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ORIGINAL CONTRIBUTIONS

Rectal Carcinoids: The Most Frequent Carcinoid Tumor

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One hundred seventy patients with gastrointestinal carcinoid tumors were treated at Ochsner Clinic from 1958 to 1990. Ninety-four rectal carcinoid tumors were diagnosed and treated during this time. Carcinoid tumors of the rectum represented the most frequent primary site (55 percent), followed by carcinoids of the ileum (12 percent), appendix (12 percent), colon (6 percent), stomach (6 percent), jejunum (2 percent), pancreas (2 percent), and other (5 percent). One-half of rectal carcinoids were discovered during anorectal examination of asymptomatic patients. The remainder were found primarily by examination of patients for symptoms of benign anorectal conditions. The diagnosis of rectal carcinoid was made at the time of initial examination in 61 patients. This allowed definitive treatment in a single session by local excision and fulguration in 48 patients. The remainder were treated by repeat biopsy and fulguration (25 patients) or by transanal excision (12 patients). Overall, 85 carcinoid tumors of the rectum measuring <2 cm were treated by local excision and fulguration or by transanal excision, with an average five-year follow-up. There were no local recurrences. Ten patients with metastasizing rectal carcinoids averaging 4 cm were treated. All were symptomatic at presentation and fared poorly despite radical surgery. Three were alive at three years but only one survived five years. At our institution, rectal carcinoids were the most frequently detected carcinoid tumor. Small carcinoids of the rectum were adequately treated by local excision and fulguration or by transanal excision, with no local recurrence. The true incidence of rectal carcinoids is detected only with careful and complete rectal examination of the asymptomatic screening population by experienced surgeons. With more widespread screening of the well population, rectal carcinoids may become recognized as the most frequent human carcinoid tumor. [Key words: Carcinoid; Rectum; Tumor; Gastrointestinal]

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Within the past three decades, increasing numbers of rectal carcinoids have been reported.[1–4] This trend has come about as complete anorectal examination including proctosigmoidoscopy has become more widespread.

Careful, complete digital and proctosigmoidoscopic examination of 8,000 patients annually led to diagnosis of 94 rectal carcinoids at the Ochsner Clinic from 1958 to 1990. Analysis of this experience with 94 patients forms the basis of this report.

PATIENTS AND METHODS

Charts of all patients with a diagnosis of carcinoid tumor (all sites) treated at the Ochsner Clinic from 1958 to 1990 were retrospectively reviewed. Record was made of the primary tumor site. Cases of rectal carcinoid were segregated and the following were noted: symptoms and clinical findings, with attention to the surgeon's descriptive terminology of appearance and feel of the tumor, tumor size, color, location within the rectum, and initial diagnostic impression. Associated malignancy, if any, was noted. Concurrent benign anorectal conditions were recorded. A pathologic diagnosis of rectal carcinoid by Ochsner Clinic pathologists was required for inclusion. Pathologic diagnosis was

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made from standard hematoxylin and eosin preparations and characteristic histopathology. All treatments and results of treatment, including recurrent disease and survival, were recorded. Follow-up visits were noted.

RESULTS

One hundred seventy patients with a diagnosis of gastrointestinal carcinoid were treated from 1958 to 1990. This included ninety-four cases of rectal carcinoid, constituting 55 percent of all gastrointestinal carcinoid tumors.

Organ distribution of the primary gastrointestinal carcinoids was rectal carcinoid, 95 (55 percent), followed by carcinoids of the ileum, 21 (12 percent); appendix, 20 (12 percent); colon, 10 (6 percent); stomach, 10 (6 percent); jejunum, 4 (2 percent); pancreas, 4 (2 percent), and other or unknown primary, 8 (5 percent) (Table 1).

Mean age at time of diagnosis for all patients with rectal carcinoids was 52 years. There were 59 men and 35 women (1.7:1). Peak distribution by age was in the fifth, sixth, and seventh decades.

Forty-seven patients (50 percent) were asymptomatic, and diagnosis was made by routine screening digital and proctosigmoidoscopic examination. When symptoms were present, the most common were bleeding (18 percent), constipation (17 percent), rectal pain (7 percent), and pruritus ani (3 percent). However, concurrent benign anorectal conditions present included hemorrhoids, 56 patients (60 percent); anal fissure, 4 patients (4 percent); anal fistula, 2 patients; and proctalgia fugax, 1 patient. These common associated disorders were responsible for the symptoms in every instance when the rectal carcinoid was of the small, nonmetastasizing variety.

