

An Algorithmic approach to cystic neoplasms of the pancreas



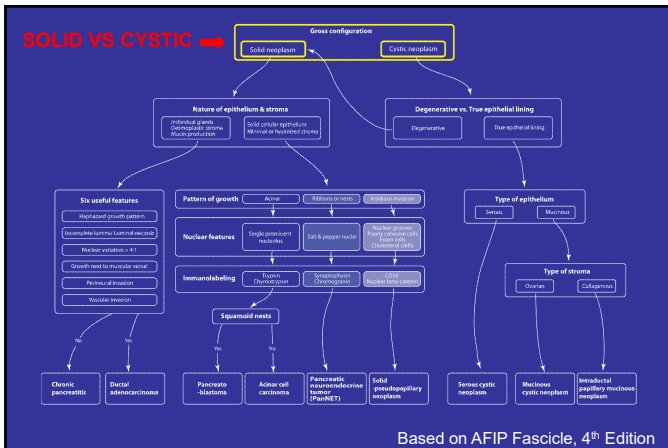
JOHNS HOPKINS
MEDICAL INSTITUTIONS

February 2017

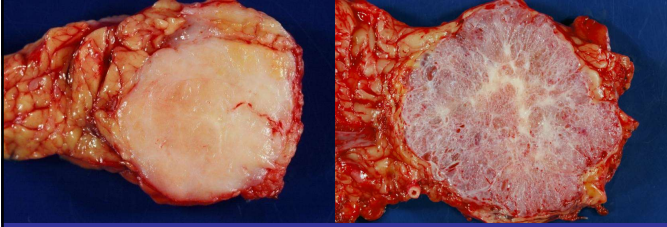
Ralph H. Hruban, M.D.
Professor and Director of Pathology
The Sol Goldman Pancreatic Cancer Research Center
The Johns Hopkins Medical Institutions

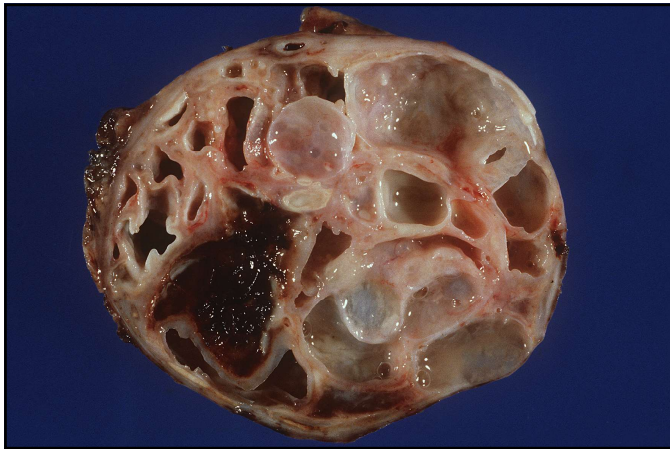
Disclosure

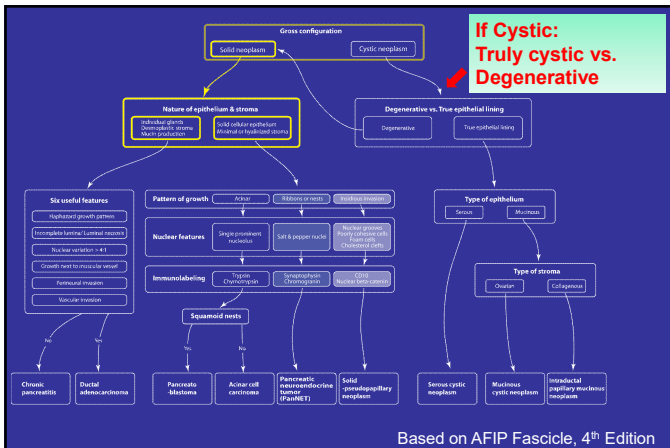
- I receive royalty payments from Myriad Genetics for the PALB2 invention.
- Selected images from the AFIP Fascicle with permission
- The pancreas pathology iPad APPs are free through the iTunes Store



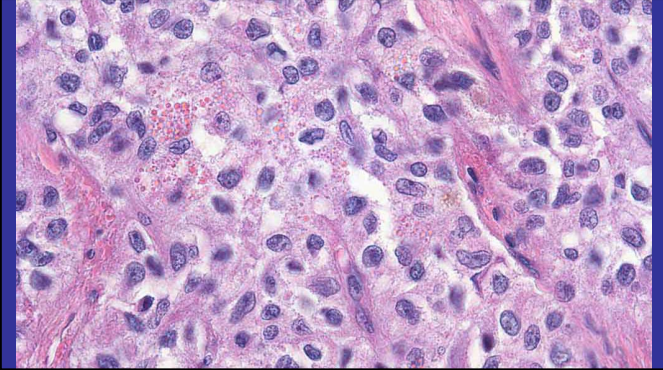
1. Is it Solid or Cystic?



