

Congenital anomalies of the pancreas

Embryologic origins | Ductal variants | Anatomic consequences

An integrated developmental pathology lecture

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Learning objectives

Trace the dorsal and ventral primordia into adult pancreatic structures.

Differentiate ductal fusion, rotation, ectopia, and agenesis.

Relate each developmental defect to anatomy, symptoms, and clinical interpretation.

Schematic — key structures or categories

Development

Anatomic phenotype

Clinicopathology

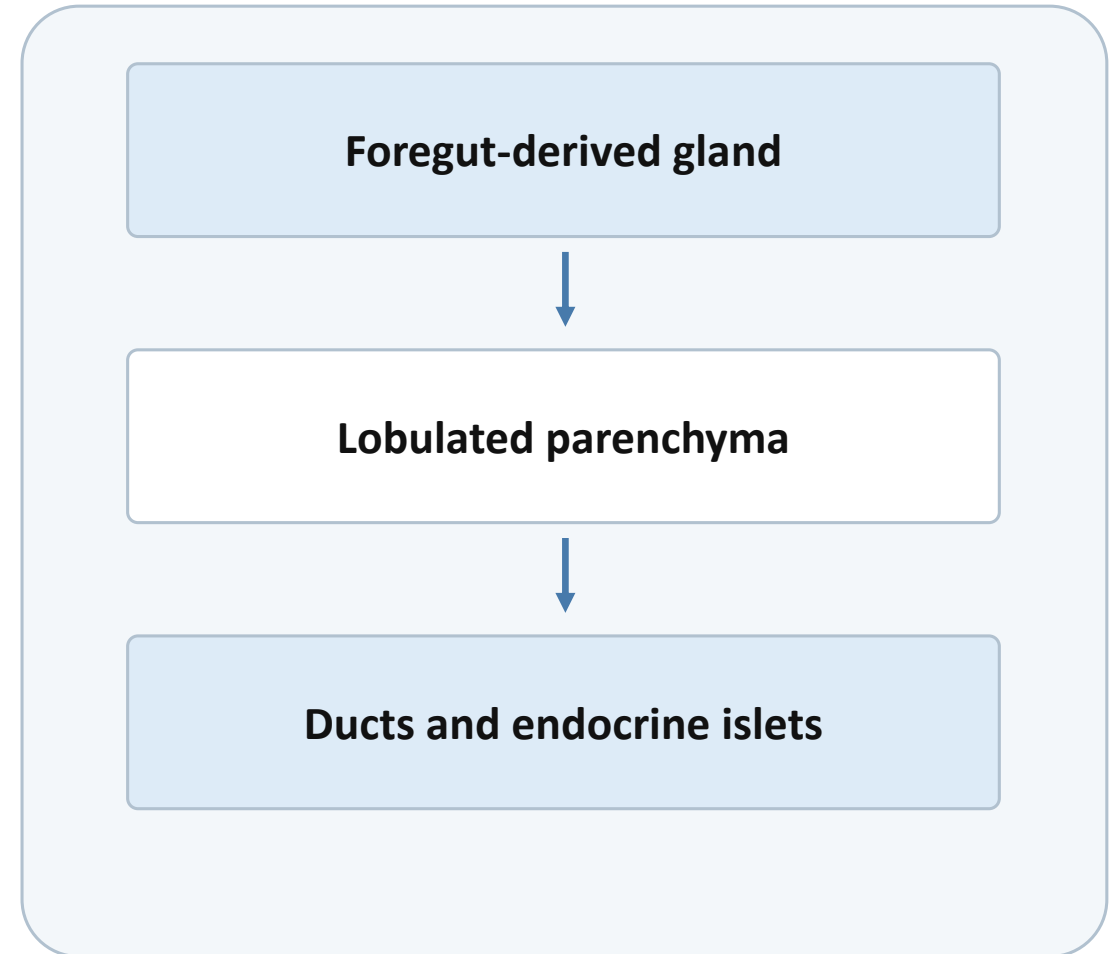
The adult pancreas: Functional framework

A retroperitoneal, transversely oriented organ from the duodenal C-loop to the splenic hilum.

Distinct exocrine and endocrine components occupy the same gland.

Congenital variants may alter position, continuity, or duct drainage.