Table 1.
Organ Distribution of Gastrointestinal Carcinoids

Organ	No.	%
Rectum	94	55.3
Ileum	21	12.4
Appendix	20	11.8
Colon	10	5.9
Stomach	10	5.9
Unknown abdominal primary with metastases	5	2.9
Jejunum	4	2.4
Pancreas	4	2.4
Small bowel, unspecified	1	0.5
Duodenum	1	0.5
Total	170	100

Ten patients with metastasizing, biologically aggressive rectal carcinoids all had symptoms, ranging from rectal pain, decreased stool caliber, and constipation to low back pain, testicular pain, dysuria, perineal fullness, and weight loss.

The surgeon's examining finger and the rigid or flexible proctosigmoidoscope were the diagnostic tools in all cases. Digital palpation yielded descriptions of submucosal (53 percent), firm (43 percent), nodular (35 percent), mobile (21 percent), hard (18 percent), smooth (14 percent), and rubbery (7 percent). Proctoscopy led to visual depictions of yellow, (56 percent), submucosal (53 percent), polypoid (39 percent), sessile (31 percent), white (6 percent), umbilicated (4 percent), and pedunculated (3 percent).

Tumor location was exactly recorded in 80 cases. Mean distance from the anal verge was 8 cm. Eight (10 percent) were between the anal verge and 5 cm. The majority (52, or 65 percent) were in the mid-rectum between 5 and 10 cm. Thus, three-fourths were within reach of the examining finger. Twenty (25 percent) were located between 10 and 15 cm. The tumors were equally distributed between anterior, posterior, and lateral rectal walls.

The diagnosis of rectal carcinoid was made at the time of initial examination in 61 of 84 (73 percent) of patients with small, nonmetastasizing tumors. In 42 (50 percent), the specific diagnosis of rectal carcinoid was made by the characteristic feel and appearance of the tumor, whereas in 19 (22 percent), an initial differential diagnosis of rectal carcinoid vs. leiomyoma, adenoma, carcinoma, or polyp was formed.

A specific diagnosis of rectal carcinoid was more elusive at initial examination in the 10 patients who presented with symptoms of biologically aggressive, metastasizing lesions. For these generally larger, more advanced tumors, the differential diagnosis included adenocarcinoma of the rectum and cloacogenic carcinoma. In only two of 10 cases was rectal carcinoid included in the differential. Biopsy and tissue diagnosis were required to define the nature of these tumors.

Of 94 rectal carcinoids, 84 were biologically nonaggressive, nonmetastasizing tumors and 10 were aggressive, metastasizing tumors. Treatment of these two groups is considered separately.

Treatment (Nonmetastasizing Tumors)

Eighty-four nonmetastasizing rectal carcinoids were treated (Fig. 1) with mean follow-up of 63.5

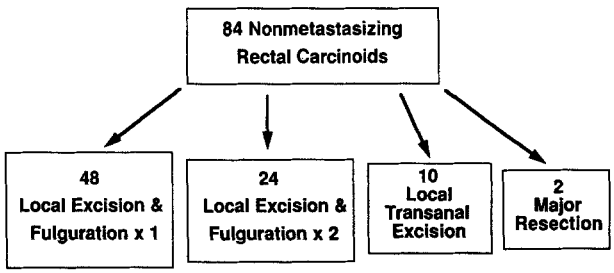

Figure 1. Treatment of nonmetastasizing rectal carcinoids.

months (range 0–248 months) and an average of 3.8 return visits (range 0–16 visits). Mean tumor diameter was 0.5 cm.

Forty-four tumors were treated by local excision with biopsy forceps and thorough, wide coagulation of the excised tumor base through a rigid proctoscope. Treatment was completed in a single session at the time of initial diagnosis with no repeat treatments. Tumors in this group had a mean diameter of 0.68 cm. Included were nine 1.0-cm tumors, one of 1.5 cm, and one measuring 2.0 cm. The patients with the 1.5-cm and 2.0-cm tumors were followed 121 months and 181 months, respectively. There were no recurrences.