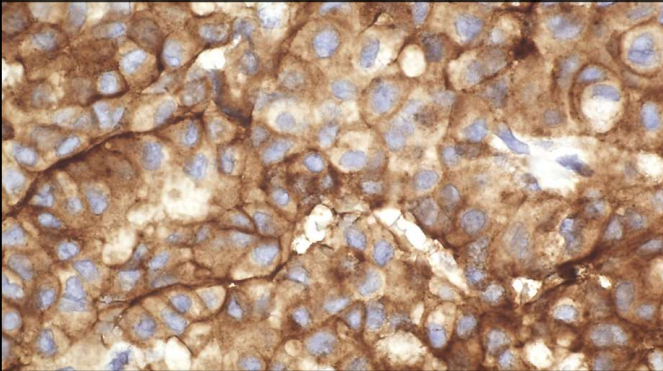




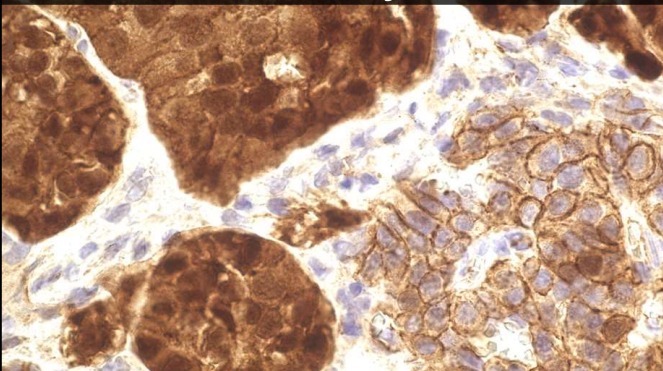
Nuclear: Grooves



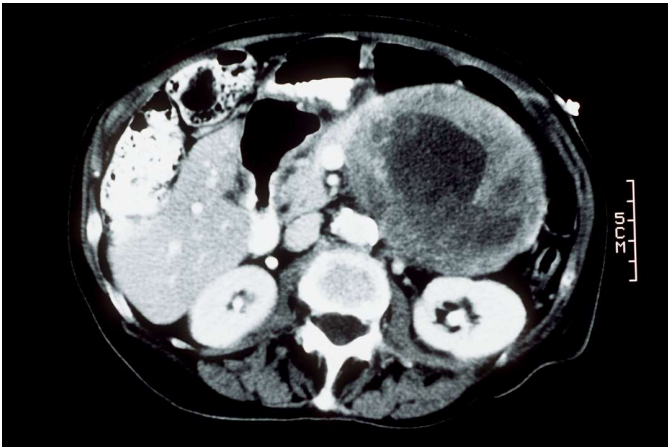
Immunohistochemistry: CD10



Immunohistochemistry: Beta-catenin

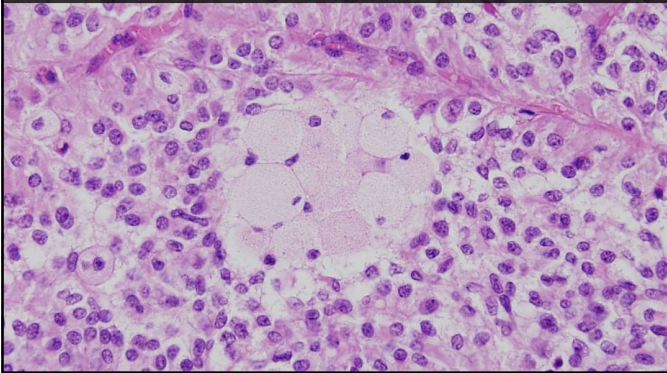


Solid- Pseudopapillary Neoplasm

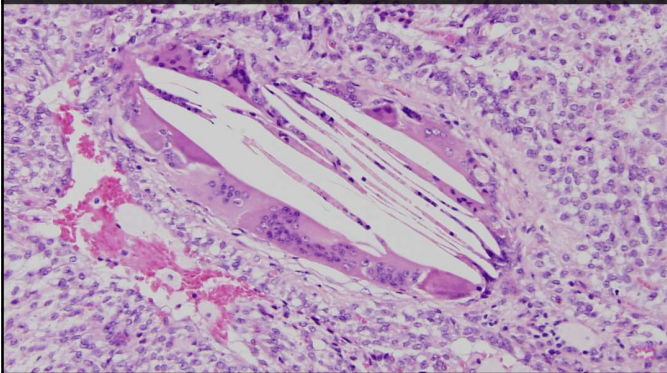




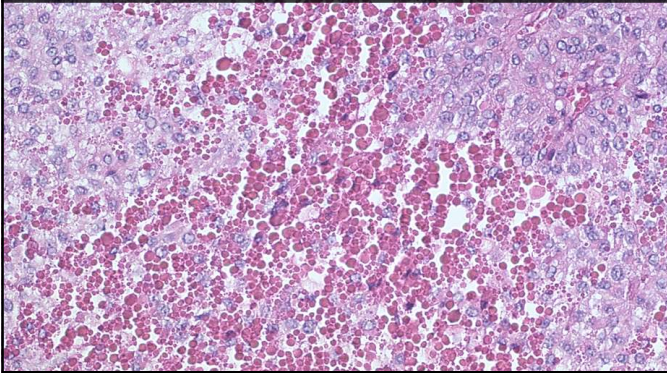
Foam Cells

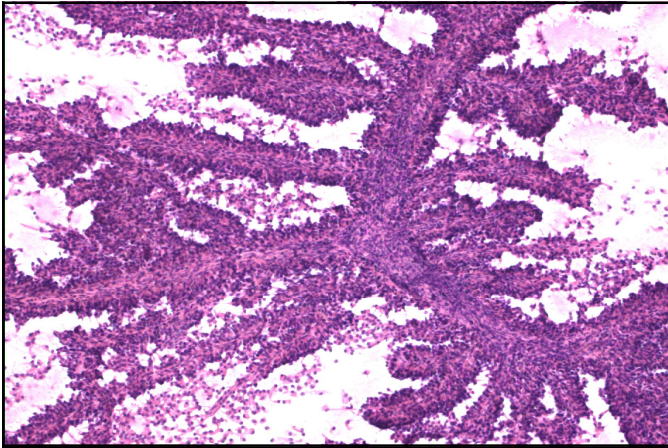


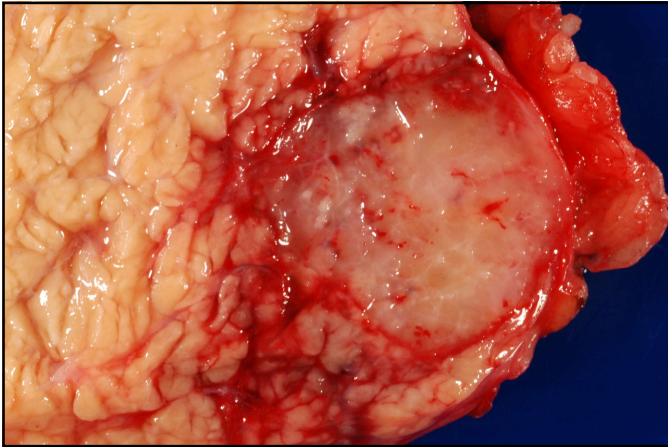
Cholesterol Clefts

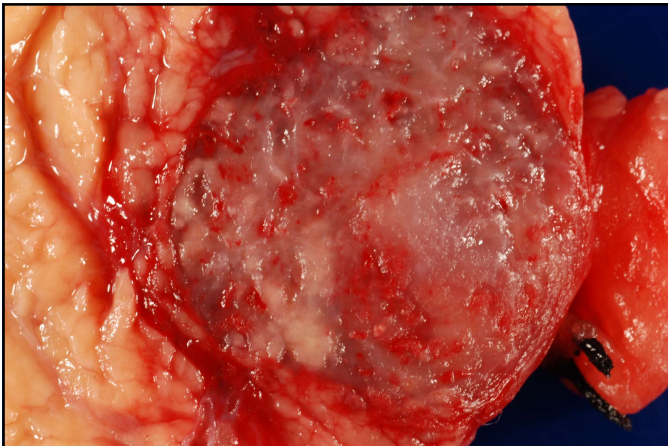


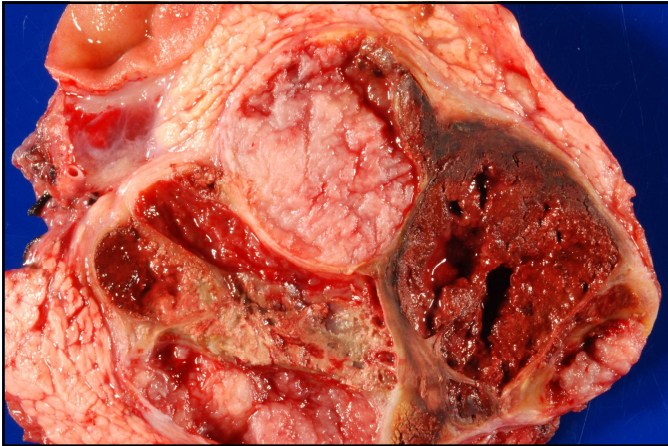
Hyaline Globules

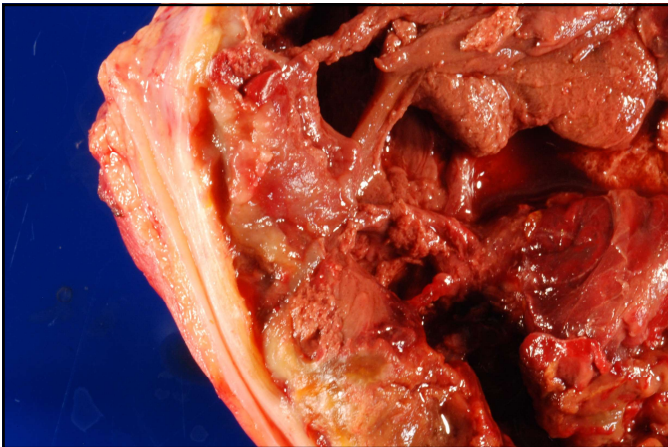






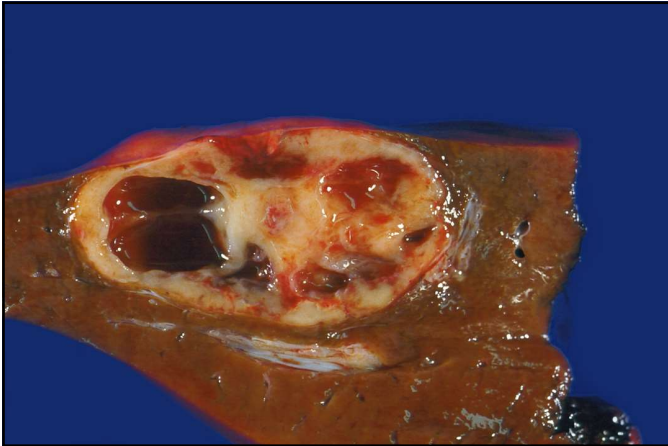






Solid-Pseudopapillary Neoplasm

- Clinically, the vast majority occur in young women (20's)
- Patients present with vague abdominal fullness or pain
- Grossly well demarcated masses. On cross section, they are cystic and solid with areas of hemorrhage and necrosis



