Nutrition therapy in patients with systemic sclerosis

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Case presentation

A 57 years-old male with weight loss of 20 kg within 10 months, fatigue, mild dysphagia, mild reflux, normal appetite, early satiety, no diarrhea, mild, but progressive dyspnea, chesty cough, no fever, pneumonia 2 months ago

Height: 178 cm, weight 57 kg, BMI 18.

What is the problem of this person with scleroderma?
What are the problems in SSc?

Systemic sclerosis is an autoimmune disease requiring more energy than healthy persons

- Cachexia, a result of a chronic inflammatory disease
  loss of muscle mass with stable fat mass

Patients with systemic sclerosis have problems with nutrition intake

- depressions with low appetite
- problem preparing meals
- money to buy healthy food
- low GI movements
- very rare malabsorption
- starvation (catabolism by pure energy deficiency)

Patients have accelerated aging due to disease or medications

- sarcopenia (age-related muscle loss in the context of dieting, physical immobility, or growth hormone deficiency)
A lot of patients have problems with their teeth: sicca syndrome, periodontosis
Upper gastrointestinal tract

Esophagus scintigraphy: delayed passage in the upper esophagus

Peritrast passage

Water melone stomach,

Reflux esophagitis

Our patient:
Caugh during investigation

JF, 03/06
our patient
Small intestinal bacterial overgrowth (SIBO) in systemic sclerosis

Marie I et al., Rheumatology 2009; 48:1314-1319

51 SSc patients were investigated
49% diffuse, 51% limited disease, 11 patients received immunosuppression
43.1% of the patients were SIBO +

<table>
<thead>
<tr>
<th>Parameter</th>
<th>SIBO +</th>
<th>SIBO -</th>
<th>P-values</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abdominal pain</td>
<td>86,4</td>
<td>31</td>
<td>0.0001</td>
</tr>
<tr>
<td>Diarrhoea</td>
<td>50</td>
<td>10,3</td>
<td>0.0034</td>
</tr>
<tr>
<td>Bloating</td>
<td>77,3</td>
<td>44,8</td>
<td>0.0246</td>
</tr>
<tr>
<td>Constipation</td>
<td>59,1</td>
<td>3,4</td>
<td>0.000001</td>
</tr>
<tr>
<td>Abd. tenderness</td>
<td>54,5</td>
<td>6,9</td>
<td>0.0027</td>
</tr>
</tbody>
</table>

administration of 50 g glucose in 250 ml sterile water
Positive test: H2/CH4> 20p.p.m above basal value, >10 p.p.m
in two consecutive measurements, and others
Small intestinal bacterial overgrowth in systemic sclerosis

Marie I et al., Rheumatology 2009; 48:1314-1319

ROC analysis
≥ 5 GI symptoms

Eradication with rotating antibiotics, 7 days/months, e.g. with 2 x 400 mg Norfloxacin/d, followed by 3 x 250 mg Metronidazol /d

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Tx +</th>
<th>Tx -</th>
<th>P-Wert</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abdominal Pain</td>
<td>27.2 %</td>
<td>90 %</td>
<td>0.008</td>
</tr>
<tr>
<td>Bloating</td>
<td>18.1 %</td>
<td>70 %</td>
<td>0.03</td>
</tr>
<tr>
<td>Diarrhoea</td>
<td>0 %</td>
<td>60 %</td>
<td>0.004</td>
</tr>
<tr>
<td>abdominal tenderness</td>
<td>9.1 %</td>
<td>50 %</td>
<td>0.06</td>
</tr>
</tbody>
</table>

Small intestine bacterial overgrowth is frequent and should be recognized
Vitamin D-Deficiency in patients with systemic sclerosis
Vacca-A et al., J. Rheumatol. 2009;36:1924-9

Detection of vitamin D between 09/07-05/08, PTH, Ca, P in 90 consecutive patients from France (north) and 66 Italy, some of these patients received vitamin D 800 IU/d

86% insufficiency 82% <30ng/ml
23% deficiency 32% < 10ng/ml

Assoziation of vitamin D deficiency with sPAP, ESR, CRP, Lung fibrosis, no associations with calzinoses, acroosteolyses
How to identify malnutrition?

