Original Paper

Digestive Surgery

Dig Surg 1998;15:42–51

Received: June 2, 1997
Accepted: September 5, 1997

# Prognostic Factors in Resected Primary Small Bowel Tumors

*B.L.D.M. Brücher*[a]
*J.D. Roder*[a]
*U. Fink*[a]
*H.J. Stein*[a]
*R. Busch*[b]
*J.R. Siewert*[a]

[a] Chirurgische Klinik and Poliklinik, und
[b] Institut für Medizinische Statistik und Epidemiologie, Klinikum rechts der Isar der Technischen Universität München, Deutschland

**Key Words**
Small bowel tumors
Prognostic factors

**Abstract**
We report a retrospective analysis of 71 patients, operated for primary small bowel tumors (SBT): 47 malignant (66.2%) and 24 benign (33.8%) tumors. Of the malignant tumors, adenocarcinomas predominated (38.3%), followed by neuroendocrine tumors (31.9%), Non-Hodgkin lymphomas (NHL) (12.8%), leiomyosarcomas (10.6%) and other rare entities (6.4%). Morbidity of surgical treatment was 16.9%, 30-day mortality 7%. The estimated 5-year survival rate in malignant lesions was 31.8%. Univariate analysis identified the presence of distant metastasis and the resection status (R status) as prognostic factors ($p = 0.034$ and $p = 0.001$). There was no influence of T, N status or grading on survival. A complete macroscopic and microscopic tumor resection has to be the aim of any curative surgical approach in patients with SBT.

## Introduction

Small bowel tumors (SBT) are remarkably rare. Constantly 320 patients died each year suffering of malignant neoplasms of the small bowel in Germany from 1990 until 1994 [1]. The surgeon is challenged in the diagnosis and the treatment of these tumors, because of their infrequency, the different histological types of these tumors and unspecific symptoms. The first report of a case of duodenal carcinoma was made by Hamburger [2] in 1746. The first collective series of malignant small bowel neoplasma was published by Leichtenstein [3] in 1876. Hearteaux [4] in 1899, was the first to describe a collective series of benign SBT. We report the surgical treatment, survival rates and prognostic factors in a series of patients with SBT, treated at a single institution between 1985 and 1995.

## Patients and Methods

Between January 1985 and December 1995, 71 patients with primary SBT were operated at the Department of Surgery. These included only primary tumors, located between the duodenum and the ileocecal valve. Tumors, found at autopsy, periampullary tumors and tumors at the ileocecal valve or arising in the mesentery were excluded. Benign lesions were mainly adenomas and leiomyomas (table 1). Malignant tumors were comprised mostly of adenocarcinomas (38.2%) and neuroendocrine tumors (31.9%) (table 2). The TNM categories, grading and the presence of residual tumor were reassessed for adenocarcinomas according to the UICC 1992 criteria [5]. Non-Hodgkin lymphoma (NHL) was classified according to the Ann-Arbor classification [6]. Leiomyosarcomas were staged according to tumor size and grading [5]. In spite of a wide variety of investigations, the correct diagnosis was made preoperatively in only 71.8% of the patients. Elective surgery was performed in 59 (83.1%) and surgical emergency surgery in 12 (16.9%) patients, 44 (93.6%) patients with malignant tumors were operated with curative, and 3 (6.4%) with palliative intention.

KARGER
Fax + 41 61 306 12 34
E-Mail karger@karger.ch
www.karger.com


0253–4886/98/0151–0042$15.00/0

This article is also accessible online at:
http://BioMedNet.com/karger

Dr. B.L.D.M. Brücher
Klinikum rechts der Isar der TUM
Ismaninger Strasse 22, D–81675 München (Germany)
Tel. 49 89 4140 2047, Fax 49 89 4140 4804
E-Mail bruecher@ntl.chir.med.tu-muenchen.de

**Table 1.** Histologic type and localization of benign small bowel tumors

| Histological entities | Duodenum | Jejunum | Ileum | Total |
|---|---|---|---|---|
| Adenoma | 13 (100%) | – | – | 13 (54.2%) |
| Leiomyoma | 1 (20%) | 3 (60%) | 1 (20%) | 5 (20.8%) |
| Fibroma | 1 (50%) | – | 1 (50%) | 2 (8.3%) |
| Ectopic pancreatic tissue | – | 1 (100%) | – | 1 (4.15%) |
| Cyst | 1 (100%) | – | – | 1 (4.15%) |
| Lipoma | 1 (100%) | – | – | 1 (4.15%) |
| Neurinoma | 1 (100%) | – | – | 1 (4.15%) |
| Total | 18 (75%) | 4 (16.7%) | 2 (8.3%) | 24 (100%) |

**Table 2.** Histologic type and localization of malignant small bowel tumors

| Histological entities | Duodenum | Jejunum | Ileum | Total |
|---|---|---|---|---|
| Adenocarcinoma | 11 (61.1%) | 7 (38.9%) | – | 18 (38.3%) |
| Neuroendocrine tumors | – | 1 (6.7%) | 14 (93.3%) | 15 (31.9%) |
| Non-Hodgkin lymphoma | 1 (16.7%) | 4 (66.6%) | 1 (16.7%) | 6 (12.8%) |
| Leiomyosarcoma | 2 (40%) | 2 (40%) | 1 (20%) | 5 (10.6%) |
| Melanoma | – | 1 (100%) | – | 1 (2.12%) |
| Schwannoma | 1 (100%) | – | – | 1 (2.12%) |
| Histiocytoma | – | 1 (100%) | – | 1 (2.12%) |
| Total | 15 (32%) | 16 (34%) | 16 (34%) | 47 (100%) |

Follow-up was performed by the oncologic outpatient clinic and personal contact with the patients, their general practitioner and/or their relatives. Survival rates were calculated according to Kaplan and Meier [7]. Statistical differences between the groups were determined using the log-rank test. $p \leq 0.05$ was considered significant.