Schematic — causal or drainage sequence



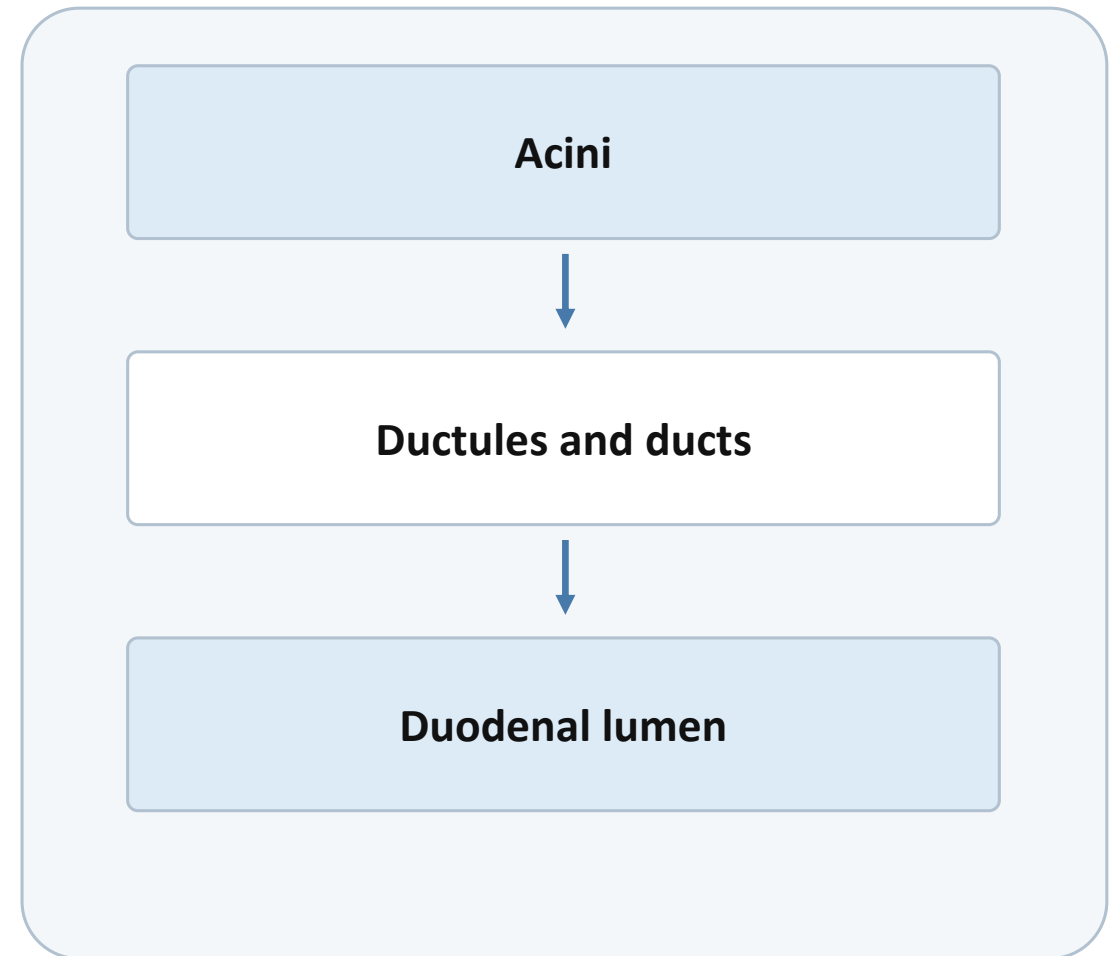
Exocrine architecture creates a drainage requirement

Acinar cells form approximately 80%–85% of the pancreas.

Zymogen-containing secretions traverse ductules and ducts into the duodenum.

Anatomic duct variants matter because they change the route of exocrine outflow.

Schematic — causal or drainage sequence



Endocrine tissue is anatomically dispersed

Islets of Langerhans are scattered throughout pancreatic parenchyma.

They contribute approximately 1%–2% of pancreatic mass.

Ectopic pancreatic rests may contain islets as well as exocrine structures.

Schematic — key structures or categories

Pancreatic acini

Ducts

Islets: Sometimes ectopic

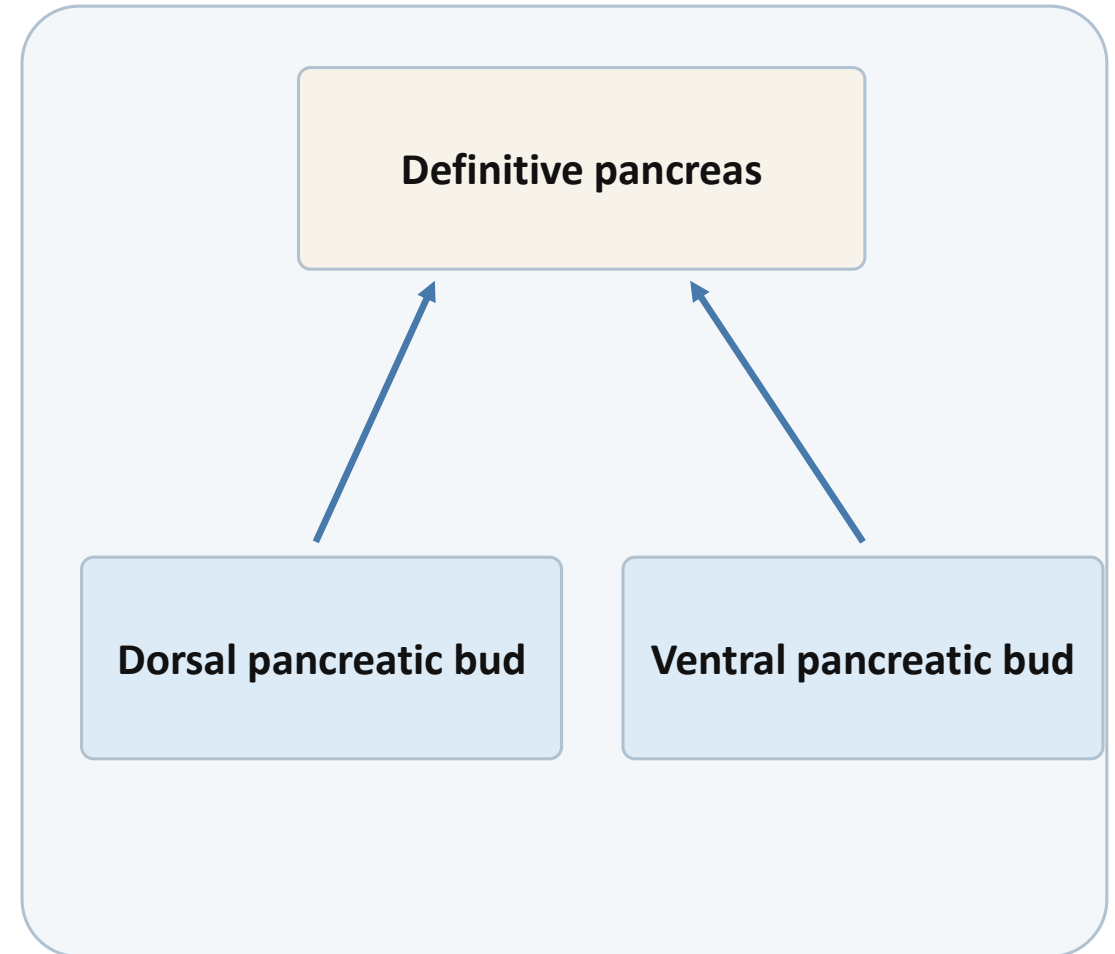
Two pancreatic primordia arise from the foregut

Schematic — developmental relationships

The developing pancreas begins as dorsal and ventral outpouchings.

