



WHO 5th edition 2019 GIT Blue Book

What's new ??





Anthony Gill MD FRCPA

Dr Gill has no conflicts of interest to disclose

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edition is the first to be led by an editorial board, with standing members and expert members we closely to evaluate the evidence underpinning the classification of tumours. As in previous edition books include numerous colour images, which provide the standards needed by pathologists to un their diagnoses. In the 5th edition, there is increased emphasis on molecular pathology and ge Other innovations include standardization of headings within sections, lists of essential diagnostic of and maps (produced by IARC scientists) illustrating the epidemiology of common tumours.

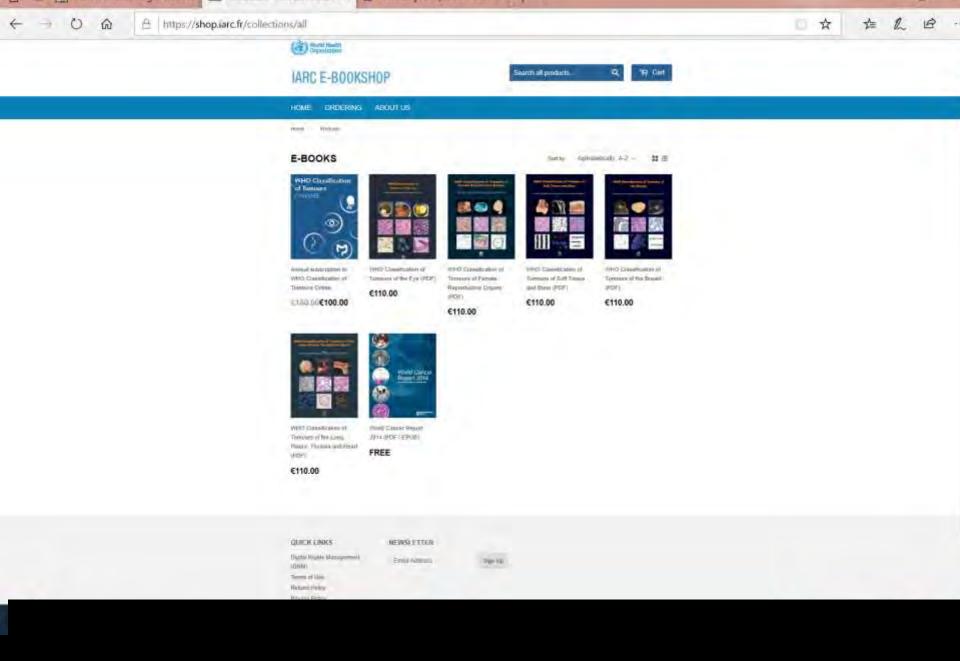
The 5th editions of Breast Tumours and Soft Tissue and Bone Tumours will be available later in 201



WHO Classification of Tumours of the Digestive System 5th Edition, Volume 1

Edited by the WHO Classification of Tumours Editorial Board Publication: 2019

More Information



https://tumourclassification.iarc.who.int/welcome/

International Agency for Research on Carcos (A) World Insulth Organization

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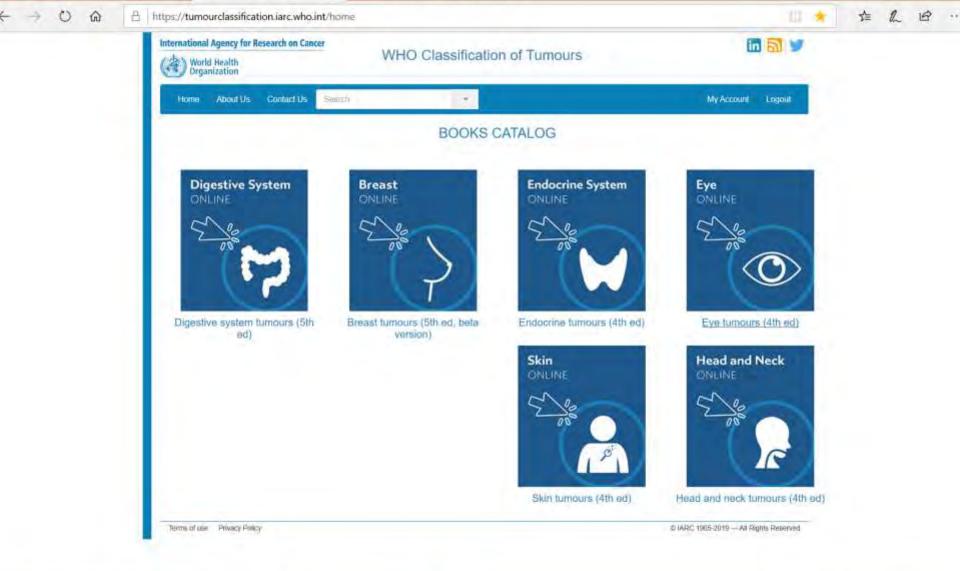
Digital pathology enhancements

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Deep zoom functionality

See images in their full detail.



World Health Organization	WHO Classification of Tumours	in <u>a</u> 1
Home About Us Contact Us Search		My Account Logout
	Digestive system tumours (5th ed)	
 WHO Classification of Tumours: Editorial Board Foreword with changes from the book, including Introduction to tumours of the digestive system Classification of neuroendocrine neoplasm Tumours of the oesophagus: Introduction Epithelial tumours Benign epithelial tumours and precursors Oesophageal squamous papilloma Barrett dysplasia Oesophageal squamous dysplasia Malignant epithelial tumours Adenocarcinoma of the oesophagus a Oesophageal adenoid cystic carcinom Oesophageal adenosquamous and m Oesophageal adenosquamous and m Oesophageal squamous cell carcinom Oesophageal neuroendocrine neoplass Tumours of the stomach Tumours of the stomach: Introduction Gastritis and metaplasia: precursors of gas Epithelial tumours Benign epithelial tumours and precursors Fundic gland polyps Gastric hyperplastic polyps Gastric dysplasia Intestinal-type gastric adenoma Foveolar-type adenoma Gastric pyloric gland adenoma Oxyntic gland adenoma Malignant epithelial tumours Gastric pyloric gland adenoma Malignant epithelial tumours 	a corrigenda s of the digestive system s and oesophagogastric junction NOS a ucoepidermoid carcinomas ha NOS ma sms tric neoplasms	

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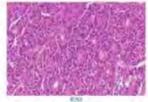
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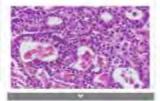
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#263 Acinar cell carcinoma



Rare patients, especially when young, can show increased blood levels of AFP (10987254 ; 25353285).

Epidemiology:-

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#263 Acinar cell carcinoma



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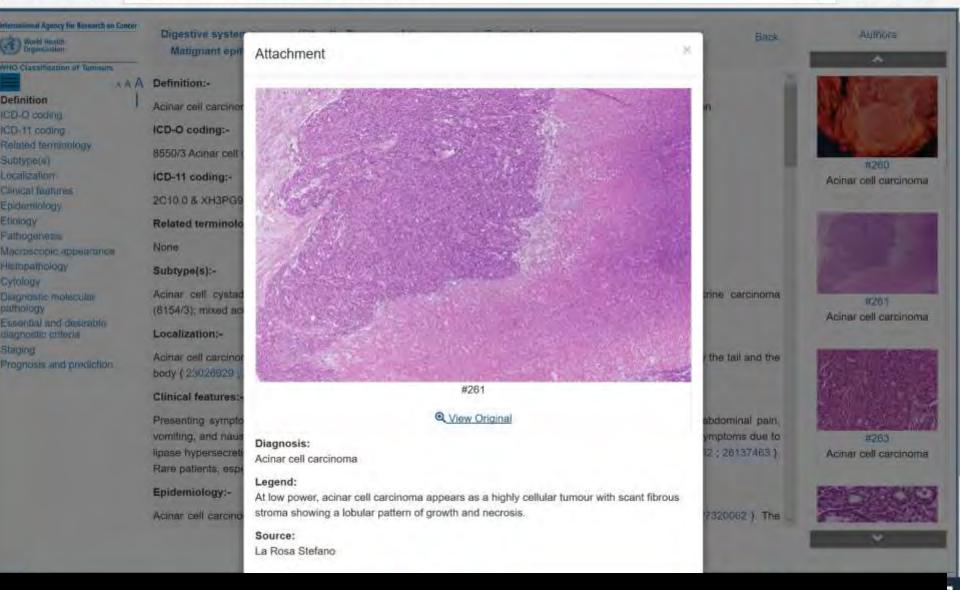
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International Agency for Research on Cancer World Health Organization	Digestive system tumours (5th ed) Tumours of the pancreas Epithelial tumours Back Malignant epithelial tumours Pancreatic ecinar cell carcinoma	Authors
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WHO Classification of Tumours

Definition

ICD-11 coding Related terminology Subtype(s) Localization Clinical features Epidemiology Etiology Pathogenesis Macroscopic appearance Histopathology Cytology Diagnostic molecular pathology

Essential and desirable diagnostic criteria Staging

Prognosis and prediction

Digestive system tumours (5th ed) Tumours of the pancreas Epithelial tumours Benign epithelial tumours and precursors Serous neoplasms of the pancreas

Definition:-

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Serous cystadenoma of the pancreas is a benign epithelial neoplasm composed of uniform cuboidal, glycogen-rich cells that often form cysts containing serous fluid. The diagnosis of malignancy in pancreatic serous neoplasms is restricted to cases with unequivocal distant metastasis beyond the pancreatic/peripancreatic bed.

ICD-O coding:-

8441/0 Serous cystadenoma

8441/3 Serous cystadenocarcinoma

ICD-11 coding:-

2E92.8 & XH8TJ0 Benign neoplasm of pancreas & Serous cystadenoma NOS

2C10.Y & XH7A08 Other specified malignant neoplasms of pancreas & Serous cystadenocarcinoma NOS

Related terminology:-

Serous cystadenoma

Acceptable: microcystic adenoma; glycogen-rich adenoma; oligocystic ill-demarcated adenoma.

Subtype(s):-

Microcystic serous cystadenoma, macrocystic (oligocystic) serous cystadenoma; solid serous adenoma; von Hippel-Lindau syndrome-associated serous cystic neoplasm; mixed serous-neuroendocrine neoplasm

Localization:-

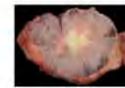
Serous cystadenomas can occur anywhere in the pancreas, but they arise most frequently (50–75%) in the pancreatic body or tail and are generally solitary { 21468008 ; 22415666 ; 26045140 ; 26559376 }. Unless associated with germline alterations in VHL, these neoplasms rarely involve the full length of the pancreas or are multifocal (22370733 ; 28697137 ; 23543325 }. Serous

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Authors



#517 Serous cystadenoma



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Serous cystadenoma

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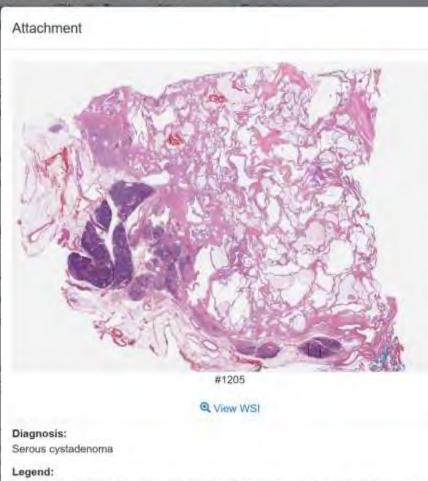
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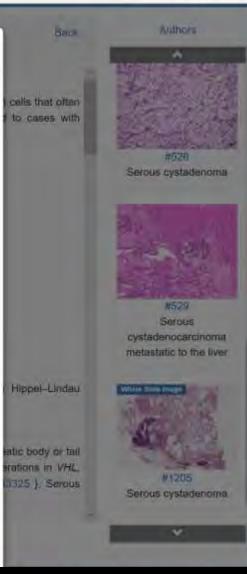
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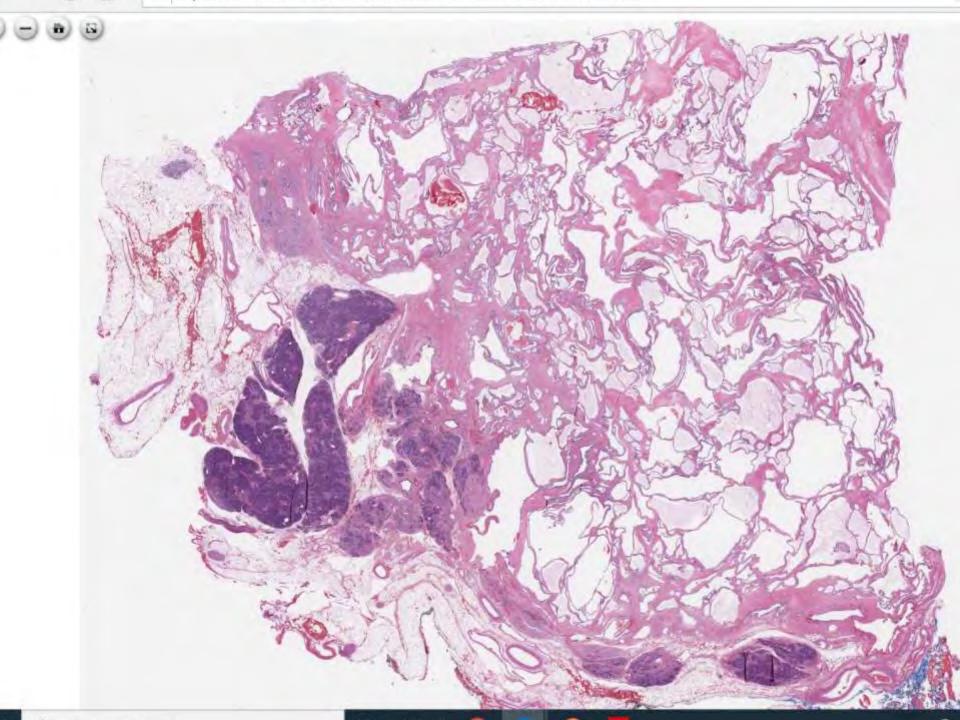
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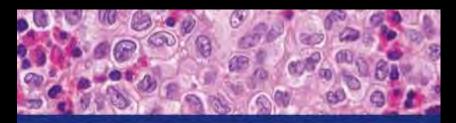


Microcystic serous cystadenoma consists of numerous tiny cysts lined by a flattened layer of epithelium with rare microscopic papillae that project into the cyst lumen. The cysts contain proteinaceous fluid and are lined by cuboidal epithelium with clear cytoplasm and uniform, round nuclei.





Uniformity Across Blue Books



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Haematolymphoid tumours of the digestive system

Edited by: Chan JKC, Fukayama M-

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Mesenchymal tumours of the digestive system

Edited by, Fukayama M, Goldblum JR, Lazar AJ, Mettinen M

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Angiosarcoma Glomus tumour Lymphangioma and lymphangiomatosia Neural tumours Schwannema Granular cell turnour Parineurioma Ganglioneuroma and ganglioneuromatosis Tumours of uncertain differentiation PEComa, including angiomyolooma Mesenchymal hamartoma of the liver Calcifying nested stromal-epithelial turnour of the liver Synovial sarcoma Gastrointestinal clear cell sarcoma / malignant gastrointestinal neuroectodermal tumour Embryonal sercome of the liver

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	Serous cystadenomas can occur anywhere in the pancreas, but they arise most frequently (50-75%) in the pancreatic body or tail and are generally solitary (21468008 ; 22415666 ; 28045140 ; 26559376). Unless associated with germline alterations in VHL, these neoplasms rarely involve the full length of the pancreas or are multifocal (22370733 ; 28697137 ; 23543325). Serous	Se	#120 erous cysta		
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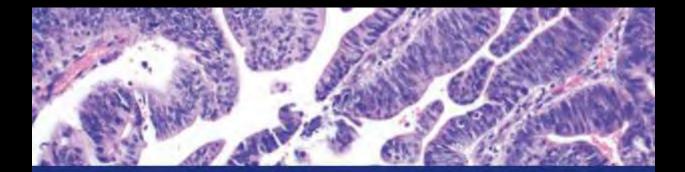
What's new ??





