Long-term results and quality of life after surgery for oesophageal achalasia: one surgeon's experience
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Long-term results and quality of life after surgery for oesophageal achalasia: one surgeon’s experience

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Abstract

Objective: To assess the long-term results and health-related quality of life in patients undergoing surgery for oesophageal achalasia.

Methods: Thirty-nine patients with achalasia (25 males, mean age 42 ± 13 years) underwent open-Heller myotomy and Dor fundoplication by the same surgeon. Long-term results were assessed by imaging, endoscopy, manometry, pH-metry, and Short Form 36 and Nottingham Health Profile questionnaires whenever applicable. Six patients were at radiological stage I, 20 were at stage II, 2 at stage III and 11 at stage IV. Dysphagia and regurgitation were scored according to the four-grade classification (1 = no symptoms; 4 = persistent symptoms). Pulmonary symptoms were present in six patients. Lower oesophageal pressure was 30 ± 34 mmHg. Six patients had undergone previous pneumatodilation.

Results: No procedure related mortality was recorded. Mean follow-up was 107 ± 30 months (54–177). Preoperative to 5-year postoperative mean decrement in lower oesophageal sphincter pressure was 12.3 ± 8.3 mmHg (P < 0.001), in oesophageal width was 11.5 ± 7.1 mm (P < 0.001), in dysphagia 1.8 ± 0.8 (P < 0.01) and regurgitation 1.4 ± 0.7 (P < 0.01). Four patients presented relapse dysphagia and two of those were re-operated upon. Three patients developed acid reflux, which was satisfactorily treated with proton-pump inhibitors. Preoperative to 4-year postoperative quality of life scores were available for 20 patients. Questionnaires showed a significant improvement (P < 0.001) especially in all psychosocial domains, which were correlated with postoperative dysphagia score and lower oesophageal sphincter pressure.

Conclusion: Heller myotomy and Dor fundoplication is a safe and effective procedure that improves symptoms, functional status and especially psychosocial aspect of quality of life in the long term in oesophageal achalasia.

Keywords: Oesophagus; Achalasia; Oesophagomyotomy; Fundoplication

1. Introduction

Oesophageal achalasia is a rare motility disorder characterized by aperistalsis or uncoordinated contractions of the oesophageal body and impaired relaxation of a frequently hypertensive lower oesophageal sphincter [1], due to irreversible degeneration of myoenteric plexus neurons. Since no currently available treatment can restore physiological oesophageal motility, the rationale for treatment is aimed at relieving symptoms by reducing the pressure of the lower sphincter [2]. Alternative treatment modalities for oesophageal achalasia, including surgical and non-surgical options, have been proposed [2–6]. So far, non-surgical options are preferred mainly due to better patients acceptance, avoidance of hospitalization and presumed minor side effects. Nevertheless, the risk of decreasing sphincter tone may often results in a gastro-oesophageal reflux and this has led to the addition of an anti-reflux technique [7].

Since the beginning of our experience on achalasia, we have added anterior partial fundoplication according to the Dor technique [8] in addition to Heller myotomy. We have gathered consistent data on patients followed for a long-term period.

The aim of this study is to analyse the long-term outcome of Heller myotomy and anterior fundoplication performed by the same surgeon as the first choice surgical treatment in patients with oesophageal achalasia. The results were evaluated according to objective and subjective
measurements using validated quality of life (QOL) questionnaires for the latter.

2. Materials and methods

Between 1987 and 1998, 39 patients with oesophageal achalasia underwent Heller myotomy and anterior fundoplication by the same surgeon (TCM) at our institution. The features and surgical data of the patients are summarized in Table 1. The diagnosis was made on the basis of clinical history, oesophagograms, endoscopy and manometry. Mean duration of symptoms was 30 ± 34 months. Six patients had respiratory symptoms including recurrent pneumonia in five patients and asthma attacks in one. Eight patients had undergone a previous pneumatic dilation.

The stage of the disease was established by evaluating the degree of oesophageal body dilation expressed in centimetres by measuring the maximum oesophageal width from standard posteroanterior projection oesophagograms. As a result, achalasia was classified in four stages: stage I, oesophageal width ≤ 3 cm (N = 6); stage II, width between 3 and 6 cm (N = 20); stage III, width > 6 cm (N = 2); stage IV, width > 6 cm and sigmoid-shaped oesophagus (N = 11).