Both primordia contribute to the definitive gland.

Correct union establishes the usual parenchymal and ductal arrangement.



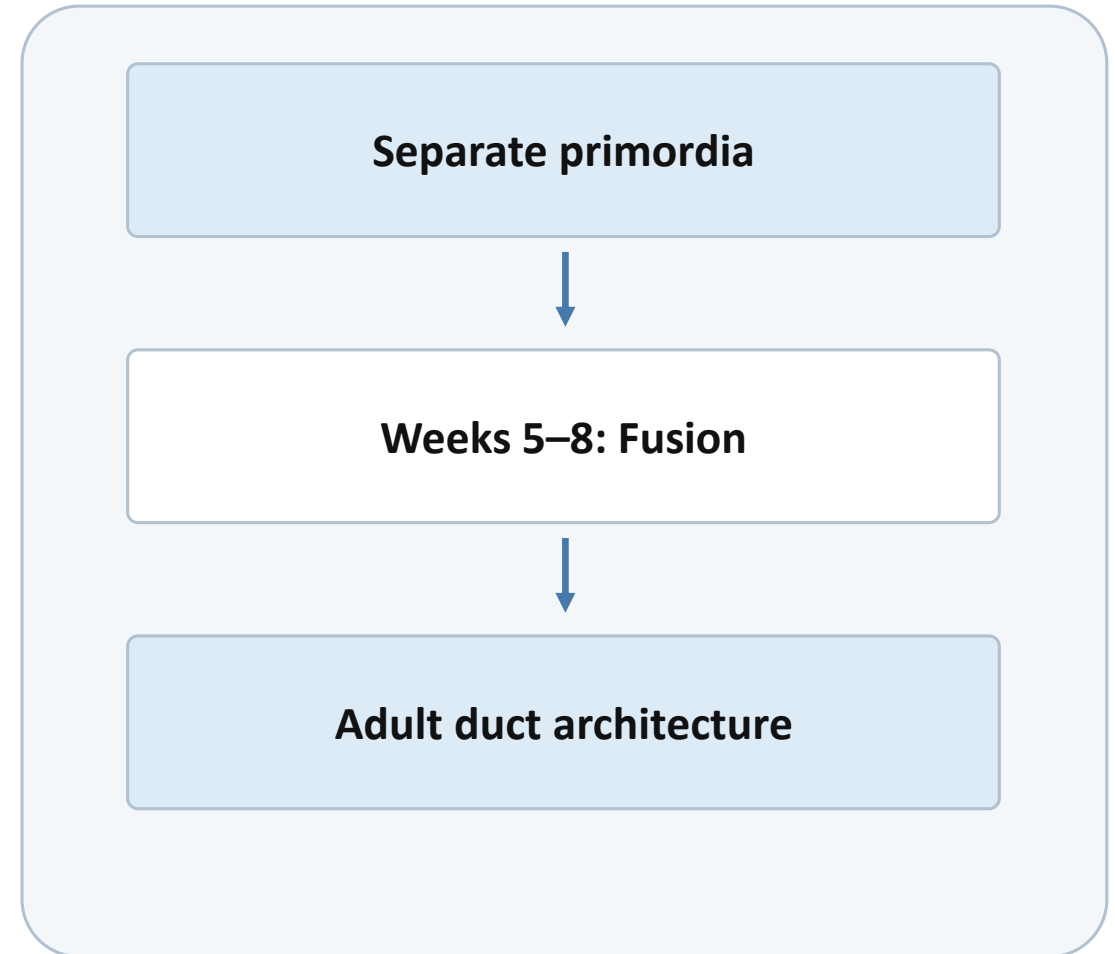
The period of fusion is developmentally vulnerable

Pancreatic fusion between the fifth and eighth gestational weeks.

This event coordinates previously separate primordia.

Imperfect fusion provides the central explanation for common ductal variants.

