PANCREATIC

CYST

NEOPLASM

→ Classification of Pancreatic cystic neoplasms:

BOX 60.1 Klöppel's Classification of Cystic Neoplasms of the Pancreas

Neoplastic	Nonneoplastic
Epithelial	**Epithelial**
Benign	Congenital cyst (in malformation syndromes)
Serous adenoma (microcystic)	Lymphoepithelial cyst
Serous adenoma (oligocystic, ill-demarcated)	Mucinous nonneoplastic cyst
MCN	Enterogeneous cyst
IPMN	Retention cyst
Acinar cell cystadenoma	Periampullary duodenal wall cyst
Dermoid cyst	Endometrial cyst
Cystic hamartoma	
von Hippel-Lindau–associated cystic neoplasm	
Borderline	
MCN	
IPMN	
Solid pseudopapillary tumor	
Malignant	
MCN-associated carcinoma	
IPMN-associated carcinoma	
Ductal adenocarcinoma, cystic	
Serous cystadenocarcinoma	
Pancreatoblastoma, cystic	
Cystic metastatic epithelial neoplasm	
Neuroendocrine carcinoma, cystic	
Nonepithelial	**Nonepithelial**
Benign neoplasm (i.e., lymphangioma)	Pseudocyst
Malignant neoplasm (i.e., sarcoma)	Parasitic cyst

IPMN, Intraductal papillary mucinous neoplasm; *MCN,* mucinous cystic neoplasm.
From Kosmahl M, et al: Cystic neoplasms of the pancreas and tumor-like lesions with cystic features: a review of 418 cases and a classification proposal. Virchows Arch 445:168-178, 2004.

* overall mc – IPMN
* clinical presentation
  - mostly incidentally detected.
  - Prevalence of asymptomatic
    PCN – 2.1.1.
    ↓
    when symptomatic
  - New onset acute pancreatitis.
    due to mucin plug in MPD
    communicating cysts.
  - Pancreatic exocrine & endocrine insufficiency
    d/t pancreatic inflammation
  - obstructive jaundice
    - d/t direct tumor compression in CBD.

→ Clinical features :

Fluid CEA cut off value
> 192 ng/mL

TABLE 60.2 Characteristics of Common Cystic Neoplasms of the Pancreas

Cystic Neoplasm	Mean Age (Years)	Gender Ratio F:M	Gross Features	Macroscopic Features	Imaging Characteristics	Cyst Fluid Characteristics	Cyst Fluid Cytology
Serous cystadenoma	61-65	7:3	Variable size Microcystic Stellate scar	Clear cytoplasm Well-defined borders, uniform nuclei Glycogen-rich cells	Microcystic characteristic honeycomb pattern Stellate scar	Low viscosity CEA low Amylase low	Cuboidal cells Clear glycogen-rich cytoplasm
Mucinous cystic neoplasm	45-55	>10:1	Large size Multilocular Thick walled	Tall columnar mucin-producing epithelium Ovarian-type stroma	Macrocystic body/tail location Does not communicate with duct Peripheral calcifications	High viscosity CEA elevated Amylase low	Variable cellularity with columnar cells with or without atypia
Intraductal papillary mucinous neoplasm	65-75	1:1	Variable size Multilocular Involves main or branch ducts	Mucin-producing epithelium with papillae	Macrocystic Ductal involvement	High viscosity CEA elevated Amylase high	Variable cellularity with columnar cells with or without atypia
Solid pseudopapillary tumor	25	9:1	Irregular cystic cavities with hemorrhage	Solid sheets of variable cells Hyaline globules Neuroendocrine features	Macrocystic Areas of hemorrhage	Bloody Necrotic debris	Polygonal cells eosinophilic cytoplasm
Ductal adenocarcinoma with cystic degeneration	60-80	1:1.3	Mass with cystic degeneration	Ductal adenocarcinoma	Cyst with solid component Ductal dilation	Bloody necrotic debris	Malignant cells
Pseudocyst	Variable	1:1	Fibrous thick-walled capsule	No epithelial lining	Unilocular Associated with pancreatitis	Low viscosity Dark CEA low CA 19-9 elevated Amylase elevated	Inflammatory cells No mucin Epithelial cells

- Guidelines for evaluation

1. Sendai consensus guideline 2006
2. International Association of Pancreas (IAP)
   Fukuoka guidelines 2017
3. American Gastroenterological Association (AGA)
4. European guideline 2018.