## Results

*Age/Sex/Symptoms and Operative Procedures*

There were 40 males (56.4%) and 31 females (43.6%). The mean age (range) of the patients was 58.9 years (20.6–84.8). 36 (50.7%) of all patients were between 50 and 70 years of age. Females were on average 10 years older than males (64.3 vs. 54.8). There were no significant difference in the age and sex distribution between patients with

**Table 3.** Operative procedures

| Operative procedures | Total | | Benign SBT | | Malignant SBT | |
|---|---|---|---|---|---|---|
| | n | % | n | % | n | % |
| Segmental small bowel resection[a] | 35 | 49.3 | 7 | 29.2 | 28 | 59.6 |
| Right hemicolectomy[b] | 8 | 11.3 | 1 | 4.2 | 7 | 14.9 |
| Wedge resection[c] | 5 | 7 | 5 | 20.8 | | – |
| Whipple-procedure | 8 | 11.3 | 2 | 8.3 | 6 | 12.8 |
| Traverso-Longmire procedure | 2 | 2.8 | | – | 2 | 4.25 |
| Billroth procedure | 2 | 2.8 | | – | 2 | 4.25 |
| Gastroenterotomy | 1 | 1.4 | | – | 1 | 2.1 |
| Endoscopic exstirpation | 5 | 7 | 5 | 20.8 | | – |
| Duodenal exstirpation | 4 | 5.7 | 4 | 16.7 | | – |
| Diagnostic laparatomy | 1 | 1.4 | | – | 1 | 2.1 |
| Total | 71 | 100 | 24 | 100 | 47 | 100 |

[a] Including: 1 patient with additional sigmoidectomy and 3 with additional excision of hepatic metastasis.
[b] Including: 1 patient with hysterectomy and excision of hepatic metastasis, and 1 patient with additional excision of diaphragmal metastasis.
[c] Including: 3 patients with partial duodenal resections.

**Table 4.** Morbidity/treatment/outcome and mortality

| | n | Morbidity/treatment | Outcome | Mortality |
|---|---|---|---|---|
| Benign SBT | 1 | duodenal fistula → drain and antibiotics | secondary healing | – |
| | 1 | anastomotic leak → drain and antibiotics | secondary healing | – |
| | 1 | abscess → drain and antibiotics | died on sepsis | † 30th postop. day |
| | 1 | cardiac failure → ICU | died | † 21st postop. day |
| Total | – | 4/71 (5.6%) | – | 2/24 (8.4%) |
| Malignant SBT | 3 | wound dehiscence | secondary healing | – |
| | 1 | postoperative hemorrhage → relaparatomy | anastomotic leak sepsis | † 7th postop. day |
| | 1 | severe pneumonia | antibiotics | † 30th postop. day |
| | 1 | postoperative hemorrhage → conservative treatment | pancreatic fistula | † 8.2 months |
| | 1 | stricture of gastroenterostomy → dilatation | secondary healing | – |
| | 1 | subphrenic abscess → drain and antibiotics | secondary healing | – |
| Total | – | 8/71 (11.3%) | – | 3/47 (6.4%) |
| Total of all SBT | – | 12/71 (16.9%) | – | 5/71 (7%) |

benign or malignant SBTs. The most frequent symptoms in decreasing order were sickness (76%), abdominal pain of less than 6 months' duration (60.6%), vomiting (49.3%), weakness and weight loss (45%). Although no symptom/complex was specific for a histological lesion, 93.4% of neuroendocrine tumors presented with flush – especially after consumption of red wine – and diarrhea (not significant). 11.3% of all SBT had a low hemoglobin and bleeding symptoms. The operative procedures performed are listed in table 3.

*Morbidity/Mortality*

Overall, 12 patients (16.9%) (8 malignant [11.3%], 4 benign [5.6%]) had postoperative complications. The overall hospital mortality was 7% (n = 5), 6.4% (n = 3) in malignant lesions, 8.4% (n = 2) in benign lesions (table 4).

*Tumor Types and Localization*

Of the 71 patients, 33 (46.5%) had lesions in the duodenum, 20 (28.2%) in the jejunum and 18 (25.4%) in the ileum. 75% of the benign tumors were located in the duodenum, 16.7% in the jejunum and 8.3% in the ileum (table 1). Pathohistological analysis showed 13 (54.2%) adenomas, 5 (20.8%) leiomyomas, 2 (8.3%) fibromas and each 1 case (4.175%) of ectopic pancreatic tissue, embryonal diverticula, lipoma and neurinoma. All adenomas were localized in the duodenum, 60% of the leiomyomas in the jejunum. Malignant SBT showed no predominant localization: 32% in the duodenum, 34% in the jejunum and 34% in the ileum (fig. 1, table 2). The most common malignant tumors of the small intestine were adenocarcinomas (38.3%) and neuroendocrine tumors (31.9%), followed by NHLs (12.8%), leiomyosarcomas (10.6%) and other rare entities (6.4%). Adenocarcinomas occurred in 61.1% in the duodenum, 38.9% in the jejunum and none in the ileum. 93.3% of neuroendocrine tumors were located in the ileum, 6.7% in the jejunum and none in the duodenum. Two thirds of NHL were found in the jejunum. 4 of 5 leiomyosarcomas were located in the duodenum and the jejunum.

