CHAPTER 85 — REFERENCES

1. Yao JC, Hassan M, Phan A, et al. One hundred years after "carcinoid": epidemiology of and prognostic factors for neuroendocrine tumors in 35,825 cases in the United States. *J Clin Oncol* 2008;26:3063–3072.
2. Surveillance, Epidemiology, and End Results (SEER) Program. SEER*Stat Database: Incidence—SEER 9 Regs Research Data, Nov 2011 Sub (1973–2010) <Katrina/Rita Population Adjustment>—Linked To County Attributes—Total U.S., 1969–2010 Counties, (ed released April 2013), National Cancer Institute, DCCPS, Surveillance Research Program, Cancer Statistic Branch, 2013. http://www.seer.cancer.gov. Accessed June 2013.
3. Lam KY, Lo CY. Pancreatic endocrine tumour: a 22-year clinico-pathological experience with morphological, immunohistochemical observation and a review of the literature. *Eur J Surg Oncol* 1997;23:36–42.
4. Butler AE, Campbell-Thompson M, Gurlo T, et al. Marked expansion of exocrine and endocrine pancreas with incretin therapy in humans with increased exocrine pancreas dysplasia and the potential for glucagon-producing neuroendocrine tumors. *Diabetes* 2013;62:2595–2604.
5. Yao JC, Eisner MP, Leary C, et al. Population based study of islet cell carcinoma. *Ann Surg Oncol* 2007;14:3492–3500.
6. Surveillance, Epidemiology, and End Results (SEER) Program. SEER*Stat Database: SEER 17 Regs Nov 2005 sub (1973–2004), (ed released April 2006), National Cancer Institute, DCCPS, Surveillance Research Program, Cancer Statistic Branch, 2006. http://www.seer.cancer.gov. Accessed June 2006.
7. Rindi G, Leiter AB, Kopin AS, et al. The "normal" endocrine cell of the gut: changing concepts and new evidences. *Ann N Y Acad Sci* 2004;1014:1–12.
8. Hanahan D. Heritable formation of pancreatic beta-cell tumours in transgenic mice expressing recombinant insulin/simian virus 40 oncogenes. *Nature* 1985;315:115–122.
9. Crabtree JS, Scacheri PC, Ward JM, et al. A mouse model of multiple endocrine neoplasia, type 1, develops multiple endocrine tumors. *Proc Natl Acad Sci U S A* 2001;98:1118–1123.
10. Bertolino P, Tong WM, Galendo D, et al. Heterozygous Men1 mutant mice develop a range of endocrine tumors mimicking multiple endocrine neoplasia type 1. *Mol Endocrinol* 2003;17:1880–1892.
11. Dor Y, Brown J, Martinez OI, et al. Adult pancreatic beta-cells are formed by self-duplication rather than stem-cell differentiation. *Nature* 2004;429:41–46.
12. Anlauf M, Schlenger R, Perren A, et al. Microadenomatosis of the endocrine pancreas in patients with and without the multiple endocrine neoplasia type 1 syndrome. *Am J Surg Pathol* 2006;30:560–574.
13. Vortmeyer AO, Huang S, Lubensky I, et al. Non-islet origin of pancreatic islet cell tumors. *J Clin Endocrinol Metab* 2004;89:1934–1938.
14. Bosman FT, Carneiro F, Hruban RH, et al. *WHO Classification of Tumours of the Digestive System*. 4th Ed. Lyon, France: International Agency for Research on Cancer; 2010.
15. Rindi G, Kloppel G, Alhman H, et al. TNM staging of foregut (neuro)endocrine tumors: a consensus proposal including a grading system. *Virchows Arch* 2006;449:395–401.
16. Edge SB, Byrd DR, Compton CC, et al. *AJCC Cancer Staging Manual*. 7th ed. New York: Springer; 2010.
17. Pape UF, Jann H, Muller-Nordhorn J, et al. Prognostic relevance of a novel TNM classification system for upper gastroenteropancreatic neuroendocrine tumors. *Cancer* 2008;113:256–265.
18. Ekeblad S, Skogseid B, Dunder K, et al. Prognostic factors and survival in 324 patients with pancreatic endocrine tumor treated at a single institution. *Clin Cancer Res* 2008;14:7798–7803.