Dysphagia, regurgitation, chest pain and heartburn were classified preoperatively and postoperatively into four grades: grade 1 = no symptoms; grade 2 = mild symptoms occurring less than once weekly; grade 3 = moderate symptoms occurring more than once weekly but not every day; grade 4 = severe and persistent symptoms. The patients’ satisfaction with the operation was assessed by asking them to choose one of the five possible responses: none = 1, poor = 2, medium = 3, much = 4, total = 5.

In addition, since 1995, QOL was pre- and postoperatively assessed in all patients by means of the Nottingham Health Profile (NHP) [9] and Short-Form-36-Items (SF-36) [10]. We chose these questionnaires because both QOL tests were re-proposed to the patients yearly.

2.1. Surgical Technique

In the present study group the procedure was performed through a xiphoumbilical laparotomy incision. Laparoscopic procedures, which now represent the first choice approach in our department, were anyway excluded from the study due to the lack of long-term results for a consistent sample size. After visualization of the oesophagogastric junction, the phrenoesophageal membrane was incised anterolaterally using a cautery and only the anterior aspects of the oesophagus were separated bluntly from the diaphragmatic crura. The vagus nerves were identified.

Oesophageal dissection was completed in the mediastinum 5–6 cm above the oesophagogastric junction by pulling the lower oesophagus through the hiatus. Oesophageal myotomy was carried out on the central-left anterior wall of the oesophagus, starting from the midpoint

### Table 1

<p>| Preoperative characteristics of the study group classified according to the radiological stage |</p>
<table>
<thead>
<tr>
<th>I (N = 6)</th>
<th>II (N = 20)</th>
<th>III (N = 2)</th>
<th>IV (N = 11)</th>
<th>Total (N = 39)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (years)</td>
<td>40 ± 14</td>
<td>41 ± 14</td>
<td>50 ± 13</td>
<td>43 ± 10</td>
</tr>
<tr>
<td>Sex (male/female)</td>
<td>4:2</td>
<td>13:7</td>
<td>2:0</td>
<td>6:5</td>
</tr>
<tr>
<td>Symptoms duration (months)</td>
<td>13 ± 9</td>
<td>18 ± 12</td>
<td>53 ± 11</td>
<td>56 ± 53</td>
</tr>
<tr>
<td>Max oesophageal width (cm)</td>
<td>3.9 ± 0.7</td>
<td>4.7 ± 0.5</td>
<td>6.5 ± 0.7</td>
<td>7.1 ± 0.8</td>
</tr>
<tr>
<td>LOS pressure (mmHg)a</td>
<td>25 ± 5</td>
<td>33 ± 10</td>
<td>33 ± 10</td>
<td>37.8 ± 10</td>
</tr>
<tr>
<td>Dysphagia score (1–4)</td>
<td>3.2 ± 0.4</td>
<td>3.7 ± 0.5</td>
<td>3.5 ± 0.7</td>
<td>3.7 ± 0.5</td>
</tr>
<tr>
<td>Regurgitation score (1–4)</td>
<td>1.7 ± 0.8</td>
<td>2.1 ± 0.8</td>
<td>1.5 ± 0.7</td>
<td>4.0 ± 0</td>
</tr>
<tr>
<td>Chest pain score (1–4)</td>
<td>1.3 ± 0.8</td>
<td>1.4 ± 0.7</td>
<td>1.0 ± 0</td>
<td>1.2 ± 0.4</td>
</tr>
<tr>
<td>Heartburn (1–4)</td>
<td>1.0 ± 0</td>
<td>1.2 ± 0.4</td>
<td>1.0 ± 0</td>
<td>1.1 ± 0.3</td>
</tr>
<tr>
<td>Respiratory symptoms (n)</td>
<td>–</td>
<td>1</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>Preoperative pneumatic dilation (n)</td>
<td>1</td>
<td>4</td>
<td>–</td>
<td>3</td>
</tr>
<tr>
<td>Weight loss (kg)</td>
<td>0.7 ± 1.0</td>
<td>1.0 ± 1.3</td>
<td>–</td>
<td>1.6 ± 2.0</td>
</tr>
</tbody>
</table>

a LOS, lower oesophageal sphincter. Available for 33 patients.
and completing muscle dissection 6 cm upwards and at least 3 cm downwards onto the stomach. During oesophageal myotomy the longitudinal and circular fibres were divided by using cautery and scissors. An anterior Dor fundoplication was then performed suturing the anterior wall of the gastric fundus to the edges of the myotomy with three non-absorbable stitches on each side, with the upper stitch including the diaphragmatic crus. A nasogastric tube was left in the stomach.