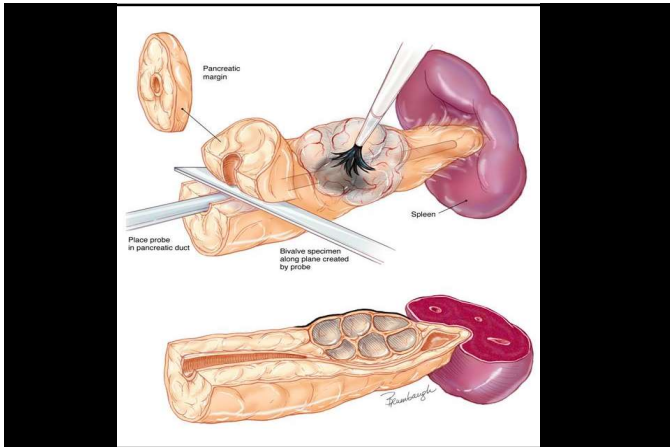
Solid-Pseudopapillary Neoplasms: Outcome

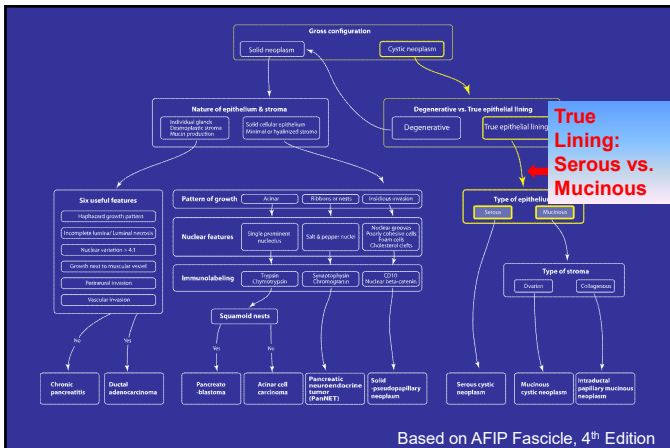
- All are classified as malignant.
- Some may be locally aggressive, but most are surgically cured.

Truly Cystic Neoplasms

- Gender
- Tail vs. Head
- Relationship to larger pancreatic ducts
- Character of cyst fluid
- Lining (serous vs. mucinous vs. none)
- Stroma (ovarian-type)

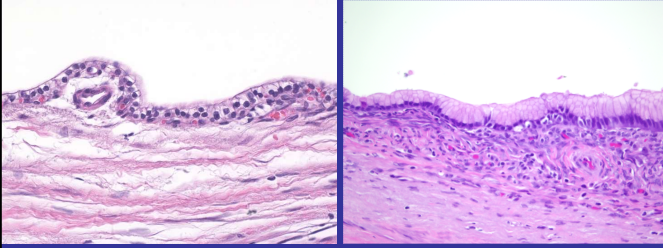
	Mucinous Cystic Neoplasm	Intraductal papillary Mucinous Neoplasm	Solid-pseudopapillary Neoplasm	Serous Cystic Neoplasm
Gender (F:M)	20:1	1:1.5	10:1	7:3
Head/Tail	Tail	Head	Tail=Head	Tail=Head
Relation to Duct	None	Always	None	None
Cyst Contents	Mucinous	Mucinous	Necrotic/Hemorrhagic	Serous
Epithelium	Mucinous	Mucinous	Non-cohesive	Serous
Stroma	Ovarian	None	None	None