19. La Rosa S, Klersy C, Uccella S, et al. Improved histologic and clinicopathologic criteria for prognostic evaluation of pancreatic endocrine tumors. *Hum Pathol* 2009;40:30–40.
20. Zerbi A, Falconi M, Rindi G, et al. Clinicopathological features of pancreatic endocrine tumors: a prospective multicenter study in Italy of 297 sporadic cases. *Am J Gastroenterol* 2010;105:1421–1429.
21. Scarpa A, Mantovani W, Capelli P, et al. Pancreatic endocrine tumors: improved TNM staging and histopathological grading permit a clinically efficient prognostic stratification of patients. *Mod Pathol* 2010;23:824–833.
22. Khan MS, Luong TV, Watkins J, et al. A comparison of Ki-67 and mitotic count as prognostic markers for metastatic pancreatic and midgut neuroendocrine neoplasms. *Br J Cancer* 2013;108:1838–1845.
23. Boninsegna L, Panzuto F, Partelli S, et al. Malignant pancreatic neuroendocrine tumour: lymph node ratio and Ki67 are predictors of recurrence after curative resections. *Eur J Cancer* 2012;48:1608–1615.
24. Sorbye H, Welin S, Langer SW, et al. Predictive and prognostic factors for treatment and survival in 305 patients with advanced gastrointestinal neuroendocrine carcinoma (WHO G3): the NORDIC NEC study. *Ann Oncol* 2013;24:152–160.
25. Fischer L, Kleeff J, Esposito I, et al. Clinical outcome and long-term survival in 118 consecutive patients with neuroendocrine tumours of the pancreas. *Br J Surg* 2008;95:627–635.
26. Bilimoria KY, Bentrem DJ, Merkow RP, et al. Application of the pancreatic adenocarcinoma staging system to pancreatic neuroendocrine tumors. *J Am Coll Surg* 2007;205:558–563.
27. Jiao Y, Shi C, Edil BH, et al. DAXX/ATRX, MEN1, and mTOR pathway genes are frequently altered in pancreatic neuroendocrine tumors. *Science* 2011;331:1199–1203.
28. Rindi G, Villanacci V, Ubiali A, et al. Endocrine tumors of the digestive tract and pancreas: histogenesis, diagnosis and molecular basis. *Expert Rev Mol Diagn* 2001;1:323–333.
29. Speel EJ, Scheidweiler AF, Zhao J, et al. Genetic evidence for early divergence of small functioning and nonfunctioning endocrine pancreatic tumors: gain of 9Q34 is an early event in insulinomas. *Cancer Res* 2001;61:5186–5192.
30. Rigaud G, Missiaglia E, Moore PS, et al. High resolution allelotype of nonfunctional pancreatic endocrine tumors: identification of two molecular subgroups with clinical implications. *Cancer Res* 2001;61:285–292.
31. Moore PS, Beghelli S, Zamboni G, et al. Genetic abnormalities in pancreatic cancer. *Mol Cancer* 2003;2:7.
32. Floridia G, Grilli G, Salvatore M, et al. Chromosomal alterations detected by comparative genomic hybridization in nonfunctioning endocrine pancreatic tumors. *Cancer Genet Cytogenet* 2005;156:23–30.
33. Jonkers YM, Claessen SM, Perren A, et al. Chromosomal instability predicts metastatic disease in patients with insulinomas. *Endocr Relat Cancer* 2005;12:435–447.
34. Karnik SK, Chen H, McLean GW, et al. Menin controls growth of pancreatic beta-cells in pregnant mice and promotes gestational diabetes mellitus. *Science* 2007;318:806–809.
35. Heaphy CM, de Wilde RF, Jiao Y, et al. Altered telomeres in tumors with ATRX and DAXX mutations. *Science* 2011;333:425.
36. Yao JC, Shah MH, Ito T, et al. Everolimus for advanced pancreatic neuroendocrine tumors. *N Engl J Med* 2011;364:514–523.
37. Larsson LI. Letter: Human pancreatic polypeptide, vasoactive intestinal polypeptide, and watery diarrhoea syndrome. *Lancet* 1976;2:149.