Schematic — causal or drainage sequence



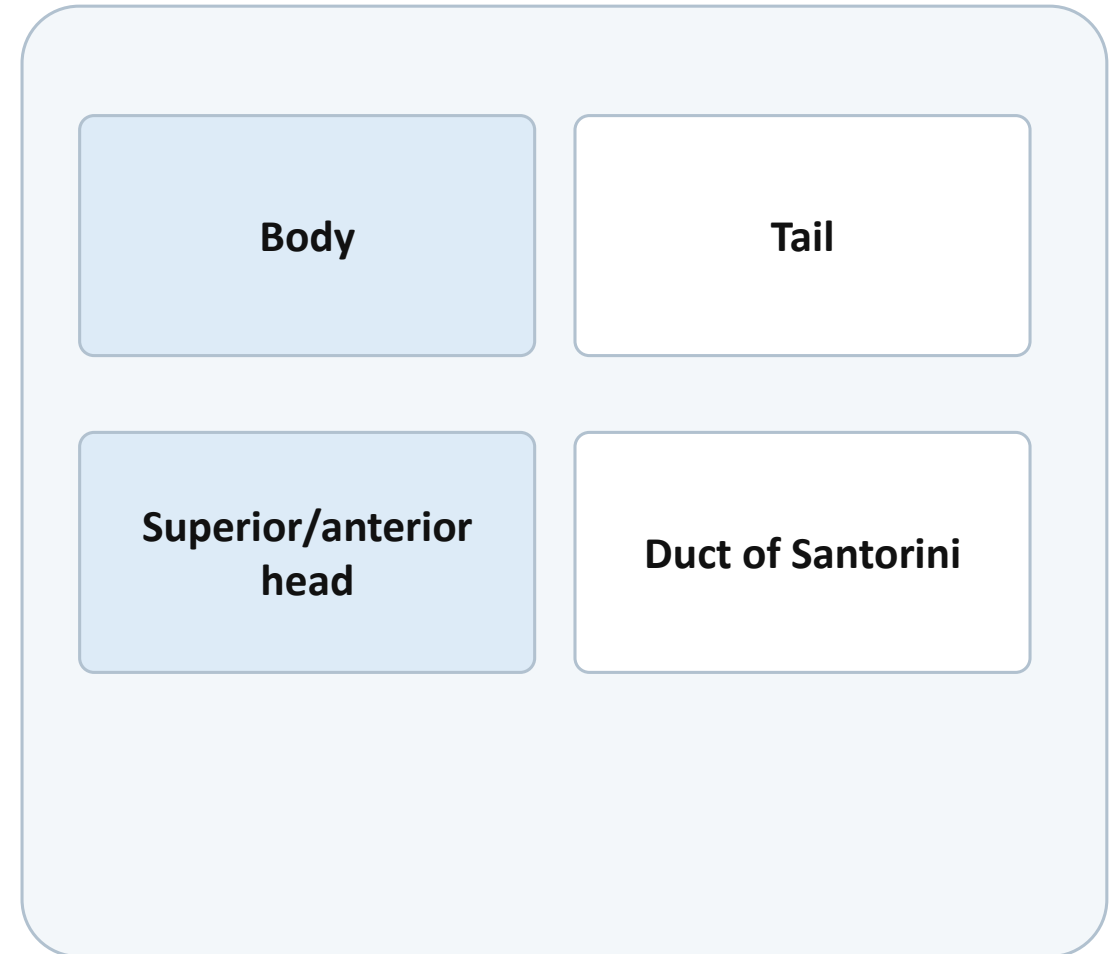
Dorsal bud: Body and tail

Schematic — key structures or categories

The dorsal primordium gives rise to the pancreatic body and tail.

It also forms the superior/anterior part of the pancreatic head.

The dorsal ductal contribution includes the duct of Santorini.



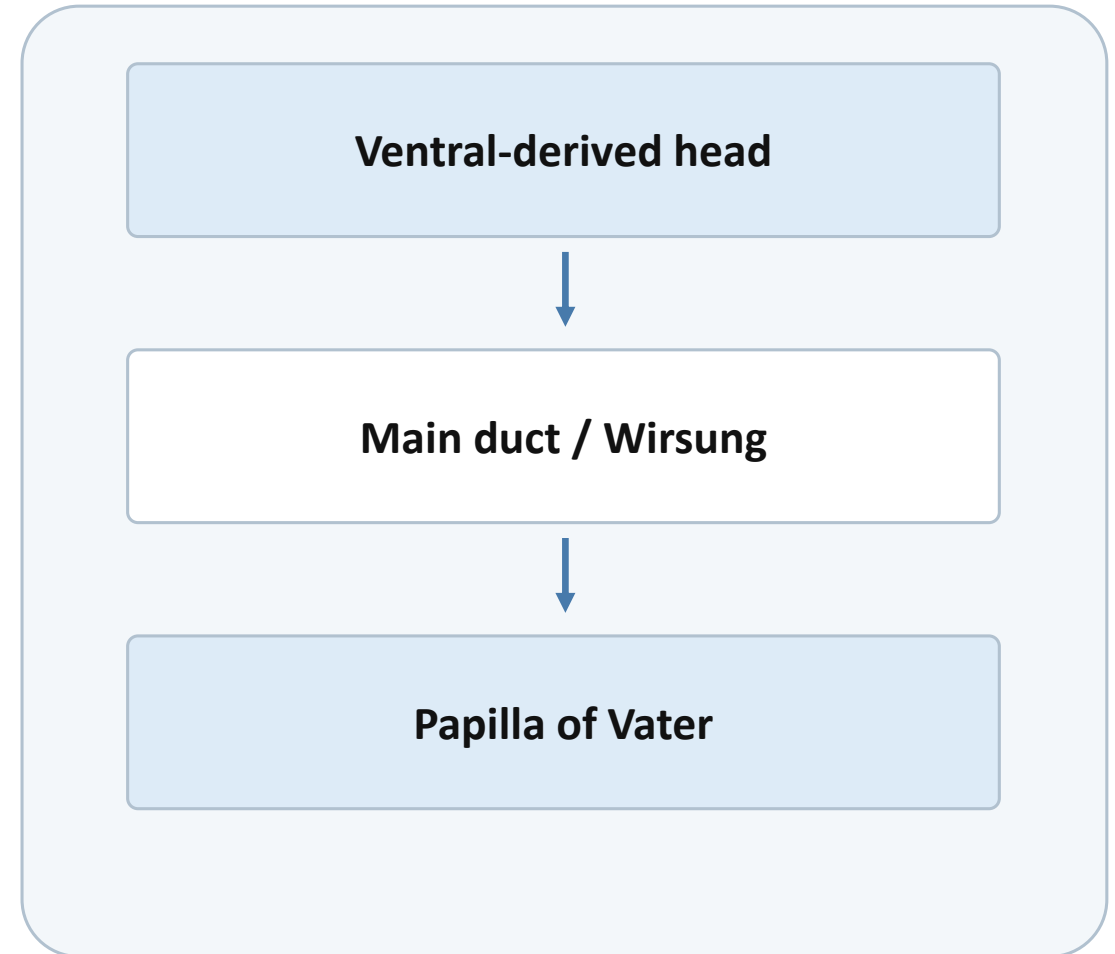
Ventral bud: Part of the head

The ventral primordium forms the posterior/inferior part of the pancreatic head.

Its usual outflow is through the main pancreatic duct.

The principal drainage pathway exits at the papilla of Vater.

Schematic — causal or drainage sequence



Adult pancreatic regions retain developmental origins

Schematic — embryologic contributions

Body and tail are primarily dorsal derivatives.

The pancreatic head receives contributions from both primordia.

Knowing the territories explains why an unfused duct system can drain unequal portions of the gland.

Dorsal: Body, tail, upper head

Ventral: Lower/posterior head

Normal ductal anatomy has two outlets

The duct of Wirsung is the main pancreatic duct.