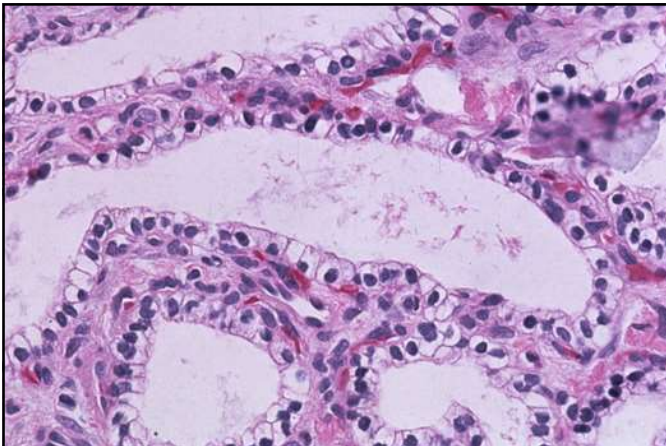


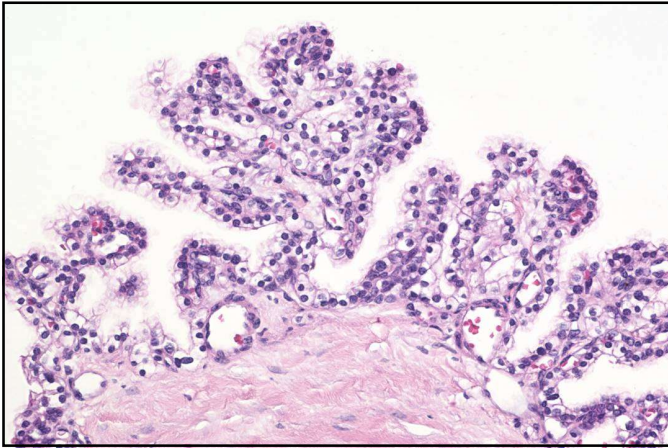
Based on AFIP Fascicle, 4th Edition

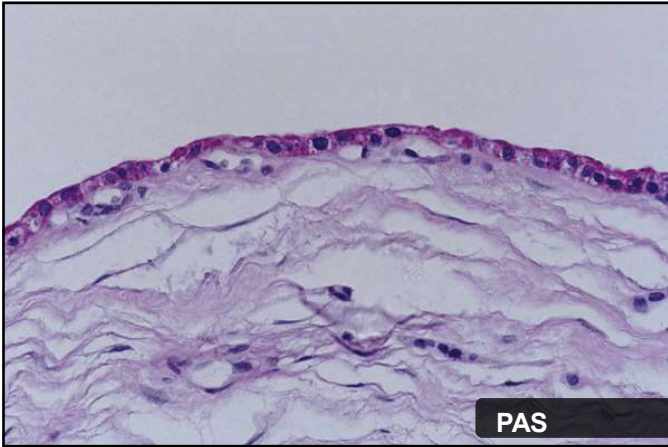
Serous vs. Mucinous

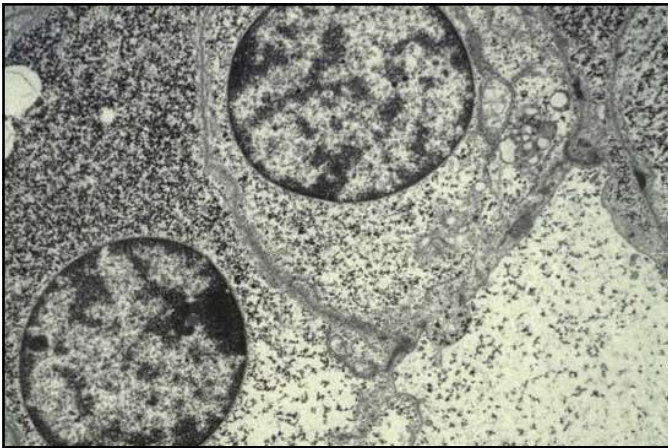


Serous Neoplasms







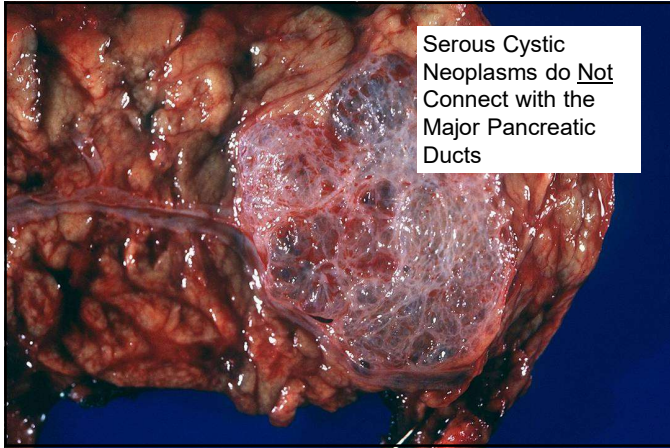


Serous Cystadenomas Clinical

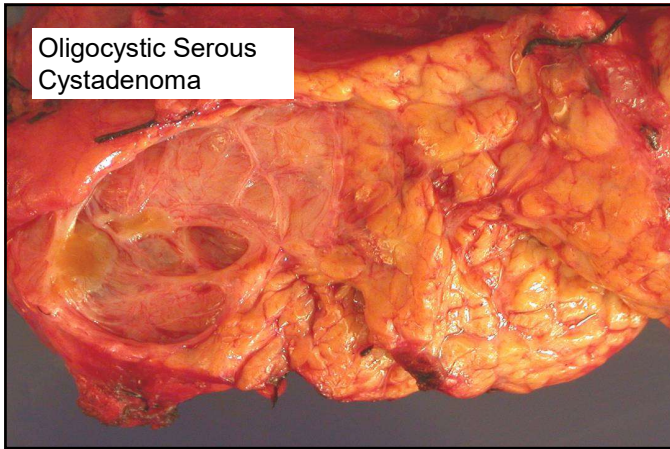
- More common in women than in men
- Average age at diagnosis: 61-68 years
- Presenting signs and symptoms include:
 - abdominal pain
 - weight loss
 - palpable abdominal mass



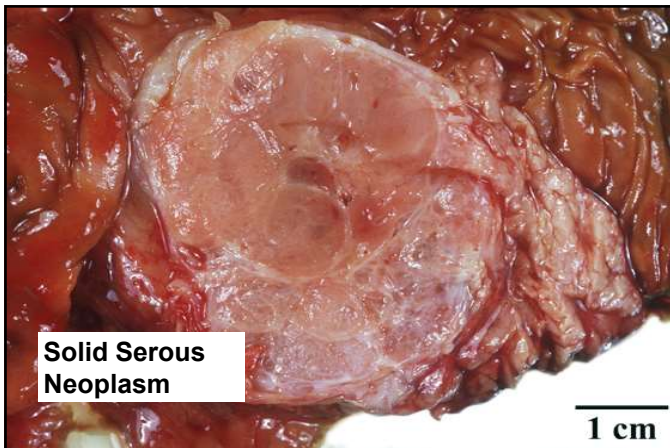




Serous Cystic Neoplasms do **Not** Connect with the Major Pancreatic Ducts

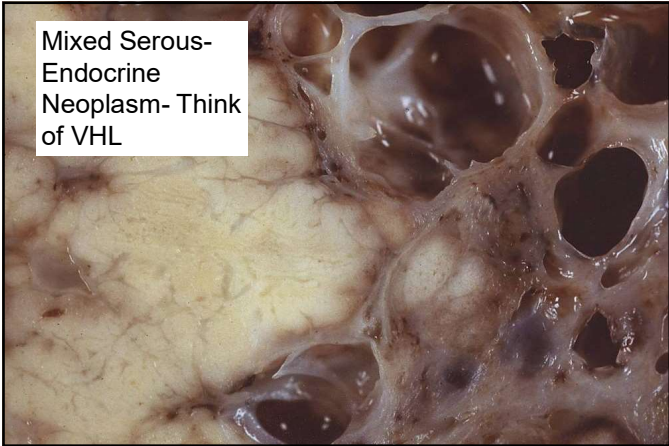


Oligocystic Serous Cystadenoma



Solid Serous Neoplasm

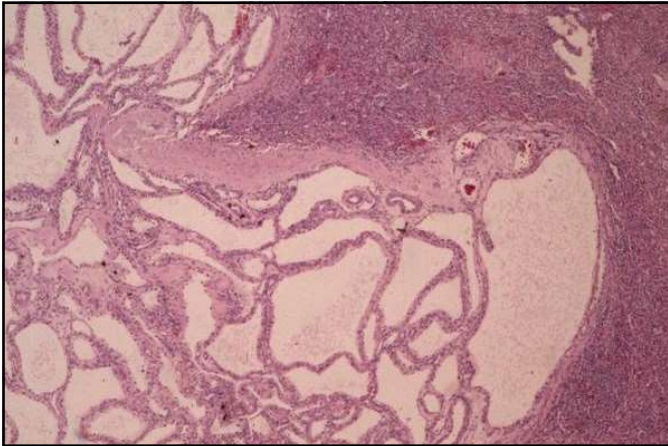
1 cm



Serous Cystadenoma Outcome

- Several case reports of “multifocal” disease
- One or two cases of malignant serous neoplasms
- The vast majority of serous cystadenomas are benign, even if incompletely resected





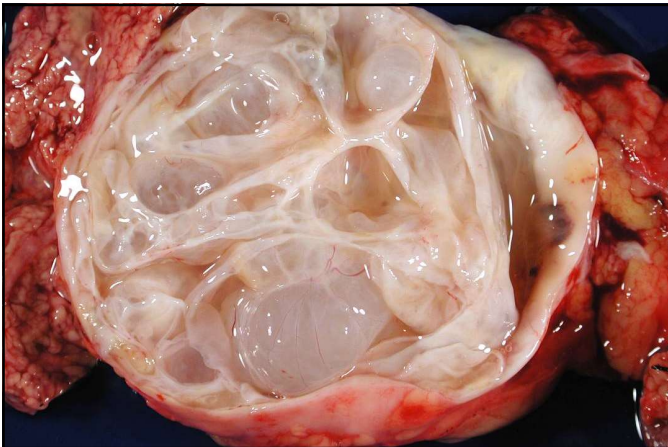
Serous Cystadenoma Differential Diagnosis

	<u>Serous cystadenoma</u>	<u>Lymphangioma</u>
Cytokeratin	+	-
Glycogen	+	-
Lymphocytes in the wall	-	+

Mucin-producing Neoplasms (MCNs and IPMNs)

