Perianal Crohn Disease

A New Scoring System to Evaluate and Predict Outcome of Surgical Intervention

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Background: Conventional Crohn disease activity indices do not reflect perianal disease activity or allow prognostic implications from surgery.

Hypothesis: A new scoring system, based on the patient’s disease history and physical examination findings, will allow more accurate use of surgical intervention for perianal Crohn disease.

Methods: A standardized scoring questionnaire was developed and applied to a consecutive group of patients before surgical treatment of perianal Crohn disease. The scoring system included abscess, fistula, ulcer and fissure, stenosis, incontinence, and concomitant disease. Weighted factors included acuity vs chronicity, de novo vs recurrent disease, and concomitant intestinal disease. The scoring system was validated against the surgical outcome, which was classified as poor, satisfactory, or good.

Results: Twenty-eight patients with Crohn disease who underwent 33 surgical procedures had a mean score of 16.5 (range, 3-37; possible range, 0-55). Using the Spearman nonparametric correlation test, the scoring system was accurate in predicting the outcome of surgical intervention (correlation coefficient, 0.78, 95% confidence interval, 0.57-0.89; P<.001) at mean follow-up of 20.8 months (range, 6-40 months). Correlation was further validated using a linear regression model (r=0.75, slope best-fit value, 3.8; 95% confidence interval, 2.46-5.14; P<.001). All patients with a score of 10 or less had a good outcome, whereas all those with a score of 20 or greater had a poor outcome.

Conclusions: The proposed scoring system correlated well with the short-term outcome of surgical intervention in patients with perianal Crohn disease and allowed prediction of surgical success. Ultimately, it may be possible to alter therapy based on preoperative prediction of the expected postoperative outcome.

Arch Surg. 2002;137:774-777

Symptomatic perianal disease requiring therapy occurs in 30% to 40% of patients with Crohn disease (CD) and is often associated with colonic inflammation. Perianal disease includes cutaneous manifestations such as tags and ulcerations; anal canal lesions such as fissures, ulcers, and stenoses; and the septic sequelae of abscesses and/or fistulas.

See Invited Critique at end of article

Conventional disease activity indices, such as the Crohn’s Disease Activity Index and the simple Bradshaw Activity Index, measure intestinal and extraintestinal manifestations of the disease. However, these indices do not reflect the severity of perianal CD (PCD) or offer any prognostic information about the expected course after surgical treatment of these conditions. The PCD activity index developed by Irvine was designed to assess the response to medical management and the overall well-being of the patient, excluding any anatomical or surgical considerations. The inherent problem with any scoring system for any disease is that the ease of its use is often inversely related to its clinical accuracy. “User-friendly” systems may not include enough data to allow an accurate assessment. Conversely, complex systems are labor intensive but may allow better prognostication. These latter scoring systems can be cumbersome because of the necessity to obtain the results of multiple ancillary studies. The “ideal” scoring system should be simple and accurate. Therefore, the aim of this study was to develop a facile but accurate objective scoring system, based only on the patient’s history and physical examination findings, to grade the severity of PCD and to help predict postoperative outcome.
MATERIALS AND METHODS

SCORING SYSTEM

A standardized scoring system (Perianal Crohn’s Disease Activity Index [PCDAI]) was developed to address 6 readily identifiable frequent features of PCD: abscess, fistula, fissure and/or ulcer, stenosis, incontinence, and concomitant disease (Table 1). Each feature was rated on a point scale according to severity and complexity. A score of zero indicates the absence of that feature. Uniformly, a score of 1 represented de novo acute disease; 2, chronic disease; and 3, recurrent disease. An incontinence score of 1 to 6 was graded as 1 point, 7 to 14 as 3 points, and more than 14 as 5 points. Concomitant disease in the rectum, colon, and small bowel was graded as none, moderate, or severe. Because of its effect on perianal disease, rectal disease received a higher score than colonic and small-bowel disease. Overall, a patient without PCD would have a score of zero, and a patient with the most advanced disease would have a score of 35.