*Tumor Stage*

Of the 18 adenocarcinomas, 1 patient had stage I, 1 had stage III disease, 9 had stage II and 7 had stage IV tumors. NHLs were classified according to the Ann-Arbor classification: 4 patients presented in stage IVb and 2 in stage IVa. 2 patients had low-grade and 4 high-grade lymphomas. Of the patients with leiomyosarcomas, 2 had a T1 and 3 a T2 tumor.

*Distant Metastasis*

Of the 47 malignant small bowel tumors, 19 (40.4%) had distant metastasis, including 7 adenocarcinomas, 8 neuroendocrine tumors, 1 NHL, 1 leiomyosarcoma, 1 melanoma and 1 histiocytoma. Liver metastasis were most common (13 patients). Solitary metastasis of the liver were found in 8 patients, while the other 5 had additional metastases in the peritoneum, the diaphragma and the lymph nodes.

*R Status*

Regarding the primary tumor, a local R0 resection could be achieved in 37 of 47 small bowel tumors, 4 had microscopic residual tumor (R1), 6 patients had a local R2 resection.

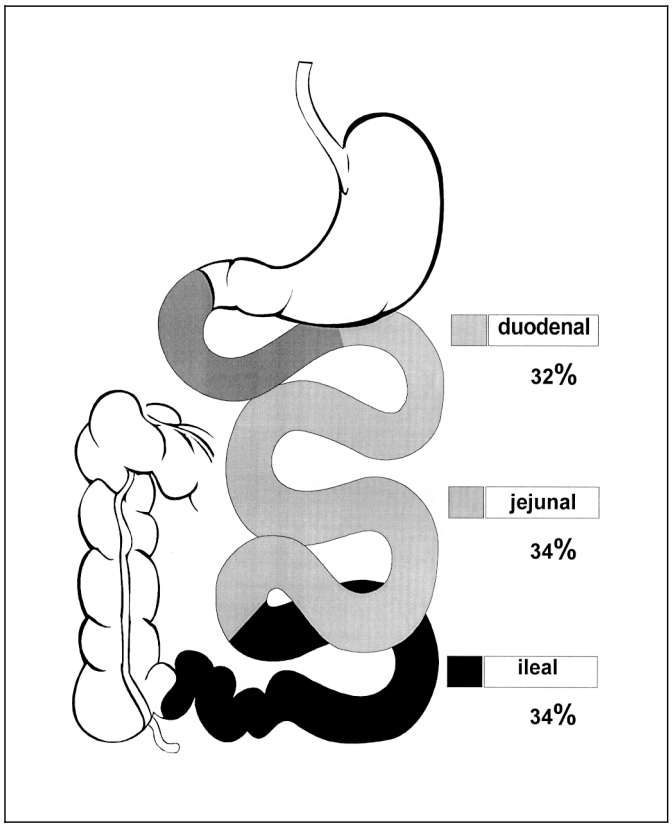


**Fig. 1.** Distribution of malignant small bowel tumors in the present series (n = 47).

*Survival*

The estimated median survival time for patients with malignant SBT was 31.8 months (range 11.9–51.8). In benign tumors the median survival was not reached, because within the period of observation merely 12.5% patients died (fig. 2) (none of these patients died because of the tumor). 40% of those with neuroendocrine tumors, 50% with adenocarcinomas, 66.6% with NHLs and 80% with leiomyosarcomas died during follow-up. There was a significant difference in survival between those with and without distant metastasis ($p = 0.034$) (fig. 3). The estimated median survival time of patients with metastasis (M1) was 12.8 months, while the median survival time was not reached for patients without metastasis (M0). 68.4% of the patients with metastasis (M1) and 28.6% without died during follow-up. There was no correlation between T, N status or grading and survival. Patients after R1 and R2 resection had a statistically lower estimated median survival of 10 months in comparison to those with local R0 resection, $p = 0.001$ (fig. 4). 43.3% of R0