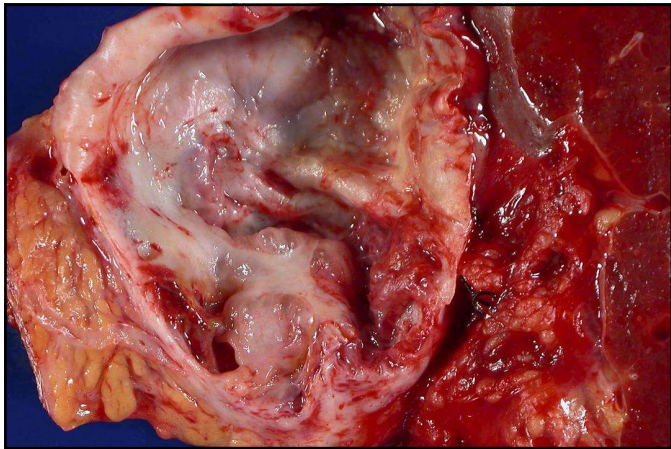
Mucinous Cystic Neoplasms

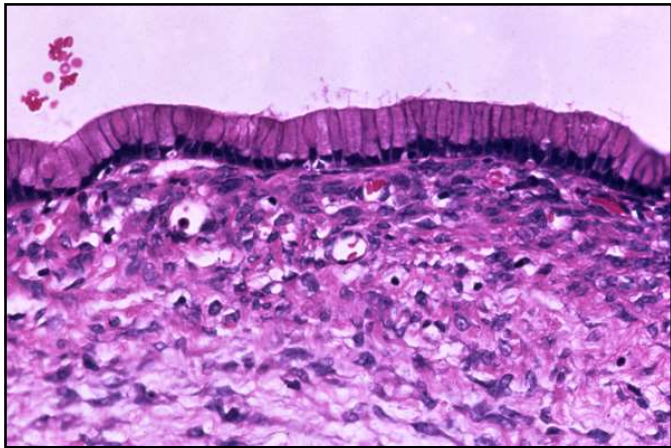
- Much more common in women than in men
- Mean age at diagnosis: ~50 (younger than for patients with serous cystadenomas)
- Tail > Head

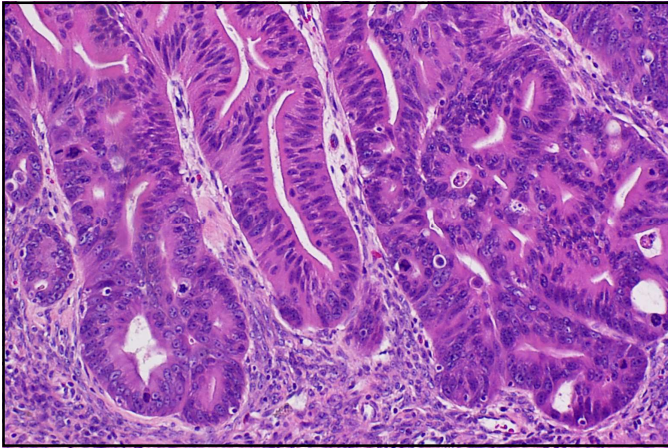


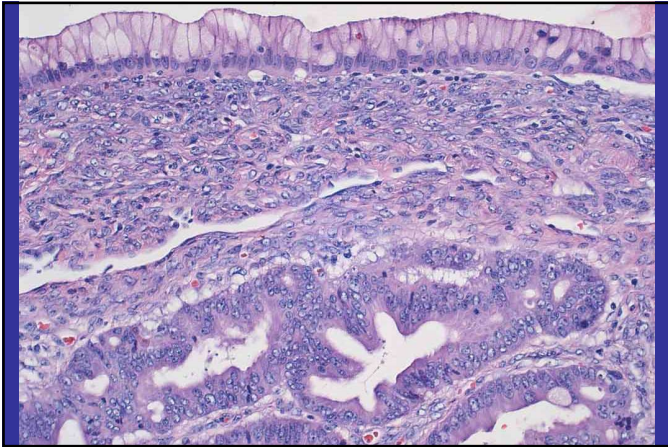


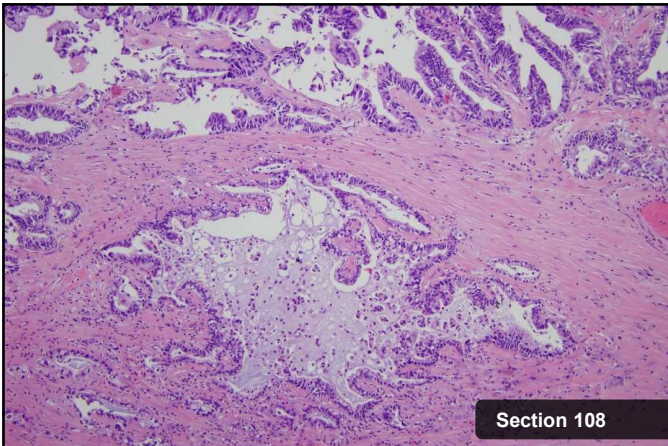










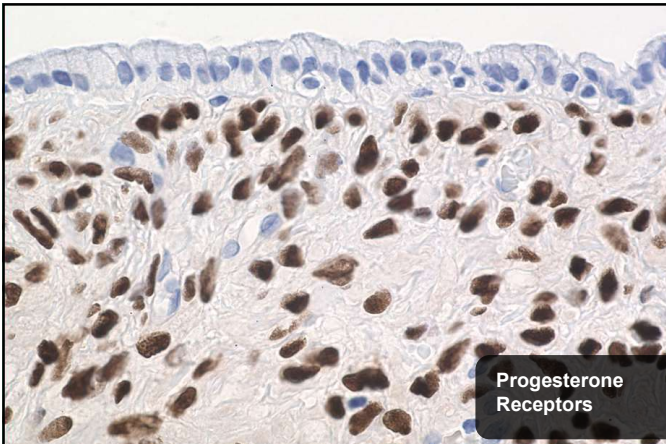


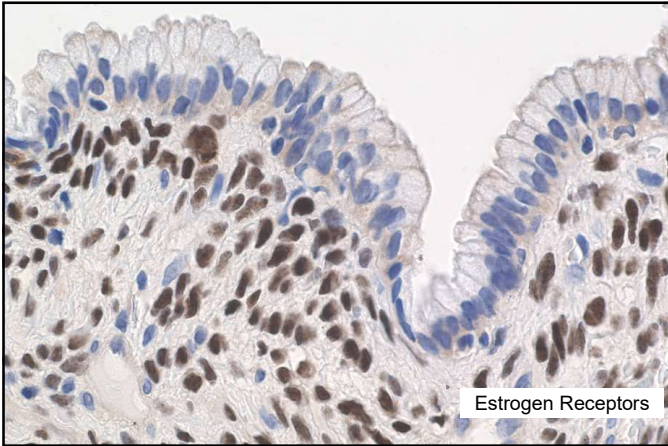
Mucinous Cystic Neoplasms

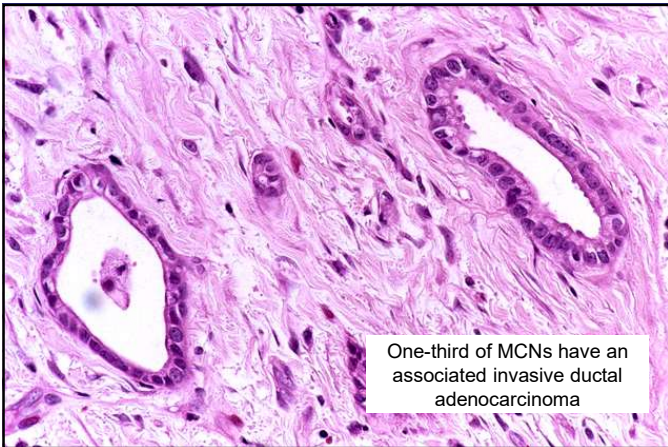
- Low-grade Dysplasia
 - Intermediate grade Dysplasia
 - High-grade Dysplasia
-
- Invasive Carcinoma