SURGICAL INTERVENTION

Specific surgical intervention for PCD was decided on by the staff surgeon based on conventional history and physical examination findings and, when clinically indicated, results of additional evaluations. These ancillary studies included endoanal ultrasound, fistulography, manometry, sigmoidoscopy, colonoscopy, histopathologic examination, computed tomography, and intra-anal coil magnetic resonance imaging. All patients were evaluated at routine intervals as determined by the surgeon. Specifically, all patients had a minimum of 3 office visits: 4 weeks, 3 months, and 6 months after surgery. Staff surgeons were not familiar with the PCDAI, mean scores, or correlation of scores with outcome of surgery until completion of the study. However, subjective success was measured by the surgeon and recorded in the patient’s medical record at each follow-up visit. To facilitate validity testing of the PCDAI, outcome was ranked as good (complete resolution of the primary problem), moderate (partial resolution with a reduction in persistence of symptoms and subjective patient satisfaction), or poor (no resolution of the primary pathologic condition or associated symptoms). To further assess correlation between the new scoring system and surgical treatment, procedures were divided into corrective (an attempt was made to cure the specific perianal disease) and conservative (perianal sepsis was treated but no attempt was made to cure the perianal pathology).

STATISTICAL ANALYSIS

To determine whether the PCDAI was measuring outcome after surgical intervention, validity testing was performed. Validity was assessed using the nonparametric (Spearman) correlation of the PCDAI score with subjective assessment of surgical outcome, as data did not show a normal distribution. Spearman correlation is based on ranking the 2 variables, and thus no assumption was made regarding the distribution of the values. To further determine whether the scoring system was accurate in predicting surgical outcome, a linear regression analysis was subsequently performed. Both tests were performed using Instat 3 for Windows (GraphPad, San Diego, Calif). P<.05 was considered sufficient to reject the null hypothesis. The correlation coefficient r ranges from –1.0 to 1.0. An r value of 0 indicates that the 2 variables do not correlate at all. A positive fraction indicates that the 2 variables increase or decrease together, 1.0 being perfect correlation. A negative fraction indicates that the 2 variables change inversely.

RESULTS

Twenty-eight consecutive patients with CD (14 women and 14 men; mean age, 43.3 years; age range, 21-74 years) were assessed between January 1, 1997, and December 31, 1999. All patients had at least 1 feature of active PCD: 26 fistulas and/or abscesses, 4 rectovaginal fistulas, 2 anal stenoses, and 1 rectourethral fistula. Thirty-three surgical procedures were performed (Table 2). Mean follow-up was 20.8 months (range, 6-40 months). Overall, the outcome of surgery was considered to be good in 11 patients (39%), moderate in 3 (11%), and poor in 14 (50%) (Figure 1). In the good outcome group, the mean ± SD PCDAI score was 6.5±2.44 (range, 3-10), whereas in the moderate and poor groups, these values were 17.0±6.42 (range, 15-20) and 24.3±2.84 (range, 16-37), respectively. There was no correlation between length of follow-up and outcome (Spearman rank correlation, r=0.1; 95% confidence interval [CI], −0.28 to 0.46; P=.59). A similar correlation was found in the subgroup of patients undergoing corrective procedures. In this subgroup, outcome was considered good in 7 patients (44%), moderate in 1 (6%), and poor in 8 (50%) (Figure 2). Mean PCDAI scores were 5.1 (range, 3-8) in the good outcome group and 16.9 (range, 14-22) in the poor outcome group. The single patient with a moderate outcome had a score of 15.0.