The duct of Santorini is the accessory pancreatic duct.

Their different papillary destinations are essential to identifying divisum.

Schematic — simplified drainage only

Dominant route

Pancreatic bulk



Wirsung



Major papilla

Smaller / accessory route

Accessory route



Santorini



Minor papilla

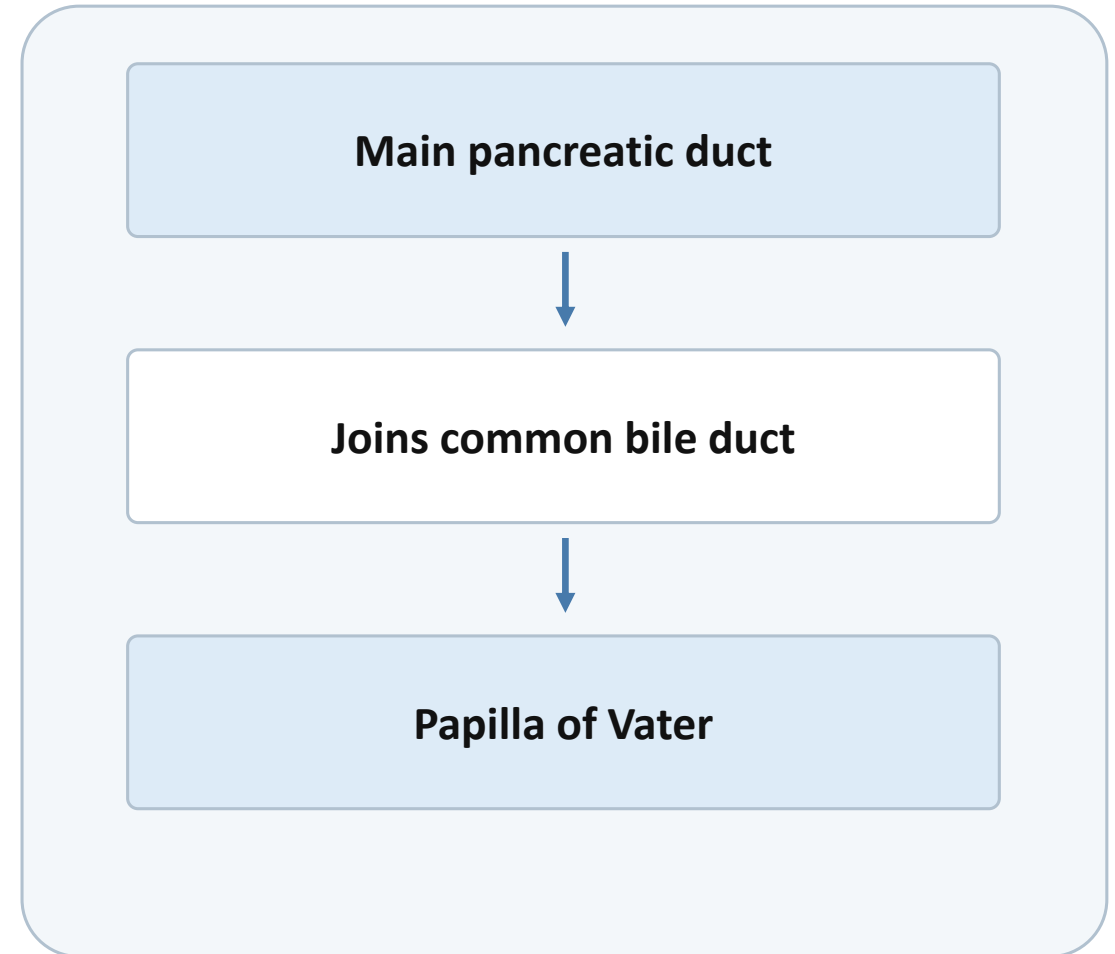
Major papilla: Main duct and common bile duct

The main pancreatic duct generally joins the common bile duct proximal to the papilla of Vater.

This convergent drainage defines the usual major-papilla route.

A separate accessory pathway may drain through the minor papilla.