Four patients with small carcinoids averaging 0.4 cm were treated in a single session by local excision with biopsy forceps alone, without fulguration. There were no recurrences. In all, there was no recurrence in 48 patients who had definitive treatment in a single session at the initial examination by local biopsy forceps excision with or without fulguration.

Among this group was one patient with 0.5-cm yellow submucosal, rectal nodule who was scheduled at initial examination to return for local excision and fulguration. The patient failed to keep the appointment and returned nine years later with an unchanged clinical examination (by the same examiner). The 0.5-cm rectal carcinoid, which had not grown in the nine-year interval, was locally excised and the base fulgurated, without recurrence.

Twenty-four patients were treated in two sessions by combinations of local excision with biopsy forceps and fulguration. These tumors averaged 0.72 cm in diameter (range 0.2–1.5 cm).

Seven of the 24 were treated first by forceps biopsy alone, then returned for either fulguration of the excised base (five patients) or repeat biopsy with fulguration (two patients). Both repeat biopsies were negative for residual tumor. There were no recurrences.

Seventeen of the 24 had local excision and fulguration followed by *repeat* fulguration (11 patients) or by biopsy with fulguration (six patients) within weeks of the initial treatment. One of six biopsies showed a fragment of residual carcinoid, which was treated by fulguration in the second session. In this group, there were no further treatments and no recurrences.

Ten patients were treated in the operating room by transanal procedures. These tumors averaged 1.0 cm (0.3–2.0 cm), and half were suspected preoperatively of being rectal adenocarcinomas.

Four of the 10 had local transanal excision and fulguration of small tumors (0.4–1.0 cm) in the operating room after initial office treatment. There were no recurrences.

Five of the 10 underwent formal transanal excision with suture closure. These tumors measured 1.0 to 2.0 cm. There were no recurrences in these five patients with follow-up of 72 months.

One of the 10 patients had a 0.3-cm rectal carcinoid as an incidental finding within a transanally excised villous adenoma of the rectum.

Two patients underwent major operative resections. In one, a small rectal carcinoid was an incidental finding in the upper rectum of a specimen from low anterior resection for rectal cancer.

One patient had a pull-through procedure for a sessile 1.75-cm rectal lesion at 8 cm, which on clinical examination was infiltrative and judged to be a rectal cancer. The operative specimen revealed a rectal carcinoid confined to the submucosa, without muscle invasion. The patient was alive and free of disease 20 years later.

Considered as a group, local recurrence was not observed among the 84 patients with nonmetastasizing rectal carcinoids treated by the various modalities.

A single complication resulted from treatment. A young male bled from the rectum after repeat fulguration of the excised base of a 1.0-cm tumor. The patient was hospitalized and observed with a 1-unit blood loss. No transfusions were needed and bleeding stopped spontaneously.

Treatment (Metastasizing Tumors)

Ten patients with metastasizing rectal carcinoids were treated (Table 2). Mean age was 53.4 years. All had symptoms, 60 percent with vague, deep-seated pain of the rectum, perineum, lower abdomen, lower back, or urogenital tract. These were symptoms of advanced disease at time of presen-