A PCDAI score of 15 or less correlated well with a good surgical outcome, whereas a score of 20 or greater accurately predicted failure of surgical intervention. Although 11 patients with a PCDAI score of 10 or less had a good outcome, all 12 patients with a score of 20 or greater had a poor outcome. All 5 patients with PCDAI scores between 10 and 20 had either moderate or poor outcomes. Using the Spearman nonparametric correlation test, the PCDAI score correlated with the outcome of surgical intervention (correlation coefficient, 0.78; 95% CI, 0.57-0.89; P<.001). The validity was further supported using a linear regression model. Outcome was correlated with the scoring system (r=0.75; slope best-fit value, 3.8; 95% CI, 2.46-5.14; P<.001). Similar findings were found when evaluating only patients undergoing corrective procedures (Spearman correlation coefficient, 0.85; 95% CI, 0.62-0.93; P<.001; linear regression model, r=0.92, 95% CI, 0.77-0.97; P<.001).

COMMENT

Giant cells on histologic examination of tissue from patients with perianal fistulas without evidence of tuber-
culosis were first described in 1921 by Gabriel.11 In retrospect, this case report may have been the first description of PCD. The first description of regional enteritis by Crohn et al12 made no mention of perianal manifestations. Bis-

sell,13 in 1934, was the first researcher to emphasize the high incidence of fistula in anoanal and perianal ab-
scesses in patients with small-bowel CD. It was another 3 decades before surgeons recognized that PCD could be the only presenting symptom, often preceding the intes-
tinal manifestations.14

Symptomatic PCD occurs in 30% to 40% of pa-
tients with CD, and it is strongly associated with co-
lonic and rectal involvement.15,16 Exacerbation of PCD
may occur as a consequence of increased intestinal in-
flammation, but it is also isolated from the intestinal dis-
ective activity activity. As noted by Garrett and Drossman,17 ac-
tivity indices currently used for CD do not adequately assess perianal manifestations. The Crohn’s Disease Ac-
tivity Index considers fissure, fistula, or abscess as a single feature, which increases the total score by 20 points; an increase of 100 points is required to denote a “signifi-
cant” increase in disease activity. The Crohn’s Disease Activity Index does not consider these perianal manifes-
tations as distinct and diverse entities. Similarly, it does not address recurrence, persistence, or disease dura-
tion.5

The only activity index directed at PCD currently
available is that of Irvine.9 This scoring system is com-
posed of 5 features—discharge, pain and activity restric-
tions, sexual activity, degree of induration, and type of

| Table 1. Perianal Crohn’s Disease Activity Index |
| Feature | Score |
| Abscess | None or first occurrence, single abscess or first occurrence, multiple abscesses or first recurrence, single or multiple abscesses or multiple recurrence, single or multiple abscesses maximum abscess score 8 |
| Fistula | None 0 short-term (<30 d) fistula or long-term (>30 d) fistula or persistent postsurgery fistula or recurrent fistula multiple fistulas rectovaginal/rectourethral fistula or recurrent rectovaginal/rectourethral fistula maximum fistula score 14 |
| Ulcer and Fissure | None 0 short-term (<30 d) ulcer/fissure or long-term (>30 d) ulcer/fissure or single ulcer/fissure or multiple ulcers/fissures maximum ulcer/fissure score 4 |
| Stenosis | None 0 short-term (<30 d) stenosis or long-term (>30 d) stenosis or recurrent stenosis maximum stenosis score 6 |
| Incontinence Score | None 0 incontinence score of 1-6 or incontinence score of 7-14 or incontinence score >14 maximum incontinence score 5 |
| Concomitant Disease* | None 0 moderate 3, 2, 1 severe 4, 3, 2 active fistula 4, 3, 2 maximum concomitant disease score 18 |

*Scores are for rectal, colonic, and small-bowel disease, respectively.

| Table 2. Corrective and Conservative Surgical Procedures Performed in 28 Patients With Crohn Disease |
| Procedure | Corrective | Conservative |
| Advancement flap | 7 | Abscess drainage |
| Fistulotomy | 5 | Seton placement |
| Anoplasty | 2 | Subtotal |
| Gracilis interposition | 2 | 17 |
| Fibrin glue instillation | 1 | |

![Figure 1. Correlation between Perianal Crohn’s Disease Activity Index (PCDAI) score and outcome in all 28 patients undergoing surgical intervention. A score of less than 15 correlated well with a good outcome.](chart1.png)

![Figure 2. Correlation between Perianal Crohn’s Disease Activity Index (PCDAI) score and outcome in a subgroup of 17 patients undergoing corrective procedures.](chart2.png)
perianal disease. Only the last feature has potential implications regarding outcome after surgical intervention. However, type of perianal disease is only 20% of the total score; thus, the Irvine score, although useful in evaluating patient satisfaction after conservative medical management, is not a prognostic tool when considering surgical intervention.