Anthony Gill MD FRCPA

Dr Gill has no conflicts of interest to disclose



Tumours of the small intestine and ampulla

Edited by: Klimstra DS, Nagtegaal ID, Rugge M, Salto-Tellez M

Benign epithelial tumours and precursors Non-ampullary adenoma Ampullary adenoma Malignant epithelial tumours Non-ampullary adenocarcinoma Ampullary adenocarcinoma Neuroendocrine neoplasms





Tumours of the small intestine and ampulla

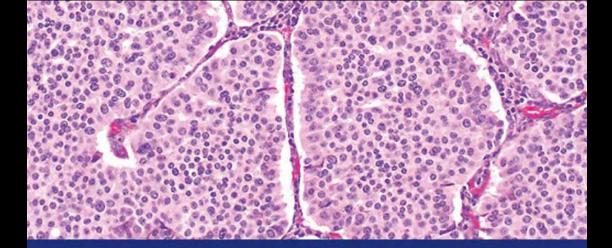
Edited by: Klimstra DS, Nagtegaal ID, Rugge M, Salto-Tellez M

Benign epithelial tumours and precursors Non-ampullary adenoma Ampullary adenoma Malignant epithelial tumours Non-ampullary adenocarcinoma Ampullary adenocarcinoma Neuroendocrine neoplasms The nomenclature in lesions from pancreatic and biliary ducts, the term "intra-ampullary papillary-tubular neoplasm" is now used for preinvasive neoplasms (adenomas and non-invasive papillary neoplasms) occurring almost exclusively within the ampulla. Fundamentally, these are intra-ampullary versions of intraductal papillary neoplasms or intraductal tubulopapillary neoplasms of the pancreas and bile ducts.

For intestinal adenomas that arise (and grow) predominantly on the duodenal surface of the ampulla ("periampullary duodenum"), the term "adenoma" (of intestinal phenotypes) is retained, in parallel with the terminology used in the intestinal tract.

In the section on ampullary adenocarcinoma, we adopt the classification of ampullary carcinomas into four anatomically based subtypes, which also have some degree of histological correlation: intra-ampullary papillary-tubular neoplasm–associated carcinoma (carcinomas arising from intraluminal-growing preinvasive neoplasms), ampullary-ductal carcinoma (arising and growing along the walls of intra-ampullary ducts), (peri)ampullary-duodenal carcinoma (growing on the duodenal surface of the ampulla), and ampullary carcinoma NOS.

This subdivision is driven by the anatomical complexity at that site and also affects the difficulty in tumour staging



Tumours of the pancreas

Edited by: Gill AJ, Klimstra DS, Lam AK, Washington MK

Benign epithelial tumours and precursors Acinar cystic transformation Serous neoplasms Intraepithelial neoplasia Intraductal papillary mucinous neoplasm Intraductal oncocytic papillary neoplasm Intraductal tubulopapillary neoplasm Mucinous cystic neoplasm Malignant epithelial tumours Ductal adenocarcinoma Acinar cell carcinoma Pancreatoblastoma Solid pseudopapillary neoplasm

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WHO 2010 Grading System

World Health Organization Classification 2010 for Neuroendocrine Neoplasms

Well differentiated NENs	Ki67index	Mitotic index		
Neuroendocrine tumour (NET) G1	≤ 2 %	<2/10 HPF		
Neuroendocrine tumour (NET) G2	3-20 %	2-20/10 HPF		
Poorly differentiated NENs				
Neuroendocrine carcinoma (NEC) G3*	>20 %	>20/10 HPF		

Mixed adenoneuroendocrine carcinoma (MANEC)

*"NET G3" has been used for this category but is not advised since NETs are by definition well differentiated

WHO 2017 Grading System

World Health Organization Classification 2010 for Neuroendocrine Neoplasms

Well differentiated NENs Neuroendocrine tumour (NET) G1 Neuroendocrine tumour (NET) G2	< <u>3%</u> 3-20 %	Mitotic index <2/10 HPF 2-20/10 HPF
Neuroendocrine tumour (NET) G3	>20%	>20/10 HPF
POORLY DIFFERENTIATED NENS		
Neuroendocrine Carcinoma (NEC) G3	>20%	>20/10 HPF

MENEN (mixed endocrine neuroendocrine carcinoma)

*"NET G3" has been used for this category but is not advised since NETs are by definition well differentiated

WHO 2017 Grading System

TABLE 1

World Health Organization Classification 2017 for Pancreatic Neuroendocrine Neoplasms

Well differentiated NENs	Ki67index*	Mitotic index
Neuroendocrine tumour (NET) G1	<3 %	<2/10 HPF
Neuroendocrine tumour (NET) G2	3-20 %	2-20/10 HPF
Neuroendocrine tumour (NET) G3	>20 %	>20/10 HPF
Poorly differentiated NENs Neuroendocrine carcinoma (NEC) G3 Small cell type Large cell type	>20 %	>20/10 HPF

Mixed neuroendocrine-nonneuroendocrine neoplasm (MiNEN)

* Ki67 index is based on at least 500 cells in areas of higher nuclear labeling ("hot spots"); mitoses in 50 high power fields (HPF, 0.2mm²) in areas of higher density and expressed per 10 HPF (2.0 mm²); the final grade based on which ever index (mitotic rate or Ki67) places the tumor in the highest grade category. For assessing Ki67, casual visual estimation ("eyeballing") is not recommended; manual counting of printed images is suggested {25412850}.

WHO 2019 Grading System for NETs

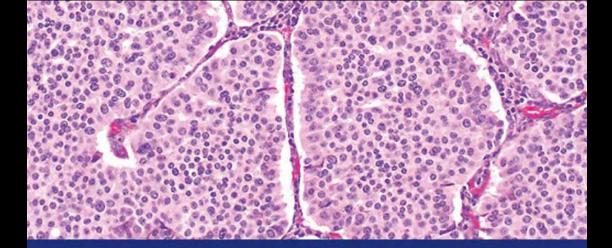
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Benign epithelial tumours and precessors Acinar cystic transformation

Intraepithelial neoplasia Intraductal papillary mucinous neoplasm Intraductal oncocytic papillary neoplasm Intraductal tubulopapillary neoplasm Mucinous cystic neoplasm Malignant epithelial tumours Ductal adenocarcinoma Acinar cell carcinoma Pancreatoblastoma Solid pseudopapillary neoplasm

Acinar Cystic Transformation



1011

Acinar cystic transformation of the pancreas Sinchi AD Adsay NV Headka N Tierrie B

Definition

Aprial cystic transformation of the panoresal to a non-neoplessic cystic lesion fined by beingn epipeaning acrise and ductal epithetiam

ICD-O coding

ICD-11 coding 2E92 8 Banjon neoplasm of punctane

Related terminology Acceptible schur cill createnorm

Subtype(s) North

Localization

These instants can octive throughout tion punctions, but they are intere common in the punctease head, some diffuently involve the entire grand (3734,1566,0061, 3569).

Clinical features

Fever than 50 cases have been paintfact, with a mean age at prevention of 40 years pange 4–85 years) and a tensit precentioner of 31 (3754.97.569.468,1187.2112.368.1566, 3061,3509.3759). Cases are divided into two categories pinicatly recognized microscopic testaria and inclustral microscopic tridings. Palants with microscopic testaria autostantial preparion are skymptomatic (1566.3061.3509.3759). Inodantial cases an only directed upon pathylogical lewine of paraneysta removed for other indications.

Epidemology Unknown

Eticlogy

The enclogy is unknown, but some pastering occar becauge of obstruction (3758)

Patriogenesis

Recent evidence suggests that this reside operations a new vacplance situation of the acrient and ductal opthesium (2061,307). Characteoremial gains, but not labole, were reported to one calcoby only comparative optionsic hybridization and sluggest a consister recipiteritis process (1960). However, a subsequent ducy bound a random X-characteoremic reactivation pattern for 5 cases, which would support these leasens as non-responsible (2061). Unlike in patternise, ductate elevence and the spirite process to the plasme, atterations in KRAS, GMAS, AWA3, TRS3, CDAA26, and SMAD4 may rob bees recorded in frees takans.



Hg. 10.XX. Active cyclic transformidem. CT of an active cyclic transformation trylly ing the participatic test



FIG. 10 XX Acres spate bandoesables. With multiplane searce breaking the entry length of the panoreal.

Macroscopic appearance

Clinically incogrisuor instance instances 1.5–19.7 cm (moder 5.8 cm) in dismission and form incidiocator or unlocator byttic matalase (3558,1506,3561,35538). Multicommotily a common and troug diffusely incident the writes gland. The cycli and in typically then, introduct and translationer, and third acts class watery faud incidentally detected cases are usually = 1.0 cm and unfocular, and they may not fac apparent grashy. Communication with the matery parameters, cast a rates.

Acinar Cystic Transformation



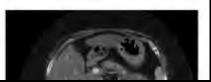
Acinar cystic transformation of the pancreas

Shidhi AD Acts av NV Hridka N Tintrin Fil.

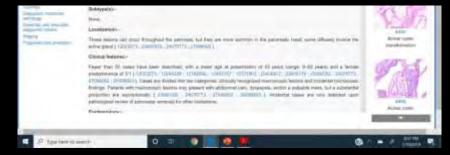
Definition

Aprilar cystic transformation of the panetosa is a non-neoplassic cystic lesion finkt by beingn appearing acinar and ductar epi-Theilarri

ICD-O coding Nicinia



Acinar cystic transformation of the pancreas is a non-neoplastic cystic lesion lined by benign-appearing acinar and ductal epithelium



These assists can only throughout the partonals, but they are more common in the panomatic hand, some diffusily muchathe online grand (3734,1566,3061,3569)

Clinical features

Fewer than 50 cases have been concribed, with a mean age al provenidon of 40 years (lange 9-89 years) and a lense predominance: pl 31 (3734 97.569.685 (651 2102 3588 1568. 3061,3509,3759). Casto are divided into two categories clinically recognized machinecopic lesions and incidential microscopic indrigs. Palants with macroacopic lesions may present with abcominal pain, dyspepsia, and/or a papable mass, but a substantial prepartion are asymptomatic (1566.3061.3509.3750). Inordivital-cases an only detected upon asthviogical leving of panoreata removed for other indications.

Epidemology Linknown

Eticlogy

The enclopy is unknown, but some pasts may occar because of obstruction 137586

Pathogenesis

Bocart evidence success that the water standards a ren sucplassic diatation of the adviar and ductal optimium (3081,307). Chromosomal Baini, but not loades, were reported for one calse EV array comparative genomic hybridization and suggest a possible neoplastic process (1566). However, a subsequent study bund a random X-chitomosome mactivation pattern for 5 cases, which would support these leaders as non-nacchartic 13061. Unlike in penmain dude eleverantume and te cyair, precurso reoplasmin, interations in KRAS, GNAS, RNF43, TR53, CDIO424, and SMAD4 Have not been reported in these lesions (3058)

Tartica.cli M. Con Luction page

He 10.32 Active route transformation CT of an even profile Vandormation Really ing Magancondic lad

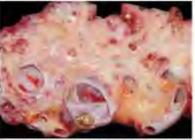


Fig. 10 XX Across could banaforeador. WP: multiplated segment involving the entry terruth of the planoweak.

Macroscopic appearance

Cirically nicogravo inciona measure 1.5-19.7 cm maan 5.8 erri in diameter and fami multilocular or unlocular ovelic mataias (3588,1506, 3081-3609). Multicommonly is common and itory diffusely involve the entire gaind. The cylit wal is typically thin, smooth and transludent, and filled with claim watery fuld. Incidentally detected cases are usually < 1.0 cm and unlocular, and they may not be apparent greasly. Communication with the PERFORMANCE AND ADDRESS OF ADDRESS OF ADDRESS ADDRESS

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Authors

#504

Acinar cystic transformation

ternational Agency for Research on Cancer	
World Health Organization	Digestive system tumours (5th ed) Tumours of the pancreas Epithelial tumours Back Back Benign epithelial tumours and precursors Acinar cystic transformation of the pancreas
HO Classification of Tumours	
AAA	Definition:-
Definition CD-O coding	Acinar cystic transformation of the pancreas is a non-neoplastic cystic lesion lined by benign-appearing acinar and ductal epithelium
CD-11 coding	Conservation
Related terminology	ICD-O coding:-
Subtype(s) localization	None
Clinical features	ICD-11 coding:-
pidemiology tiology	DC30.0 Cyst of pancreas
Pathogenesis	Related terminology:-
Aacroscopic appearance listopathology	Acceptable: acinar cell cystadenoma.
Cytology Diagnostic molecular	Subtype(s):-
athology	None
issential and desirable liagnostic criteria	Localization:-
Staging Prognosis and prediction	These lesions can occur throughout the pancreas, but they are more common in the pancreatic head; some diffusely involve the entire gland { 12023573 ; 23060352 ; 24076773 ; 27086062 }.
	Clinical features:-

Fewer than 50 cases have been described, with a mean age at presentation of 43 years (range: 9-83 years) and a female predominance of 3:1 { 12023573 ; 12004359 ; 12162680 ; 12483157 ; 10721803 ; 20438912 ; 23605178 ; 23060352 ; 24076773 ; 27086062 ; 28599853 }. Cases are divided into two categories: clinically recognized macroscopic lesions and incidental microscopic findings. Patients with macroscopic lesions may present with abdominal pain, dyspepsia, and/or a patpable mass, but a substantial proportion are asymptomatic (23060352; 24076773; 27086062; 28599853). Incidental cases are only detected upon pathological review of pancreata removed for other indications.