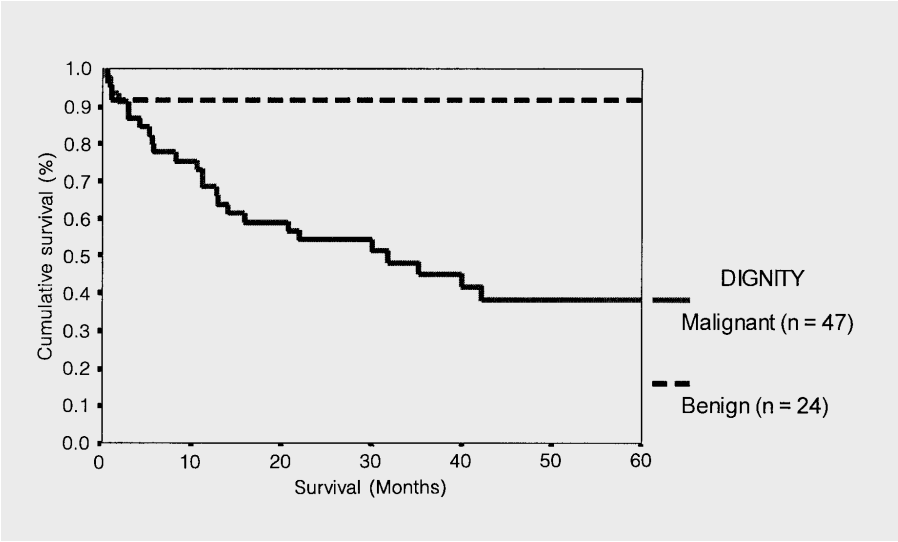


**Fig. 2.** Survival of benign and malignant small bowel tumors.

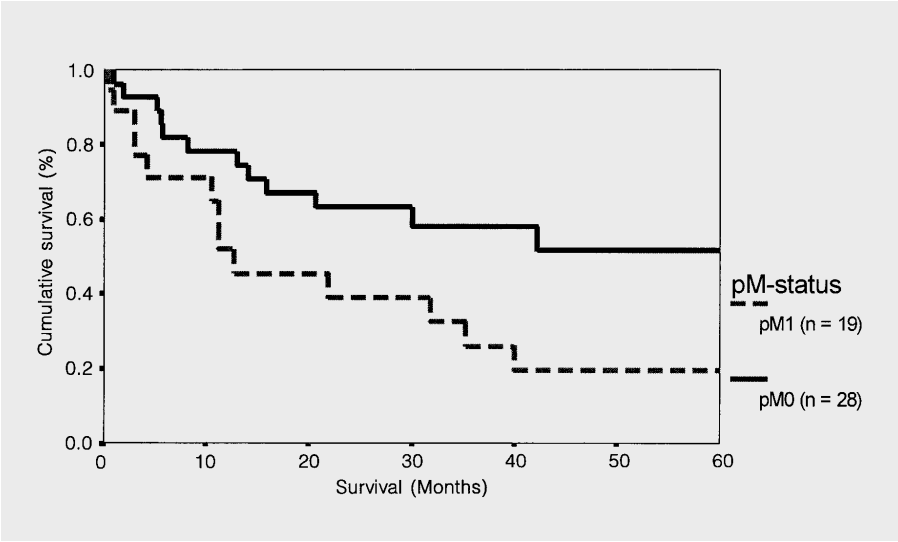


**Fig. 3.** Survival according to pM status in 47 malignant small intestine neoplasms.

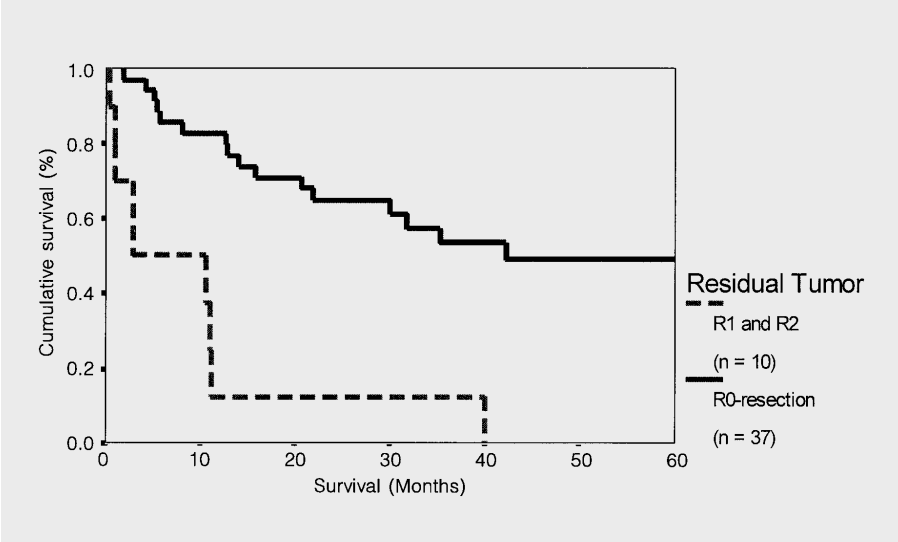


**Fig. 4.** Survival according to residual tumor in 47 malignant small intestine neoplasms.

and 90% of R1 and R2 resected patients died during follow-up. All patients who had a palliative resection died during follow-up.

## Discussion

Merely 1–3% of all gastrointestinal malignant neoplasms are localized in the small intestine [8–14]. Our retrospective series of 71 patients with primary SBTs included 24 benign and 47 with malignant tumors. In agreement with other authors, the highest incidence of tumors in the small intestine was noted in patients between 50 and 70 of age [13, 15–20] with a slight male predominance [12, 20, 21]. 93.4% of the patients with neuroendocrine tumors presented with flush and diarrhea – especially after consumption of red wine, otherwise symptoms were unspecific. A correct preoperative diagnose was made in 71.8% compared with 17–69% in other series [10, 11, 18, 20, 22–25]. This figure may be misleading, since in our series many patients were seen at a later stage in their illness when they had obvious symptoms.