Schematic — causal or drainage sequence



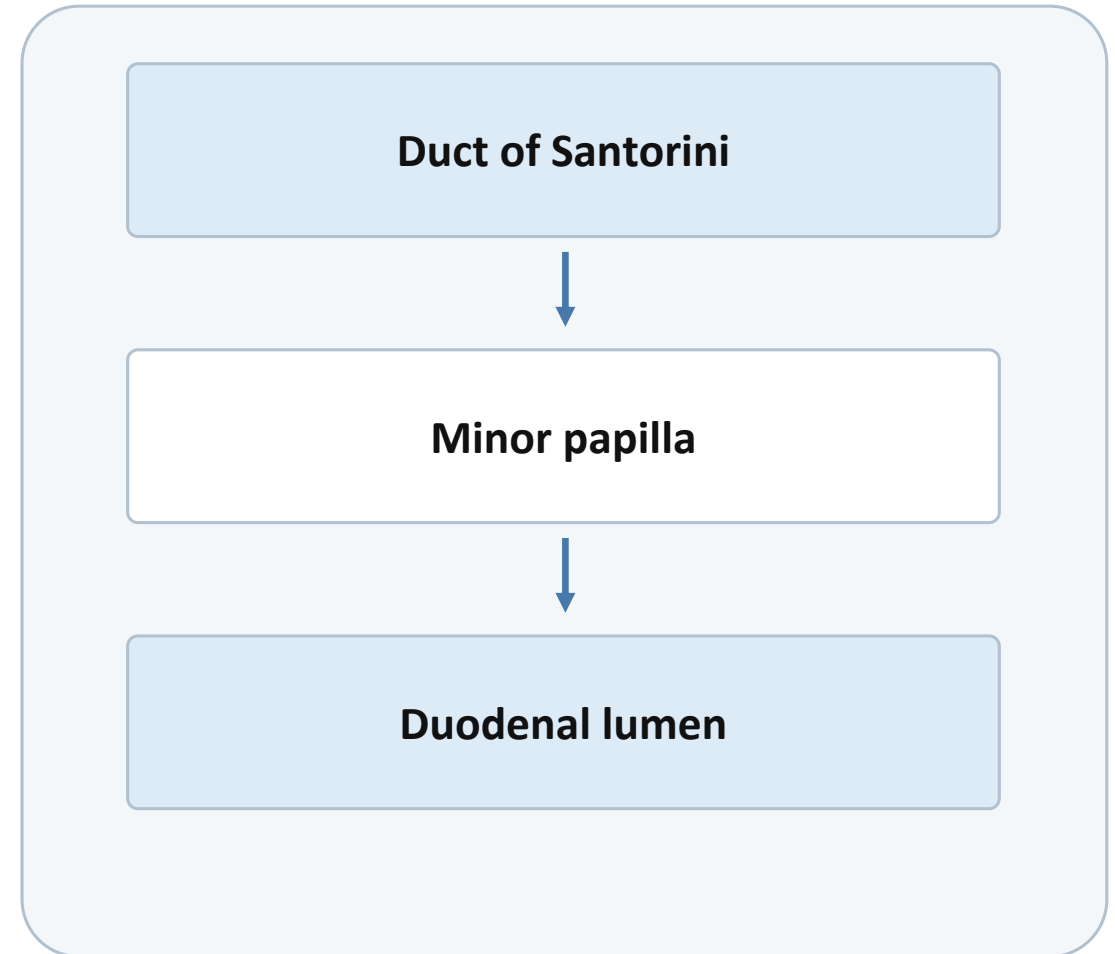
Minor papilla: The accessory outlet

Santorini normally provides an accessory drainage route.

It enters the duodenum through the minor papilla.

In pancreas divisum, the minor papilla becomes the major drainage outlet for dorsal pancreatic tissue.

Schematic — causal or drainage sequence



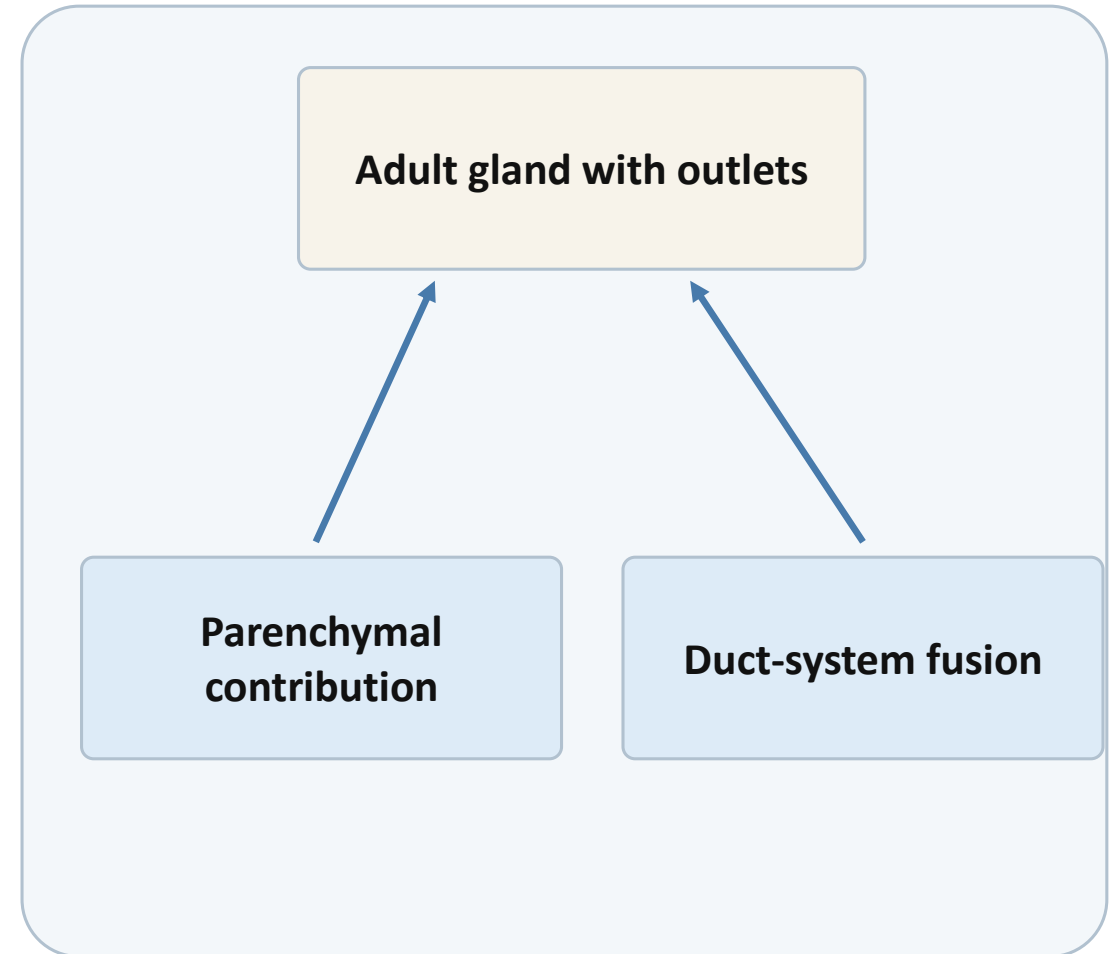
Fusion shapes both tissue and duct topography

The two primordial components contribute distinct adult territories.

Ductal fusion normally redirects most pancreatic secretions to the major papilla.

A fusion failure changes drainage without necessarily preventing formation of parenchyma.

Schematic — developmental relationships



Congenital anomalies: Four mechanisms

Failed duct fusion → pancreas divisum.

Incomplete ventral-bud rotation → annular pancreas.

Aberrant tissue location → ectopic pancreas.