Table 2.
Clinical Course of Metastasizing Rectal Carcinoids

Age	Sex	Symptoms	Clinical Description	Tumor Size	Stage at Diagnosis	Treatment	Result
48	M	Severe rectal pain for 2–3 yr	Smooth, rubbery, tender presacral mass	10 cm	10/20 lymph node +; no liver metastasis	APR	Liver metastasis at 27 months; alive at follow-up to 56 months
45	F	Yes, but unknown: referral	Unknown: referral	Unknown	Regional lymph node +	APR with pelvic x-ray therapy	Liver and diffuse metastasis at 20 months. Dead of disease at 44 months (metastasis to liver, lungs, heart, adrenal, kidney, bone)
32	F	Perineal "fullness"	Hard, fixed ulcerated irregular	6 cm	Regional lymph node +; ovarian metastasis; no liver metastasis	Preoperative x-ray therapy and APR	Alive at follow-up to 17 months
66	F	Yes, but unknown: referral	Sessile, umbilicated	1 cm	2/20 lymph node + (synchronous cloacogenic carcinoma)	APR	No follow-up (synchronous diagnosis of cloacogenic carcinoma)
55	M	Low back and iliac pain	Friable	Unknown	Regional lymph node +; liver metastasis; metastatic to bone marrow	APR with pelvic x-ray therapy	Dead of disease at 14 months
62	M	Constipation	Firm, mobile, central ulceration	4 cm	3/14 lymph node +; no liver metastasis	APR	Alive without recurrence at follow-up to 17 months
59	M	Rectal bleed, constipation, decreased stool caliber	Irregular, fungating	4.5 cm	1/12 lymph node +; liver metastasis	LAR and fluorodesoxyuridine via hepatic artery	Alive at follow-up to 34 months
59	M	Testicular and low abdominal pain	Firm, exophytic, broad-based	2 cm	Metastases to sigmoid mesentery and retroperitoneum	Exploratory laparotomy and biopsy (unresectable, rectal fulguration)	Alive at follow-up to 24 months
62	M	Weight loss, right upper quadrant liver fullness	Infiltrative firm, sessile central ulceration	4 cm	Liver metastasis (massive)	Palliative rectal fulguration, actinomycin D	Alive at follow-up 27 months
46	M	Back pain for 8–9 months, constipation	Hard, mobile, central ulceration	1 cm	Localized to rectum (no operative staging)	Local excision and fulguration twice	Liver metastasis at 37 months follow-up. No local recurrence; 5-fluorouracil/methotrexate/cytoxan for metastatic disease

APR = abdominoperineal resection. LAR = low anterior resection.

tation, proven in 9 of 10 patients (eight with lymph node metastases, three with liver metastases). A possible exception was one patient who presented with a 1.0-cm mobile rectal carcinoid with central umbilication. The patient had some complaint of lower back pain. Local therapy alone was performed, so the stage of disease at presentation is not known. This patient developed liver metastases at 37 months.

Treatment consisted of major resection in seven patients (six abdominoperineal resections, one low anterior resection). Two patients were deemed unresectable due to locally advanced disease or hepatic metastases and had palliative rectal fulguration. The single patient with a 1.0-cm lesion was treated by local excision and fulguration in two sessions.

Complete follow-up was available for only two patients, as most returned to the care of referring physicians. Median follow-up was 25.5 months (0–56 months). Among seven patients who underwent major resection, two died of disease and only one was disease-free at last follow-up (17 months). Two patients treated by palliative fulguration were alive at 24 and 27 months, respectively. The single patient with a 1.0-cm tumor treated by local excision and fulguration developed liver metastases at 37 months without local recurrence.

Overall known survival, based on limited follow-up of nine patients, was 29 months from time of diagnosis (range 5–56 months). Liver metastases appeared in three patients an average of 28 months after initial treatment. Adjuvant x-ray therapy (three patients) and chemotherapy (two patients) were used without observed effect. Clinical courses of the 10 patients with metastasizing tumors is recorded in Table 2.

Twelve of 94 patients (12.8 percent) had another cancer diagnosed either prior to or within a mean five-year follow-up period after diagnosis of rectal carcinoid (Table 3). Four patients (4.2 percent) had associated colon or rectal adenocarcinoma. None of the 94 patients had ulcerative colitis.

Colonoscopy was not performed frequently until the later years of this series, so data concerning the incidence of proximal colon polyps in patients with rectal carcinoid are unavailable.

Urine for 5-HIAA was not routinely submitted. Seventeen specimens in 11 patients with rectal carcinoid, including three with liver metastases, were all negative for 5-HIAA. No patient displayed the carcinoid syndrome.

No instance of multiple or multicentric rectal carcinoid was found in this series of 94 patients.

Size of rectal carcinoid was examined with regard to metastatic behavior (Table 4). Of 56 tumors of less than 1 cm, none metastasized. A single 1.0-cm carcinoid among 20 tumors of at least 1.0 cm but less than 1.5 cm metastasized. Five tumors between 1.5 and 1.9 cm were all nonmetastasizing. Of three tumors measuring 2 cm, one metastasized. All of five very large tumors (4–10 cm) were metastatic at initial presentation.

DISCUSSION

The classic 1959 description by Hanley *et al.*[1] of rectal carcinoids and their treatment leaves scant room for improvement. Since the report by Hanley *et al.* of 26 cases, an additional 94 cases have accrued at Ochsner Clinic, some of which were included in a prior report.[5] This forms the largest experience since Caldarola and colleagues[3] reported 133 cases in 1964.