#1204

#1019 Acinar cystic

transformation

Acinar cystic transformation of the pancreas

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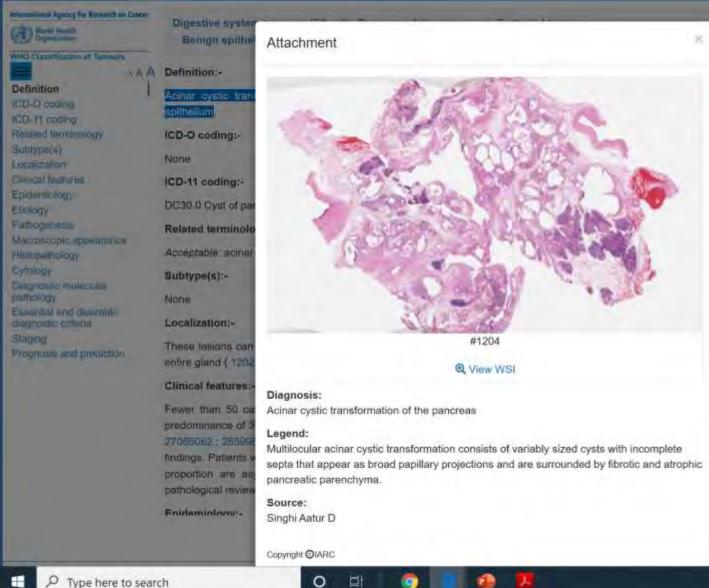
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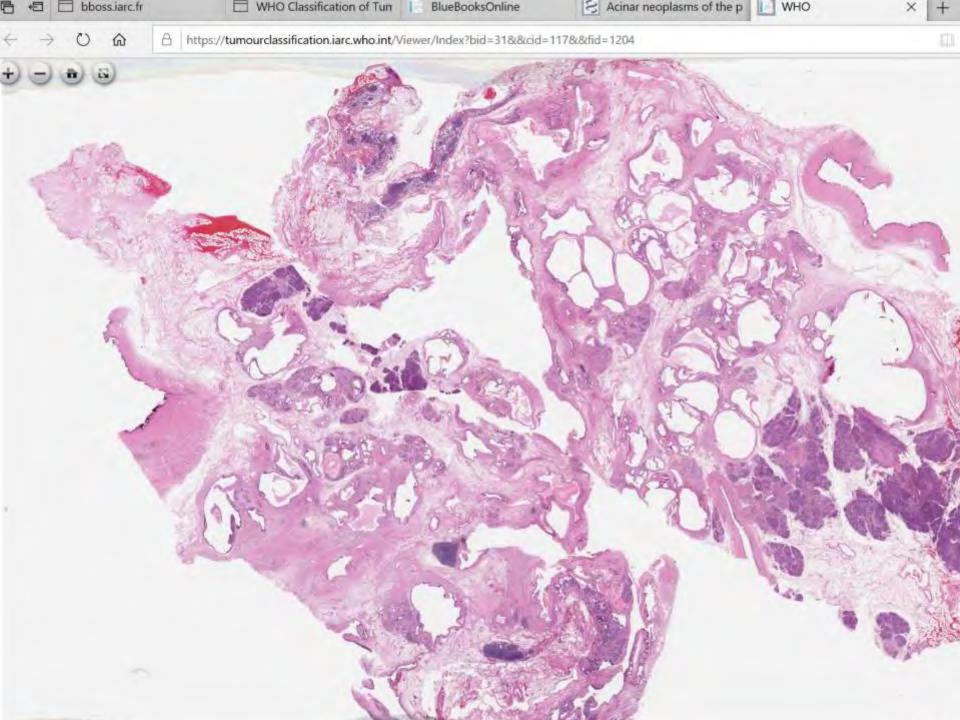
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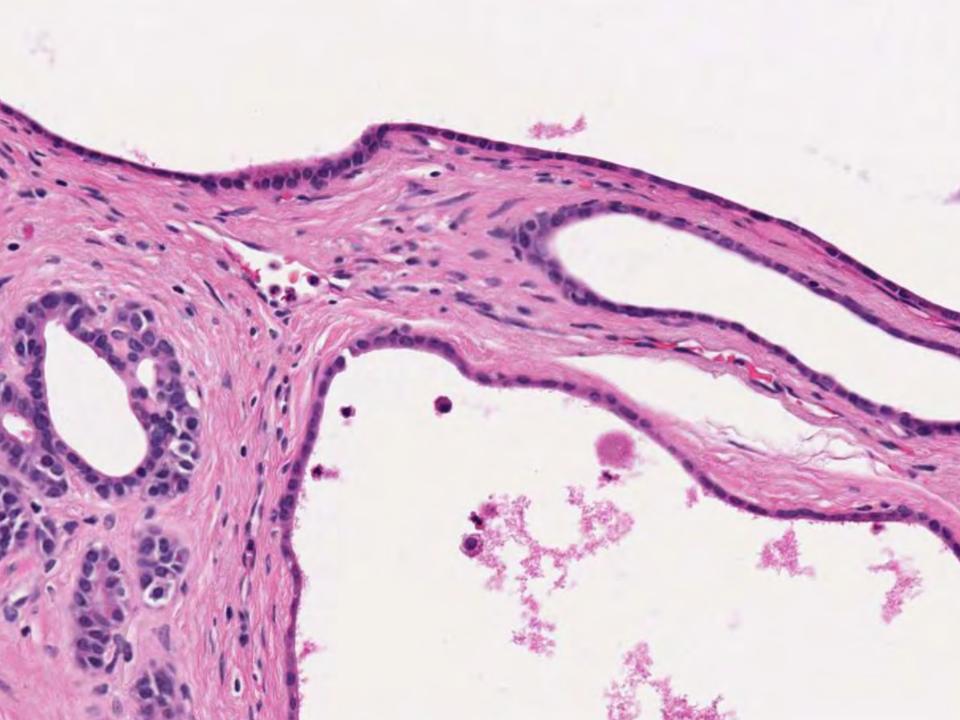
Acinar cystic

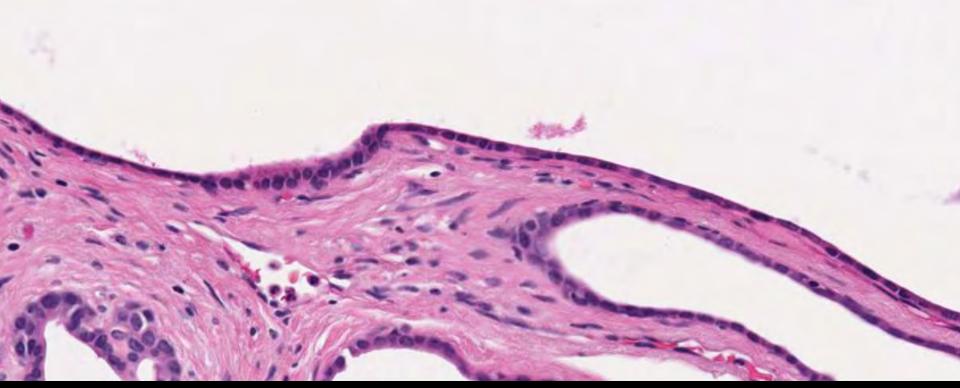


transformation #1019 Acmar cystic transformation aely involve the and a female 52 : 24076773 //1204 ttal microsconio Acinar cystic ut a substantial transformation of the detected upon pancreas 9-01 PM \bigcirc 7/10/2019

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includes serous cystadenomas (pariculatly diffuse type associated with VHL), squamoid of the pancreatic ducts, intraductal papillary mucinous neoplasms, and mucinous cystic asms.

recognition of both acinar and ductal differentiation by morphology and IHC in, chymotrypsin, BCL10, CK19)



Tumours of the pancreas

Edited by: Gill AJ, Klimstra DS, Lam AK, Washington MK

Benign epithelial tumours and precursors Acinar cystic transformation Serous neoplasms Intraepithelial neoplasia Intraductal papillary mucinous neoplasm Intraductal oncocytic papillary neoplasm Intraductal tubulopapillary neoplasm Mucinous cystic neoplasm Malignant epithelial tumours Ductal adenocarcinoma Acinar cell carcinoma Pancreatoblastoma Solid pseudopapillary neoplasm Neuroendocrine neoplasms Non-functioning neuroendocrine tumours Functioning neuroendocrine tumours Insulinoma Gastrinoma VIPoma Glucagonoma Somatostatinoma ACTH-producing neuroendocrine tumour Serotonin-producing neuroendocrine tumour Neuroendocrine carcinoma MiNENs

Tumours of the pancreas

Edited by: Gill AJ, Klimstra DS, Lam AK, Washington MK

Benign epithelial tumours and precursors

Acinar cystic transformation Serous neoplasms

Intraductal papillary mucinous neoplasm Intraductal oncocytic papillary neoplasm Intraductal tubulopapillary neoplasm Mucinous cystic neoplasm Malignant epithelial tumours Ductal adenocarcinoma Acinar cell carcinoma Pancreatoblastoma Solid pseudopapillary neoplasm Neuroendocrine neoplasms Non-functioning neuroendocrine tumours Functioning neuroendocrine tumours Insulinoma Gastrinoma VIPoma Glucagonoma Somatostatinoma ACTH-producing neuroendocrine tumour Serotonin-producing neuroendocrine tumour Neuroendocrine carcinoma MiNENs

Serous Cystadenoma (serous neoplasms)

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Serous neoplasms of the bancreas

ICD-0 coding

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ICD-11 coding. 25928 & XHETJO Benjon mechanis of parenum & Serous contraductoria MOS

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Cirical Instance

Ware Strategy lief of

The mean age at preparation is 56 years (lamps . 18-91 years). wen a lavouili predictionance of 0.1 (0710)1410(0000) Patherm may isofable symployinis minibild to local mains attack, with at itoropecific abcominal and track pains a parpathe mass, recently wed vorsible, classess, and weight can (659 3007,3710,1410). Jacobia caused by adamsterior of the chickel comprom biller (buch in University), even in association with recopleans within the percentric head. However, 40% of patients gis assimptionate al pinical presonantes (\$60,1608,1419).

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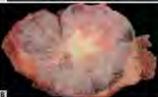


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mailly hyserklenias on 12-weighted images and hysometicae on 71-weighteit images (1772-623-1502). Decatornalis detries separatly harmoritage) is the cys alters this arged milersity partient. This seats of the stopplesm are well theplitted are 72-selphted images, but this central scar is not EUS reveise acachicitatio mails with numbrous cysts, which produce a charackenetic honeycome pattern (1602). The percelivity of EUS our beincreated by using it in conjunction with raised e-based confoand leaves intellocations, but the techniques is leaved to larger oversi 20140.17051. Trasis in no umbar coversultation behavior. the cash shift the panchosite dealty system. Dur upstatem deloted (Batation has been documented in 11% of capes (1419) Despite the typical reducerspirical appearance of most classe, the pirchsolidy of a methagevisit based on precisentive importo a high 1410,16082 Securi turnay manage an garagely when compa Arrest.

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mational Agency for Research on Cancer World Health Organization	Digestive system tumours (5th ed) Tumours of the pancreas Epithelial tumours. Benign epithelial tumours and precursors Serous neoplasms of the pancreas	ck	_	Authors	
O Classification of Tumours	Definition:-	2			
finition D-O'coding D-11 coding fated terminology	Serous cystadenoma of the pancreas is a benign epithelial neoplasm composed of uniform cuboidal, glycogen-rich cells that off form cysts containing serous fluid. The diagnosis of malignancy in pancreatic serous neoplasms is restricted to cases w unequivocal distant metastasis beyond the pancreatic/peripancreatic bed.			10	R
btype(s)	ICD-O coding:-			Star Star	
calization nical features	8441/0 Serous cystadenoma		10.5	#518	
idemiology	8441/3 Serous cystadenocarcinoma		Sei	ous cystaden	oma
ology	ICD-11 coding:-		25	auro	ind
ecroscopic appearance	2E92.8 & XH8TJ0 Benign neoplasm of pancreas & Serous cystadenoma NOS		-1	A PAR	and a
stopathology tology	2C10.Y & XH7A08 Other specified malignant neoplasms of pancreas & Serous cystadenocarcinoma NOS		1	Bar	1.
agnostic molecular	Related terminology:-		1		15
thology sential and desirable	Serous cystadenome		184	Participanti -	37
ignostic criteria aging	Acceptable: microcystic adenoma; glycogen-rich adenoma; oligocystic ill-demarcated adenoma.		Set	#520 ous cystaden	oma
ognosis and prediction	Subtype(s):-				
	Microcystic serous cystadenoma; macrocystic (oligocystic) serous cystadenoma; solid serous adenoma; von Hippel-Lind syndrome-associated serous cystic neoplasm; mixed serous-neuroendocrine neoplasm	au	3	S.	2
			22.0	and the second	1 A A

Localization:-

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Serous cystadenomas can occur anywhere in the pancreas, but they arise most frequently (50–75%) in the pancreatic body or tail and are generally solitary { 21468008 ; 22415666 ; 26045140 ; 26559376 }. Unless associated with germline alterations in VHL, these neoplasms rarely involve the full length of the pancreas or are multifocal { 22370733 ; 28697137 ; 23543325 }. Serous cystadenocarcinomas often arise in the body and/or tail of the pancreas.



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	https://tumourclassification.iarc.who.int/chaptercontent/31/118	
ational Agency for Basearch en Cancer World Health Organization	Digestive system tumours (5th ed) Tumours of the pancreas Epithelial tumours. Back Benign epithelial tumours and precursors Serous neoplasms of the pancreas	Authors
Classification of Tumours	Definition:-	
inition -O coding -11 coding ated terminology	Serous cystadenoma of the pancreas is a benign epithelial neoplasm composed of uniform cuboidal, glycogen-rich cells that often form cysts containing serous fluid. The diagnosis of malignancy in pancreatic serous neoplasms is restricted to cases with unequivocal distant metastasis beyond the pancreatic/period of means benign	
type(s)	ICD-0 coding:-	and the second
alization ical features	8441/0 Serous cystadenoma	#518
demiology	8441/3 Serous cystadenocarcinoma	Serous cystadenoma
hogenesis	ICD-11 coding:-	SALLTIN
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ology	2C10.Y & XH7A08 Other specified malignant neoplasms of pancreas & Serous cystadenocarcinoma NOS	1 Partie
gnostic mélécular hology	Related terminology:-	M. 555
ential and desirable gnostic criteria	Serous cystadenoma	#520
ging citizna	Acceptable: microcystic adenoma; glycogen-rich adenoma; oligocystic ill-demarcated adenoma.	Serous cystadenoma
gnosis and prediction	Subtype(s):-	the second second
	Microcystic serous cystadenoma; macrocystic (oligocystic) serous cystadenoma; solid serous adenoma; von Hippel-Lindau syndrome-associated serous cystic neoplasm; mixed serous-neuroendocrine neoplasm	2 Star
	Localization:-	The second
	Serous cystadenomas can occur anywhere in the pancreas, but they arise most frequently (50–75%) in the pancreatic body or tail and are generally solitary { 21468008 ; 22415666 ; 26045140 ; 26559376 }. Unless associated with germline alterations in VHL, these neoplasms rarely involve the full length of the pancreas or are multifocal { 22370733 ; 28697137 ; 23543325 }. Serous cystadenocarcinomas often arise in the body and/or tail of the pancreas.	#521 Serous cystadenoma
		and the second se
-		

Code	Neoplasm
/0	Benign
/1	Uncertain whether benign or malignant
	Borderline malignancy
	Low malignant potential
	Uncertain malignant potential
/2	Carcinoma in situ
	Intraepithelial
	Noninfiltrating
	Noninvasive
/3	Malignant, primary site
/6*	Malignant, metastatic site
	Malignant, secondary site
/9*	Malignant, uncertain whether primary or metastatic site

* Not used by cancer registries (used by some pathologists in some parts of the world)

sms

Malignancy in Serous Neoplasms

10.1.2

Serous neoplasms of the pancreas

Singh AD Adapy NV Historia N Terris S

Definition

Securi cyclastinomia of the plantmar in a benign spill with meblain composed of unitum suboldal, specigen with over the ben icom cyclic contemp securities that the disprotein of halprancy in parchadic aerosis respectively is relationed to classe. Whi uniquicical dispet metastatic beyond the perchaduct approximate bei

CD-O coding

54410 Serous cystaderionia 14410 Serous cystadirocercentria

ICD-11 coding

ERCH X XHBT/0 Bengs musikam of partness & Santaa cystelenome/NOS

Related terminology

Acceletable mercervere whereane grycegen rest educative, religiosystic il domarcasi d'advocima.

Sublype(s)

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Localization

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Chrical features

Seircus cyntechnomia

Most versus replantmentias an discovingl inclaimaily by abdominal trapping A statute CT trating as well circumstrated and multicodal cystemicrocyclic mean (223,625). Approxinetary 20% of cases demonstrate a contral con with a sursury calcillation getown. On 1965, assure, providentmes and





Hig. NO.65 Service contractments: 4 CT of a management encour cyliteterature to the parameteristic beact new the presence of a contral size. 6 Grows approximate to a microcylitic because installencing metal the contral tradeds approach program. He appearance of the contract program.

usually hyperimetes on 12-weighted images and hypometes on T1-weightert images (3772,629 (5011). Octorsonally, centres, (especially haemorthage) in the cyst allom this signal intensity patient. The septe of the neighborn are well departed on T2-weighted images, but the control and is hot. EUS reveals an echagenic mails with numerous bysts, which produce a characsecond horwycramb pattern (1006). The penatyley of EUS can be recreased by using it in conjunction with readle based confo cal least endomicroscopy, but this lechnopse is finited in larger cytta (2310-1705). Thinki is no visible communication between the Dyst and the printminic ducial system, but upbream dricks character has been donuminated to 17% of cases 114191. Decold the typical set/ophical appearance of most deare. the probability of a mischarchoals based on procperative magina is fact \$F10.10088. Serum tumpur mankets all penerally within normal **Onits**

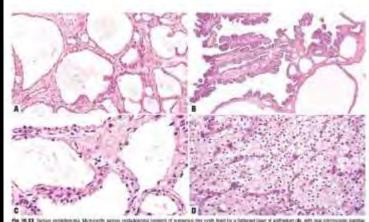


Fig. IRSE server aprelations developing environment of memory in memory to be by a tablewart pay of apprelation de, whit was internative payment de, whit was internative payment de and the environment of the server apprelation and white (3). It finds server a server was explained and well apprelation and environment apprelation and environment apprelation and apprelation and environment apprelation and apprelation apprelation and apprelation apprelation and apprelation and environment apprelation and environment apprelation a

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Cytokogy

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Disgnostic molecular pathology

Serous cystadenome: Genomic attentions in VML can be detected in precipitative pancrostic cyst fluid and used difcelly for disprartic parposes (VFM) 5004. Meestows in XFA3. ENAS, CDNASA and SMADA have not been reported in terous cystationoma. university in pancrostic ducibal identicalitymma. and thit cettic precipit mergitisms (22971).