2.2. Statistical analysis

Group descriptive statistics are presented as the mean ± SD. The Wilcoxon or the Mann–Whitney tests were used for paired and unpaired data, respectively. Internal consistency of the two questionnaires was evaluated for groups and for global scores and specific domains by the Alpha Cronbach’s test: the highest score documented from 1 to 5 and 10 years (Table 2). Interestingly, no correlation existed between the residual dysphagia score and oesophageal width increment.

Oesophageal pH manometry was repeated yearly for the first 5 years and data were available in 33 patients. None showed significant reflux even in the presence of symptoms. Lower oesophageal sphincter pressure decreased significantly from 33 ± 10 to 19.1 ± 5.9 mmHg at 1 year. Postoperative pressure decrement was significant at all stages, but although greater in the advanced stages it was not correlated with the stage. Mild and non-significant increments were recorded at 5 years (20.3 ± 5.7) and correlated with dysphagia ($\rho = -0.66$, $P = 0.001$) and regurgitation ($\rho = -0.40$, $P = 0.04$) scores. At 5 years as many as 16 patients presented pressure lower than 20 mmHg. In the nine patients assessed at 10 years, pressure was still significantly lower (23.4 ± 6.7, $P < 0.001$) than the basal value.

Oesophagoscopy was routinely performed postoperatively at 1 year even in the absence of symptoms. Afterwards, oesophagoscopy was performed yearly in all patients at stage III and IV and in the presence of heartburn. Grade I oesophagitis (according to Savary–Miller) was diagnosed in three symptomatic patients at 9, 13 and 33 months belonging to stage III, IV and I, respectively. In these patients gastro-oesophageal reflux was confirmed by 24-h pH-metry.

We carried out multiple biopsies of the oesophageal mucosa in only 10 patients at stage III and IV after a mean of 26 months; only one showed mild squamous dysplasia. No patient eventually required oesophagectomy or developed oesophageal carcinoma.

3. Results

3.1. Objective assessment

There was no perioperative mortality or major morbidity. Mean hospital stay was 7.2 ± 2.2 days. A regular diet was always resumed after an oesophagogram confirmed the absence of leaks at the myotomy site. Mean follow-up was 107 ± 30 months and ranged from 54 months to 177 months. Data are available for 37 patients at 5 years and 12 patients at 10 years (Table 2).

All 15 patients who had loss weight returned at least to their ideal body weight within a maximum period of 12 months after the operation and remained stable for the following years. In the first year mean oesophageal width decreased from 5.3 ± 1.4 to 3.7 ± 1.4 cm ($P = 0.0001$); pressure reduced by 10 mm or more in 29 patients and by 20–30 mm in 14 patients, while in five there was no change. The decrement in oesophageal width was statistically significant at all radiological stages, but it was not correlated with the above-mentioned stages, or with the duration of the symptoms. Repeated postoperative X-ray oesophagograms were performed in all patients yearly for 5 years. A non-significant increment of the oesophageal width was documented from 1 to 5 and 10 years (Table 2).

Postoperative pressure decrement was significant at all stages, but although greater in the advanced stages it was not correlated with the stage. Mild and non-significant increments were recorded at 5 years (20.3 ± 5.7) and correlated with dysphagia ($\rho = -0.66$, $P = 0.001$) and regurgitation ($\rho = -0.40$, $P = 0.04$) scores. At 5 years as many as 16 patients presented pressure lower than 20 mmHg. In the nine patients assessed at 10 years, pressure was still significantly lower (23.4 ± 6.7, $P < 0.001$) than the basal value.