Our series represents the largest experience with operated primary malignant SBTs (47 within 10 years) treated at a single institution, except the work of Miles [17] 1978, who reported 79 cases within 16 years time of observation. Brookes 1968 reviewed 168 patients within 9 years, all documented in the Birmingham Regional Cancer Registries [26]. Barclay 1983 reported 209 surgical *and* autopsied cases [9]. 174 patients within a 38-year period were documented by Martin [13] in 1986. The distribution of the anatomical localization was statistically equal (fig. 1) and comparable to the data of Coit, Likely and Zollinger [27–29]. Other authors showed a predominance of ileal [9–11, 13–15, 17, 18, 22, 24, 26, 30–32] and jejunoileal lesions [33–36]. Only Kelm et al. [37] described a predominance of malignant small bowel lesions in the jejunum. Differences in the distribution of the histologic entities in the various series make a comparison difficult (table 5).

The main histological entities in malignant SBTs in this series are adenocarcinomas (38.3%), neuroendocrine tumors (31.9%), NHLs (12.8%) and leiomyosarcomas (10.6%). Regarding these categories, our data are comparable to those of Martin [13] and Broll et al. [22], but there are substantial differences when considering each histological entity on its own. In the literature, adenocarcinomas are reported in more than 50% in the small intestine [18, 19, 25, 30, 35, 36] and in less than 30% by other authors [15, 16, 31, 33, 34, 37] (table 6). As known from the literature [38], adenocarcinomas are predominant in the duodenum (table 2). None of our adenocarcinomas were localized in the ileum in comparison to Coit [27] who describes approximately 22% of the adenocarcinomas, localized in the ileum. According to the data of Peck et al. [39], 93.3% of neuroendocrine tumors were localized in the ileum, while large series report neuroendocrine tumors as ileal lesions in 57–84.7% [19, 21]. Some series found neuroendocrine tumors to be *the* most common entity in small bowel neoplasms (34.2–45%) [9, 10, 17, 37], while in other series these tumors occurred within 30% in the small intestine [13, 15, 17, 18, 24, 25, 28]. In the gastrointestinal tract malignant lymphoma are usually localized in the stomach and only within 10% in the small bowel [40]. Outstanding are the data of Gupta and Gupta [31] and Freund et al. [34] with 63.4% and 67.8% malignant lymphomas localized in the small bowel. This is in contrast to our data and to others from the western world – geographic differences seem to be important. 62.5% sarcomas have been reported to be in the small intestine [33], while generally less than 20% are reported to be in the small intestine [41]. The rate of elective/emergency surgery was 83.1%/16.9% compared to 55%/45% reported by Feil et al. [42]. The rate of resection for cure of 93.6% compares favourable to the literature [18, 24, 25]. Postoperative mortality in all patients with malignant SBT was 6.4% – reported data in the literature are between 13 and 16% [22, 24, 43]. 40.4% of patients had metastasis at the time of diagnosis. These findings compare the current literature, with 8–62% of all operated patients presenting with metastatic disease at the time of operation [9, 12, 18, 23, 24]. Location of metastasis was mainly in the liver, as reported by other authors [22].

The overall survival of malignant SBTs is poor. Reported 5-year survival rates for malignant small bowel tumors range from 30 to 40% [18, 44] in contrast to 48.8% in this series. The estimated median survival time for adenocarcinomas in our series was 58.8 months, while other series report survival rates between 16 and 43% [14, 18, 45, 46]. Merely Scott-Coombes reported a 5-year survival rate in adenocarcinomas of the duodenum of 50% [47]. Classified according to the UICC criteria [5], a trend toward decreasing survival with advanced stages is evident: 6 of 7 patients in stage IV died. Neuroendocrine tumors had the best 5-year survival with 60% in comparison to adenocarcinomas (50%), NHLs (33.3%) and leiomyosarcomas (20%). These findings resemble the survival data of neuroendocrine tumors from other series [11, 17, 24, 25, 31, 48]. Our survival rate in neuroendocrine tumors is comparable to the data of Goel et al. [12] and