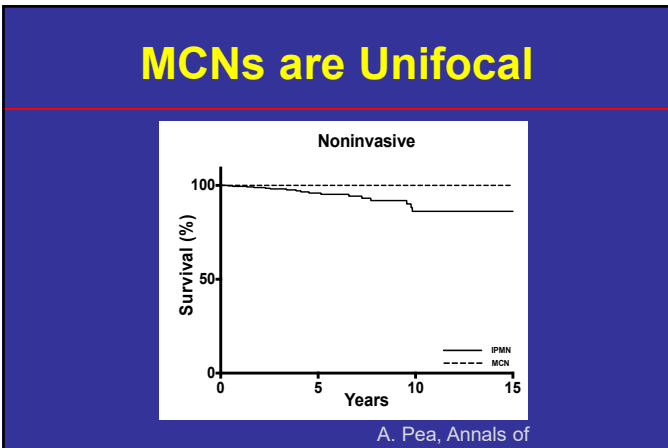
Mucinous Cystic Neoplasms- Proposed

- Low-grade Dysplasia
 - High-grade Dysplasia
-
- Invasive Carcinoma









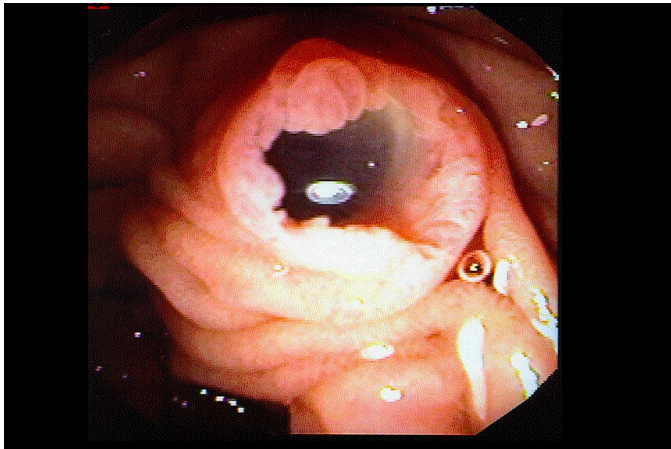
Intraductal Papillary Mucinous Neoplasm

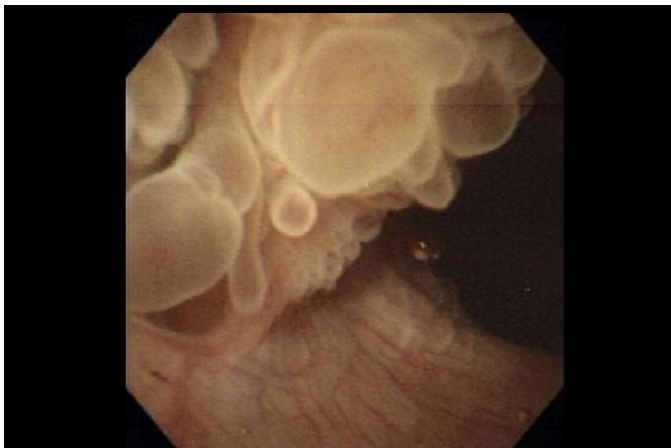


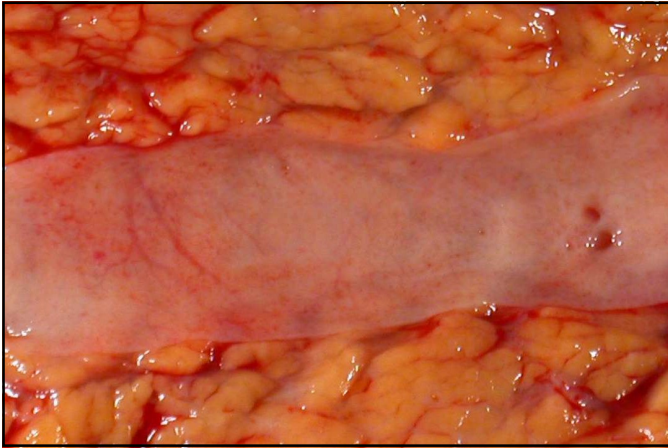
Intraductal Papillary Neoplasms

- Long history of symptoms
- Incidence in men equals that in women
- Head > Tail
- Mucin oozing from the ampulla of Vater