Carcinoids of the rectum are unique among carcinoids of the gastrointestinal tract, as Hanley[1] observed, since a preoperative diagnosis may readily be made early in the disease. The accessibility of the rectum to two simple tools—the examining

Table 3.
Rectal Carcinoid-Associated Cancers*

Cancer	No.
Breast cancer	2
Lung cancer	2
Malignant lymphoma	1
Malignant melanoma	1
Squamous cell carcinoma of vulva	1
Cloacogenic carcinoma	1
Adenocarcinoma of colon†	1
Adenocarcinoma of rectum†	1
Carcinoma *in situ* (rectal polyp)†	1
Carcinoma *in situ* (colon polyp)†	1

* Data for 12 of 94 patients, or 12.8%.
† Associated colorectal adenocarcinoma, in 4 of 94 patients, or 4.2%.

Table 4.
Size* of Rectal Carcinoid *vs.* Metastatic Behavior

Size	No. of Tumors	No. Metastasizing
<1.0 cm	56	0
1.0 to 1.4 cm	20	1
1.5 to 1.9 cm	5	0
2.0 cm	3	1
>2.0 cm	5	5

* Size data available for 89 tumors.

finger and the proctoscope—make this possible. These remain the best diagnostic tools.

Rectal carcinoids have a characteristic feel and appearance which permit diagnosis and treatment at the initial examination. Contrary to the recent survey of Burke *et al.*,[6] in which only one of 35 rectal carcinoids was described as "yellow" and just three as "submucosal," the majority in our series were described as yellow (56 percent) and/or submucosal (53 percent).

Proper recognition of the typical feel and appearance of these tumors leads to correct diagnosis and treatment. Just one of 35 cases of Burke *et al.*[6] was diagnosed preoperatively, whereas 73 percent (61 of 84) in the present series were diagnosed at initial examination, which lead to definitive treatment in a single session for 57 percent (48 of 84) of those with nonmetastasizing rectal carcinoids.

We have found that definitive treatment is achievable at the first examination by recognition of the distinctive yellow, submucosal appearance of these tumors along with their very firm or hard, discrete, smooth, mobile feel on digital examination. Treatment of rectal carcinoids may be dictated to a degree, but not absolutely, by their size. Asymptomatic, nonulcerated tumors of less than 1 cm rarely metastasize and are best treated by local biopsy forceps excision and wide, thorough fulguration of the excision base in one or two sessions. This method of treating small rectal carcinoids was first championed by Hanley *et al.*[1] over 30 years ago and has successfully withstood the test of time.

Although trends in office practice have the flexible fiberoptic sigmoidoscope supplanting the rigid proctoscope for routine examination, rectal carcinoids are best diagnosed and treated via the traditional instrument. An advantage of the rigid proctoscope is more complete air insufflation and expansion of the rectum. This stretches the rectal mucosa over the firm submucosal tumor and augments its typical appearance.

Treatment, consisting of biopsy forceps excision and fulguration, is best achieved through the rigid instrument. The firm, rubbery tumor is best grasped and fulgurated with rigid forceps directed through the wide channel of the proctoscope. When a rectal carcinoid is seen in our office on routine flexible fiberoptic sigmoidoscopy, the flexible scope is removed and a rigid proctoscope inserted for treatment.

Local recurrence was never observed in 72 patients treated in this fashion over a 32-year period. Where local recurrence has been reported by others due to incomplete initial excision,[6,7] repeat local excision still resulted in complete cure.

For larger tumors measuring 1 to 2 cm, local transanal excision in the operating room is the preferred treatment. This approach yields a better specimen for the pathologist, with attention to invasion of the muscularis propria. Among 10 tumors so treated in this series there were no recurrences.

We have found that with complete local excision of the tumor, by whatever means, local recurrence is not a problem, whether the tumor is of the nonmetastasizing or metastasizing variety. This experience agrees with another recent report.[8] The true difficulties come with identifying those rectal carcinoids that are at greatest risk to metastasize and determining how best to treat patients with such tumors.