Mean dysphagia score assessed at 1 year decreased significantly from 3.6 ± 0.5 to 1.3 ± 0.7 and remained stable at 5 and 10 years; regurgitation showed the same trend (Table 2). Radiological stage IV presented the worst mean values, although they were not significant.

Four patients re-presented with dysphagia at 48 months for the patient at stage III, 56 months at stage IV, 71 months at stage II and 88 months at stage I. In the last three the discomfort was as intense as preoperatively. Two were re-operated (I and II stage) and one underwent periodical dilations (III). The patient at stage IV was satisfied and refused any further procedure. We were able to dismount the Dor plasty in the two re-operated patients and redo it. All these patients are well at 34 and 16 months after the second procedure. According to the re-operative findings the reason for failure was attributed to progressive inflammatory stricture.

Chest pain and heartburn mean scores did not significantly increase. Significant heartburn appeared in the three above-mentioned patients with endoscopic evidence of grade I oesophagitis, which was satisfactorily treated with proton-pump inhibitors. Fig. 1 shows the Kaplan–Meier curve for dysphagia relapse and heartburn development.
with a different pattern characterized by the earlier onset of the latter.

At 5 years the procedure result was scored as total in 12 (32%) of the operated patients, high in 15 (41%), medium in 5 (14%), poor in 1 (2%) and none in 4 (11%). At 10 years the score was total in 2 (16%), high in 5 (42%) and medium in 5 (42%).

3.3. Quality of life assessment

Analysis was possible in 20 subjects for a 4-year period; in the earlier period of the study, questionnaires were not available. Internal consistency was higher for general NHP scores at 1 and 4 years (0.91–0.92) than SF-36 (0.84–0.88); also the rate of answered questions was higher for NHP
baseline NHP and SF-36 domains scores are reported in Table 3. Fig. 2 shows a greater impairment of the psychosocial domains than physical ones when matched with a large sample size population of the same age. At postoperative assessment both physical and mainly psychosocial domains improved (data not shown) and remained stable up to 4 years, approximating values scored by a normal population of the same age. In NHP domains a significant long-lasting improvement was found in energy \( (P < 0.01) \), social isolation \( (P < 0.001) \) and emotional reactions \( (P < 0.01) \). Also in many SF-36 domains we observed a significant 4-year improvement: general health \( (P < 0.01) \), social function \( (P < 0.001) \), vitality \( (P < 0.001) \), mental health \( (P < 0.01) \), physical role \( (P < 0.05) \), and emotional role \( (P < 0.05) \).

No significant difference was found when comparing the earlier and more advanced stages (I–II versus III–IV).

Four-year postoperative dysphagia score was inversely correlated with postoperative changes in social isolation \( (\rho = 0.41, P = 0.04) \), physical role \( (\rho = -0.42, P = 0.04) \) and mental health \( (\rho = -0.45, P = 0.02) \) domains, while postoperative lower oesophageal sphincter pressure was inversely correlated with changes in social isolation \( (\rho = 0.40, P = 0.04) \), emotional reactions \( (\rho = 0.43, P = 0.03) \), physical role \( (\rho = -0.52, P = 0.01) \), emotional role \( (\rho = -0.42, P = 0.03) \) and mental health \( (\rho = -0.52, P = 0.01) \).

4. Discussion

Normal motility in the oesophageal achalasia cannot be restored with the medical or surgical therapies currently available. The rationale for care is symptomatic treatment by decrement of lower oesophageal sphincter pressure. In the short term, both pneumatic dilation and surgical Heller cardiomyotomy can relieve dysphagia in most patients [1–4]. However, for longer observations, only 26–49% of the patients, who underwent dilation [1,4,11–13] and selected for surgery,

![Fig. 1. Kaplan–Meier curve for postoperative symptom-free survival. Dysphagia and heartburn present a different pattern with earlier onset of the latter.](image)

Table 3
Baseline and positive changes in quality of life scores according to the radiological stage