Failure of gland development → pancreatic agenesis.

Schematic — key structures or categories

Fusion

Rotation

Ectopia

Development absent

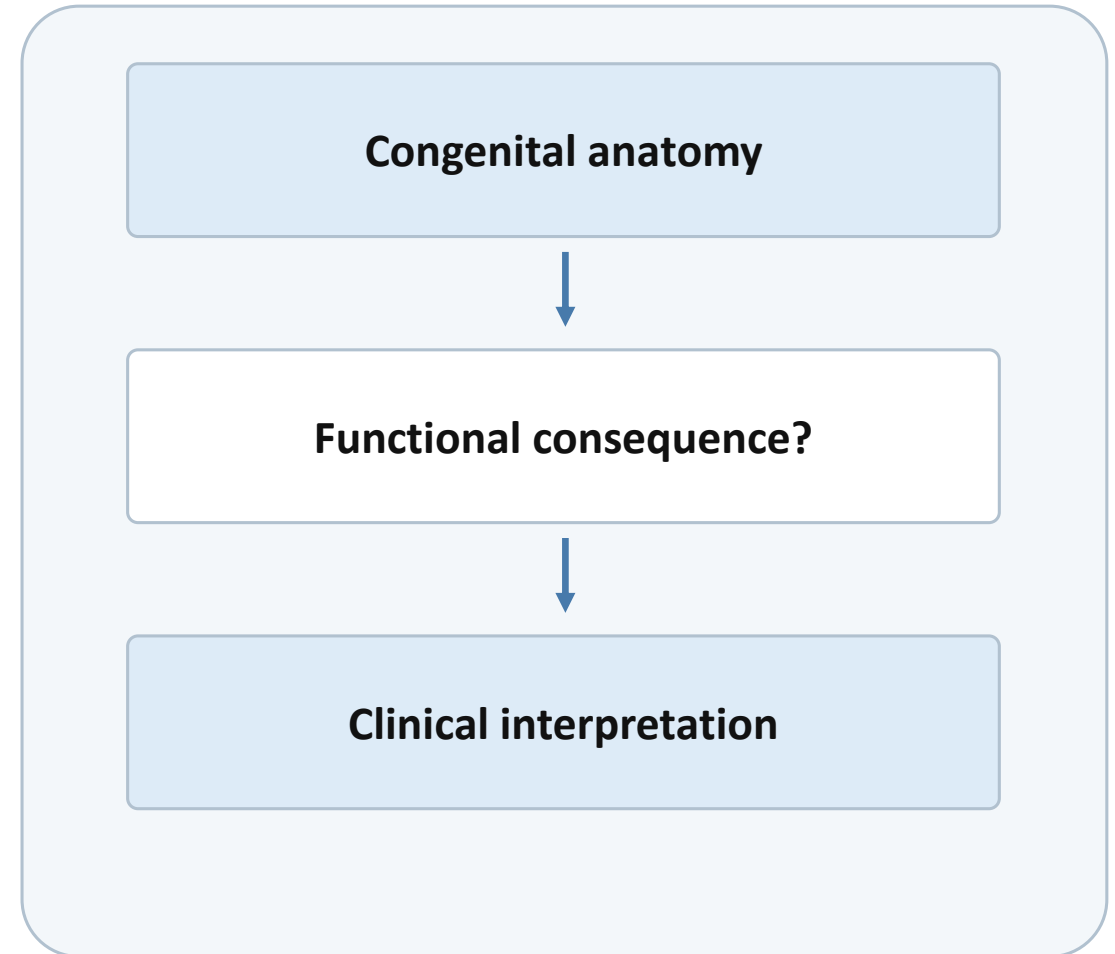
Anatomic variant is not synonymous with disease

Many congenital pancreatic variants are incidental findings.

Clinical consequences depend on outflow obstruction, local tissue effects, or absent development.

A visible anomaly does not by itself establish the cause of symptoms.

Schematic — causal or drainage sequence



Pancreas divisum: Failure of ductal union

Pancreas divisum is caused by failure of fetal dorsal and ventral duct-system fusion.

The resulting pancreatic tissue remains divided in its ductal drainage.

It is the most common congenital anomaly described in the chapter.