There are different methods …

- Questionnaires: Malnutrition Universal Screening Tool score (a score analysing weight loss) other

- Body Mass Index kg/m²: often used for the decision to give nutrition therapy

- BioImpedance Analysis: analysis of the electrical resistance
Symptoms of malnutrition

- Unexplained 10% or more weight loss over a 3 months period
- Weakness and muscle wasting
- Excessive or new onset fatigue
- Increased susceptibility to infection
- Delayed wound healing
- Brittle nails and excessive hair loss
- Excessively dry and flaky skin
UCLA SCTC 2.0 – 5-minute screen and Rx plan

The UCLA SCTC GIT 2.0 Questionnaire

Your total GIT score is **0.89**. Your individual section scores are detailed in the table below and after each section in the questionnaire.

Click here for a printable version of your results.

<table>
<thead>
<tr>
<th>Category</th>
<th>Your Score</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Reflux</strong></td>
<td><strong>1.13</strong></td>
</tr>
<tr>
<td><strong>Dilation/Distention</strong></td>
<td><strong>0.5</strong></td>
</tr>
<tr>
<td><strong>Sedative</strong></td>
<td><strong>0</strong></td>
</tr>
<tr>
<td><strong>Diarrhea</strong></td>
<td><strong>1</strong></td>
</tr>
<tr>
<td><strong>Insomnia</strong></td>
<td><strong>0</strong></td>
</tr>
<tr>
<td><strong>Social Function</strong></td>
<td><strong>1.17</strong></td>
</tr>
<tr>
<td><strong>Emotional Well-Being</strong></td>
<td><strong>2.44</strong></td>
</tr>
<tr>
<td><strong>GIT Score</strong></td>
<td><strong>0.89</strong></td>
</tr>
</tbody>
</table>
UCLA SCTC 2.0 – 5-minute screen and treatment plan

- Reflux scale → anti-reflux and PPIs
- Distention/Bloating scale → Malabsorption or delayed gastric emptying
  - PLUS Diarrhea → trial of antibiotics +- promotility agents
  - No Diarrhea → trial of promotility agents
- Constipation → stimulant laxatives, good bowel regimen
- Fecal soilage → refer to colorectal surgeon
- “Out of proportion” emotional symptoms → Irritable Bowel Syndrome

A useful approach: [www.thenuttynutritionist.com](http://www.thenuttynutritionist.com)
[linakaminski@thenuttnutritionist.com](mailto:linakaminski@thenuttnutritionist.com)

As suggested by Denish Kahan
Why it malnutrition and why it is important to treat?

**Definition:**
- Malnutrition is a deficiency of energy, protein, or other nutrition elements including vitamins leading to changes of the body compositions.
- Consequently, malnutrition does not mean absence of fat, fat people also can be malnourished.

**why treatment?**
- Energy deficiency can counteract with defense mechanisms of the body to receive a healthy status.
- Protein and energy deficiency can impair healing e.g. of digital ulcers.
Both people could have the same weight and the same energy uptake.

Energy uptake without exercise

Energy uptake with exercise
Bioelektrische Impedanzanalyse (BIA):

- fast
- reproducible
- non invasive
- easy to use

Sensitivity to change, high reliability

Method is used in oncology, gastroenterology, nephrology
**Nutrition status measured by phase angle values compared to age-and sex-matched persons**

<table>
<thead>
<tr>
<th>Age (years)</th>
<th>18-29</th>
<th>30-59</th>
<th>60-69</th>
<th>&gt; 70</th>
<th>all patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Good Nutrition status</td>
<td>28.6</td>
<td>41.2</td>
<td>57.1</td>
<td>35.7</td>
<td>44.4</td>
</tr>
<tr>
<td>Abnormal nutrition status</td>
<td>71.4</td>
<td>58.8</td>
<td>42.9</td>
<td>64.3</td>
<td>55.6</td>
</tr>
</tbody>
</table>

In addition: 50% of the patients revealed energy deficiency, in 20% of the patients energy uptake lower than basal metabolism.
## Associations between nutrition status and SSc features