→ USG Abdomen – most appropriate initial imaging

→ Cross sectional imaging – incidental finding on CT – 2-3%.
on MRI / MRCP – 13-45%

- MRI vs CT :

MRI preferred for detection & follow up

CT indicated in
- Parenchymal calcification, underlying chronic pancreatitis.
- PCN & malignant pancreatic tumor. for vascular assessment.
- Suspicion of postoperative recurrence of Cancer.

- Role of EUS in PCN :
  - As an adjunct to other imaging

- disadvantage of EUS
  - inter observer variability
  - inability to diff benign & malignant cysts.
  - inability to identify exact type of cyst.

- EUS guided cyst fluid FNA & cytology
  - Not indicated always. only when imaging shows concerning features.

Indications - only if additional info will change management.

Contraindications
- if diagnosis already established on imaging.
- if surgical excision is anyway required
- distance of >10 mm btw cyst & transducer of EUS.
- high risk of bleeding.

• EUS + FNB from solid component of cyst.

Concerning features
- diameter > 3 cm
- enhancing mural nodule
- PD dilated.

SUMMARY INDICATIONS OF EVALUATING PCN WITH EUS

2015 AGA[48] guideline
- At least two of the following concerning features:
  - Cyst diameter >30 mm
  - Nodule
  - PD dilatation

2017 IAP[3] guideline
- Growth rate ≥5 mm over 2 years
- Increased levels of serum CA19-9
- PD dilatation between 5 and 9 mm
- Cyst diameter ≥30 mm
- Acute pancreatitis (caused by IPMN)
- Enhancing mural nodule (<5 mm)
- Abrupt change in calibre of PD with distal pancreatic atrophy

2018 European[4] guideline
- EUS–(FNA) should only be performed when the results are expected to change clinical management.
- EUS–(FNA) is recommended if the PCN has either clinical or radiological features of concern identified during the initial investigation or surveillance

→ Management of Serous cystic Neoplasm: (Elderly women, mostly benign)

- Asymptomatic SCN ± confirmed diagnosis — Observation only.
- Sx indicated when (European guidelines)
  - malignancy is a concern
  - Symptomatic related to compression of stomach, bile duct.
  - Large SCN > 4 cm.

→ Management of mucinous cystic Neoplasm:

European guidelines
- Sx in MCN when
  - size ≥ 4 cm
  - High risk factors like mural nodules
  - Symptomatic

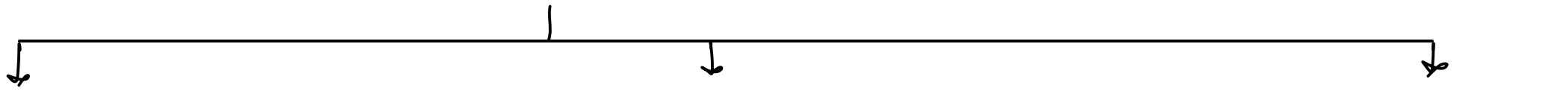
IAP / AGA → all MCN in surgically fit patients irrespective of size / symptoms should undergo Sx.

→ Management of Solid pseudopapillary Neoplasm (SPEN)

- Rare
- Young female, body / tail of pancreas
- Radical resection in all patient, even in recurrent / metastatic SPEN.
- R0 resection in metastatic disease also curative.

→ Management of IPMN: (spectrum from dysplasia to invasive malignancy)

- mc in elderly men.



- Segmental/diffuse dilation of MPD > 5 mm without other cause of obstruction.

- Pancreatic cyst > 5mm communicating with Branch ducts.

- mixed features of both.

→ management of MD-IPMN:

→ worrisome features of IPMN: (MD)

- cyst ≥ 3cm
- Enhancing mural nodule < 5mm
- Thick cyst wall
- MPD - 5-9 mm
- Abrupt cut off in MPD with distal atrophy.
- Lymphadenopathy
- elevated Serum CA 19.9
- Rapid rate of cyst growth > 5mm/2 years

Evaluate with EUS for further high risk stratification

→ High risk stigmata in IPMN - MD

- obstructive jaundice in PCN in head of pancreas.
- Enhanced mural nodule ≥ 5mm
- MPD ≥ 10 mm

Any feature +ve should undergo surgical resection in fit patients without further testing.

☆ MD - IPMN surgical management

- Segmental type
- Diffuse type / focal lesions

→ Easy to determine type of resection proximal vs distal pancreatectomy / intraop frozen section.