38. Larsson LI, Schwartz T, Lundqvist G, et al. Occurrence of human pancreatic polypeptide in pancreatic endocrine tumors. Possible implication in the watery diarrhea syndrome. *Am J Pathol* 1976;85:675–684.
39. Mortenson M, Bold RJ. Symptomatic pancreatic polypeptide-secreting tumor of the distal pancreas (PPoma). *Int J Gastrointest Cancer* 2002;32:153–156.
40. Bordi C, Togni R, Baetens D, et al. Human islet cell tumor storing pancreatic polypeptide: a light and electron microscopic study. *J Clin Endocrinol Metab* 1978;46:215–219.
41. Choksi UA, Sellin RV, Hickey RC, et al. An unusual skin rash associated with a pancreatic polypeptide-producing tumor of the pancreas. *Ann Intern Med* 1988;108:64–65.
42. Kent RB 3rd, van Heerden JA, Weiland LH. Nonfunctioning islet cell tumors. *Ann Surg* 1981;193:185–190.
43. Wermers RA, Fatourechi V, Wynne AG, et al. The glucagonoma syndrome. Clinical and pathologic features in 21 patients. *Medicine (Baltimore)* 1996;75:53–63.
44. Loyer EM, David CL, Dubrow RA, et al. Vascular involvement in pancreatic adenocarcinoma: reassessment by thin-section CT. *Abdom Imaging* 1996;21:202–206.
45. Appel BL, Tolat P, Evans DB, et al. Current staging systems for pancreatic cancer. *Cancer J* 2012;18:539–549.
46. Evans DB, Christians KK, Foley WD. Pancreaticoduodenectomy (Whipple operation) and total pancreatectomy for cancer. In: Fischer JE, Jones DB, Pomposelli FB, et al, eds. Fischer's Mastery of Surgery. 6th ed. Philadelphia: Lippincott Williams & Wilkins; 2012:1445–1465.
47. Christians KK, Riggle K, Keim R, et al. Distal splenorenal and temporary mesocaval shunting at the time of pancreatectomy for cancer: initial experience from the Medical College of Wisconsin. *Surgery* 2013;154:123–131.
48. Christians KK, Pilgrim CH, Tsai S, et al. Arterial resection at the time of pancreatectomy for cancer. *Surgery* 2014;155:919–926.
49. Goebel SU, Serrano J, Yu F, et al. Prospective study of the value of serum chromogranin A or serum gastrin levels in the assessment of the presence, extent, or growth of gastrinomas. *Cancer* 1999;85:1470–1483.
50. Bernini GP, Moretti A, Ferdeghini M, et al. A new human chromogranin "A" immunoradiometric assay for the diagnosis of neuroendocrine tumours. *Br J Cancer* 2001;84:636–642.
51. Nehar D, Lombard-Bohas C, Olivieri S, et al. Interest of Chromogranin A for diagnosis and follow-up of endocrine tumours. *Clin Endocrinol (Oxf)* 2004;60:644–652.
52. Bajetta E, Ferrari L, Martinetti A, et al. Chromogranin A, neuron specific enolase, carcinoembryonic antigen, and hydroxyindole acetic acid evaluation in patients with neuroendocrine tumors. *Cancer* 1999;86:858–865.
53. Kouvaraki MA, Ajani JA, Hoff P, et al. Fluorouracil, doxorubicin, and streptozocin in the treatment of patients with locally advanced and metastatic pancreatic endocrine carcinomas. *J Clin Oncol* 2004;22:4762–4771.
54. Yao JC, Lombard-Bohas C, Baudin E, et al. Daily oral everolimus activity in patients with metastatic pancreatic neuroendocrine tumors after failure of cytotoxic chemotherapy: a phase II trial. *J Clin Oncol* 2010;28:69–76.
55. Friesen SR, Tomita T, Kimmel JR. Pancreatic polypeptide update: its roles in detection of the trait for multiple endocrine adenopathy syndrome, type I and pancreatic polypeptide-secreting tumors. *Surgery* 1983;94:1028–1037.
56. Partelli S, Gaujoux S, Boninsegna L, et al. Pattern and clinical predictors of lymph node involvement in nonfunctioning pancreatic neuroendocrine tumors (NF-PanNETs). *JAMA Surg* 2013;148:932–939.