**Table 5.** Summary of the localization of malignant small bowel tumors in literature

| Study | PY | Time | Y | n | Duodenum | Jejunum | Ileum |
|---|---|---|---|---|---|---|---|
| Likely – New York | 1948 | 1920–1947 | 27 | 17 | 5 (29.4%) | 6 (35.3%) | 6 (35.3%) |
| Darling – Boston | 1959 | 1913–1957 | 44 | 86 | 11 (12.8%) | 35 (40.7%) | 40 (46.5%) |
| Rochlin – Los Angeles | 1961 | 1940–1959 | 19 | 40 | 10 (25%) | 12 (25.6%) | 18 (45.8%) |
| Ostermiller – Los Angeles[a] | 1966 | 1937–1965 | 28 | 122 | 36 (29.5%) | 25 (20.5%) | 55 (45.1%) |
| Brookes – Birmingham[b] | 1968 | 1950–1959 | 9 | 168* | 32 (19%) | 43 (25.6%) | 77 (45.8%) |
| Wilson – New York | 1973 | 1932–1972 | 40 | 96 | 30 (31.2%) | 21 (21.9%) | 45 (46.9%) |
| Croom – Chapel Hill | 1975 | 1952–1973 | 21 | 41 | 6 (14%) | 15 (36%) | 20 (50%) |
| Sager – Maine[c] | 1977 | 1955–1976 | 21 | 30 | 2 (6.6%) | 10 (33.4%) | 16 (53.4%) |
| Freund – Jerusalem | 1978 | 1956–1974 | 18 | 31 | 2 (6.4%) | 14 (45.2%) | 15 (48.4%) |
| Miles – Memphis | 1978 | 1960–1970 | 10 | 79 | 13 (16.5%) | 25 (31.6%) | 41 (51.9%) |
| Mittal – Michigan[d] | 1980 | 1967–1977 | 10 | 38 | 7 (18.5%) | 11 (28.9%) | 20 (52.6%) |
| Gupta – Varanasi | 1982 | 1966–1979 | 13 | 30 | 5 (16.7%) | 9 (30%) | 16 (53.3%) |
| Barclay – Miami | 1983 | 1950–1979 | 19 | 209 | 37 (17.7%) | 41 (19.6%) | 131 (62.7%) |
| Martin – Houston | 1986 | 1944–1982 | 38 | 174 | 45 (25.9%) | 42 (24.1%) | 87 (50%) |
| Zollinger – Cleveland | 1986 | 1960–1980 | 20 | 38 | 12 (31.6%) | 15 (39.5%) | 11 (28.9%) |
| Cicarelli – Hartford | 1987 | 1969–1983 | 14 | 51 | 6 (11.8%) | 15 (29.4%) | 30 (58.8%) |
| Cunningham – New York[e] | 1997 | 1970–1991 | 21 | 73 | 18 (25%) | 27 (37%) | 28 (38%) |
| MRI | 1997 | 1985–1995 | 10 | 47 | 15 (32%) | 16 (34%) | 16 (34%) |

PY = Publication year; Y = years; n = number of patients.

[a] 6 (4.9%) inadequately classified.

[b] 16 (9.6%) inadequately classified.

[c] 2 (6.6%) not classified.

[d] Inclusive 1 carcinoid, localized in a Meckel'-diverticula.

[e] It is not mentioned, if ileocoecal tumors were also excluded.

**Table 6.** Summary of the histologies of malignant SBT in literature

| Study | PY | n | AC | CC | SR | LY | MM | MS | HM | NC |
|---|---|---|---|---|---|---|---|---|---|---|
| Likely – New York | 1948 | 17 | 8 (47%) | 6 (35.3%) | 3 (17.7%) | – | – | – | – | – |
| Darling – Boston | 1959 | 86 | 33 (38.4%) | 15 (17.4%) | 9 (10.5%) | 29 (33.7%) | – | – | – | – |
| Rochlin – Los Angeles | 1961 | 40 | 18 (45%) | 17 (42.5%) | 5 (12.5%) | – | – | – | – | – |
| Ostermiller – Los Angeles[a] | 1966 | 122 | 64 (52.5%) | 21 (17.2%) | 35 (28.7%) | – | 1 (0.8%) | – | – | 1 (0.8%) |
| Brookes – Birmingham[b] | 1968 | 168 | 55 (32.7%) | 32 (19%) | 56 (33.4%) | – | – | – | – | 25 (14.9%) |
| Wilson – New York | 1973 | 96 | 48 (50%) | 37 (38.5%) | 11 (11.5%) | – | – | – | – | – |
| Croom – Chapel Hill | 1975 | 41 | 12 (29.3%) | 14 (34.2%) | 10 (24.4%) | 4 (9.8%) | – | – | – | 1 |
| Sager – Maine[c] | 1977 | 30 | 14 (46.7%) | 12 (40.9%) | 4 (13.3%) | – | – | – | – | – |
| Freund – Jerusalem | 1978 | 31 | 5 (16.1%) | 3 (9.7%) | 1 (3.2%) | 21 (67.8%) | 1 (3.2%) | – | – | – |
| Miles – Memphis | 1978 | 79 | 16 (20.2%) | 31 (39.2%) | 11 (13.9%) | 15 (19%) | – | – | – | 6 (7.7%) |
| Mittal – Michigan[d] | 1980 | 39 | 21 (53.8%) | 9 (23.1%) | 3 (7.7%) | 5 (12.8%) | 1 (2.6%) | – | – | – |
| Gupta – Varanasi | 1982 | 30 | 9 (30%) | 1 (3.3%) | 1 (3.3%) | 19 (63.4%) | – | – | – | – |
| Barclay – Miami | 1983 | 209 | 74 (35.4%) | 94 (45%) | 41 (19.6%) | – | – | – | – | – |
| Martin – Houston | 1986 | 174 | 77 (44.3%) | 64 (36.8%) | 27 (15.5%) | 6 (3.4%) | – | – | – | – |
| Zollinger – Cleveland | 1986 | 38 | 18 (47.4%) | 10 (26.3%) | 10 (26.3%) | – | – | – | – | – |
| Cicarelli – Hartford | 1987 | 51 | 17 (33.4%) | 20 (39.2%) | 8 (15.7%) | 6 (5.7%) | – | – | – | – |
| Cunningham – New York | 1997 | 73 | 29 (40%) | 18 (25%) | 8 (10%) | 18 (25%) | – | – | – | – |
| MRI | 1997 | 47 | 18 (38.3%) | 15 (39.9%) | 5 (10.6%) | 6 (12.8%) | 1 (2.12%) | 1 (2.12%) | 1 (2.12%) | – |

PY = Publication year; Y = years; n = number of patients; AC = adenocarcinomas; CC = carcinoids/neuroendocrine tumors; SR = sarcomas; LY = lymphomas; MM = melanoma; MS = malignant schwanoma; HM = histiocytoma; NC = not classified.