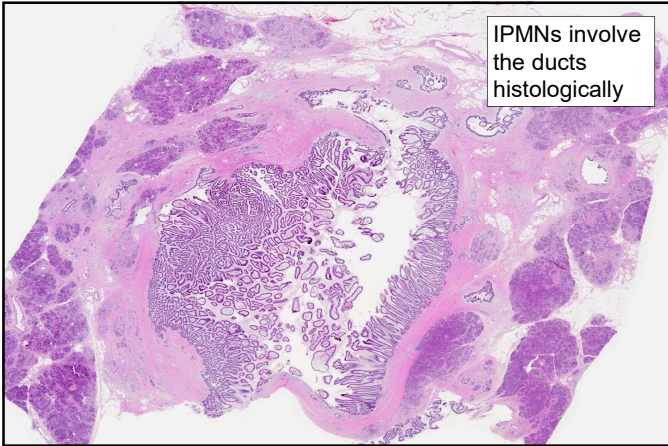


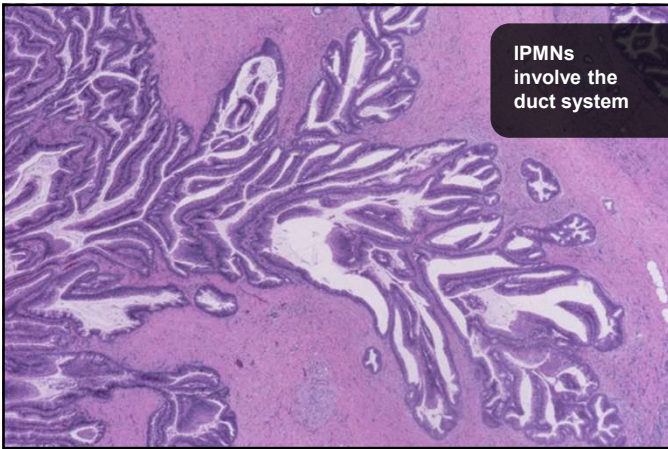


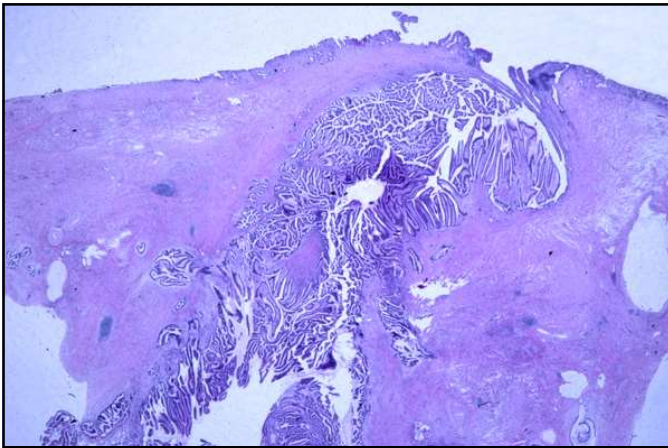










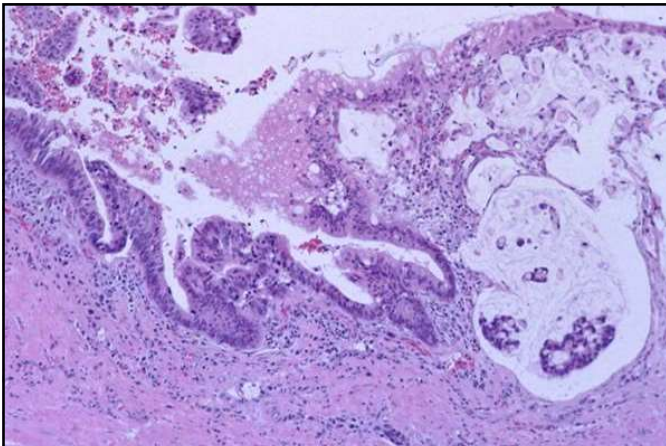


Intraductal Papillary Neoplasms

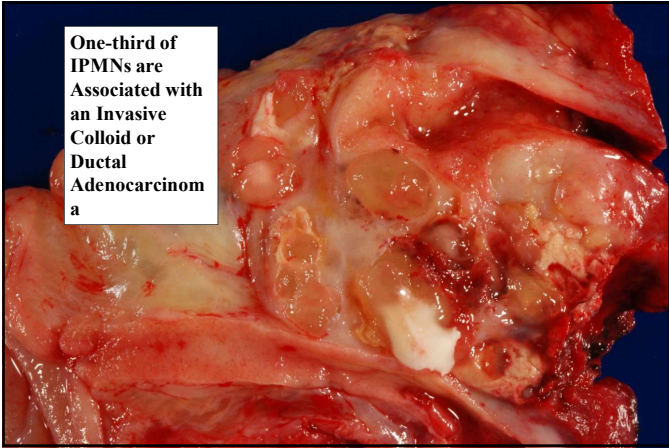
- IPMN with low-grade dysplasia
 - IPMN with intermediate dysplasia
 - IPMN with high-grade dysplasia
-
- IPMN with an invasive carcinoma
 - Colloid
 - Tubular/ductal

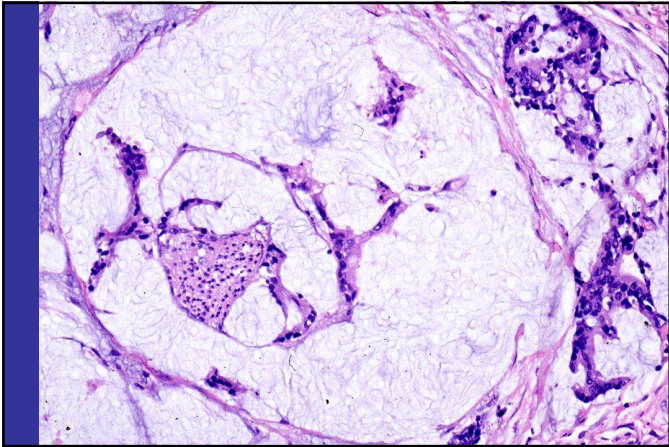
Intraductal Papillary Neoplasms Proposed

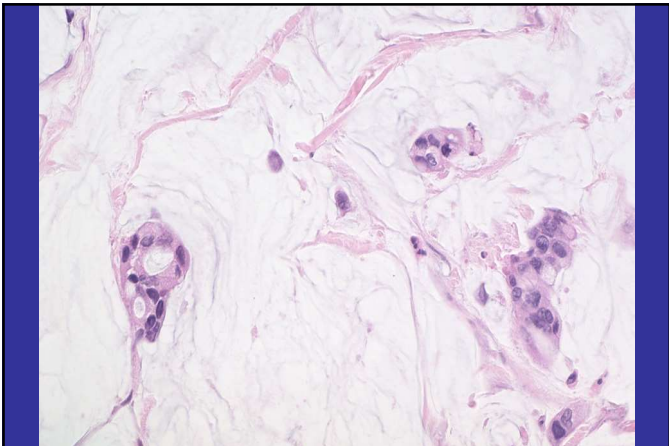
- IPMN with low-grade dysplasia
 - IPMN with high-grade dysplasia
-
- IPMN with an invasive carcinoma
 - Colloid
 - Tubular/ductal

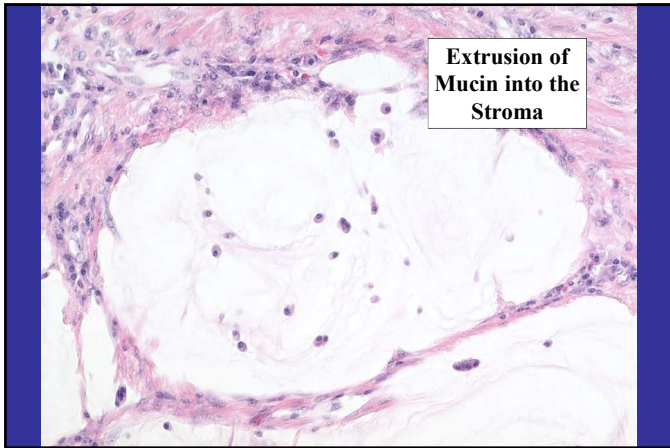


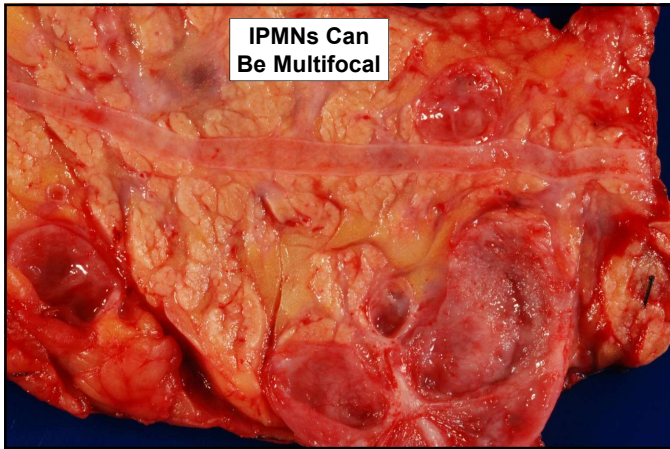
One-third of IPMNs are Associated with an Invasive Colloid or Ductal Adenocarcinoma

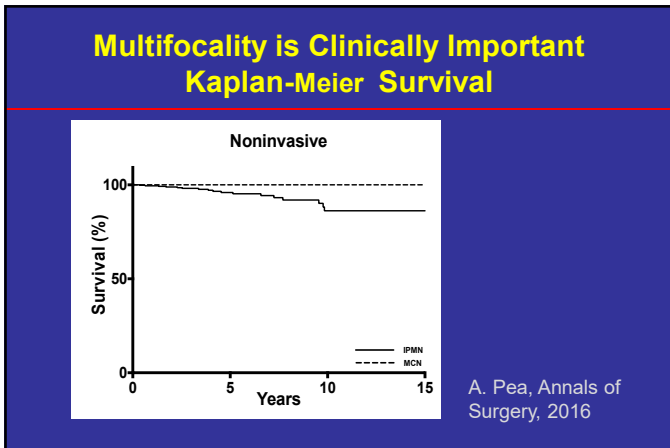




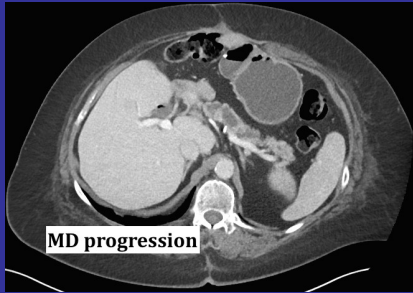








Recurrence of Disease in the Remnant Pancreas is a Big Problem for Patients with an IPMN