57. Falconi M, Bartsch DK, Eriksson B, et al. ENETS Consensus Guidelines for the management of patients with digestive neuroendocrine neoplasms of the digestive system: well-differentiated pancreatic non-functioning tumors. *Neuroendocrinology* 2012;95:120–134.
58. Gaujoux S, Partelli S, Maire F, et al. Observational study of natural history of small sporadic nonfunctioning pancreatic neuroendocrine tumors. *J Clin Endocrinol Metab* 2013;98:4784–4789.
59. Lee LC, Grant CS, Salomao DR, et al. Small, nonfunctioning, asymptomatic pancreatic neuroendocrine tumors (PNETs): role for nonoperative management. *Surgery* 2012;152:965–974.
60. Bacchetti S, Bertozzi S, Londero AP, et al. Surgical treatment and survival in patients with liver metastases from neuroendocrine tumors: a meta-analysis of observational studies. *Int J Hepatol* 2013;2013:235040.
61. Solorzano CC, Lee JE, Pisters PW, et al. Nonfunctioning islet cell carcinoma of the pancreas: survival results in a contemporary series of 163 patients. *Surgery* 2001;130:1078–1085.
62. Mayo SC, Herman JM, Cosgrove D, et al. Emerging approaches in the management of patients with neuroendocrine liver metastasis: role of liver-directed and systemic therapies. *J Am Coll Surg* 2013;216:123–134.
63. Bruns C, Lewis I, Briner U, et al. SOM230: a novel somatostatin peptidomimetic with broad somatotropin release inhibiting factor (SRIF) receptor binding and a unique antisecretory profile. *Eur J Endocrinol* 2002;146: 707–716.
64. Rinke A, Muller HH, Schade-Brittinger C, et al. Placebo-controlled, double-blind, prospective, randomized study on the effect of octreotide LAR in the control of tumor growth in patients with metastatic neuroendocrine midgut tumors: a report from the PROMID Study Group. *J Clin Oncol* 2009;27:4656–4663.
65. Caplin M, Ruszniewski P, Pavel ME, et al. A randomized double-blind placebo-controlled study of lanreotide antiproliferative response in patients with enteropancreatic neuroendocrine tumours (CLARINET), The European Cancer Congress. Amsterdam, EJC, 2013, pp abstract E17-7103.
66. Missiaglia E, Moore PS, Williamson J, et al. Sex chromosome anomalies in pancreatic endocrine tumors. *Int J Cancer* 2002;98:532–538.
67. Yao JC, Phan AT, Chang DZ, et al. Efficacy of RAD001 (everolimus) and octreotide LAR in advanced low- to intermediate-grade neuroendocrine tumors: Results of a phase II study. *J Clin Oncol* 2008;26:4311–4318.
68. Yao JC, Shah MH, Ito T, et al. Everolimus versus placebo in patients with advanced pancreatic neuroendocrine tumors (pNET) (RADIANT-3). *Ann Oncol* 2010;21.
69. Kulke MH, Lenz HJ, Meropol NJ, et al. Activity of sunitinib in patients with advanced neuroendocrine tumors. *J Clin Oncol* 2008;26:3403–3410.
70. Raymond E, Raoul J, Niccoli P, et al. Phase III, randomized, double-blind trial of sunitinib versus placebo in patients with progressive well-differentiated pancreatic islet cell tumours. *Ann Oncol* 2009;20:vii11.
71. Moertel CG, Hanley JA, Johnson LA. Streptozocin alone compared with streptozocin plus fluorouracil in the treatment of advanced islet-cell carcinoma. *N Engl J Med* 1980;303:1189–1194.
72. Moertel CG, Lefkopoulo M, Lipsitz S, et al. Streptozocin-doxorubicin, streptozocin-fluorouracil or chlorozotocin in the treatment of advanced islet-cell carcinoma. *N Engl J Med* 1992;326:519–523.
73. Broder LE, Carter SK. Pancreatic islet cell carcinoma. II. Results of therapy with streptozotocin in 52 patients. *Ann Intern Med* 1973;79:108–118.