Serout cystaderocarcinoma: Linked indecutar data and available due to the paucity of reported cares. However, new cases of carcetorpa we recrocyteld submorne wee wildlype for KRAS (3750).

Essential and desirable diagnostic criteria

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Staging (TNM)

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Fig. 10.22. Lienus opriadorecasonami netanalis is the Iver. Note the land optitopol features that are indefinguidants from those of an anthrary series opticale

Malignancy in Serous Neoplasms

The diagnosis of malignancy in pancreatic serous neoplasms is restricted to cases with unequivocal distant metastasis beyond the pancreatic/peripancreatic bed



Malignant behaviour of serous neoplasms has been reported (serous cystadenocarcinoma), but it is extraordinarily rare, and the diagnosis of malignancy in pancreatic serous neoplasms is restricted to cases with unequivocal distant metastasis beyond the pancreatic/peripancreatic bed





Although atypical and potentially a sign of aggressive behaviour, vascular, perineural, and adjacent organ and lymph node involvement by direct spread is insufficient for the diagnosis of serous cystadenocarcinoma, which requires metastasis (almost always to the liver)





Of note, some serous neoplasms reported as being cytologically malignant have not behaved aggressively and overt cytological features of malignancy have not been reported in truly malignant (metastatic) cases.

Tumours of the pancreas

Edited by: Gill AJ, Klimstra DS, Lam AK, Washington MK

Benign epithelial tumours and precursors Acinar cystic transformation Serous neoplasms Intraepithelial neoplasia Intraductal papillary mucinous neoplasm Intraductal oncocytic papillary neoplasm Intraductal tubulopapillary neoplasm Mucinous cystic neoplasm Malignant epithelial tumours Ductal adenocarcinoma Acinar cell carcinoma Pancreatoblastoma Solid pseudopapillary neoplasm Neuroendocrine neoplasms Non-functioning neuroendocrine tumours Functioning neuroendocrine tumours Insulinoma Gastrinoma VIPoma Glucagonoma Somatostatinoma ACTH-producing neuroendocrine tumour Serotonin-producing neuroendocrine tumour Neuroendocrine carcinoma MiNENs

Tumours of the pancreas

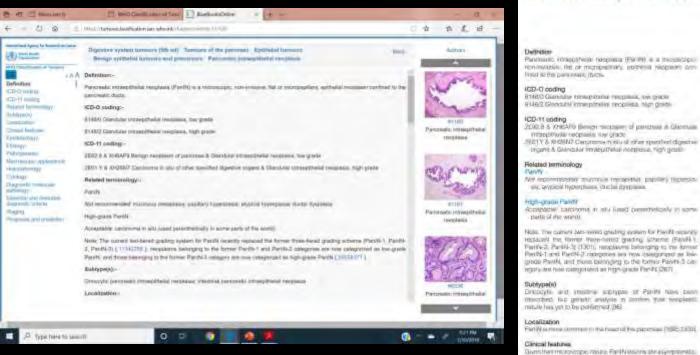
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Benign epithelial tumours and precursors Acinar cystic transformation

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Pancreatic Intra-Epithelial Neoplasia (PanIN)



Paricreatic intraepithelia/ neoplasia

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Epidemology

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Eliciogy

Line (TDAC) Pre-MI one over appreciate to be anticoalide with advanced age, clicking, acrestence later, information, and ages in militar (1928, 1353, 2513 article) 2018; Mac, Pre-MI interna are more numericus and of a lingtim graduit in the parameters (impatients with a line land article) product (2018).

Pathogenesis

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The histological programmich of Parith to investing satorhoma. minimal by uniatic programmical mandaulting as airen an remains frequency of morecular advertisions riversit, or tritlend towards in Notice Segments Containly for any individual emblant. la abwedon in nigh-gade. Parilil may re low glade Parily 100001. For example > 50% of Figs2N lesson of all cracket Tartics/ KRAS muldoling Towards, the VILLARY allele Robberto as cheature of cioneity) is Adodentially fedfor in Not-books Paul N1510L Attuach the preciae accusion of appendices of les was defined torials points apromation, auch as the more shortening and settiating mitations of the ARAS since game, and harty changes, tablar year in loss-cruide Pariful witt pitchanky coordinate to disease initializity in contrast, wide igneed clonel copy-number alteratives, as well as bishing load iveligits of CEWINEA (PHI), are closerved in high-prade Parith suggissting an association behavior these are unargen and cleasae progression (1957/1084). In contrast to viter had been research months, included during loads share that instations 37 TPSS-alli late if not absord, and there are no mittlions of remarygius delemental SARDA et liptaged high-geade handy seigen, ridbying that nacivation of treas two genes prodomhardly oddure in bone field invalves parcheetine (1264.1187) Airc. softe michaele suggests a sightwey to invasie card norm on citrarofrigat-lin avera abronoutra andarry will chartic reasonably, which causes subdwill catablephic

Pancreatic Intra-Epithelial Neoplasia (PanIN)

WHO 2010

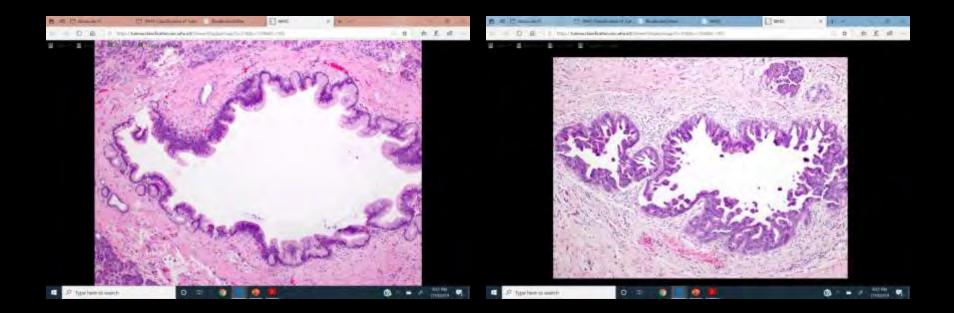
PanIN1

PanIN2

PanIN3

Pancreatic Intra-Epithelial Neoplasia (PanIN) WHO 2010 WHO 2019 PanIN1 Low Grade Previously PanIN 1 PanIN2 PanIN2 PanIN3 High Grade Previously PanIN3

Pancreatic Intra-Epithelial Neoplasia (PanIN)



Low Grade

High Grade

Pancreatic Intra-Epithelial Neoplasia (PanIN)

- Binary grading is in keeping with broad approach across entire WHO GIT 5th Edition blue book
- Applying:
 - Mucinous Cystic Neoplasms (MCN)
 - Intraductal Papillary Mucinous Neoplasms (IPMN)





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Malignant epithelial tumours Ductal adenocarcinoma Acinar cell carcinoma Pancreatoblastoma Solid pseudopapillary neoplasm Neuroendocrine neoplasms Non-functioning neuroendocrine tumours Functioning neuroendocrine tumours Insulinoma Gastrinoma VIPoma Glucagonoma Somatostatinoma ACTH-producing neuroendocrine tumour Serotonin-producing neuroendocrine tumour Neuroendocrine carcinoma MiNENs

IPMN, IOPN and ITPN are separated

Pancreatic intraductal papillary mucinous neoplasm

Detrailion

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CD-O coding

54530 Intraductal papillary muchave reconstrem with low-grade dysplastic

#450/2 Inhaductal papilary nacessar resplann with Eghtorado destilitata

ICD-11 coding

"Eliz # 6. %H8M02 Benign relignment of panchase 6. Intractactial papellary-muchous tumour with tox-practe dysplases 2E611 & XH3MB3 Carcinoma in eiu of other specified diges the organic & installucial papillary muchous neoplasts with high-grade dysplasic

Related terminology

Might-grade #W/N

Acceptable cardinoma in alta talled paramhetically in some Datts of the lendel.

Note: The maneric lac-cience: grading system for PMN recently. replaced the former three-torisci grading softemer, neoplasme belonging to the former categories of "#NN with Izw-grade dyaplasie (LGDF and "IPWN with warmediate-grade dyspiesie" (1001,1336) alls now categorized as knw-grade (PMN. Those belonging to the former dategory of 18MN with high-grade dysplassa (HGD)* are new passpotood as high-grade IPMN (282).

Subtycentri

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Concords: Noe Intraductal matellary mappings receipters is now recognized as a distinct entry [29,275,2048,285]. The designations "main ductinype intraductal papilitary much ous intoplasm", "tranch duct-type intraducts papillary mucinous reoplasm", and "mosed duct-lyple impaducial papillary mucheus. recolary" are imaging lents used by plinicate take Crinical leanives, tiskiwij raitnir triwn pathological subtypes.

Encation.

PMhill can occur anywhere in the main percentate ouct and or its branches: however, most any located in the head of the parchise (212 1663;266). Multipartricity is observed in as many ate 40% of cases (35.3241 1938 3091 2568.1429).

Hong SM Esposito I Moppel G Fukustenia N Mettra A FERMINA T Zumicini (II)

Bahnark O.

Pancreatic intraductal tubulopapillary neonlasm

Definition

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CD-O coding

8503/2 Wittaductal Suturiopupiliary Peoplemin 8553/3 Estinctuotal iudukapapiking (selphann with associated) Theather catterists

CD-11 coding

2E61.V 5 XH8457 Carcivorus in situ ill offen screet/wet departive organa & Intrahuctal Usular gapiliary Respisant, high plade CIGE & HHRWI Adonocationome of partomate & Internation Fig. 18, XX 1 placifiery noopieter with associated investors and IPANC (bacts

tion he was Related terminology filled with the

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Clinical R Subtype(s)

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Patients in About hart of all TIPTAs locul in that head of the pontrose and a 3-5 years theti menin the gard skillionity (200). pinomia t

sters [18] Clinical features

chibric p. (TPhia account bit < 1% of ell printmald explaine requirement dice (333 and 3% or intraducity resolution of the patiential (2643) (TPS). often delt anv slightly more common in Impains. Patient age tengen from By analy 25 to 84 years impact 55 years) (263). Pataging present with non-1663,321 genetic symptoms including apcommalizian vominia, weight by pitting long also interesting and database mobiles. Obstraction is relevant mental or Invincemmon Roma TPNs are detected moder/w/y (263) moles Tr

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Pathogenesia

have are tax term to pathogeness. The perceic features of (TPNs offer from those of ducted adonatelichomas and other strainantal neoplasme of the parageau. Meal of the reported attentions mater to ductal admosechemic and intraductal pupitary machine mophens IIPMIN, statuting KR42 matalione. n/o lebeare in ITPMs 13643.3611.3642.1201. However,

Pancreatic Intraductal oncocytic papillary neoplasm

> Battati D Hong BM ENcontro T Forgravel Ta Fuls minima N Marria A. Furnkows 1 Zamboni G

certain crearatio immobiliogi genesi (KMIC23 (MLL/I, AMIC29) [AUL2] KM72G [AULU], BARI) and PUK paltway gained (PIK3CA, PTEN) can be indused. A maximi of issues fullbase FGFHZ historic (28-II), which much be targetable. (remetace)

Macrosorpic appearance

(TPha form acid, firstly to habbury, redular masses within (Sinted periorbatic docts (579 (244,9160,3645), but the virus. Illary maccluck/ growth may be related to encodente. Cycl formation is often less evident than in PMRs. Musingue appellons are not present. The evening I/PW is 4.5 cm in channel trange. mm0.25-8:0

Histopethology

Microscopically, ITHIG form nodules of back to-back e-parkness the pland luturar glanda, resultag in large orbinisment imactures (283,3208,5843) The Vitraductal Excelor of it least some of the rockies is evenencial by contrally of the tetroletic operation from with monormation rate that protriate on However, must reliaclustral tumous rook any obliginate the dustral lumon, appearing as sharply conumeritied resits europunders by Utantic eldens. Although most (TPNs are pseudominantly tubular paperes may bid seen (3543). (TPRis are architecturally complex and hypically have high-matic constant. The lation could are constructively cuboidal, with modest impures of econophilo to amphophilo and rarray citial cylochaen 57,282). In some classe, installeneral dentitions may be seen. However, intecellate much is typically her enterchical in is remained (1763,5643). The nuclei are resard to coal and anyolal but unline. Attob: Name are offen multip sterrithings (\$14.3). Multiproves white into it iterment within the textules, often with a miniate-like patient.

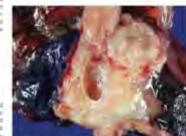


Fig. 10.22 Introducted approximation constants. Grad approximate new Top score the conjugate within the dilated man paycolding term.

symptome attributed to photolic pancinghtis ender to the make effect of the receptaem, such as jeundos (28,2048) Endoscopic biopsy or cylology may provide historigicial confirmation. 13712,706,26001

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Pathogenesis

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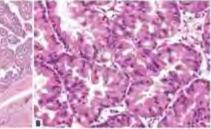
There are live data on pathogenesis. HWWartypically tack the absentions wounded to be related to during adoptionaria used intraductal parallery machines resplaym, such as mutations in RHAS GRAS and RMF4310022 205 2196 22927 31181 in conlight, paries including ARHGAPSE, ASIL1, EPHA8, and ERBEA are recurrently mutated in some XOPNs, but there are no entitydefining genomic attractors present in most cases (270)

Macroscopic appearance

Groesly, KOPNix typically form large (everage size) 5.5 cm), tenbioen, histle papilary projections of acid rod and within cystcally dilated panereatic ducts, with little intraductal much accumastion (26.3487). Occasionally, the connection of the cysts to the chuckle's system may real by apparent procedu-

Histopathology

prot. ct. the Microscopically. The summurs form complex and arboriting papil ion. Paperti ties inth delicate fibrovancular-tories. Sometimes the estachable insi (3520). growth may be difficult to recountas, but at least local involveminiard with mero of the ductal solitery can be demonstrated [28.35,3520]



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Pancreatic intraductal papillary mucinous neoplasm

Definition

Intraductal pepiliary mucinous neoplasm (IPMN) of the pancreas is a grouply visible (typically ≥ 1.0 cm) intraductal epithelial neoplasm of mucin-producing cells, arising in the main pancreatic duct and/or its branches.

ICD-O coding

- 8453/0 Intraductal papillary mucinous neoplasm with low-grade dysplasia
- 8453/2 Intraductal papillary mucinous neoplasm with highgrade dysplasia

ICD-11 coding

- 2E92.8 & XH8MD2 Benign neoplasm of pancreas & Intraductal papillary-mucinous tumour with low-oracle dysplasia
- 2E61.Y & XH3MB3 Cercinoma in situ of other specified digestive organs & intraductal papillary mucinous neoplasm with high-grade dysplasia

Related terminology High-grade IPMN

Acceptable: carcinoma in situ lused parenthetically in some parts of the world).

Note: The current two-tiered grading system for IPMN recently replaced the former three-tiered grading achienc; neoplasma belonging to the former categories of "PMN with inve-prade dysplasia (LGD)" and "IPMN with intermediate-grade dysplasia" (1301;1306) are now categorized as low-grade IPMN. Those belonging to the former category of "IPMN with high-grade dysplasia (HGD)" are now categorized as high-grade IPMN [257].

Subtype(s)

Gastric-type intraductal papillary mucinous neoplasm; intestinal-type intraductal papillary mucinous neoplasm; pancreatobiliary-type intraductal papillary mucinous neoplasm.

Oncocytic-type intraductal papillary mucinous neoplasm is now recognized as a distinct entity (28,270,2048,265). The deslignations "main duct-type intraductal papillary mucinous neoplasm", and "mixed duct-type intraductal papillary mucinous neoplasm" are imaging terms used by clinicians (see *Clinical features*, below) rather than pathological subtypes.

Localization

IPMNs can occur anywhere in the main pancreatic duct and/ or its branchos: however, most are located in the head of the pancreas [212,1663,266]. Multicentricity is observed in as many as 40% of cases [35,3241,1939,3091,2560,1423]. Basturk O Hong SM Esposito I Klöppel G Fukushima N Maitra A Furukawa T Zamboni G





Fig. 10.XX intraductal papiliary macinous neoplasm (PMN). A Main doct-type PMR; the duct is diffusely diabet and dran tibed with toky mucin. It IPMN involving both the main and secondary parcenaits ducts; markedly dialed parcenaits ducts are fixed with toket papiliary transmission.