[a] 6 (4.9%) inadequately classified.

[b] 16 (9.6%) were inadequately classified.

[c] 2 (6.6%) not classified.

[d] Inclusive 1 carcinoid, localized in a Meckel diverticula.

Feil and Schulz [42] but in contrast to Zeitels et al. [49], who analyzed 101 patients and showed a survival of more than 80%. Generally survival rates of neuroendocrine tumors range from 99% (appendix) to 33% (sigmoid colon) [21]. 66.7% of NHLs died. As reported by Feil and Schulz [42], we found in 5 of 6 patients high-grade NHLs and in contrast to the work of Dragosics et al. [40], all patients had stage IV (2 stage IVA, 4 stage IVB). In leiomyosarcoma 80% died. In contrast to the work of Chiotasso and Fazio [41], we did not find a correlation between a long history in leiomyosarcomas prior to operation and a better prognosis, but our number of such cases is to small, to allow a valid comparison.

The presence of metastasis was found to be a statistically significant prognostic factor, but there was no significant correlation between nodal metastasis and survival, as described by Adler et al. [38], Bridge and Perzin [50] and Rotman et al. [51]. The median survival of patients with metastasis (12.8 months) was analogous to other data [52]. The radicality of surgical procedures was another prognostic factor: 90% of patients with malignant lesions, who had a R1 or R2 resection, died during the follow-up. Though a multivariate analysis is the ideal way of investigation of prognostic factors, this was omitted due to the small amount of patients.

In conclusion, in patients with unclear abdominal symptoms and after exclusion of more common abnormalities, a SBT should be suspected. Surgical treatment is the therapy of choice. Unfortunately, most patients with malignant small bowel tumors reach the surgeon in an advanced stage, which is reflected by the high rate of systemic metastasis at the time of presentation. Our data indicate that the M and R status are the only prognostic factors. Both are dependent on an early diagnosis. An aggressive diagnostic approach in patients with unclear abdominal symptoms and extended surgical resection of all tumors is therefore the only way to improve the prognosis of patients with small bowel tumors.

## References


1 Statistisches Bundesamt Wiesbaden, 1996.
2 Hamburger G: De ruptura intestine duodeni. Jena Ritterianis, 1746.
3 Leichtenstern O: Handbuch der speciellen Pathologie und Therapie. Leipzig, Vogel, 1876, pp 523–524.
4 Hearteux A: Note sur les tumeurs benignes de l'intestin. Arch Prov Chir 1899;8:701–709.
5 Hermanek P, Sobin LH (eds): UICC TNM classification of malignant tumors, ed 4, 2nd revision. Berlin, Springer, 1993.
6 Carbone PP, Kaplan HS, Musshoff K, Smithers DW, Tubiana M: Report of the committee on Hodgkin's disease staging procedures. Cancer Res 1971;31:1860–1861.
7 Kaplan EL, Meyer P: Nonparametric estimation from incomplete observations? J Am Stat Assoc 1958;75:457–487.
8 Ashley SW, Wells SA Jr: Tumors of the small intestine. Semin Oncol 1988;15:116–128.
9 Barclay THC, Schapira DV: Malignant tumors of the small intestine. Cancer 1983;51:878–881.
10 Cicarelli O, Welch JP, Kent GG: Primary malignant tumors of the small bowel: The Hartford hospital experience, 1969–1983. Am J Surg 1987;153:350–354.
11 Darling RC, Welch CE: Tumors of the small intestine. N Engl J Med 1959;260:397–408.
12 Goel IP, Didolkar MS, Elias EG: Primary malignant tumors of the small intestine. Surg Gynecol Obstet 1976;143:717–719.
13 Martin GM: Malignant tumors of the small intestine. Surg Clin North Am 1986;6:779–785.
14 Rochlin DB, Longmire WP Jr: Primary tumors of the small intestine. Surgery 1961;50:586–592.
15 Croom RD, Newsome JF: Tumors of the small intestine. Am Surg 1975;41:160–167.
16 Delcore R, Thomas JH, Forster J, Hermreck AS: Improving resectability and survival in patients with primary duodenal carcinoma. Am J Surg 1993;166:626–631.
17 Miles RM, Crawford D, Duras S: The small bowel tumor problem: An assessment based on 20 year experience with 116 cases. Ann Surg 1978;189:732–740.
18 Mittal VK, Bodzin JH: Primary malignant tumors of the small bowel. Am J Surg 1980;140:396–399.
19 Ouriel K, Adams J: Adenocarcinoma of the small intestine. Am J Surg 1984;147:66–71.
20 Cunningham JD, Aleali R, Aleali M, Brower S, Aufses AHI: Malignant small bowel neoplasms: Histopathological determinants of recurrence and survival. Ann Surg 1997;225:300–306.
21 Godwin JD: Carcinoid tumors: An analysis of 2837 cases. Cancer 1975;36:560–569.
22 Broll R, Bruch H-P, Daniel D, Schiedeck Th: Maligne Dünndarmtumore: Diagnostische Probleme und differenzierte chirurgische Therapie. Chirurg 1994;65:451–456.
23 Ebert P, Zuidema GD: Primary tumors of the small intestine. Arch Surg 1965;91:452–455.
24 Norberg KA, Emas S: Primary tumors of the small intestine. Am J Surg 1981;142:569–573.
25 Wilson JM, Melvin DB, Gray GF, Thorbjarnarson B: Primary malignancies of the small bowel: A report of 96 cases and review of the literature. Ann Surg 1974;180:175–179.
26 Brookes VS, Waterhouse JAH, Powell DJ: Malignant lesions of the small intestine: A ten year survey. Br J Surg 1968;55:405–410.
27 Coit DG: Cancer of the small bowel, chap 29; in Cancer: Principles and Practice of Oncology, ed 4. Philadelphia, Lippincott, 1993, pp 915–926.
28 Likely DS, Lisa JR, Stitch MH, Stein HD: Primary malignant disease of the small bowel. Arch Intern Med 1948;82:206–216.
29 Zollinger RM, Sternfeld WC, Schreiber H: Primary neoplasms of the small intestine. Am J Surg 1986;151:654–658.
30 Buchholz J, Strosche H, Krawzak H-W, Kahl N: Primäre Dünndarmmalignome – Symptomatik, Diagnostik und Therapie. Leber Magen Darm 1988;4:197–203.
31 Gupta S, Gupta S: Primary tumors of the small bowel: A clinicopathological study of 58 cases. J Surg Oncol 1982;20:161–167.
32 Sager GF: Primary malignant tumors of the small intestine. Am J Surg 1978;135:601–603.
33 Bindewald H: Dünndarmtumoren: Eine häufig vergessene Differentialdiagnose. Chirurg 1982;53:638–640.
34 Freund H, Lavi A, Pfeffermann R, Durst AL: Primary neoplasms of the small bowel. Am J Surg 1978;135:757–759.