No single criterion, other than the documented presence of existing metastases, has allowed prediction of the biologic behavior of these enigmatic tumors. Size is the most often used guide and is reliably predictive for tumors under 1 cm (which rarely metastasize) and for tumors over 2 cm (which nearly always are metastatic at the time the patient presents). For tumors of intermediate size (1–2 cm), size is not as predictive. This report includes one case of a 1-cm tumor which metastasized, and other reports exist of small rectal carcinoids with metastatic behavior.[2,4] The risk of a tumor smaller than 1 cm metastasizing is estimated at 3 percent or less.[7] A rectal carcinoid between 1 and 1.9 cm carries an 11 percent risk of metastasizing, as estimated by Naunheim *et al.*[7] This figure closely agrees with the risk seen in the present series (2 of 28 tumors 1 to 2 cm metastasized, or seven percent) and with the 13 percent rate (3 of 28 cases) seen by Sauven *et al.*[8] It may thus be seen that 2 cm, often quoted as the cutoff point to discriminate between innocuous and aggressive tumors, is not a magic number. All carcinoids are malignant tumors, and patients should be informed of this. Reassurance of the miniscule risk with small lesions should accompany extended anorectal surveillance for these patients.

Clinical observations may be as valuable as tumor size in judging the nature of a rectal carcinoid. The presence of symptoms is an ominous finding. Sauven *et al.*[8] demonstrated a highly significant de-

crease in survival for symptomatic patients when compared with asymptomatic patients. Mucosal involvement, manifesting as central ulceration or umbilication, is also an ominous feature and was present in 4 of 10 patients in this series with metastasizing tumors. Central umbilication was seen in three nonmetastasizing tumors in this series, but frank ulceration was present only in lesions which metastasized. Any rectal carcinoid, despite its size, should be viewed with increased suspicion if the patient has symptoms which could be related to the tumor or if there is mucosal involvement.

The presence of muscle invasion has also been used as an index of tumor aggressiveness. For tumors of intermediate size (1–1.9 cm), Naunheim *et al.*[9] found that 46 percent of lesions with invasion of the muscularis propria metastasized.[7] Based on this, radical surgery was proposed for such patients. However, since rectal carcinoids by nature incite an intense desmoplastic reaction and are locally infiltrative, muscle invasion is difficult to interpret and use of this as a criterion for radical surgery may result in unneeded major resection.

Sauven *et al.*[8] at the Memorial Sloan-Kettering Cancer Center have recently challenged the notion that patients with biologically aggressive rectal carcinoids should undergo radical surgery. This stance is based on an experience with 16 patients treated by radical surgery. Despite such operation, all of their patients with tumors penetrating to the subserosa or beyond (T3 or T4), or with involved lymph nodes, or with tumors larger than 2 cm, eventually died of metastatic disease. These authors suggest that radical surgery does not cure patients with advanced locoregional disease (deeply penetrating tumor or involved lymph nodes), and that local excision may be the treatment of choice even for aggressive lesions if the local tumor can be completely excised.

Sauven *et al.*[8] may be entirely correct in their conclusion that advanced locoregional disease precludes a surgical cure. Certainly, the survival rates in patients with aggressive lesions have not been much affected by major operation in their series or in ours. We are reluctant on several counts, however, to adopt a policy which would withhold the option of extended resectional surgery from patients without evidence of distant metastatic disease. A surgical cure is the only possible resolution for rectal carcinoid. Radiation and chemotherapy have had no impact on this tumor.

The poor survival results following major resection for biologically aggressive rectal carcinoids may be due more to late discovery and treatment than to the absolute impotence of surgical therapy. To suggest that radical surgery be withheld for tumors with deep local invasion or nodal involvement is to imply that such tumors have spread beyond the reach of the scalpel. While this may be so, it is not illogical to speculate that, at some stage in the growth and spread of these tumors, disease may be locally limited and surgically treatable for cure. Unless aggressively behaving rectal carcinoids are systemic at their inception, there should be hope for a surgical cure if the tumor is found in time.

Techniques of extended curative resection, advocated by Heald[9] and Enker and associates,[10, 11] have been demonstrated to improve the cure rate of rectal adenocarcinoma. These methods might effect a cure at certain early stages of aggressive rectal carcinoid.

Rectal carcinoids are generally perceived to be slow-growing tumors. They may be present for many years before they are discovered, and patients have lived for many years with metastatic disease. We have shown that rectal carcinoids are imminently discoverable by the examining finger and proctoscope. Perhaps as screening for the more common disease, colorectal adenocarcinoma, becomes more prevalent, earlier aggressive rectal carcinoids may be discovered before symptoms develop. Such patients may benefit from extended operative resection.