Clinical features

IPMNs are fairly common, particularly in eldenly people, in consecutive CT scans, the prevalence was reported as 1.7%, image to 6.7% in people in their eighth docade of life (563). Patients with IPMNs with an associated invasive carcinoma are 3-5 years older from those without an associated invasive carcinoma, suggesting that progression occurs over a period of years (1689,3091). Clinical symptoms include epigastric pein, chronic pancreatitis, weight loss, diabetes mellitus, and jeundice (3338,1649,667,2837,3091). Branch duct-type IPMNs are often detected incidentally (3240).

By imaging, three distinct types of IPMN can be discerned: (1663.3239.3240.42). Main duct-type IPMNs are characterized by primary involvement of the main pancreatic duct with segmental or diffuse dilatation. Branch duct-type IPMNs typically involve the smaller, secondary ducts without affecting the main pancreatic duct (2837.3170.3238.3239). Mixed duct-type IPMNs is a combination of the other two types (3239.3240.42.1640). Mural inclules and/or irregularities in the duct contours may correspond to HGD or involve carcinoma (995.1697).

Pancreatic intraductal papillary mucinous neoplasm



D Hong SM I Klöppel G sa N Maitra A a T Zambori G

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Localization

IPMNs can occur anywhere in the main pancreatic duct and/ or its branches; however, most are located in the head of the pancreas [212,1663,266]. Multicentricity is observed in as many as 40% of cases [35,3241,1939,3091,2560,1423].





Fig. 10.XX intraductal papiliary mucinous neoplasm (PMN). A Main doct-type (PMR) file duct is diffusely diabet and often filed with tricky mucin. B IMM involving both the main and secondary parcenaitic ducts; markedly dialed parcenaitic ducts are filed with triated pagillary transmission.

Clinical leatures

IPMNs are fairly common, particularly in eldenly people. In consecutive CT scans, the prevalence was reported as 1.7%, riang to 6.7% in people in their eighth decade of life (563). Patients with IPMNs with an associated invasive carcinoma are 3-5 years older from those without an associated invasive carcinoma, suggesting that progression occurs over a period of years (1689,3091). Clinical symptoms include epigastric pein, chronic pancreatitis, weight loss, diabetes mellitus, and jeundice (3338,1649,667,2837,3091). Branch duct-type IPMNs are often detected incidentally (3240).

By imaging, three distinct types of IPNN can be discerned. (1663.3239.3240.42). Main duct-type IPMNs are characterized by primary involvement of the main pancreatic duct with segmental or diffuse dilatation. Branch duct-type IPMNs typically involve the smaller, secondary ducts without affecting the main pancreatic duct (3837.3170.3238.2339). Mixed duct-type IPMNs is a combination of the other two types (3239.3240.42.1640). Mural inclules and/or irregularities in the duct contours may correspond to HGD or involve acciona. 1965;16671.

D/Dx with PanIN

PanIN is a microscopic, usually < 5 mm in diameter (almost all gastric foveolar differentiation)

IPMNs are > 5 mm in diameter and can have varying differentiation

The term "incipient IPMN" or "incipient intraductal oncocytic papillary neoplasm" can be applied to lesions 0.5–1.0 cm in diameter with long finger-like papillae, intestinal or oncocytic differentiation, or a GNAS mutation.

Pancreatic intraductal papillary mucinous neoplasm

Basturk O Hong SM Esposito I Klöppel G Fukushima N Maitra A Furukawa T Zamboni G

Definition

Intraductal papiliary mucinous neoplasm (IPMN) of the pancreas is a grouply visible (typically ≥ 1.0 cm) intraductal epithelial neoplasm of mucin-producing cells, arising in the main pancreatic duct and/or its branches.

ICD-O coding

- 8453/0 Intraductal papillary mucinous neoplasm with low-grade dysplasia
- 8453/2 Intraductal papillary mucinous neoplasm with highgrade dysplasia

ICD-11 coding

- 2E92.8 & XH8MD2 Benign neoplasm of pancreas & intraductal papillary-mucinous tumour with low-grade dysplasia.
- 2E61Y & XH3MB3 Carcinoma in situ of other specified digestive organs & intraductal papillary mucinous neoplasm with high-grade dysplasia

Related terminology High-grade IPMN

Acceptable: carcinoma in situ lused parenthetically in some parts of the world).

Note: The current two-tiered grading system for IPMN recently replaced the former three-tiered grading achience, neoplasma belonging to the former categories of "PMN with low-gnide dysplania (LGD)" and "IPMN with intermediate-grade dysplania" (1301;1306) are now categorized as low-grade IPMN. Those belonging to the former categorized as low-grade IPMN. Those plania (HGD)" are now categorized as high-grade IPMN (257).

Subtype(s)

Gastric-type intraductal papillary mucinous neoplasm; intestnal-type intraductal papillary mucinous neoplasm; pancrestobiliary-type intraductal papillary mucinous neoplasm.

Oncocytic-type intraductal papillary mucinous neoplasm is now recognized as a distinct entity (28,270,2048,265). The deslignations "main duct-type intraductal papillary mucinous neoplasm", and "mixed duct-type intraductal papillary mucinous neoplasm" are imaging terms used by clinicians (see *Clinical* features, below) rather than pathological subtypes.

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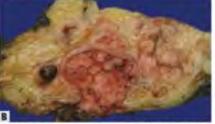


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Grade

IPMN with low-grade dysplasia (LGD) IPMN with intermediate-grade dysplasia

Low grade IPMN

IPMN with high-grade dysplasia (HGD)

High grade IPMN

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Molecular Pathology

KRAS mutations in 60–80% of IPMNs

GNAS mutations 50–70% of IPMNs, particularly in the intestinal subtype (rare in PDAC)

RNF43 mutated in 50% of IPMN

Pancreatic intraductal papillary mucinous neoplasm

Basturk O Esposito I Fukushima N Furukawa T Hong SM

Klöppel G

Zamboni G

Maitra A

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Subtypes Gastric Intestinal Pancreaticobiliary

Table 10.XX Immunohistochemical profile of intraductal papillary mucinous neoplasm (IPMN), intraductal oncocytic papillary neoplasm (IOPN), and intraductal tubulopapillary neoplasm (ITPN)

	CK7/CK8/CK18/CK19	CK20	EMA (MUC1)	MUC2	MUC5AC	MUC6	CDX2
IPMN							
Gastric	+	-	-	-	+	-/+	-
Pancreatobiliary	+	-	+	-	+	+	-
Intestinal	+	+	-	+	+	-	+
IOPN	+	+ in goblet cells	+	+ in goblet cells	+	+	+ in goblet cells
ITPN	+	-	+	-	-	+	-

Pancreatic intraductal papillary mucinous neoplasm

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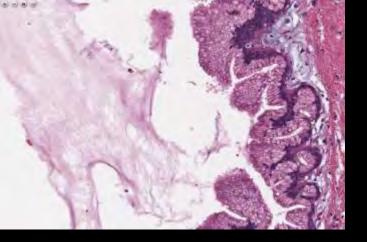


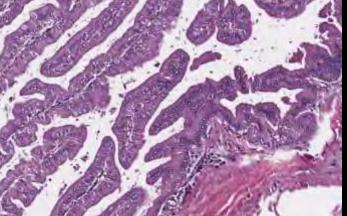


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IPMN							
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Pancreatobiliary	+	-	+	-	+	+	-
Intestinal	+	+	-	+	+	-	+
IOPN	+	+ in goblet cells	+	+ in goblet cells	+	+	+ in goblet cells
ITPN	+	-	+	-	-	+	-





Gastric -

Intestinal – MUC2/CDX2

Pancreaticobiliary - MUC1

Intraductal oncocytic papillary neoplasm (IOPN)

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Pancreatic intraductal oncocytic papillary neoplasm

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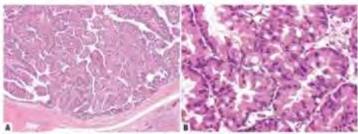
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Macroscopic appearance

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Intraductal oncocytic papillary neoplasm (IOPN)

102.3

Pancreatic intraductal oncocytic papillary neoplasm

Bastum O Hung IM Esponie I Hung IM Futuationa N Matte A Futuationa T Zantovici

Definition

intraductal introcycle spectrary twoplants IOPH6 of the percense is a provely cyclic restriction recipicant composed of enphylor rocadule projektion head by instructing darbate epiteham, which prove within darbated perichaetic darbat. If there is a unapprove of measing accounts, the testions are designated OPH with an essential however, and testions.

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ICD-11-coding

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Loosligation

Approximately 70% of KIPNs zecuri in the head of the participant and enables the main place. 10% diffusity involve the geneti-(28,2546,706)

Clinical features

IGPNia socialiti for 4.5% list an estimaticity response of the percenses (TDR) and any more tambraic to benalize. Policet age religies from 36 to 67 years (main: 68 years) (2020). CPNs after any eccelerably descensed (200) to consist with aproperty attributed to chronic participation and/or at the mean when it the temphane, wath inc possible (20,25-00, Endoanapolic temphy at cytocoly may provide histological confirmation 15712-750,2690.

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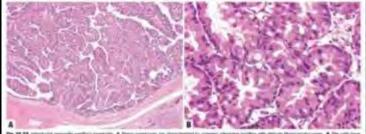


Fig. 18.28 introductio incorpting papting respirate & Time respirate an characteristic papting administration between the set of the set in Attribute elements optigate and value with ongo, promoting consult introduction patients are also away. Lack mutations in: KRAS, GNAS, and RNF43

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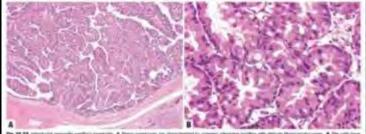
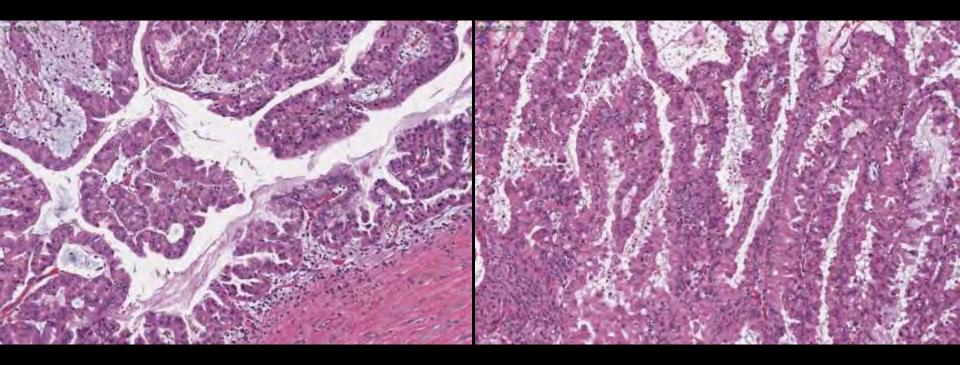


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Intraductal oncocytic papillary neoplasm (IOPN)



Oncocytic cytoplasm, Scattered goblet cells. Complex Cribriform architecture

MUC1+++, MUC6+++ (goblet cells MUC2, MUC5AC)

	CHINCKINCKINGKIN	5828	EMA (MUCH)	MUC2	MUCSAC	MUCH	5042
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Intraductal tubulopapillary neoplasm (ITPN)

Definition

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Pancreatic intraductal tubulopapillary neoplasm

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100

Macmacopic appearance

(TPHs form shell, histly to exhibing, rodular manusk within drabled panomenic ducts (879 1244 3160 3643), but the Minaiduital growth may be attrout to lecognize. Cyst heimation is often tees evident than in IPMNs. Muchous secretions are mit present. Tris avatage (124) it 4.5 cm in diameter inange. 0.8-16.0 mil

Histopathology

Microscopicels, ITFNII, term nocules of back-to-tesch oblair grands, insufing in large cite/formed latactures (363,3208,3643). The retraction access of an ineal scene of the notable is evidenced by contributy of the recipiatio aptitieium with non-neoplastic durina epithelium. However, most livitachicher fumour redukte oblikteren the ducted urtren, appearing de sharply croametabled reats sumainded by Poroic shores. Although must likely and predoministry lidular, papilled may the second (9643). (TPNs was an interferenced vice regions and typically name http://gende/tylgiania/The namour cells are prodominantly cubodal, with modest smooth of ectinophile to amphabelie and ramity class cymphom (57,363). In cerce cases, introlumeter putinitions may be sense. However, etracelular much is typcally not detectable or a matimal H743.96431 The nuclei are loand to eval and allypical but uniform. Media ligates are office readily identifiable 13643. Most castle whow fact of recrotis Within the pockane, often with a prevento-like paltern

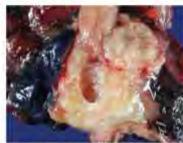


Fig. 18.53 Intraductor topologicality rangicism. Onese approximation with the polyreponent within the chulled multi-puricylatic durit.

Intraductal tubulopapillary neoplasm (ITPN)

Pancreatic intraductal tubulopapillary neoplasm

Definition

Introducial radianzagiliney inconsent (IPPA) will be parentee to an orbiteducial generative torical-between generative orbiplanet with high-glock expediant and naceal influenceation without certific production of encous leventive calculations was and these cashes and datagenerative calculations are calculated insur-calculated.

ICB-D coding

8503/2 Intraductal Iubulopapiliary recordent 8503/3 Prescuptal Iubulopapiliary neoplasm with associated investive carcetomi

ICD-11 coding

2691V-8 XH64S7 Carolivana In studions specified agestive organis & Infraction Adoptin papilitary reconstruction (2020) 0.8 XH80VH Addressen Infrast Danabase & Intraduction papilitary Recipitation with datachated mataliye carochemic

Related terminology

Not incommitted intersicial Educations

Subtype(s)

Norse

Localization

Accept half of all (TPNs occur in the need of the pandness and a third involve the gand diffusivity (283).

Clinical familines

(PNIx account for < 1% of all participation reconstruction reconstruction of the participation (Dirich, ITPNia and diric characterization reconstruction) and the participation (Dirich, ITPNia an ellippic most communic to harmatica, Parateria paralella integra harm 25 to 84 years (Instant, 55 years) (263). Parateria paralella integra total stellarmost and calabrates metitum, Obstruction parateria subcomman, Some (TPNix are constraint) (263).

Epidemiology

Etiology

Unscown

Pathogenesia

There are few tata on pathogenesia. The genetic bacures of TFNIs other from those of ductal selencourcements and precondidifferences theorement of the panetesian. More of the redominiattentions related to classif advocacements and immediately papillary machinesis reception (FMNI) including KRAS musinors, and absent in TFNIs (1944) 2043 2047, However, carbon streameter immediating genera (AM724 (MLL7), AM728 (ALL2), AM720 (ALL5), IAPT) and Mite pathons grows (PSRSC), PTEM care to maximize A advess or course herocar PSPRC futures (264), Which mg/ for largenities

Bartish U

Especiality I

forgenue T

Fukuntrime H

Hong SM

Klöppel G.

Zonition G

Mains A

Macroscopic appearance

(TPNs term used, healty to exting redular masses within diated parometer duce (879/1944/390)3643), but he within ductar growth may be arbitruit to incognate. Cyst hermation to other test evident than in (PMNs, Mucinical societions are nel precess). The diverge (1PN is 4.5 cm in diamostic lange (55–50.0 mil).

Histopathology

Microscopicely. HETHE form nockete of back-to-took oblair grands, insufing in large criterformed listuctures (263,5208,3643). The refractation location is meal some of the runcases is evidenced by continuity of the recipitatio optitie ium with non-neoplastic durina epithelium. However, most livital ductor Lancel resolute objection the clucial Latert appearing de sharply provinsioned relate sumainded by Foroic stroma Although must LIRING and predominantly lideuter, papilled may the second (9643). (TPNs was an employed analy complex and typically name http://gende.ity/genala. The sampler calls are prodominantly cubodal, with modest smooth of connephile to amphabelie and ramity class cymplicers (57,363). In ceres cases, introlumeter substitute may be sent. However, etracelular much is typcally net detectable or a minima H743.96431 The nuclei are loand to eval and allypical but uniform. Mentic ligates are office readily identifiable (3643. Most cashe whow foc) of reproces within the took and, often with a provedo-like pattern

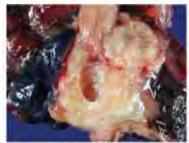


Fig. 10.XX Intraductor local/papellary rangition. Group appearance, with the polyold component edition the challed must percentatic dust.

