Autoimmune pancreatitis (AIP) was first classified as a defined disease entity in 1995. It accounts for approximately 2% of cases of chronic pancreatitis (western world prevalence 36-41/100,000 inhabitants) and AIP is diagnosed in 2.4% of pancreas resection specimens. Presentation of strategies for diagnosis and treatment with focus on differentiation of AIP and pancreatic carcinoma. Selective literature research in PubMed regarding pathogenesis, diagnosis and treatment of AIP. Key characteristics of AIP are recurrent jaundice due to obstructed bile ducts, histological evidence of fibrosis, a lymphoplasmocytic or granulocytic infiltrate and the response to steroid therapy. There are two distinctive forms of AIP: type I or lymphoplasmocytic sclerosing pancreatitis and type II or idiopathic duct centric pancreatitis. The IgG4 positive AIP type I belongs to the group of IgG4-related systemic diseases. Diagnosis of AIP is established according to the international consensus diagnostic criteria (ICDC) or HISORT (mnemonic standing for histology, imaging, serology, other organ involvement and response to therapy) criteria. Differentiation from pancreatic adenocarcinoma can be challenging. The standard treatment consists of corticosteroids and in some cases azathioprine can be added. In refractory disease rituximab is a further option. Treatment is indicated in patients with jaundice, systemic
manifestation or persistent pain. Although AIP is increasingly being identified, the differentiation from pancreatic adenocarcinoma still remains difficult and in cases of a suspicion of neoplasia, resection should be favored. It can successfully be treated conservatively with steroids and rituximab.