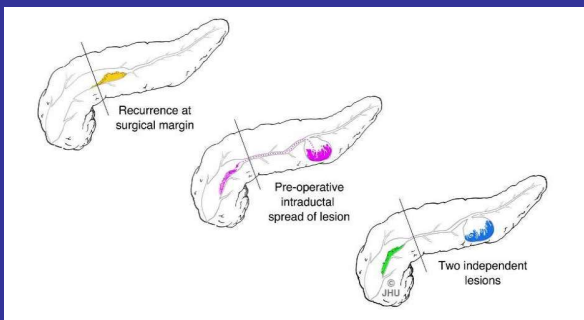


A. Pea, Annals of

Intraductal Papillary Mucinous Neoplasms

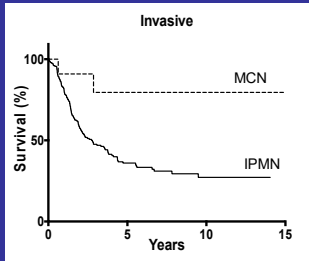
Surgically resected non-invasive IPMNs have a 90% 5-year survival rate. Most of the recurrences in patients with non-invasive IPMNs appear to be from multi-focal disease, because patients who undergo total pancreatectomy for a non-invasive IPMN have a close to 100% 5-year disease free survival

Clinical Implications



A. Pea, Annals of

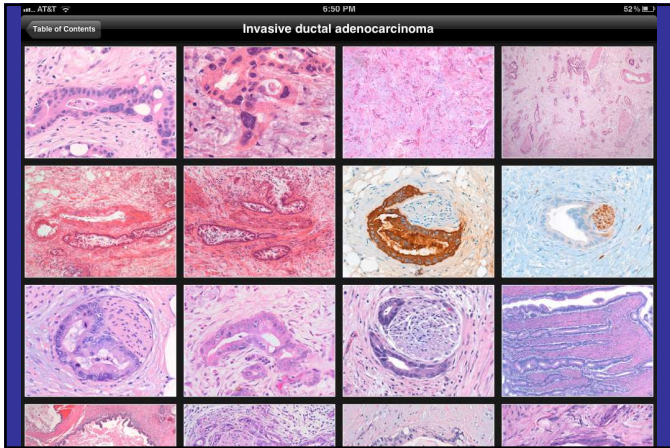
Invasive Carcinoma Arising in IPMN vs. MCN

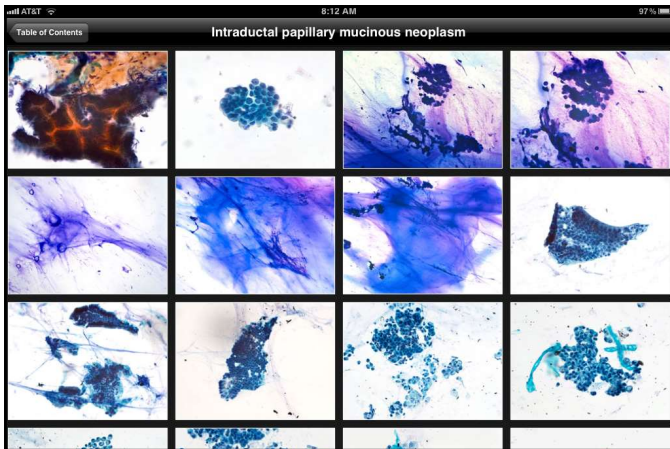


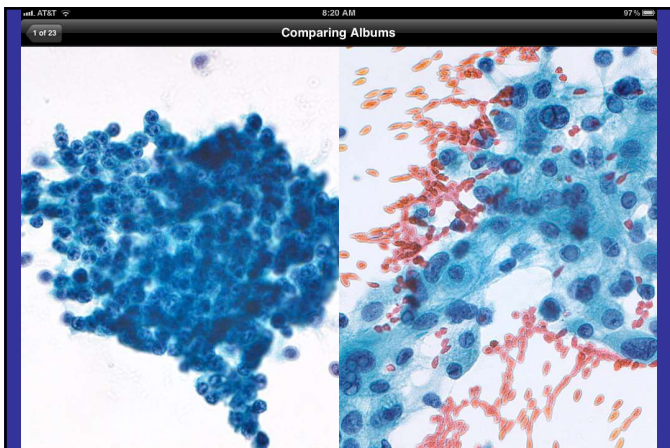
A. Pea, Annals of Surgery, 2016

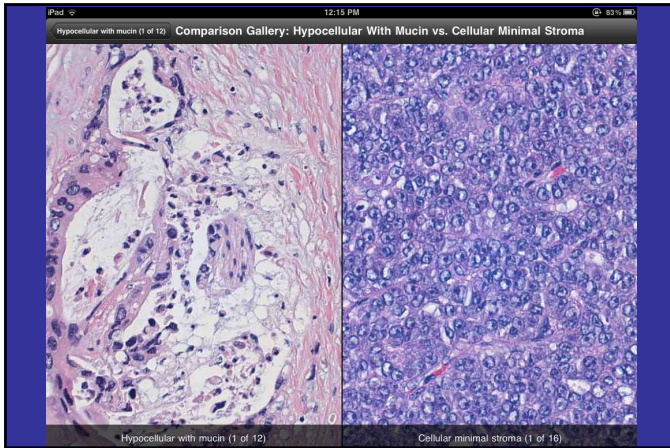
	Mucinous Cystic Neoplasm	Intraductal Papillary Mucinous Neoplasm
Age	40-50 years	60's
Gender	Female>>male	Male>female
Head vs. body/tail	Body/tail	Head
Connectivity to large ducts	Usually not	Always
Cyst Contents	Mucoid	Mucoid
Mucin oozing from ampulla	No	Yes
Stroma	Ovarian-type	Collagen
Multifocal disease	Very rare	20-30%

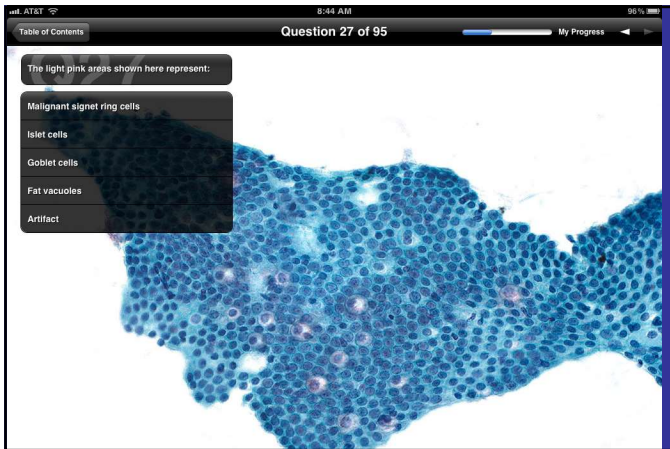


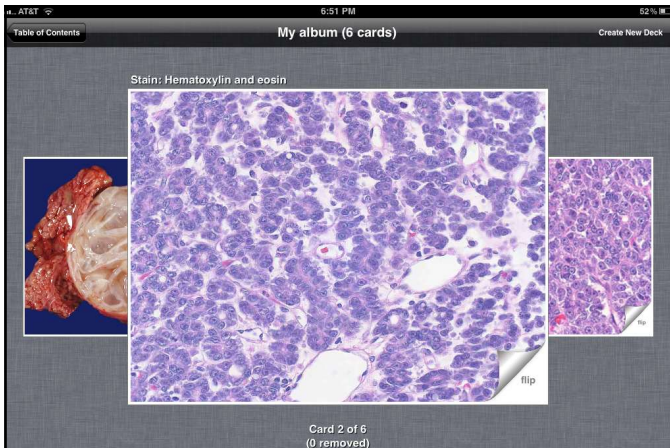












Follow us on Twitter- @Hopkins_GI_Path