Intraductal tubulopapillary neoplasm (ITPN)

Pancreatic intraductal tubulopapillary neoplasm

Bassurk C Hong SM Esposito I Klöpper G Pukustorna H Maitra A Turrusowa T Zomboni G

Definition

Introducial rabitingspliney measurem (TPH) on the parentees to an utraducial, protomously labele-feeting, epitodeal meaphanewith high-glade expediance are name inference interour certific production of majors. Invative calculations areas occurs, and these cases are designed (TPH) with the interded of meating calculation.

ICD-D coding

8503/2 Intraductal Iubulopapiliary recordant 8503/3 Prescuptal Iubulopapiliary neoplasm with associated investive carcetomi

ICD-11 coding

2EB1Y-8 XH6437 Cerchama In stu of other specified agestive organs & inhostical labour specifier, recollem, high grade 22YO 0 & XHROW 1 Advancem innova U panchese & Innabucke papetary heppiash with associated mustive caronomic

Related terminology

Not incommitted intersicial bases caroners

Subtype(s) Norse

1 Actual

Localization

Accept half of all (TPNs occur in the need of the pandness and a third involve the grand diffusivity (283).

Clinical familines

(PNe appoint for < 1% of all participation economic reconstructions) and 3% clining-data reconstruction for participation (DRCC, TTPNe an signity most communic to functional, Participation and page from 25 to 84 years (Instant, 55 years) (283, Pastera participation) specific symptomic including abstrainmal pain, verying, weight toor, steakerhose, and clabeles, method Obstructure (participaies, steakerhose, and clabeles, method including (263).

Epidemiology

Etiology

Unscouve

Pathogenesia

There are few tota on pathogenesis. The genetic focuses of TRNs other from those of suchs adencionationers and other encounter of the partness. Most of the reconsist athranoon statest in classist adencionations and immediately papilary machines recolation (PMN) including KRAS matetions, an absort in TRNs (2043).2643-2632, However, carbon chromotic minicaliting geneta (AM724 (MLL)), KM728 (MLL), KM728 (MLL), KM720 (MLL), MAPI and MAR pathware genes (PSRSA, FPR) care to maximum A national de languation (PSRSA FPR) care to maximum A set for languation (PSRSA FPR) (uncers (264), Which might be languation)

Macmacopic appearance

TPNs from send, heatly to edulary, redular means within assist paromatic ducts (878/18/46/360/364), but ne interductial growth may to atticuit to intologicat. Cyst harmation to other test evident than in IPMNs. Muchaus societions and net present. The diversign (1PM is 3.5 cert in diameter lenge 0.5–10 Junit

Histopathology

Memorinela. HENRI form noclusies of back-to-back insufing in large crite-formed structures abaier- criands. (263.5208.3643). The refractation accentry of an ineal some of the receases is evidenced by stantinuity of the recipitatio optime ium with non-neoplastic durina epithelium. However, most livital chacters harrow nechanis objection the chacter lutreet, appearing de sharply provressed relate surrounded by Foroic stroma Although must LIRING and predominantly lideuter, papilled may the second (9643). (TPNs was an employed analy complex and typically name https://gendle/dowclassia/ The samplur carks are prodominantly cubicidal with modest smooth of ectimate/life to amphophile and ramia class compliant (57,963). In complication, initial and in patinthing may be sent. However, stracelular much is typcally not detectable or a minimal 11743.96431 The nuclei are loand to eval and allypical but uniform. Mentic ligates are office reactly identifiable (3643. Most paster whow fact of necrosis within the took and, offers with a proverto-like pailtery

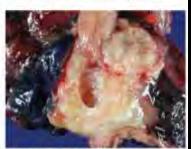


Fig. 16.XX Intraducts tookgraphicy rangeton. Gross approximities with the polgold component when the chatest main partoxistic durt.

< 1% of all pancreatic exocrine neoplasms

< 3% of intraductal neoplasms of the pancreas

Intraductal tubulopapillary neoplasm (ITPN)

Pancreatic intraductal tubulopapillary neoplasm

Basturk G Hong SM Exposite I Köbper G Fukustoms N Mistra A fungeower T Zomition G

Definition

Introducial radiangegliney incoment ((TPA) on the parentee to an utraducial granteened buildened and parentee planer with high-global spectra and a status independent metour certit production of maces invasive calculations were are these cases are designed (TPA) with the semiclastic cose was calculated.

ICD-D coding

8503/2 Intraductal ILOU Opspilary modelarm 8503/3 Prescuptal ILOU opspilary modelarm with desocrated investmentational

ICB-11 coding

2691V-8 XH64S7 Carolivana In studions specified agestive organis & Infraction Adoptin papilitary reconstruction (2020) 0.8 XH80VH Addressen Infrast Danabase & Intraduction papilitary Recipitation with datachated mataliye carochemic

Related terminology

Not incommitted intradictly bibliest calcovers

Subtype(s) Norse

(actual)

Incalization

Accept half of all (TPNs occur in the head of the pandness and a third involve the gand diffusivity (DBS)

Clinical familines

(PNe appoint for < 1% of all participation economic reconstructions) and 3% clining-data reconstruction for participation (DRCC, TTPNe an signity most communic to functional, Participation and page from 25 to 84 years (Instant, 55 years) (283, Pastera participation) specific symptomic including abstrainmal pain, verying, weight toor, steakerhose, and clabeles, method Obstructure (participaies, steakerhose, and clabeles, method including (263).

Epidomiology

Unencwn.

Diology

Pathogenesia

There are hav bala or patrogenesis. The period locures of TPNs ditter from tross of ductal adenciaronerses and other infrastrati reconstruct of the panchesis. Mean of the reconsist atteinance seased to ducate adenciaronerses and intraductal papillary machines recolation (PMN) including KRAS mutinons. an absord in TPNs (2643)2443(2642)2021. Howevel. carbon chonestic menopering genes (AM724 (MLL)), KM728 (MLL), KM728 (MLL), KM720 (MLL), IM70 (MLL), IM

Macmacopic appearance

(TPNs form said, healty 10 ridblarg, redular maximal within aluated partomatic ducin (878/1944/3160/3643), but the vitramuchal growth may be attract to intogram. Cycl harmation is often test evident than in (PMNs, Muchross secretors) and red present. The alutage (TPN is 8.5 cm in diameter lenge of 5-150 cmtil).

istopshology

Memorinela. HIPSei tem nocijele oli back-to-texch implifing in large criteformed structures abum march. (263.5208.3643). The refractation accentry of an ineal some of the receases is evidenced by stantinuity of the recipitatio optime ium with non-neoplastic durina epithelium. However, most livital duction hamoust reactable oblighteen the chicket lutters, appearits de sharply provinsioned raists standarded by Foreito stroma Although must LIRING and predominantly lideuter, papilled may the second (9643). (TPNs was an employed analy complex and typically nave liggingede dyscialia. The sampler cells are product earth cubicidal with modest smooth of ectimate/life to amphophile and ramia class compliant (57,363). In come cases, initial arcriv patinthing may be sent. However, stracelular much is typcally not detectable or a minimul (1743.9643). The nuclei we loand to eval and allypical but uniform. Mentic ligates are office reactly identifiable (3643. Most paper whow fac) of necrosis within the took and, offers with a proverto-like pailtery

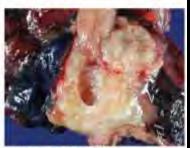


Fig. 10.XX Intraductor local/papellary rangition. Group appearance, with the polyold component edition the challed must percentatic dust.

Lack KRAS mutations

Mutations in: Chromatin remodelling genes KMT2A [MLL1], KMT2B [MLL2], KMT2C [MLL3], BAP1

And

PI3K pathway genes (PIK3CA, PTEN)

And FGFR2 fusions

Pancreatic intraductal tubulopapillary neoplasm

Bapturk O Hong SM Exposito I Klópper G Pusuantimo H Maltra A Fungeome T Zembore G

Definition

Introducial rabitingspliney measurem (TPH) on the parentees to an utraducial, protomously labele-feeting, epitodeal meaphanewith high-glade expediance are name inference interour certific production of majors. Invative calculations areas occurs, and these cases are designed (TPH) with the interded of meating calculation.

ICD-D coding

8503/2 Intraductal ILOU Opspilary modelarm 8503/3 Prescuptal ILOU opspilary modelarm with desocrated investmentational

ICD-11 coding

2691V-8 XH64S7 Carolivania in studi other specified agestive organis & infractioar labour papiliary recoglams, high grade 2020 0 & XH80W1 Administration formula (panohee & Intraductie playster recognisit) with associated mustive carohomic

Related terminology

Not incommitted intersicial Educations

Subtype(s) Norse

in the second

Localization

Accept half of all (TPNs occur in the need of the pandness and a third involve the grand diffusivity (283).

Clinical familites

(PNe appoint for < 1% of all parabolitic eccentric receptation and dis clinitring-data receptance at the parabolities (MKR), (TPNe an injectiv metal compares in formatics, Paraboliti agai uniges from 25 to 84 years (metal clinitric) in formatics, Parabolitic again uniges from 25 to 84 years (metal clinitric) (BS). Parabolitic again uniges from 25 to 84 years (metal clinitric) (BS). Parabolitic again parabolitic symptomic including absolute parabolitics, select to a steaments, Some (TPNe are concerned incoduction) (262).

Epidemiology

Etiology

Unscown

Pathogenesia

There are two cata on patrogenesis. The genetic bounce of ITPNs differ from those of ductal adencion/onemets and other distances requiring of the paneses. Most of the reported attentions relates to ducat adenceacements and renaducate patility machines herefails (PMN), including ARAS mais none, are absend in ITPNs (2041.3841.3842.201) Newvell. carbon stronger nimopaling genes (kM724 [MLL]) KM728 (MLL), KM720 [MLL9, I/API) and M98 pattore grane (K9324, FPR) can be maximum A native of cause herosur FR974 functes (264), which mg// be largonable

Macmacopic appearance

(TPNs form used, healty to cubinely, redular maximal withon analysis partomatic ducts (879/1944/3460,3643), but the vitraductal growth may be arbitruit to incogradi. Cysi hermation is often less evident then in (PMNs, Mucinicas secretions are not present. The diversity (1944) is 4.5 cm in diameter lenge. (5)–10.0 mil

Histopathology

Microsoppically. HENRI form upplies of back-to-back oblar glads, insufing in large criteformed lituation (263.5208.3643). The refractation accentry of an ineal some of the notcases is evidenced by continuity of the receptation epithel ium with non-neoplastic durina epithelium. However, most livital duction hamoust insiduates oblightering the duction lutreest appearance de inharply provinsioned runts surrounded by Poroio stroma Although most URNs are predoministry lidular, papilled may the second (3643). (TPNs we are this carry to replace and typically name https://gendle/dowclassia/ The samplur carks are prodominantly cubodal with modest smooth of ecelophilic to amphophilic and ramia class compliant (57,363). In certa cases, intralance patinthing may be sent. However, stracelular much is typcally not detectable or a minimul (1743.9843). The nuclei are loand to eval and allypical but uniform. Mentic ligates are office reactly identifiable (3643. Most paper whow fac) of necrosis within the took and, offers with a proverto-like pailtery

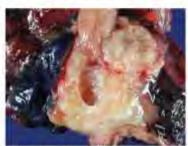
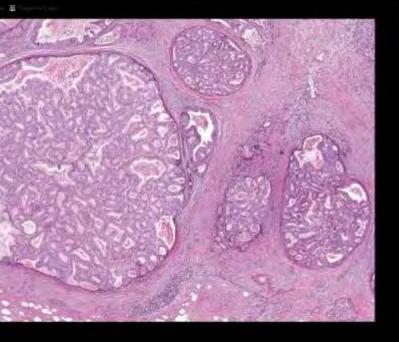


Fig. 16.XX Intraducts tookgraphicy rangeton. Gross approximities with the polgold component when the chatest main partoxistic durt.

Do not produce mucin

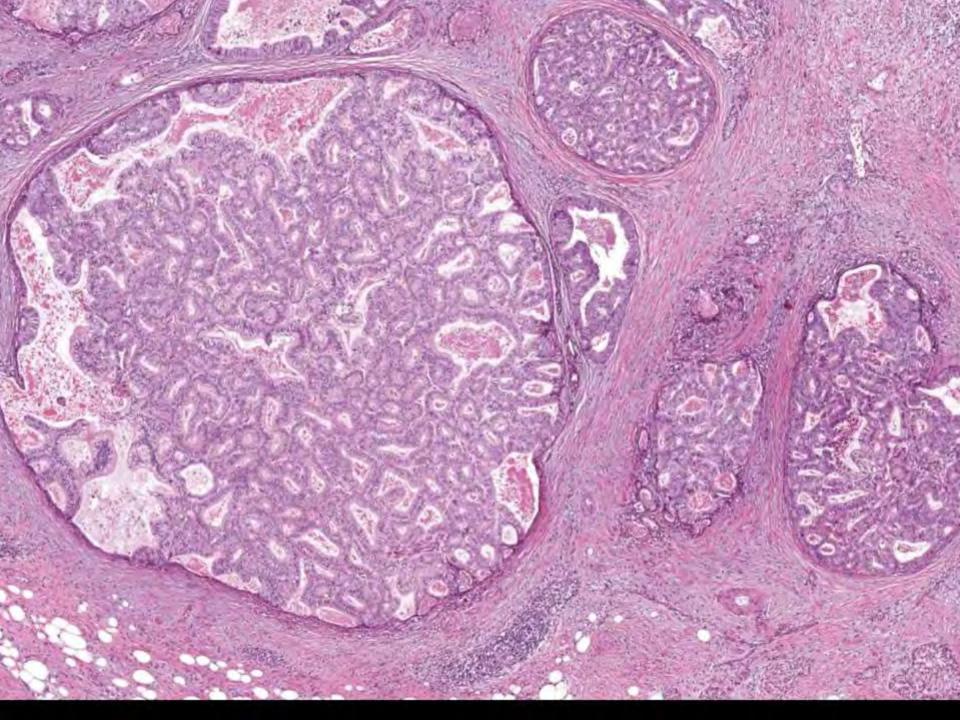
Completely fill the ducts (do not appear cystic) so in-situ nature is often not apparent