<table>
<thead>
<tr>
<th>Stage</th>
<th>I–II ( (N = 10) )</th>
<th>III–IV ( (N = 10) )</th>
<th>Total ( (N = 20) )</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Baseline</td>
<td>4-year change</td>
<td>Baseline</td>
</tr>
<tr>
<td>NHP domain</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Motility</td>
<td>11 ± 6</td>
<td>5 ± 9*</td>
<td>22 ± 10</td>
</tr>
<tr>
<td>Energy</td>
<td>13 ± 18</td>
<td>8 ± 15**</td>
<td>23 ± 10</td>
</tr>
<tr>
<td>Sleep</td>
<td>3 ± 6</td>
<td>1 ± 12</td>
<td>19 ± 11</td>
</tr>
<tr>
<td>Pain</td>
<td>10 ± 12</td>
<td>3 ± 10</td>
<td>18 ± 12</td>
</tr>
<tr>
<td>Isolation</td>
<td>23 ± 26</td>
<td>15 ± 18***</td>
<td>26 ± 17</td>
</tr>
<tr>
<td>Emotion</td>
<td>16 ± 14</td>
<td>11 ± 15***</td>
<td>18 ± 9</td>
</tr>
<tr>
<td>SF-36 domain*</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>PF</td>
<td>85 ± 10</td>
<td>5 ± 6</td>
<td>84 ± 14</td>
</tr>
<tr>
<td>RP</td>
<td>63 ± 18</td>
<td>20 ± 15**</td>
<td>61 ± 17</td>
</tr>
<tr>
<td>BP</td>
<td>87 ± 11</td>
<td>6 ± 10</td>
<td>75 ± 16</td>
</tr>
<tr>
<td>GH</td>
<td>61 ± 20</td>
<td>11 ± 11**</td>
<td>63 ± 9</td>
</tr>
<tr>
<td>SF</td>
<td>56 ± 19</td>
<td>17 ± 7**</td>
<td>50 ± 7</td>
</tr>
<tr>
<td>VT</td>
<td>60 ± 11</td>
<td>16 ± 3***</td>
<td>53 ± 14</td>
</tr>
<tr>
<td>RE</td>
<td>74 ± 15</td>
<td>12 ± 17**</td>
<td>74 ± 23</td>
</tr>
<tr>
<td>MH</td>
<td>72 ± 12</td>
<td>12 ± 11**</td>
<td>63 ± 14</td>
</tr>
</tbody>
</table>

PF, physical functioning; RP, role physical; BP, bodily pain; GH, general health; SF, social functioning; VT, vitality; RE, role emotional; MH, mental health. Intragroup Wilcoxon test significance *\( P < 0.05 \); **\( P < 0.01 \); ***\( P < 0.001 \).

* Available for 12 patients.
33–79% after surgical myotomy [4,11–15] are completely free from dysphagia. Dilation or myotomy may very likely facilitate a gastro-esophageal reflux that can be better faced by prophylactic surgical anti-reflux procedure than chronic consumption of proton-pump inhibitors. Heller’s oesophagomyotomy relieves dysphagia but does not restore oesophageal peristalsis. According to this rationale the addition of a too tight fundoplication may jeopardize the relief induced by myotomy and predispose to relapse achalasia. Comparison between series with no fundoplication, partial or total fundoplication presented different results [16–20]. The automatic relationship between myotomy and reflux has been recently denied by Sharp et al. [16] are now conducting a randomized trial to compare the effective need for a Dor fundoplication with a Heller myotomy. On the contrary, Ponce et al. [17] found a relatively frequent presence of reflux despite open myotomy and Dor fundoplication. Falkenback et al. [18], in a recent prospective randomized study compared results with and without floppy Nissen fundoplication and found a significantly lower 24-h reflux in the latter group without adding dysphagia. Oelschlager et al. [19] compared anterior Dor and posterior Toupet fundoplication associated with standard and extended myotomy, respectively: sphincter pressure and dysphagia were both less frequent and less severe after the second combination, thus improving the results of surgical therapy for achalasia for dysphagia without increasing the rate of abnormal gastro-oesophageal reflux. Finally, Yau et al. [20], in an experimental model, found that 90° wrap is as effective as total fundoplication restoring adequate competence to the gastro-oesophageal junction in the early postoperative period.

In our opinion, partial fundoplication can prevent reflux after myotomy. This procedure represents a good compromise between the need to prevent otherwise inevitable reflux without re-creating an obstacle too important for an aperistaltic oesophagus. We have experience with the Dor procedure that we associate with an extended myotomy with good long-term results.