74. Cheng PN, Saltz LB. Failure to confirm major objective antitumor activity for streptozocin and doxorubicin in the treatment of patients with advanced islet cell carcinoma. *Cancer* 1999;86:944–948.
75. McCollum AD, Kulke MH, Ryan DP, et al. Lack of efficacy of streptozocin and doxorubicin in patients with advanced pancreatic endocrine tumors. *Am J Clin Oncol* 2004;27:485–488.
76. Eriksson B, Skogseid B, Lundqvist G, et al. Medical treatment and long-term survival in a prospective study of 84 patients with endocrine pancreatic tumors. *Cancer* 1990;65:1883–1890.
77. Ramanathan RK, Cnaan A, Hahn RG, et al. Phase II trial of dacarbazine (DTIC) in advanced pancreatic islet cell carcinoma. Study of the Eastern Cooperative Oncology Group-E6282. *Ann Oncol* 2001;12:1139–1143.
78. Kulke M, Hornick J, Frauenhoffer C, et al. O6-methylguanine DNA methyltransferase deficiency and response to temozolomide-based therapy in patients with neuroendocrine tumors. *Clin Cancer Res* 2009;15: 338–345.
79. Kulke MH, Stuart K, Enzinger PC, et al. Phase II study of temozolomide and thalidomide in patients with metastatic neuroendocrine tumors. *J Clin Oncol* 2006;24:401–406.
80. Forrer F, Valkema R, Kwekkeboom DJ, et al. Peptide receptor radionuclide therapy. *Best Pract Res Clin Endocrinol Metab* 2007;21:111–129.
81. Bushnell DL Jr, O'Dorisio TM, O'Dorisio MS, et al. 90Y-edotreotide for metastatic carcinoid refractory to octreotide. *J Clin Oncol* 2010;28:1652–1659.
82. Kwekkeboom DJ, de Herder WW, Kam BL, et al. Treatment with the radiolabeled somatostatin analog [177 Lu-DOTA 0,Tyr3]octreotate: toxicity, efficacy, and survival. *J Clin Oncol* 2008;26:2124–2130.
83. Gupta S, Johnson MM, Murthy R, et al. Hepatic arterial embolization and chemoembolization for the treatment of patients with metastatic neuroendocrine tumors. *Cancer* 2005;104:1590–1602.
84. Schnirer II, Yao JC, Ajani JA. Carcinoid: A comprehensive review. *Acta Oncol* 2003;42:672–692.
85. Marrache F, Vullierme MP, Roy C, et al. Arterial phase enhancement and body mass index are predictors of response to chemoembolisation for liver metastases of endocrine tumours. *Br J Cancer* 2007;96:49–55.
86. Rhee TK, Lewandowski RJ, Liu DM, et al. 90Y Radioembolization for metastatic neuroendocrine liver tumors: preliminary results from a multi-institutional experience. *Ann Surg* 2008;247:1029–1035.
87. Kennedy AS, Dezarn WA, McNeillie P, et al. Radioembolization for unresectable neuroendocrine hepatic metastases using resin 90Y-microspheres: early results in 148 patients. *Am J Clin Oncol* 2008;31:271–279.
88. Andrews JC, Walker SC, Ackermann RJ, et al. Hepatic radioembolization with yttrium-90 containing glass microspheres: preliminary results and clinical follow-up. *J Nucl Med* 1994;35:1637–1644.
89. Sarmiento JM, Heywood G, Rubin J, et al. Surgical treatment of neuroendocrine metastases to the liver: a plea for resection to increase survival. *J Am Coll Surg* 2003;197:29–37.
90. Treut YP, Delpero J, Dousset B, et al. Results of liver transplantation in the treatment of metastatic neuroendocrine tumors. *Ann Surg* 1997;225: 355–364.
91. Berber E, Flesher N, Siperstein AE. Laparoscopic radiofrequency ablation of neuroendocrine liver metastases. *World J Surg* 2002;26:985–990.
92. Ito T, Igarashi H, Jensen RT. Zollinger-Ellison syndrome: recent advances and controversies. *Curr Opin Gastroenterol* 2013;29:650–661.