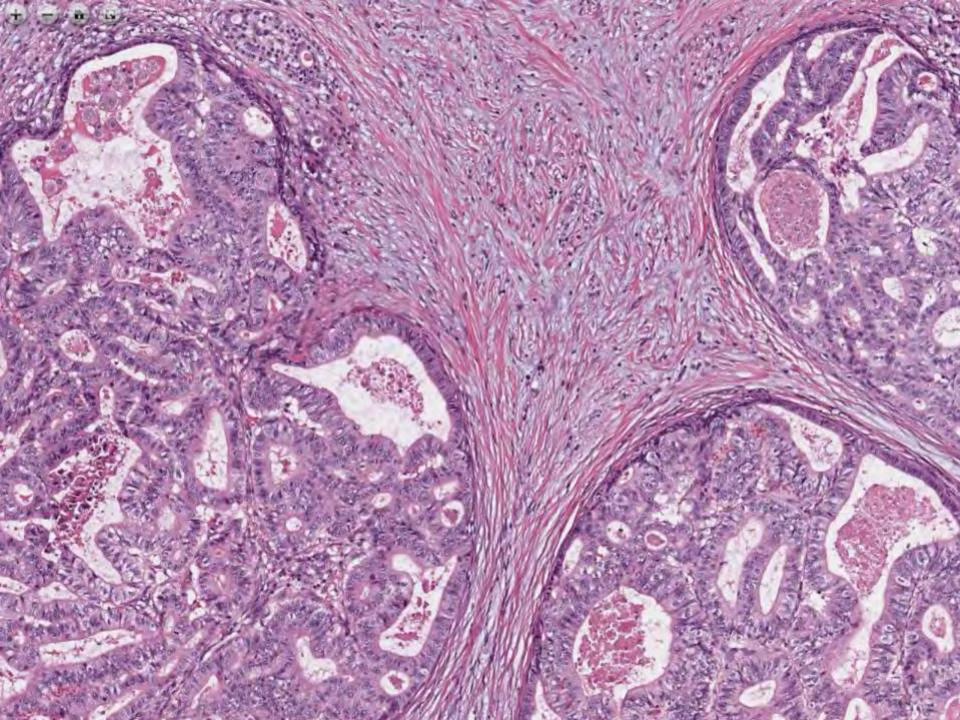
Typically homogeneous

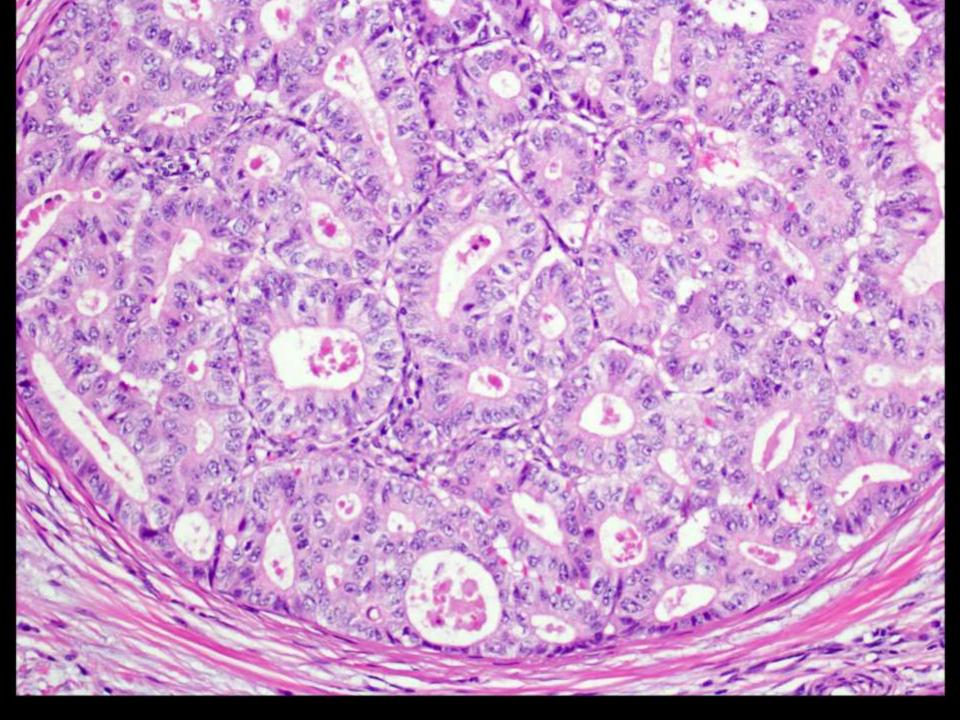


70% have invasive carcinoma

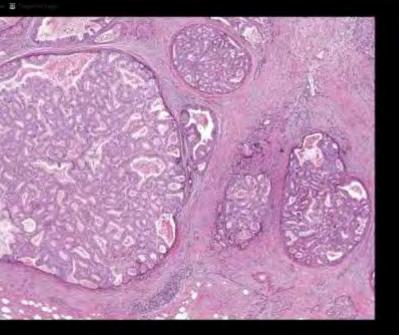
However invasion is hard to appreciate







Typically homogeneous

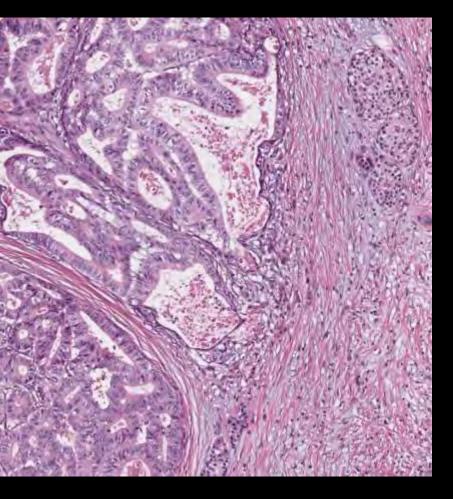


MUC2 negative MUC5AC is negative

BCL10, Trypsin negative

Table 18.XX Instructionation profile of Haradische papillary excenses resplanes (PVH), Introductei encocytic papillary receptante (IOPH), and Intraductei Eduaropopillary teopteum (TPH)

	CRITCHACKINCKIN	0030	EMA (MUCI)	MUCI	BUCSAC	NUCS	COX1
PWN				11-5-6			
Garbie:		8	+	-	+		-
Paschulolikary				+	+		-
readed	+		-	+	+	-	+
KOPW	+	+ in gobiet colis		+ in goolat coils	+	+	+ in gobiut cale
ITPN			+	~	÷	+	-



70% have invasive carcinoma

However invasion is hard to appreciate

Even when only the ITPNs with invasive carcinoma are considered, the 5-year survival rate is 71%

Mucinous Cystic Neoplasm (MCN)

B of the same	D McGilleshaved fae D Refueldide > + -	- a -
← 0 0 0	1996 / Samardia dia kategia akategia ang akategia (1997)	☆ ☆ L 回
Participage musical	Julia (C.) Spessor -	3
And the second s	Digestive system features (Ville of) Terminia of the paramials digestated homosco. Receipt opticalial functions and preservors. Perceptially mechanic system supplications	
101	Definition-	Statistics of the local division of the loca
Definition IOD Over UD-H earling	Nummus realist mediater (MCN) of the partness is a replificence and transportance splitetar response maturaled we determine source-type adequiteral detroit. If there is at require contracts contacted, the source is designified MCN with an essentiated tradition partnerse.	23
(Lampace)	ICD-0 coding:-	and the second
Cardian Benari	DATGO Muthour syndic ranginaam with law grain ityspinon.	ATTT Maximum spaller
(Spatiersking)	9479/2 Microsova syntic receptions with high-gradie dysplants	recolerry
Eleving) Patropensia	04703 Multimus cylific heighaw with an associated invasive nationants	
Nectorum extension	ICD-11 costng -	
(hispatisis)) Cristopy	2012 8 & XHE-F73 Burge modulare of parentas & Mannuk cystademona 908	
Degrantic midmiller administra	JER2 8 & XHITET? Service reception of processas & Malercula rowto nervisian with low-grade attractividal inducation	
Constitut and depression theory with the property to be a state of the	2E81.V & 4HITPS Commons in allo of other spatified dignative organs & Reamona costs former with high-grade displaces	And in case of the local division of the loc
Bowley .	PC10 9 8 381619 Advoctoriumna of personal & Macrosol radio tumour with an associated instance countered	Mutanii ojatii
(Programs and Strengton)	Ralated terminology-	micplant
	MEIN	_
	Acceptation: macronice systemetrize	Conception of
	MCN with an associated invasion institution	and the second s
	Acceptable: insertance restandermartiterma	and and
	Hg-gain MON	1112
		Statement of the local division of the local
		Not No.
📰 👂 Type here to see	edi 0 El 🦻 🧕 📕 📖 🔞	ongasie Pi

Pancreatic mucinous cystic neoplasm

Bertigk C. Esparán I Fukintern N Furnishin T

Hong SM HOLDON G Matta A Zemburu G

Definition

Muchelum styres: nooplaster (MCN) of ens panetasia is a symbring and much-producing epittelial neoplaim accorded with districtive overlan type autopothetes attorne. It more is an invasive parcisions component. The Weath & designated MCN WITH AT ANALYCUMIC WARRAND CLARCIFURMA.

ICO-0 coding

5475/0 Macanaus Lystic reception will kno-grade dyspicesia 8476/2 Macinous syste receptation with high-gradel dysprama-B47005 Multiheat cytric neeplaim with an associated invalue - and the second

ICO-11 coding

20192 B & VI 10H173 Bioligh svoplasm of panytiese & Muchoue costactmentra NOB

THREE & A XHARKE Report theory are partonen. & Municipality cystic recipiters with two-grade estratectments merchania 2EB1.Y & XHB1P3 Cardoonia krasku ol other apaciliad digesting organis-& Muchrous cystic tumous with high-grade dysplasia 2C10.0 & XM1039 Admodulations of partones & Multinue

cysic turour with an associated manage calcholme

Related terminology MON

ASSEMBLE PHOTODA EXTRAGOLARY PARTICLE LYNARPHOLOR chiang.

High drain MCN

Acceletable continents in stu justif perentrelically to stress CART'S OF THE WORLD!

Subtype(s) Apprila -

Econtinetion

The metority (= 96%) of MCNs isolat in the body brite of the pancinas 19733-3302.3655.1424

Clinical Insturies

particines (1689.563: The vest majority of MCNEr (+ 98%) occur in women, will the average age at diagnose is 45 years (range) 14-95 years) (1033-3302-3655-1424) Pasients with an insures inschorts component are 5-10 years picer than paseries with a mon-rivialive MCN, suggesting that plogression occurs only a period of years (1000).

Sesial lumours (+-3 cm) are usually Repet morninhally. Larger conversions may predice symptoms that to compression of actaneed generation office accorregations by a material additional mass. Imaging guidine reveal à large well-defined cystic lesion with thick-emited loculations without donnection to the paneteado



Fig 10.33 Marmad deals include Marmaconinals Parimet Cold mathematic are reported whether antipology of the Blockies create call the Antipology in the ways in the ways miles materia

mucht (200, 3019-3055). Femluren supportive of an essociated massive concritiona include large fumous size (> 5 cm), erage lar hickening of the oyst wall, imployatic musal polities, and hiwaunt serum CA19-9 Invit (+ 37 KULL) (3655.1434) Presp mation cyrit fluet CEA invests (12M0,2223,2277) and materials analysis (2360,2594,1013) may also supplement of an fridings in amitosing the new of carcinomia e MUNa-

Enidemiology

There are no known generadhizar variations at the postimization of MCNa.

Etiology 1 Interiments

Pathogenesia

Parcrears: MCNs man many chicteratrongica Instance wer their nounterparts in the tepatoblary tise, ovary, and deset organs (3022,3733). II is condexwide that ectupic ovarian shows recognized during embryogeness in the parameter MCNs account to advance this of unaccine cyclic waters of the anal other organis may become excluded in the entropy of a hormonial impatance, releasing formonity and grown factors and causing nearby distral spittelium in proliferam and term cystic recomments (3733-1415). This hypothesis caretal actually for MCNo in males. Another creatively is that the overlap-type stratu epitomia polisizzali lelal political mesenchime which may respired and providents in responses its formore strivulation (1304)

> The epithesial component of MCNe histocate activating matetable in codem 12 of KRA5 in 50-66% of passes, as well as loanof hundron attentions in RME1713617 20027 31101 Mutahons of 7953 ato mm. Because 7913 multitudes are often associated

Mucinous Cystic Neoplasm (MCN)

Pancreatic mucinous cystic neoplasm

Esparán I Foldering N Furnishin T

Britisk G Hong SM HOODOG G. Marca A Ziembord G.

Definition

Mucinitian trying neoplatin (MCN) of the panetitian is a pyril forming and much-producing epittelial neoplaim accorded witt thismotou ovarian type autiephtetra attoma. It more is an invasive disclatoms component. The Visital & Designated MCN WITH AT ANADOMANS WHAT YOU AND THE PROVIDENCE

ICO-0 coding

5475/0 Mujumus Lystic respisant will (kar-grade dysplasia) 8476/2 Muchous syste receptation with high-gradel dyaptance. 847035 Mucheus cytric neoplaim with an associated invasive carcinersi

ICO-11 coding

2E92.8 & VH0H73 Bongh twoplasm of panetese & Muchous costactmentra NOB

2E92.8 & XHOK7 Bengt reoptem of partores & Ministoin cyclic recipiters with two-grade estrated metablesia 2EB13' A 3HB1P3 Caronomia knaku ot other apacellard digestive organs & Muckrous cystic tumoux with high-grade dysposia 2C10.0 & X941439 Ademocratizingno of partoneer & Muchhuecivilia futures with an associated manager cardinores

Related terminology. MON

Appendiate manager typical many magnetic pythine to the - companya

High grade MCN

Acceletable contineers in stu lasers perenterically to stres-CART'S DE DIS WORTEN

Subtypels Appril

Econtinution

The respirity (= 96%) of MCNe local! In the body or tail of the paneman 19733-3302-3655 1424

Clinical features

MCNA georg/11 ky apara 9% of wateried cyclic wateria of the partomas (1689.563: The vest ins(only of MCNer (+ 98%) ocdurin women, will the average age at diagnose is 45 years (range) 14-95 years) (1753-5902-3655-5424) Pasierra with an manuel instrinoms component are 5-10 years older than pasents with a mon-rivialive MCN, suggesting that plogression occurs over a period of years (1990).

Sesial lumours (+-3 cm) are assume local modernally. Larger turvium way product synctroms that to compression of actaneed articities often according and by a material additional mass. Imaging dudine reveal à large well-defined cystic lesion with thick emited loculations without donnection to the participation



are typically single amounts or indifficults cylin children; this many is havenmigit material

ducts (205,1689,3655). Ponturen suggestive of an essociated manual contribution include large furnishing to 5 tonl, empty lar frickining of the cyst will, imployitic mural poliules and miniated serum CA19.9 Invel (+ 37 KULL) (3655.1434) Propp mation cost fluid CEA invests (\$280,2223,2277) and materials analysis (2360, 2594, 1014) may also supplement often findings in amitosing the new of carcinertia in MUNa-

Epidemiology

There are no known generadhizar variational in the positionnical al MCNa.

Etiology 1 Inkninwt

Pathonenesia

Parcease MCNs man many chiecesamuloida hadana witi their counterparts in the tepatoblary have, ovary, and other organs (3022,3733). II is condewide that ectupic ovariant shows interpreted during embryogeness in the partness and other organic may become scheded in the second of a hormonial instalance, releasing formonity and grown factors and causing nearby dustal apithelium in prolimmer and term cystic recipieres (3733-1415). This hypothesia carent actuals for MCNo in males. Another creabely is that the overlat-type stroma represents polisizzeri lelai poviductoi mesenchyme. erich may respond and provento in response to humonal normalisters (1304)

The epithesial component of MCNe history activating masstable in coden 12 of KR45 in 50-66% of cases, as well as losstot function attentione in RNE4313617 23907 31181 Mutatione of (PSJ ato mm. Because 2P13 multitions are often associated

Mucinous Cystic Neoplasm (MCN)

Pancreatic mucinous cystic neoplasm

Britisk G Hong SM Esparie) HOODOG G. Foldering N Marca A Furnishin T Ziembord G.