35 Nöthiger F: Erfahrung mit 50 klinisch manifesten Dünndarmtumoren. Helv Chir Acta 1980; 47:597–599.
36 Schippers E, Langer S, Flosdorff W, Hilber U: Das primäre Dünndarmmalignom. Chirurg 1982;53:364–369.
37 Kelm C, Padberg W, Zimmermann T, Hürtgen M, Buhr J: Maligne Tumoren des Dünndarms. Zentralbibl Chir 1994;119:639–644.
38 Adler S, Lyon D, Sullivan P: Adenocarcinoma of the small bowel: Clinical features, similarity to regional enteritis, and analysis of 338 documented cases. Am J Gastroenterol 1982;77: 326–330.
39 Peck JJ, Shields AB, Boyden AM, Dworkin LA, Nadal JW: Carcinoid tumors of the ileum. Am J Surg 1983;146:124–132.
40 Dragosics B, Bauer P, Radaskiewicz T: Primary gastrointestinal non-hodgkin's-lymphomas: A retrospective clinicopathological study of 150 cases. Cancer 1985;55:1060–1073.
41 Chiotasso PJP, Fazio VW: Prognostic factors of 28 leiomyosarcomas of the small intestine. Surg Gynecol Obstet 1982;155:197–202.
42 Feil W, Schulz F: Dünndarmtumoren – Diagnose, Therapie und Prognose. Langenbecks Arch Chir 1985;365:25–35.
43 Desa LAJ, Bridger J, Grace PA, Krausz T, Spencer J: Primary jejunoileal tumors: A review of 45 cases. World J Surg 1991;15:81–87.
44 Ross RK, Hartnett NM, Bernstein L, Henderson BE: Epidemiology of adenocarcinomas of the small intestine: Is bile a small bowel carcinogen? Br J Cancer 1991;63:143–145.
45 DiSario JA, Burt RW, Vargas H, McWorther WP: Small bowel cancer: Epidemiological and clinical characteristics from a population-based registry. Am J Gastroenterol 1994;89:699–701.
46 Zornig C, Klomp H-J: Das Dünndarmcarcinom. Chirurg 1989;60:603–606.
47 Scott-Coombes DM, Williamson CN: Surgical treatment of primary duodenal carcinoma: A personal series. Br J Surg 1994;81:1472–1474.
48 Ostermiller W, Joergenson EJ, Weibel L: A clinical review of tumors of the small bowel. Am J Surg 1966;111:403–409.
49 Zeitels J, Naunheim K, Kaplan EL, Strauss F: Carcinoid tumors: A 37 year experience. Arch Surg 1982;117:732–737.
50 Bridge MF, Perzin KH: Primary adenocarcinoma of the jejunum and ileum: A clinicopathologic study. Cancer 1975;36:1876–1887.
51 Rotman N, Pezet D, Lagniez P-L, Cherqui D, Celicou B, Lointier P: Adenocarcinoma of the duodenum: Factors influencing survival. Br J Surg 1994;81:83–85.
52 Hilbun BM, Block W: Primary malignant tumors of the small bowel. MSMA 1987;7:169–171.