93. Berna MJ, Hoffmann KM, Long SH, et al. Serum gastrin in Zollinger-Ellison syndrome: II. Prospective study of gastrin provocative testing in 293 patients from the National Institutes of Health and comparison with 537 cases from the literature. evaluation of diagnostic criteria, proposal of new criteria, and correlations with clinical and tumoral features. *Medicine* 2006;85: 331–364.
94. Norton JA, Jensen RT. Resolved and unresolved controversies in the surgical management of patients with Zollinger-Ellison syndrome. *Ann Surg* 2004;240:757–773.
95. Tomassetti P, Migliori M, Caletti GC, et al. Treatment of type II gastric carcinoid tumors with somatostatin analogues. *N Engl J Med* 2000;343:551–554.
96. Service FJ, McMahon MM, O'Brien PC, et al. Functioning insulinoma—incidence, recurrence, and long-term survival of patients: a 60-year study. *Mayo Clin Proc* 1991;66:711–719.
97. Niina Y, Fujimori N, Nakamura T, et al. The current strategy for managing pancreatic neuroendocrine tumors in multiple endocrine neoplasia type 1. *Gut Liver* 2012;6:287–294.
98. Doppman JL, Chang R, Fraker DL, et al. Localization of insulinomas to regions of the pancreas by intra-arterial stimulation with calcium. *Ann Intern Med* 1995;123:269–273.
99. Service FJ. Recurrent hyperinsulinemic hypoglycemia caused by an insulin-secreting insulinoma. *Nat Clin Pract Endocrinol Metab* 2006;2:467–470; quiz following 470.
100. Hirshberg B, Cochran C, Skarulis MC, et al. Malignant insulinoma: spectrum of unusual clinical features. *Cancer* 2005;104:264–272.
101. Stehouwer CD, Lems WF, Fischer HR, et al. Aggravation of hypoglycemia in insulinoma patients by the long-acting somatostatin analogue octreotide (Sandostatin). *Acta Endocrinol (Copenh)* 1989;121:34–40.
102. Aspinwall CA, Lakey JR, Kennedy RT. Insulin-stimulated insulin secretion in single pancreatic beta cells. *J Biol Chem* 1999;274:6360–6365.
103. Leibiger IB, Leibiger B, Moede T, et al. Exocytosis of insulin promotes insulin gene transcription via the insulin receptor/PI-3 kinase/p70 s6 kinase and CaM kinase pathways. *Mol Cell* 1998;1:933–938.
104. Kulke MH, Bergsland EK, Yao JC. Glycemic control in patients with insulinoma treated with everolimus. *N Engl J Med* 2009;360:195–197.
105. Fiebrich HB, Siemerink EJ, Brouwers AH, et al. Everolimus induces rapid plasma glucose normalization in insulinoma patients by effects on tumor as well as normal tissues. *Oncologist* 2011;16:783–787.
106. Bernard V, Lombard-Bohas C, Taquet MC, et al. Efficacy of everolimus in patients with metastatic insulinoma and refractory hypoglycemia. *Eur J Endocrinol* 2013;168:665–674.
107. Verner JV, Morrison AB. Islet cell tumor and a syndrome of refractory watery diarrhea and hypokalemia. *Am J Med* 1958;25:374–380.
108. Gorden P, Comi RJ, Maton PN, et al. NIH conference. Somatostatin and somatostatin analogue (SMS 201-995) in treatment of hormone-secreting tumors of the pituitary and gastrointestinal tract and non-neoplastic diseases of the gut. *Ann Intern Med* 1989;110:35–50.
109. Altimari AF, Bhoopalam N, O'Dorsio T, et al. Use of a somatostatin analog (SMS 201-995) in the glucagonoma syndrome. *Surgery* 1986;100: 989–996.
110. Norton JA, Kahn CR, Schiebinger R, et al. Amino acid deficiency and the skin rash associated with glucagonoma. *Ann Intern Med* 1979;91:213–215.
111. Bloom SR, Polak JM. Glucagonoma syndrome. *Am J Med* 1987;82:25–36.
112. Mao C, Shah A, Hanson DJ, et al. Von Recklinghausen's disease associated with duodenal somatostatinoma: contrast of duodenal versus pancreatic somatostatinomas. *J Surg Oncol* 1995;59:67–73.