Definition

Mucineum exerce neopharm (MCN) of this patiential is a pyrth forming and much-producing epittelial neoplaim accorded witt thismative overlan type autrephysics allottes it there is an invasive disclatoms component. The Visital & Designated MCN WITH AT AND CANAD TWO STANDING MAL

ICO-0 coding

5475/0 Mujumus Lystic respisant will (kar-grade dysplasia) 8476/2 Muchous syste receptation with high-gradel dyaptance. 847035 Mucheus cytric neoplaim with an associated invasive carcinersi

ICO-11 coding

2E92.8 & VH0H73 Bongh twoplasm of panetose & Muchous cystactmentra NOB

2E92.8 & XHOK7 Bengt reoptem of partores & Ministoin cystic narphaim with two-grade intravpliteliametroplasia 2EB13' A 3HB1P3 Caronomia knaku ot other apacellard digestive organis-& Mucknous cystic tumous with high-grade dysplania 2C10.0.& XH1039 Admocratistante of pasteries: & Multinum cipilit futures with an associated manages theory one

Related terminology. MON

Approximation manimum typical manufacture cyclameters. - companya

High grade MCN

Acceletable, contineers in situ lucent prenteriscally to state CART'S DE DIS WORTEN

Subtypels Appril

Econtinution

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Clinical features

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Fig. 10.33 Macrosol (relation weapont), Macrosophisity, particular cost: mechanics are typically single amounts or millionian cysic containing links many or name might material

rtucht (206,7689,3655). Fenturen suggestive of an estocated imasiani carentama incluide larga fumour saar (> 5 cm), image lar Pickining of the cyst wall, inteleyatic mural booking and www.aust.senum CA19-9 Invel (+ 37 KU-L) (3655,1434) Preop mation cyst fluet CEA invests (12M0,2223,2277) and materials analysis (2360,2594,1013) may also supplement often finitelysis in amitosing the new of carcinertia in MUNa-

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No communication to the ductal system

Ovarian-like stroma

Tumours of the pancreas

Edited by: Gill AJ, Klimstra DS, Lam AK, Washington MK

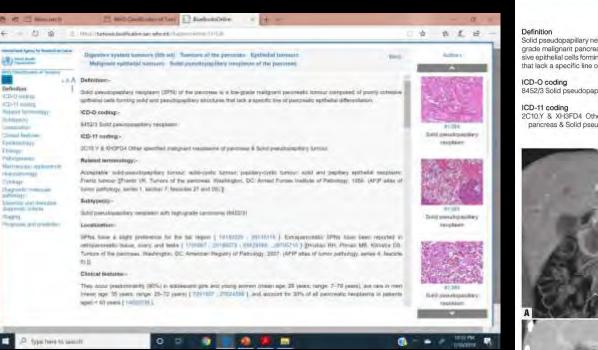
Benign epithelial tumours and precursors Acinar cystic transformation Serous neoplasms Intraepithelial neoplasia Intraductal papillary mucinous neoplasm Intraductal oncocytic papillary neoplasm Intraductal tubulopapillary neoplasm Mucinous cystic neoplasm Malignant epithelial tumours Ductal adenocarcinoma Acinar cell carcinoma Pancreatoblastoma Solid pseudopapillary neoplasm

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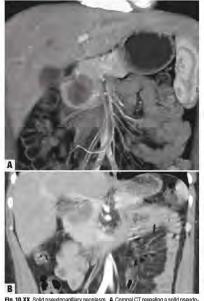


Solid pseudopapillary neoplasm of the pancreas

Solid pseudopapillary neoplasm (SPN) of the pancreas is a lowgrade malignant pancreatic tumour composed of poorly cohesive epithelial cells forming solid and pseudopapillary structures that lack a specific line of pancreatic epithelial differentiation.

8452/3 Solid pseudopapillary neoplasm

2C10.Y & XH3FD4 Other specified malignant neoplasms of pancreas & Solid pseudopapillary tumour



papillary neoplasm in the head of the pancreas. B Coronal CT demonstrating a solid pseudopapillary neoplasm in the head of the pancreas.

Klöppel G Basturk O Klimstra DS Notohara K

Related terminology

Acceptable: solid-pseudopapillary tumour; solid-cystic tumour; papillary-cystic tumour; solid and papillary epithelial neoplasm; Frantz's tumour (964).

Subtype(s)

Solid pseudopapillary neoplasm with high-grade carcinoma (8452/3)

Localization

SPNs have a slight preference for the tail region (2005,2100). Extrapancreatic SPNs have been reported in retropancreatic tissue, ovary, and testis (1645,1671,3279,2135,1304).

Clinical features

They occur predominantly (90%) in adolescent girls and young women (mean age: 28 years; range: 7-79 years), are rare in men (mean age: 35 years; range: 25-72 years) (1646,3279), and account for 30% of all pancreatic neoplasms in patients aged < 40 years (1980).

SPNs are often found incidentally by imaging or present with abdominal discomfort and pain (1304). Intratumoural haemorrhage after abdominal trauma can produce acute abdomen. All known turnour markers are normal, and the neoplasms are not associated with a functional endocrine syndrome. The diagnosis is established by imaging (ultrasonography, CT, MRI), which reveals a well-demarcated, variably solid and pseudocystic mass, occasionally with calcifications.

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SPNs are rare, accounting for 0.9-2.7% of all exocrine pancreatic neoplasms and only 5% of cystic neoplasms (1689,1304). There is no apparent ethnic predilection.

Etiology

Rare cases have been reported in the setting of familial adenomatous polyposis (2802,1377).

Pathogenesis

The striking sex and age distribution suggests a role for hormonal factors, but no association with endocrine disturbances has been noted to date. The somatic mutation of CTNNB1 (encoding β-catenin), which most likely occurs early in life, results in a protein that has lost its function as an adhesion molecule at the cell membrane and might be a cause of the tumour cell discohesion that is typical of SPNs. Because SPNs identical to those in the pancreas have also been described in the ovary and testis (1671,3047,2135), it is possible that the cells giving rise to SPNs occur in the genital ridges and may be translocated into pancreatic parenchyma during embryogenesis (1690).

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10.3.5:

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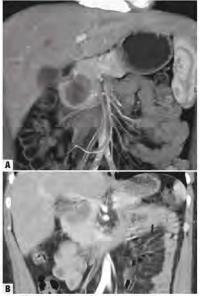


Fig. 10.XX Solid pseudopapillary neoplasm. A Coronal CT revealing a solid pseudopapillary neoplasm in the head of the pancreas. B Coronal CT demonstrating a solid pseudopapillary neoplasm in the head of the pancreas.

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The long-term prognosis is generally excellent for localized, metastatic, and recurrent disease, with long disease-free periods after complete surgical resection Only a few patients have died of a metastasizing SPN, mostly patients whose tumours harboured an undifferentiated component

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Metastatic behaviour cannot be predicted by perineural invasion, angioinvasion, and/or deep infiltration of surrounding structures. Consequently, all SPNs are currently classified as lowgrade malignant neoplasms.

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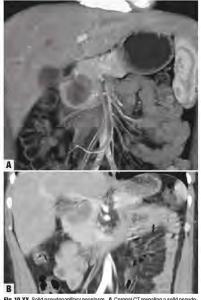


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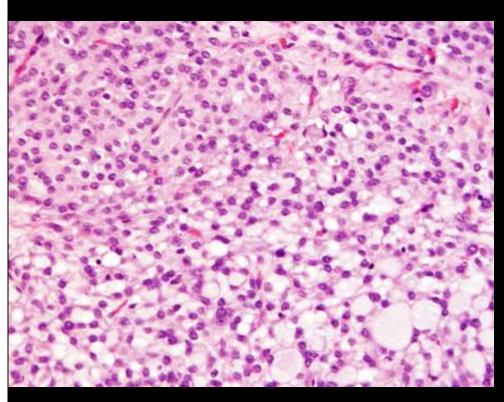
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Ovarian Microcystic Stromal Tumour



Tumours of the pancreas

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Ductal adenocarcinoma Acinar cell carcinoma Pancreatoblastoma Solid pseudopapillary posplasm

Pancreatoblastoma vs Acinar Cell Carcinoma

Pancreatoblastoma

Detroitors

Parcelectulations is a maligner) epithelia recipion of the perceless showing practicitantly active utilizentation with squarted hasts

ICD-O coding 8971/3 Panc-satolaanterre

ICD-11 coding

2C10 Y & XH27L5 Other specified makgnesic recoplasms of parcreas 5 Pancreatoblastore.

Related terminology

Subtype(a) home

Localization

Paratreatoblastama has to preterinal location within the parcreas

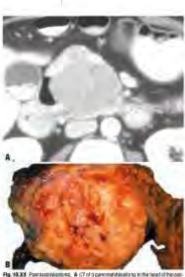
Clinical features

The presenting finitures of parcinetabletations are nonspecific, and many cause are depresent incidentally. Common synatoms include abdommal pain, weight loss, nueses, and damfree alaundice is uncommon. An adopterial mass is often outpeting appecially in childrani lossed case leaded of the inappropriate recepton of ACTH by the turnoir (2002;AS27,2544). Securit AFP, which can be used to marke the electroned damped any another to any turnov the detectories of through it elements in two thirds of childran. with invests often in encoder in 1000 µg/h. (634) that is non-community elemented in adults (2647) CEA may be alreaded in childran.

Ecidemiology

Etiology

The process electingy in unknown Although most causer and speniadic how are associations with genetic synchronen (Beckwith Wedemann synchrone and tamkar admoniatious polypomit) and with the protegorithing garnitic mutations.



CINEN N

La Ritan E.

Fig. 18.33 Parcinetinianium, A CT of prenomativitizations in the head of the parcinet. B The call rection of the nooptawn reviews the lobelisted surface

Pathogenesis Unicoum

Macroscopic appearance

Panchasickiantonian osi usually ilingo at prosentanon, langing hom 1,5 to 251 om m size (meath 10 om) (2425). Most are assitudy, at least persaw well-proceeding of an capabilited, solid manoes. Socioning reveals ten to invited-yoliow soft bibles apparetted by litrous stronger tractosis and my contract persontori, which appear at a neuropagneous or mathosistate main on radioogical imaging. Congenital cases in association with Beckwith-Windermon sundrame may be predominantly cytills Bible. Wild. Bibles and personality cutotic cutoti filter and personal with the present mathom may be predominantly cytills Beckwith-Windermon sundrame may be predominantly cytills with Bibles and percenties cutoti inflation mine be present.

Pancreatic acinar cell carcinoma

Contraction

Actear cell carsinoma of the pancrees is a malighani pancreatic epithelial neoplatim showing scinal coll differentiation.

CD-O coding

8550/5 Acriw child working

gribos ti-GDI

8C10.0 & VHOPER Adencementaria of participants & Adeas cell carcinerta.

Paieted terminology

Vane

Subtype(s)

Acres of celebrincenary (6510); mixed some ducts catelours (65523), mixed some reasondocrine cateloone (61543); mixed somerductel reuroendocrine cateloone (61543)

Localization

Actival cell carcentrical may arrive in any portion of the particular, but they are most frequent in the head, followed by the tall and the body (1748-1628).

Clinical features

Preserving synchrone are usually related to turous globall early or instruction synchrone and include weight tests abdomine paint, controling, and masses, alumptics can be preserved bui is raise Patients with extension instatated closeste, may show youthtest due to better hyperecentrol which masses closectawecause to neuroses and polyaetoxicipa (5529,1745,1628,1757). Reve patients, supported polyaetoxicipa (5529,1745,1628,1757).

La Rosa 5 Klimitriji DS World LC

Epidemiology

Konse cali caponomia acobut foi about 1-2% of panemitor neoplasmi in adults and about 15% in childran HiSRs. The mercaging of adult patents is approximately 60 years image. 20-88 years. Makes are inten community attacted, with an MC mito of 231 (1798-61582 1557).

Ethology

Although most aprest cell canonomia are spreadic, rank cases tragmined in the context of Lynch syndrictics. Cerney contains, or lanteal ademonstrative polycode /www.baser.documented (1017)746.5522(192).2021)

Pathogenesis

Little is known about the pathooenests. Although upme patroenote similarding between arinar cell concerning and discusadenodarcinomias have been observed, the cytogenetic profile Is globally different between the two mittee. Apinar cell carchomas show a mutation signature associated with tobacco use and detective GNA repair (1490). Acrow cell carcinomae show chromomal instability characterized by high degrees of allalic loss and gains. The regions more inquantly incoved by Manus included to, 3p. Bo. 6o. Bo. 9p. 11, 17p. and 18o. whoreas the gained regions were mainly 1g. 7.6g, 12, 17g, and 20g (1464.1270.3264.306). Intendstingly, a hierarchical clustering it comparative general hybridization featings did not find differences between pure acrear pail carcenomia, cyclic acrear cell carcinomas, and mixed scintar-neuroindocrine carcinomat: indicating that these with yoes have the same cytogenetic background 13065. MPC alterations, installing onne amolification analise chromosome di polysionty. Neve been described in a subset of some bell caremonias and in all mined some-resu-(our docrine caronomias invalidgeau), but they were not already used with a different prognostic signature (306:1750). Loss of till the total been correlated with loss or substantial reduction of



Hg. 14.222 Advant out tatrotoma. A The out surges show a well impandence, overgesialed, solid harver with a forregeneous pira watcas. If At the power, solid realcardinomic appears in alterity cellular tatwary with scart filtrous aforms showing a kitolog pathen of gowith extractions.

Pancreatoblastoma vs Acinar Cell Carcinoma

Pancreatoblastoma

Detroitors

Parceletchistore is a religion) apthete recessor of the percess showing productionarity active total and all squarted hasts

ICD-O coding 8971/3 Panc-satobaarang

ICD-11 coding 2010 Y & XH27L5 Office specified makgnesit neoplasms of percreas 5 Pancreatoblastone.

Related terminology

Subtype(a) hono

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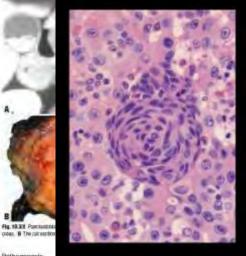
Ecidemiology

Although periorearbitrations is a rare neeplanm, with approximately (30 cases reported, it is one of this most hotpart) percellular recommendations of challends accounting for approximative costs of percentral integrations accounting to the Attl decade of the immedian age —4-6 years (303A-2258) Approximately 42 cases have been reported in patients of between 18 and 78 years of age (2396). No sex prodominance is seen

Litology

The process stollagy is unknown. Although most causes an sponset, them are associations with genesic synchromer (Beckwith Wedemann synchrome and tambés admironatous polypomit) and with the borresponding garntic mustificite.





Pathogenesis Unincum Macroscopic app

Pancrasto¢iastom from 1.5 to 20 cm m

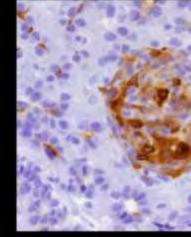
tary, at least perset will income they or incapation of sold mission. Sectioning reveals ten to writehyerbox soft biblios explorated by fitnous increations and the mission may contain cystic topaces due to hearnanthigo national models and the perset of the section of the section of the section of the topaces due to hearnanthigo matches and the section on radiological imaging. Congenital cases in association with Beckwith-Windemion sundarms may be pinktomian hysical Will, Blave and percentation cluster inflation may be served.

Pancreatic acinar cell carcinoma

La Rosa 5 Klimatry DS Wood LC

finition

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Epidemiology Acres call carcinomia acobart for about 1-2% of panematics inseptemic in adults and about 15% in children McSN. The age of adult patents is approximately 60 years image years. Make patents is approximately 60 years image years. Make patents is approximately 60 years image.

In most access cell contractings are spontation, new cases and in the contrast of Lynch synthesize. Cerrary complex, all adversersaries polycoods have based documented a63(522)(1921)(2931)

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Pancreatic Acinar Cell Carcinoma

Pancreatic acinar cell carcinoma

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Epidemiology

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Tumours of the pancreas

Edited by: Gill AJ, Klimstra DS, Lam AK, Washington MK

Benign epithelial tumours and precursors Acinar cystic transformation Serous neoplasms Intraepithelial neoplasia Intraductal papillary mucinous neoplasm Intraductal oncocytic papillary neoplasm Intraductal tubulopapillary neoplasm Mucinous cystic neoplasm Malignant epithelial tumours Ductal adenocarcinoma Acinar cell carcinoma Pancreatoblastoma Solid pseudopapillary neoplasm

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Pancreatoblastoma Solid pseudopapillary neoplasm

Pancreatic Ductal Adenocarcinoma

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Pancreatic Ductal Adenocarcinoma

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Invasive micropapillary carcinoma

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WHO 5th edition 2019 GIT Blue Book

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Tumours of the pancreas

Ediled by: Gill AJ, Kirmitra DS, Lam AK, Washington MK

Benign epithelial turnours and precursors Acros registic transformation Serous neoplasme Intraepithelial neoplasia Intraductal populary mucinous neoplasm Intraductal encogrite papillary neoplasm Intraductal hubulopapillary neoplasm Malignant epithelial turnours Ductal adenocarcinoma Acinar cel carcinoma Pancreatoblastoma Solid peeudopapillary neoplasm Neuroendocrine-reoplasms Non-functioning neuroendocrine turnours Functioning neuroendocrine turnours Insulinoma Gastrinoma VIPoma Glucagonoma Somatostatinoms ACTH-producing neuroendocrine turnour Serotonin-producing neuroendocrine turnour Neuroendocrine carcinoma MINENs

Two tiered grading (PanIN,IPMN, MCN)

E-format and uniformity

IOPN and ITPN separated from IPMN