Medsger TA. The value of the Health Assessment Questionnaire and special patient-generated scales to demonstrate change in systemic sclerosis patients over time. *Arthritis Rheum*, 1997; 40 :1984–91.
- 15. Bruce B, Fries JF. The Stanford Health Assessment Questionnaire: dimensions and

practical applications. Health Qual Life Outcomes, 2003; 1: 20.

- Stratton RJ, Hackston A, Longmore D, et al. Malnutrition in hospital outpatients and inpatients: prevalence, concurrent validity and ease of use of the "malnutrition universal screening tool" ('MUST') for adults. *Br J Nutr*, 2004; 92 : 799–808.
- 17. Nelson M, Haraldsdóttir J. Food photographs: practical guidelines II. Development and use of photographic atlases for assessing food portion size. *Public Health Nutr*, 1998; 1:231–7.
- 18. Crawley H, Mills A, Patel S. Food portion sizes. Second. London: HMSO; 1993.
- Paul AA, Southgate DAT. McCance and Widdowson's The Composition of Foods 4th Edition. 4th ed. HMSO, London.; 1978. (Medical Research Council (Great Britain). MRC special report. no. 297).
- Schofield WN. Predicting basal metabolic rate, new standards and review of previous work. *Hum Nutr Clin Nutr*, 1985; 39: 5–41.
- 21. Scheers T, Philippaerts R, Lefevre J. SenseWear-determined physical activity and sedentary behavior and metabolic syndrome. *Med Sci Sports Exerc*, 2013; 45: 481–9.
- 22. Nelson M, Atkinson M, Darbyshire S. Food photography II: use of food photographs for estimating portion size and the nutrient content of meals. *Br J Nutr*, 1996; 76: 31–49.
- 23. Timon CM, Forster SE, Barker ME, et al. A comparison of younger v. older adults' ability to estimate food portion sizes. *Proc Nutr Soc* 2011; 70: E51.
- Scagliusi FB, Ferriolli E, Pfrimer K, et al. Characteristics of women who frequently under report their energy intake: a doubly labelled water study. *Eur J Clin Nutr*, 2009; 63: 1192–9.
- Hughes M, Denton C. The critical need for patient-reported outcome measures to assess the severity and impact of systemic sclerosis. *Br J Dermatol*, 2021. doi: 10.1111/bjd.20777. [Online ahead of print].
- 26. Hvas CL, Harrison E, Eriksen MK, et al. Nutritional status and predictors of weight loss in patients with systemic sclerosis. *Clin Nutr ESPEN*, 2020; 40: 164–70.
- Hughes M, Heal C, Siegert E, et al. Significant weight loss in systemic sclerosis: a study from the EULAR Scleroderma Trials and Research (EUSTAR) database. *Ann Rheum Dis*, 2021; 79: 1123-1125.

 Meier FMP, Frommer KW, Dinser R, et al. Update on the profile of the EUSTAR cohort: an analysis of the EULAR Scleroderma Trials and Research group database. *Ann Rheum Dis*, 2012; 71: 1355-60

Details		Patients (n=36)				
Demographics						
	Median ±SD age (range)	57.9±12.2 years (32.3 - 72.9)				
	Male (%)	6 (17%)				
SSc Details						
Diffuse	dcSSc/lcSSc (%)	14 (39%)/22 (61%)				
cutaneous						
SSc						
	Median \pm SD interval from SSc onset	125±90 months (13 - 334)				
	(range)					
	Anti-topoisomerase-1 antibody (%)	7 (19%)				
	Anti-centromere antibody (%)	9 (25%)				
Nutritional Data						
	Mean BMI (range)	23.9±4kg/m ² (16.3 - 33.7)				
	Recent unintentional weight loss	5 (14%)				
	High 'MUST' (%)	4 (11%)				
	Medium 'MUST' (%)	4 (11%)				
	Low 'MUST' (%)	28 (77%)				
Clinical manifestations						
	Small bowel (%)	10 (28%)				
	Moderate to end-stage respiratory (%)	14 (39%)				
	Mild cardiac (%)	10 (28%)				
Functional scores						
	Mean HAQ-DI (0-3)	1.43±0.76 (0.0 - 3.0)				
	Mean lung SHAQ (0-3)	1.23±0.82 (0.1 - 2.9)				
	Mean global disability SHAQ (0-3)	1.48±0.79 (0.0 - 3.0)				

Table 1: Patient demographic and clinical data. Age and time presented as median \pm SD. All other data for continuous variables are presented as the mean \pm SD. Ranges are displayed in parentheses.

Predicted requirements					
All (kcal/day)	1930±265 (1453 - 2626)				
Men (kcal/day); n=6	2376±207 (2019-2626)				
Women (kcal/day); n=30	1840±164 (1452 - 2173)				
Armband data					
Time wearing					
Recording period (hh:mm)	71:08 (67:11 - 72:00)				
Time wearing (hh:mm)	69:02 (64:41 - 72:00)				
Total energy (kcal/day)	2027±476 (1221 - 3400)				
Corrected* total energy (kcal/day)	2075±481 (1283 - 3489)				
METs	1.4±0.3 (0.9 - 1.9)				
Time (min/day):					
1.1 to 2.9METs	365±194 (78 - 826)				
3.0 to 5.9METs	88±71 (2 - 291)				
>6.0METs	3±5 (0 – 19)				
Daily steps	4035±3288 (230 - 14148)				
Time (min/day) lying	491±113 (146 - 808)				
Time (min/day) sleeping	397±110 (128 – 672)				
Time (min/day) lying not asleep	93±46 (18 – 195)				
Dietary Assessment					
Energy (kcal/day)	1788±509 (958 – 3498)				
Protein (g/day)	71.3±18.9 (40.1 – 117.5)				
Total fat (g/day)	74.8±34.5 (27.5 – 196.3)				
Saturated fat (g/day)	28.4±12.5 (11.2 - 70.2)				
Carbohydrate (g/day)	212.1±53.4 (92.3 – 354.9)				

Table 2: Predicted and measured (armband data) energy requirements and energy intake.*Total energy corrected for time.

	Steps	Time >1.5	Time lying	Time sleeping			
		METS					
Demographics							
Age	s=-0.667; p<0.01	s=-0.390; p=0.02	s=-0.021; p=0.90	s=0.181; p=0.29			
BMI	s=0.009; p=0.96	s=-0.257; p=0.13	s=0.411; p=0.01	s=0.508; p<0.01			
Functional scores							
Total HAQ	s=-0.260; p=0.13	s=-0.10; p=0.57	s=0.17; p=0.32	s=0.26; p=0.13			
SHAQ	s=-0.276; p=0.10	s=-0.149; p=0.39	s=0.161; p=0.35	s=0.162; p=0.34			
global							
SHAQ lung	s=-0.291; p=0.09	s=-0.135; p=0.43	s=-0.04; p=0.98	s=-0.005; p=0.98			
Medsger Scores							
Cardiac	s=-0.313; p=0.07	s=-0.197; p=0.26	s=0.113; p=0.52	s=0.075; p=0.69			
Respiratory	s=-0.350; p=0.04	s=-0.296; p=0.08	s=0.333; p=0.05	s=0.185; p=0.28			

Table 3: Energy expenditure comparisons according to activity (Spearman's correlations).



Mean of expended and predicted expenditure (kcal/day)

Figure 1: Agreement plot of expended and predicted energies.



Figure 2: Difference between recorded energy expenditure and consumption against age.



Figure 3: Difference between expended minus consumed energy against BMI.