We would favor a treatment scheme which includes the option of major resection for rectal carcinoids with features indicating that they may be of the aggressive, metastasizing variety. Such features include size greater than 2 cm, central mucosal ulceration or umbilication, symptoms referable to the tumor, or fixation on digital examination. We would continue to consider patients with tumors invading through the full thickness of the muscularis propria, and those with involved regional nodes, as candidates for extended surgical resection until such time as further evidence accumulates to prove that such a course is universally futile. Sphincter-saving resections, as Hanley *et al.*[1] stated long ago, may have a role for intermediate-sized rectal carcinoids of the mid- to low rectum with aggressive features.

Endorectal ultrasound has been applied to few

cases of rectal carcinoid to date. This technique, perhaps combined with ultrasound-directed node biopsy, might play a role in the decision of what therapy to offer.

Tsioulias *et al.*[12] recently reported DNA aneuploidy to be an accurate predictor of metastatic behavior for rectal carcinoids. DNA analysis, in conjunction with the history of symptoms, tumor size, and physical characteristics of the tumor on examination, may aid in the decision regarding the proper therapy of intermediate-sized lesions.

Associated colorectal malignancy has been reported at increased incidence with rectal carcinoid,[7] although some studies, ours included, do not reflect this finding. We recommend that patients with rectal carcinoid initially undergo total colonoscopy. Since the long-term risk is presently unknown, the safest recommendation is for these patients to then enter a program of periodic total colonic surveillance.

Findings of the current report are at sharp variance with the extant surgical literature on the relative frequency of various gastrointestinal carcinoid tumors. Rectal carcinoids predominated in this series (55 percent of cases) more than four to one over ileal (12 percent) and appendiceal (12 percent) carcinoids. The concensus of the literature (Table 5), in contrast, has ranked rectal carcinoids as the third most common carcinoid tumor, trailing carcinoids of the appendix and ileum in frequency.[13, 14, 18] Occasional reports list rectal carcinoids as second most common,[15, 16] or equal in frequency to small bowel carcinoids.[17]

What reasonable explanations may be given for the results of the present series?

The frequency of carcinoid distribution, for one, may vary with the type of practice reporting it. Speciality referral centers will see a tumor distribution weighted toward cases of ileal carcinoid,[16] since these will most often present diagnostic and therapeutic challenge. Rectal and appendiceal carcinoids are more likely to be diagnosed and treated without referral. Reports from specialty referral centers may not represent the true relative frequency of gastrointestinal carcinoids, being skewed toward a predominance of small bowel carcinoids. Since Ochsner Clinic serves as a primary care clinic for the Greater New Orleans area and as a referral center for the Gulf South, our patient base may reasonably be argued to be a balanced representation of the populace.

Second, rectal carcinoids may be less likely than others to find their way into published series. Most are discovered and treated in the private office setting, where most of the examinations are done. Since the incidence of rectal carcinoids is roughly one per 2,500 proctoscopies,[2, 3] the individual practitioner will diagnose only a few of these tumors over many years. At Ochsner Clinic, 8,000 proctoscopic examinations per year led to diagnosis of three rectal carcinoids annually. Most of these tumors are found in settings where they are unlikely to accrue in sufficient number to be "reportable."

A third factor which may lead to inaccurate reporting of the frequency of rectal carcinoids is that most of these small, indolent, nonmetastasizing tumors are likely never discovered. Even at autopsy they are likely to escape detection since the mid and lower rectum are relatively inaccessible sites and small tumors may be overlooked.

It may be argued that the predominance of rectal carcinoids in the current series merely reflects a paucity of ileal and appendiceal carcinoids at our institution. We know of no reason why this would be so. Appendiceal carcinoids are usually asymptomatic and will be randomly diagnosed in any hospital where incidental appendectomy is performed.[8] Specialty referral centers are more likely