In 1980, Buchmann and Alexander-Williams classified the clinical features of PCD into 3 categories: (1) skin lesions, including maceration, erosion, ulceration, abscess, and skin tags; (2) anal canal lesions, including fissure, ulcer, and stenosis; and (3) fistulas, including low (anal canal to skin), high (rectum to skin), and recto-vaginal. This classification is useful for portraying the anatomical features of lesions, but it does not measure the severity or duration of perianal disease, which affects clinical handling.

The proposed clinical index of PCD was tested and was able to determine the short-term outcome of surgical intervention. The index consists of 6 items that are easily evaluated during physical examination and history taking; no ancillary studies are required. The correlation between the PCDAI score and the surgeon’s subjective assessment of outcome after surgery was high, illustrating its validity. This correlation was repeated when evaluating only patients who underwent corrective procedures. This pilot study has some limitations, mainly a small number of patients and relatively short follow-up. Further testing in a larger group of patients is required. We believe, however, that some conclusions can still be drawn. In cases where the score is 20 or greater, recurrence or persistence of symptoms after surgery can be anticipated. Thus, in these patients, conservative measures to control perianal sepsis may be preferable to more aggressive corrective procedures. Failure of these more advanced procedures may lead to sepsis, stenosis, or incontinence and to recurrence of disease. In turn, these adverse sequelae may necessitate a stoma with or without proctectomy. The ability to preoperatively identify patients who can potentially be spared this consequence of failed surgery for PCD is enticing. Patients with scores between 10 and 20 have variable outcomes and could be counseled accordingly.

The PCDAI seems to correlate well with short-term outcome after surgical intervention for PCD. This scoring system should be further prospectively tested as it may serve as an important prognostic tool in patients with PCD.

This study was supported in part by a grant from the David G. Jagelman Research Fund of Cleveland Clinic Florida, Weston.

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REFERENCES

Anorectal problems afflict one fifth of all patients with Crohn disease. Some series report 80% to 90% surgical success rates in selected patients,1-4 but postoperative complications substantially impair some patients.5-7 Thus, patient selection is critical in deciding whether to be aggressive or to perform conservative procedures such as simple drainage or seton placement. Pikarksy and coauthors propose that a simple score might predict surgical outcome. Prospectively collected data and an a priori scoring system yielded a dichotomous distribution that correlated with postoperative outcome when the results were analyzed retrospectively. The study group was small and heterogeneous regarding surgical indications and procedures. Furthermore, half of the patients underwent "conservative procedures" and therefore do not help test the hypothesis that these scores predict whether an aggressive procedure is likely to be successful. (Although potentially corrective, fibrin glue instillation does not carry the same level of postoperative risk as the more aggressive procedures described.) Concomitant medical therapy for Crohn disease is not discussed but is of particular interest because anti–tumor necrosis factor α antibody facilitates closure of some fistulas. Outcomes were categorized subjectively by surgeons, not by patients or by objective criteria. Nevertheless, half of the patients selected clinically for more aggressive procedures had poor outcomes that might have been predicted by their scores. Conversely, 6 of 16 patients selected for conservative procedures had low scores. We do not know whether these patients would have profited from more aggressive procedures or whether the score simply supplements clinical intuition. Because the proposed score thresholds were derived retrospectively, they should be validated prospectively in a larger series, and their superior predictive value to clinical intuition and other scoring systems should be tested. If this testing succeeds, this score may prove useful in reporting and comparing published clinical trials or in cueing surgeons that certain patients might or might not profit from aggressive therapy despite contrary clinical intuition.

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