The difficulty in evaluating the effectiveness of each surgical technique depends also on the subjective criteria of evaluation and mix of symptoms not always correlated with dysphagia or with reflux [16]. Objective data (i.e. oesophagus size and manometry pressure of lower oesophageal sphincter) may largely change from one patient to another and can be difficult to interpret.

The advent of QOL questionnaires gave a more reliable and comparable outcome assessment measure. To our knowledge, this is the first time that NHP has been used to assess QOL in oesophageal achalasia. On the contrary, the SF-36 questionnaire has recently been used in some small surgical series [17,21–23]. Ben-Meir and colleagues [21] evaluated 19 patients 21 months after surgery and found that physical function, bodily pain, vitality, and social function were significantly improved. Luketich and colleagues [22] studied 53 patients and found that after myotomy, all eight domains of SF-36 data scored at least equal or better than the normal US population used for comparison. However, in this series, no preoperative data were available for pre- to postoperative comparisons. These results were analogous with those obtained by Ponce et al. [17] in 28 Spanish-speaking patients who underwent open Heller myotomy plus Dor fundoplication. Katilius et al. [23] studied 15 patients after laparoscopic and eight after laparotomy myotomy. The SF-36 general health domain improved significantly in both groups although best scores in physical function and bodily pain were achieved following laparoscopic myotomy. Postoperative QOL assessment revealed no difference with the general population. Our findings support the use of the SF-36 as a reliable instrument for QOL assessment in achalasia patients. We have found greater postoperative improvements in the domains such as role limitations due to physical problems and social function. Correlation with degree of dysphagia and pressure of lower oesophageal sphincter seems not surprising since the existence of symptoms and signs leading to modification of eating
habits has also been shown to impair QOL in other benign oesophageal diseases [24].

Findings recently reported by Decker et al. [25] demonstrating significant improvements in a specific gastrointestinal QOL questionnaire (not yet available in Italian validated translation) in patients with early and advanced stage achalasia seem in accordance with ours.

We acknowledge some limitations of our study: sample size, incomplete QOL score, long time-frame, but all these difficulties arise from the relative rarity of the disease and the difficulty in gathering a greater number of patients. One supplementary bias might be represented by the difficulty in assessing objectively reflux on the basis of symptoms, pH-manometry and oesophagoscopy; periodic routine 24-h pH-metry was introduced in 2001, thus long-term data for the study population are unavailable.

On the contrary, the merits of the study can be represented by the introduction of a QOL questionnaire never used in this disease and by the homogeneous technique in selection, operating procedure and follow up of the patients.

In this study, the Heller myotomy coupled with Dor fundoplication proved highly effective in improving objective measures, subjective symptoms and QOL especially in psychosocial domains in patients with any stage of achalasic oesophagus. Such results can also be observed in the long-term thus leading to the conclusion that Heller myotomy should be considered as a first choice treatment option for oesophageal achalasia.

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References


Appendix A. Conference discussion

Dr G. Decker (Luxembourg, Luxembourg): You have a more than 5-year follow-up and there are only 6 or 7 published series with a longer follow-up than that. However, the two series, with a more than 10-year follow-up, the one from Pearson and the one from Ellis, show that after 5–10 years the results significantly deteriorate. So I would rather suggest...
that you call this intermediate-term results rather than long-term results. We have also studied the quality of life issue in achalasia and this was published in the Annals of Surgery. We used another instrument, named the Gastrointestinal Quality of Life Index. We found that in those patients who preoperatively had very low scores, they improved very much, whereas those who had almost normal scores preoperatively, these patients, at the slightest complication or when they had reflux postoperatively, those were the ones who had the scores going down at follow-up. Did you observe similar findings?

Dr. Ambrogi: Yes, we have, but this is part of another study we are trying to set up. I didn’t understand the questionnaire you use. Do you use the Gastrointestinal Quality of Life?

Dr. Decke: Yes.

Dr. Ambrogi: Unfortunately, we have this questionnaire, but unfortunately there is not a validated version for Italian-speaking people.

Dr. Decke: I don’t know for Italian, but there is a validated questionnaire for many other languages, Dutch, English, German and so on.

Dr. Ambrogi: Of course.
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