113. Norton JA, Fraker DL, Alexander HR, et al. Surgery to cure the Zollinger-Ellison syndrome. *N Engl J Med* 1999;341:635–644.
114. Norton JA, Doppman JL, Jensen RT. Curative resection in Zollinger-Ellison syndrome. Results of a 10-year prospective study. *Ann Surg* 1992;215:8–18.
115. Jensen RT. Management of the Zollinger-Ellison syndrome in patients with multiple endocrine neoplasia type 1. *J Intern Med* 1998;243:477–488.

116. Doherty GM, Olson JA, Frisella MM, et al. Lethality of multiple endocrine neoplasia type I. *World J Surg* 1998;22:581–586; discussion 586–587.
117. Norton JA, Alexander HR, Fraker DL, et al. Comparison of surgical results in patients with advanced and limited disease with multiple endocrine neoplasia type 1 and Zollinger-Ellison syndrome. *Ann Surg* 2001;234:495–505; discussion 505–506.
118. Ruszniewski P, Rougier P, Roche A, et al. Hepatic arterial chemoembolization in patients with liver metastases of endocrine tumors. A prospective phase II study in 24 patients. *Cancer* 1993;71:2624–2630.
119. Kouvaraki MA, Shapiro SE, Cote GJ, et al. Management of pancreatic endocrine tumors in multiple endocrine neoplasia type 1. *World J Surg* 2006; 30:643–653.
120. Lowney JK, Frisella MM, Lairmore TC, et al. Pancreatic islet cell tumor metastasis in multiple endocrine neoplasia type 1: correlation with primary tumor size. *Surgery* 1998;124:1043–1048, discussion 1048–1049.
121. Thompson NW. Current concepts in the surgical management of multiple endocrine neoplasia type 1 pancreatic-duodenal disease. Results in the treatment of 40 patients with Zollinger-Ellison syndrome, hypoglycaemia or both. *J Intern Med* 1998;243:495–500.
122. Thompson NW. Management of pancreatic endocrine tumors in patients with multiple endocrine neoplasia type 1. *Surg Oncol Clin N Am* 1998;7: 881–891.
123. Lindau A. Zur frage der angiomatosis retinae und ihrer hirncomplikation. *Acta Ophthal* 1927;4:193–226.
124. Charlesworth M, Verbeke CS, Falk GA, et al. Pancreatic lesions in von Hippel-Lindau disease? A systematic review and meta-synthesis of the literature. *J Gastrointest Surg* 2012;16:1422–1428.
125. Zhao J, Moch H, Scheidweiler AF, et al. Genomic imbalances in the progression of endocrine pancreatic tumors. *Genes Chromosomes Cancer* 2001;32:364–372.
126. Plank TL, Logginidou H, Klein-Szanto A, et al. The expression of hamartin, the product of the TSC1 gene, in normal human tissues and in TSC1- and TSC2-linked angiomyolipomas. *Mod Pathol* 1999;12:539–545.
127. Yao JC. Molecular targeted therapy for carcinoid and islet-cell carcinoma. *Best Pract Res Clin Endocrinol Metab* 2007;21:163–172.
128. Johannessen CM, Reczek EE, James MF, et al. The NF1 tumor suppressor critically regulates TSC2 and mTOR. *Proc Natl Acad Sci U S A* 2005;102: 8573–8578.
129. DeLellis RA, Lloyd RV, Heitz PU, et al. *WHO Classification of Tumours, Pathology and Genetics of Tumours of Endocrine Organs*. Lyon, France: IARC Press; 2004.
130. Moertel CG, Kvols LK, O'Connell MJ, et al. Treatment of neuroendocrine carcinomas with combined etoposide and cisplatin. Evidence of major therapeutic activity in the anaplastic variants of these neoplasms. *Cancer* 1991;68:227–232.
131. Hou Z, Elasmar SA, Lozano R, et al. A pilot study of irinotecan plus cisplatin in patients with metastatic high-grade neuroendocrine carcinoma. *Proc Am Soc Clin Oncol* 2003;22:1508.
132. Yao JC, Vauthey JN. Primary and metastatic hepatic carcinoid: is there an algorithm? *Ann Surg Oncol* 2003;10:1133–1135.