<table>
<thead>
<tr>
<th></th>
<th>PhA ≥ 5 good nutrition status</th>
<th>PhA 4.9-4.0 bad nutrition status</th>
<th>PhA &lt; 3.9 insufficient nutrition status</th>
<th>significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>ESR</td>
<td>22.39 ± 20.87</td>
<td>20.90 ± 15.44</td>
<td>49.39 ± 34.80</td>
<td>p &lt; .001 ***</td>
</tr>
<tr>
<td>FVC values</td>
<td>93.46 ± 17.78</td>
<td>89.12 ± 16.23</td>
<td>73.53 ± 16.77</td>
<td>p &lt; .001 ***</td>
</tr>
<tr>
<td>modified Rodnan skin score (MRSS)</td>
<td>7.49 ± 7.49</td>
<td>8.33 ± 7.30</td>
<td>14.25 ± 11.43</td>
<td>p = .009 **</td>
</tr>
<tr>
<td>cardiac involvement</td>
<td>31.9%</td>
<td>33.3%</td>
<td>75.0%</td>
<td>p = .002 **</td>
</tr>
<tr>
<td>presence of muscle atrophy</td>
<td>46.7%</td>
<td>50.0%</td>
<td>75.0%</td>
<td>p = .037 *</td>
</tr>
<tr>
<td>presence of muscle weakness</td>
<td>45.7%</td>
<td>57.9%</td>
<td>75.0%</td>
<td>p = .018 *</td>
</tr>
<tr>
<td>disease duration</td>
<td>5.16 ± 5.64</td>
<td>7.26 ± 6.07</td>
<td>3.84 ± 3.58</td>
<td>p = .041 *</td>
</tr>
<tr>
<td>age</td>
<td>53.00 ± 10.45</td>
<td>55.21 ± 15.99</td>
<td>55.44 ± 13.91</td>
<td>p = .631</td>
</tr>
<tr>
<td>BMI</td>
<td>23.36 ± 3.39</td>
<td>23.08 ± 4.30</td>
<td>23.34 ± 6.81</td>
<td>p = .953</td>
</tr>
<tr>
<td>presence of digital ulcers</td>
<td>25.5%</td>
<td>38.5%</td>
<td>42.1%</td>
<td>p = .092</td>
</tr>
<tr>
<td>nausea</td>
<td>34.8%</td>
<td>35.1%</td>
<td>35.0%</td>
<td>p = .542</td>
</tr>
<tr>
<td>reflux</td>
<td>63.0%</td>
<td>63.2%</td>
<td>75.0%</td>
<td>p = .250</td>
</tr>
<tr>
<td>constipation</td>
<td>28.3%</td>
<td>34.2%</td>
<td>20.0%</td>
<td>p = .391</td>
</tr>
</tbody>
</table>
Cumulative survival months after first BIA years after first diagnosis

Cumulative survival months after first BIA

Cumulative survival months after last BIA

Cumulative survival months after first BIA

BMI
How to improve malnutrition in patients with SSc?

- There are no guidelines in SSc patients, no special diet
- There are different reasons for malnutrition
- Nutrition is part of the life and of the individual education
- Nutrition is a lot of habit and religion
- Patients differently tolerate foods
- General recommendations such as to eat more fruits and vegetables are often not helpful
- You need knowledge, time (and sometimes money)
What do we already know from other diseases as well as from SSc?

Patients with SSc have a 10-fold increased risk for arteriosclerosis

one glass alcohol/day is useful

chocolate is useful, at least 100g/week

smoking is harmful, stop it

Creatine 0.15g/kg daily can increase muscle function

most SSc patients have not increased cholesterol levels, fat is rich of energy and vitamin (EDEKA)
General nutrition recommendations according to ESPEN guidelines

This is for healthy people, not for diseased people
Nutrition and exercise, not only „healthy food“

This is something for freaks and artists
Our approach to address malnutrition

There are international (ESPEN) S3 guidelines for enteral nutrition therapy in malnourished patients using a step-wise approach.

1. First step: dietary counselling (DC) by a nutrition expert based on the individual nutrition, energy and protein deficiency, and body composition assessed by BIA.
2. Regular food will be improved to reach normal energy and protein uptake (1.2-1.5 g/kg body weight/d, protein uptake, 35 kcal/kg body weight/d) and to improve PhA values above the 10th percentile of age-, sex-, and BMI-matched controls.
3. If no significant changes in PhA values are reached, patients will receive oral high-caloric and protein-rich nutritional supplements (ONS) 200ml once or more often a day as needed with 1.3-1.5 kcal/ml and 10g/protein/100 ml.
4. If no improvement, thinking about parenteral nutrition
Summary

Recognition of malnutrition and its reasons is crucial in SSc

A lot of patients have pure energy deficiency and the disease requires energy, too

Best method to identify malnutrition in Bioimpedance analysis

Malnutrition is common, but can only been treated in an individual approach

A useful approach: www.thenuttynutritionist.com
lindakaminski@thenuttinutritionist.com