Table 5.
Frequency of Organ Distribution in Gastrointestinal Carcinoids

Report	No. of Patients	Organ Frequency (%/Rank)		
		Appendix	Ileum	Rectum
Beaton *et al.*[13]	59	27/1	25/2	22/3
Wareing & Sawyers[14]	94	35/1	22/2	14/3
Godwin[15]	2,456 (collected review)	44/1	11/3	15/2
Thompson *et al.*[16]	154	11/3	43/1	30/2
Saha *et al.*[17]	112	21/2	27/1	27/1
Present series	170	12/3	12/2	55/1

to see large numbers of small bowel carcinoids. Hospitals reporting on carcinoids saw 10.8 cases (Mayo), 8.1 cases (Vanderbilt), 1.9 cases (Tulane-Charity), and 1.5 cases (The New York Hospital–Cornell) of nonrectal gastrointestinal carcinoid per year.[16] We saw 2.5 cases per year of nonrectal gastrointestinal carcinoid at Ochsner Clinic. We would state then that the expected number of nonrectal carcinoids was seen at our institution. The greater frequency of rectal carcinoids which we report must be due to the fact that more rectal carcinoids were found and diagnosed.

We suggest that the frequency of distribution of gastrointestinal carcinoids uncovered by this report, with rectal carcinoids being far and away the most frequently diagnosed, represents the true distribution of carcinoid tumors in the human gastrointestinal tract. If the trend toward more widespread screening of the well population continues, rectal carcinoids may become recognized as the most frequent human carcinoid tumor.

REFERENCES

1. Hanley PH, Hines MO, Ray J, Armstrong R. Carcinoid tumors of the rectum: experience with 26 cases. J R Soc Med 1959;52:113–7.
2. Quan SH, Bader G, Berg JW. Carcinoid tumors of the rectum. Dis Colon Rectum 1964;7:197–206.
3. Caldarola VT, Jackman RJ, Moertel CG, Dockerty MB. Carcinoid tumors of the rectum. Am J Surg 1964; 107:844–9
4. Bates HR Jr. Carcinoid tumors of the rectum. Dis Colon Rectum 1962;5:270–80.
5. Grablowsky OM, Ribaudo J, Ray JE. Carcinoid tumors of the rectum: experience with 38 cases. Dis Colon Rectum 1974;17:532–5.
6. Burke M, Shepherd N, Mann CV. Carcinoid tumors of the rectum and anus. Br J Surg 1987;74:358–61.
7. Naunheim KS, Zeitels J, Kaplan EL, *et al.* Rectal carcinoid tumors—treatment and prognosis. Surgery 1983;94:670–6.
8. Sauven P, Ridge JA, Quan SH, Sigurdson ER. Anorectal carcinoid tumors: is aggressive surgery warranted? Ann Surg 1990;211:67–71.
9. Heald RJ. The 'Holy Plane' of rectal surgery. J R Soc Med 1988;81:503–8
10. Enker WE, Laffer UT, Block GE. Enhanced survival of patients with colon and rectal cancer is based upon wide anatomic resection. Ann Surg 1979;190; 350–60.
11. Enker WE, Heilweil ML, Hertz RE, *et al. En bloc* pelvic lymphadenectomy and sphincter preservation in the surgical management of rectal caner. Ann Surg 1986;203:426–33.
12. Tsioulias G, Muto T, Kubota Y, *et al.* DNA ploidy pattern in rectal carcinoid tumors. Dis Colon Rectum 1991;34:31–6.
13. Beaton H, Homan W, Dineen P. Gastrointestinal carcinoids and the malignant carcinoid syndrome. Surg Gynecol Obstet 1981;152:268–72.
14. Wareing TH, Sawyers JL. Carcinoids and the carcinoid syndrome. Am J Surg 1983;145:769–72.
15. Godwin JD II. Carcinoid tumors: an analysis of 2837 cases. Cancer 1975;36:560–9.
16. Thompson GB, van Heerden JA, Martin JK Jr, Schutt AJ, Ilstrup DM, Carney JA. Carcinoid tumors of the gastrointestinal tract: presentation, management, and prognosis. Surgery 1985;98:1054–63.
17. Saha S, Hoda S, Godfrey R, Sutherland C, Raybon K. Carcinoid tumors of the gastrointestinal tract: a 44-year experience. South Med J 1989;82:1501–5.
18. Brennan MF, MacDonald JS. Cancer of the endocrine system: carcinoid tumors. In: DeVita VT Jr, Hellman S, Rosenberg SA, eds. Cancer: principles & practice of oncology. 2nd ed. Philadelphia: JB Lippincott, 1985:1223–8.