- mid duct dilation – Right sided resection preferred.
- Diffuse dilation – Total pancreatectomy in selected individuals.

↓

Low grade dysplasia at margin does not require further resection.

→ Management of mixed duct - IPMN

- Should be resected when diagnosed.

→ Management of BD - IPMN

- guidelines evolving for conservative management as they have more indolent behaviour compared to MD-IPMN.

- Surgical management

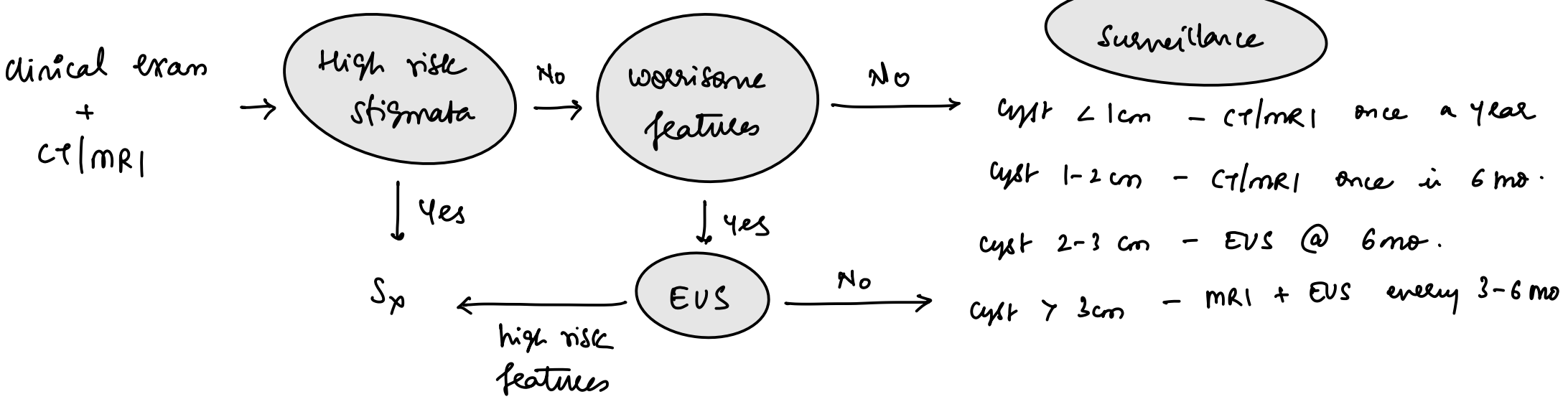
Absolute indications

- Mural nodule enhancing ≥ 5mm
- solid mass
- jaundice
- cytology +ve for HGD / cancer. on EUS

Relative indication

- Growth rate ≥ 5mm/year
- ↑ S. CA 19.9
- MPD 5–9.9 mm
- cyst ≥ 4 cm
- mural nodule < 5mm
- symptomatic.

→ Fukuoka guidelines



→ Extent of surgery in IPMN

- Invasive IPMN
  - oncologic resection + lymphadenectomy + adjuvant therapy.
- Non invasive IPMN
  - Parenchyma sparing procedures

→ Summary

Cystic neoplasm of Pancreas

↓

Triphasic CT / MRI

↓

- Symptomatic
- mural nodules
- MPD dilated
- MD - IPMN, SPEN

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Surgery

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- Open / minimally invasive
- small cyst – Enucleation when feasible
- Head tumors – PD, PPPD
- Body tumors – median / Subtotal Pancreatectomy.
- Tail tumors – distal pancreatico splenectomy.

Asymptomatic

Absence of dilated MPD

No Radiological features to malignancy.

↓

Size > 3cm → Risk of Sx

Size < 3cm → Observe

↓

Low risk → Sx

High risk → EUS / FNA, ERCP, cyst fluid CEA

malignant → Sx

negative → Observe

→ Surgical approach in PCN

Conservative approach
(Organ preserving Pancreatic Resections OPPAR)

- Enucleation
- Central Pancreatectomy
- Spleen preserving distal Pancreatectomy

↓

Early morbidity ↑ - higher incidence of Post operative pancreatic fistula.

Late morbidity ↓ - low incidence of exocrine & endocrine insufficiency.

Radical Resection

- Whipples Pancreatico duodenectomy
- Distal Pancreatico Splenectomy.