# Pancreatic Cystic Neoplasms: Diagnosis and Management

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- Pancreatic cystic neoplasm
- Intraductal papillary mucinous neoplasm
- Mucinous cystic neoplasm • Serous cystic neoplasm
- Solid-pseudopapillary neoplasm

Pancreatic cystic lesions are being detected with increasing frequency, at least partly because of the increased use of cross-sectional imaging for the evaluation of abdominal complaints or screening for other conditions.[1] Image-based studies report prevalences of pancreatic cystic lesions ranging from 1.2% to 19.6%.[2–4] In a study of 1064 pancreatic cystic neoplasm (PCN) patients pathologically confirmed over a 12.5-year period, 108 (10.5%) of the patients had pancreatic lesions that were detected while undergoing workup for other diseases.[5]

PCNs are reported to account for up to 60% of all pancreatic cystic lesions, followed by injury-related and inflammation-related cysts (30%).[6] PCNs include intraductal papillary mucinous neoplasm (IPMN), mucinous cystic neoplasm (MCN), serous cystic neoplasm (SCN), solid-pseudopapillary neoplasm (SPN), cystic neuroendocrine neoplasm, ductal adenocarcinoma with cystic degeneration, and acinar-cell cystic neoplasm.[7] The proportion of PCNs varies with population. In the Western Hemisphere, SCNs account for 32% to 39%, MCNs for 10% to 45%, IPMNs for 21% to 33%, and SPNs for less than 10% of all PCNs.[7] A nationwide survey from Korea reports the proportions of PCN which are composed of IPMNs (41.0%), MCNs (25.2%), SPNs (18.3%), SCNs (15.2%), and others (0.3%).[5]

## THE MAJOR PANCREATIC CYSTIC NEOPLASMS

The 4 major types of PCNs are IPMN, MCN, SCN, and SPN. Distinguishing among the 4 most common types of cysts is important, since the management varies with each type of cyst.

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### *Intraductal Papillary Mucinous Neoplasms*

IPMNs are defined as intraductal grossly visible (typically ≥1.0 cm) epithelial neoplasms of mucin-producing cells, arising in the main pancreatic duct or its branches.[8] The first report was 4 cases with the triad of mucus secretion, main pancreatic duct dilatation, and a swollen duodenal papilla.[9] Since then, the number of IPMN cases reported has been increasing significantly. This may be due to a true increase in the incidence, improvement in the understanding of IPMN, and/or increased use of cross-sectional imaging in clinical practice.[1,7]

IPMNs are usually diagnosed in the elderly, often diagnosed in the seventh decade of life.[5,10–13] There is a slight male preponderance.[5,10,11,13] Although the true incidence of IPMN is unknown, IPMNs are reported to be the most common PCN. IPMN accounts for approximately 1% to 3% of pancreatic exocrine neoplasms and 20% to 50% of all PCNs.[5,7,8,14, 15] Most patients diagnosed with IPMN are asymptomatic and are usually incidentally diagnosed. Symptomatic patients present with abdominal pain, pancreatitis, weight loss, diabetes mellitus, and jaundice.[5,10–13]

The World Health Organization (WHO) classification of tumors of the pancreas, including IPMN, was modified in 2010. In the 2000 WHO classification, IPMNs were separated according to the degree of dysplasia into intraductal papillary mucinous adenoma; intraductal papillary mucinous neoplasm with moderate dysplasia; intraductal papillary mucinous carcinoma, noninvasive; and intraductal papillary mucinous carcinoma, invasive.[16] In the 2010 WHO classification, IPMNs were classified into IPMN with low- or intermediate-grade dysplasia (which includes the former intraductal papillary mucinous adenoma and intraductal papillary mucinous neoplasm with moderate dysplasia), IPMN with high-grade dysplasia (which is equivalent to the intraductal papillary mucinous carcinoma, noninvasive), and IPMN with an associated invasive carcinoma (changed from intraductal papillary mucinous carcinoma, invasive).[8]

Macroscopically, depending on the pancreatic ductal system involved, IPMNs are classified as either main-duct IPMN (MD-IPMN), branch-duct (BD-IPMN), or combined-type IPMN.[17] The clinicopathologic behavior of combined-type IPMN is similar to that of MD-IPMN.[13] Intraductal proliferation of columnar mucin-producing cells is the main histologic characteristics of IPMN. The neoplastic epithelium may show diverse architecture and cytology. Four subtypes of IPMNs have been characterized: gastric, intestinal, pancreatobiliary, and oncocytic.[18] In a recent report, the 4 subtypes of IPMNs were associated with significant differences in survival. Patients with gastric-type IPMN had the best prognosis, whereas those with pancreatobiliary type had the worst prognosis.[19]

Most IPMNs are diagnosed incidentally. Even symptomatic patients present with nonspecific symptoms such as abdominal pain, malaise, nausea/vomiting, and weight loss. In patients with an associated invasive carcinoma, symptoms and signs of pancreatic ductal adenocarcinoma such as weight loss, diabetes mellitus, and/or painless jaundice may be observed.[11,20,21]

Routine blood tests, such as complete blood count, liver function test, amylase, and lipase, are usually within normal limits or show nonspecific changes.[22] Serum CA 19-9 and carcinoembryonic antigen (CEA) are generally not of diagnostic value. However, there is a report that serum CA 19-9 was elevated in 74% of patients with invasive IPMN and in 14% of patients with noninvasive IPMN. Furthermore, it is reported that 80% of patients with invasive IPMN had elevated serum levels of CA 19-9 and/or CEA, compared with 18% of patients with noninvasive IPMN.[23]