Schematic — simplified drainage only

Dominant route

Dorsal bulk



Santorini



Minor papilla

Smaller / accessory route

Ventral portion



Wirsung



Major papilla

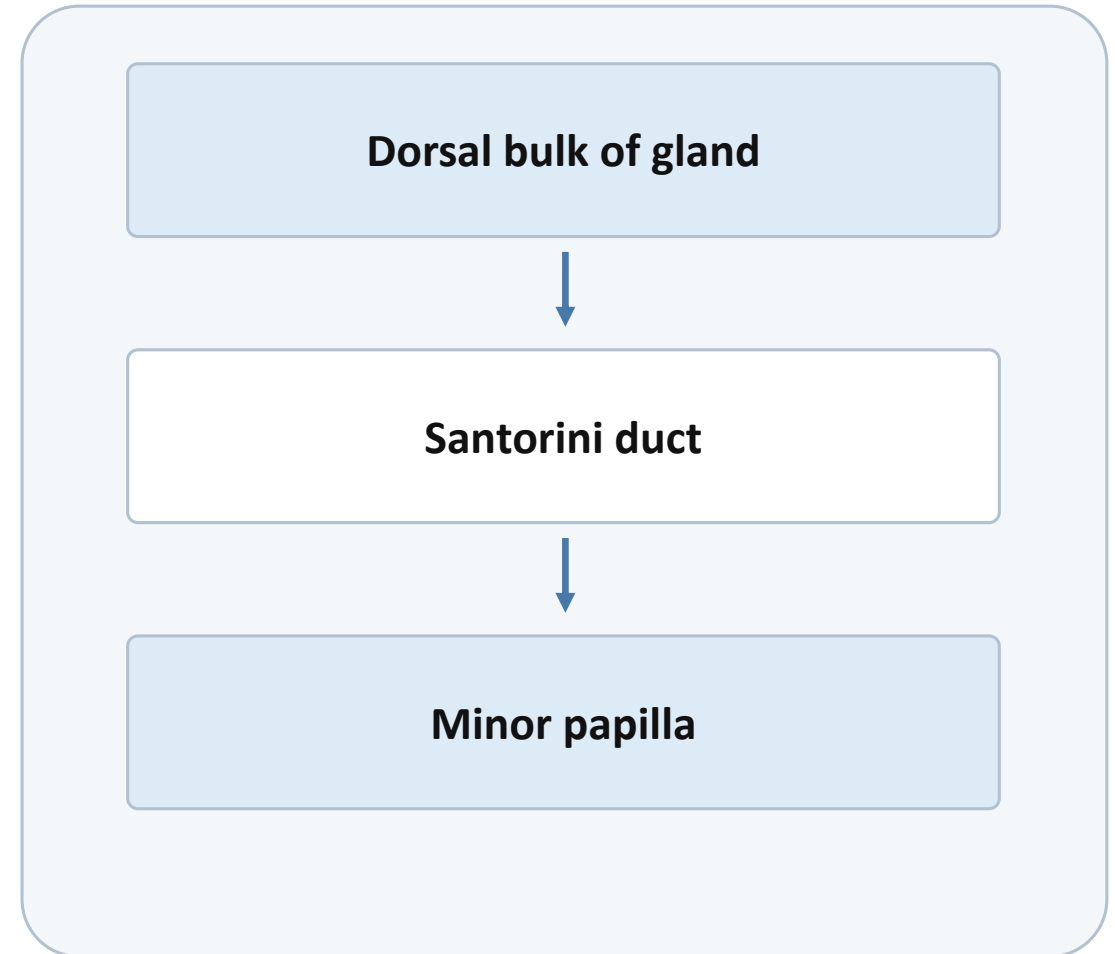
Divisum is a drainage anomaly

The bulk of the pancreas is derived from the dorsal primordium.

In divisum, this dominant volume drains via the duct of Santorini.

The minor papilla carries the larger share of secretion.

Schematic — causal or drainage sequence



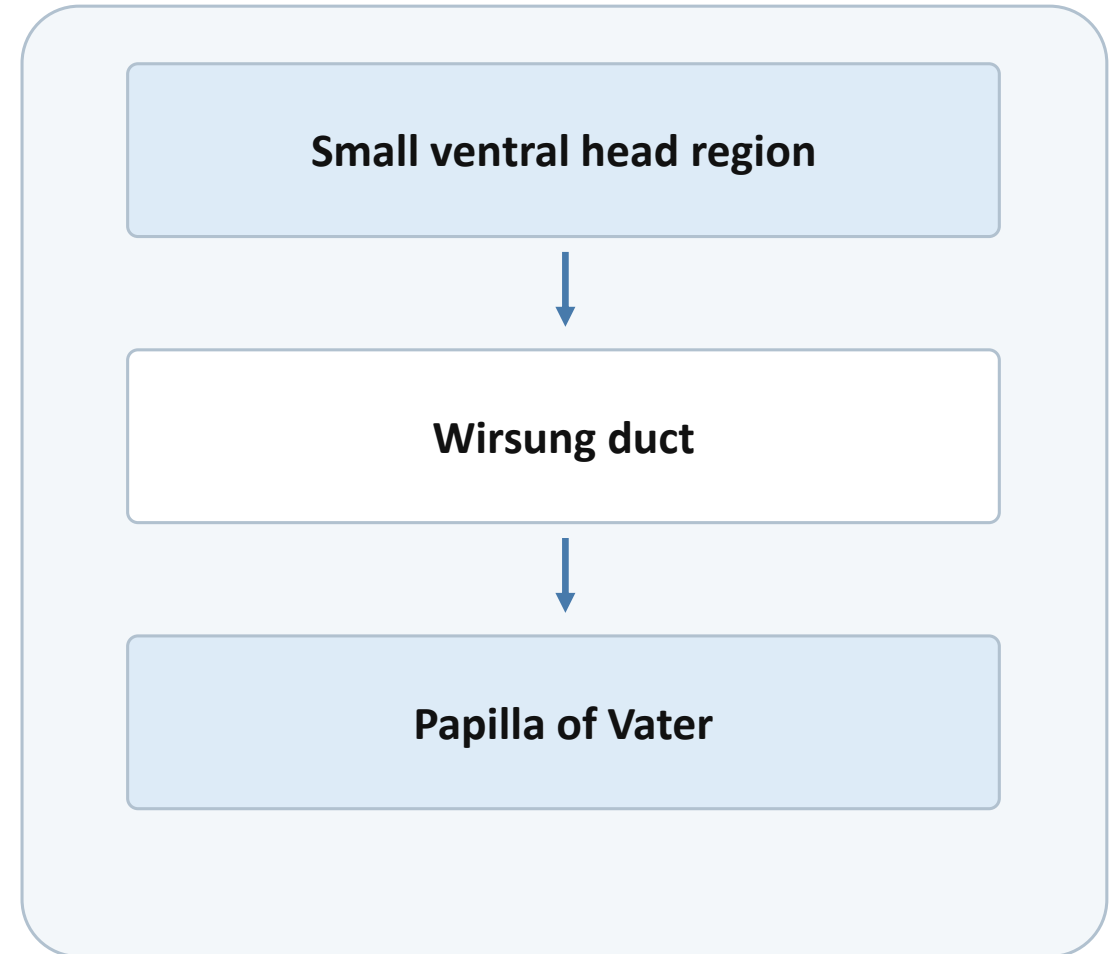
Ventral drainage remains comparatively small

The duct of Wirsung drains only a small portion of the pancreatic head in divisum.

It exits via the papilla of Vater.

Thus major-papilla drainage is not the dominant exocrine route in this variant.

Schematic — causal or drainage sequence



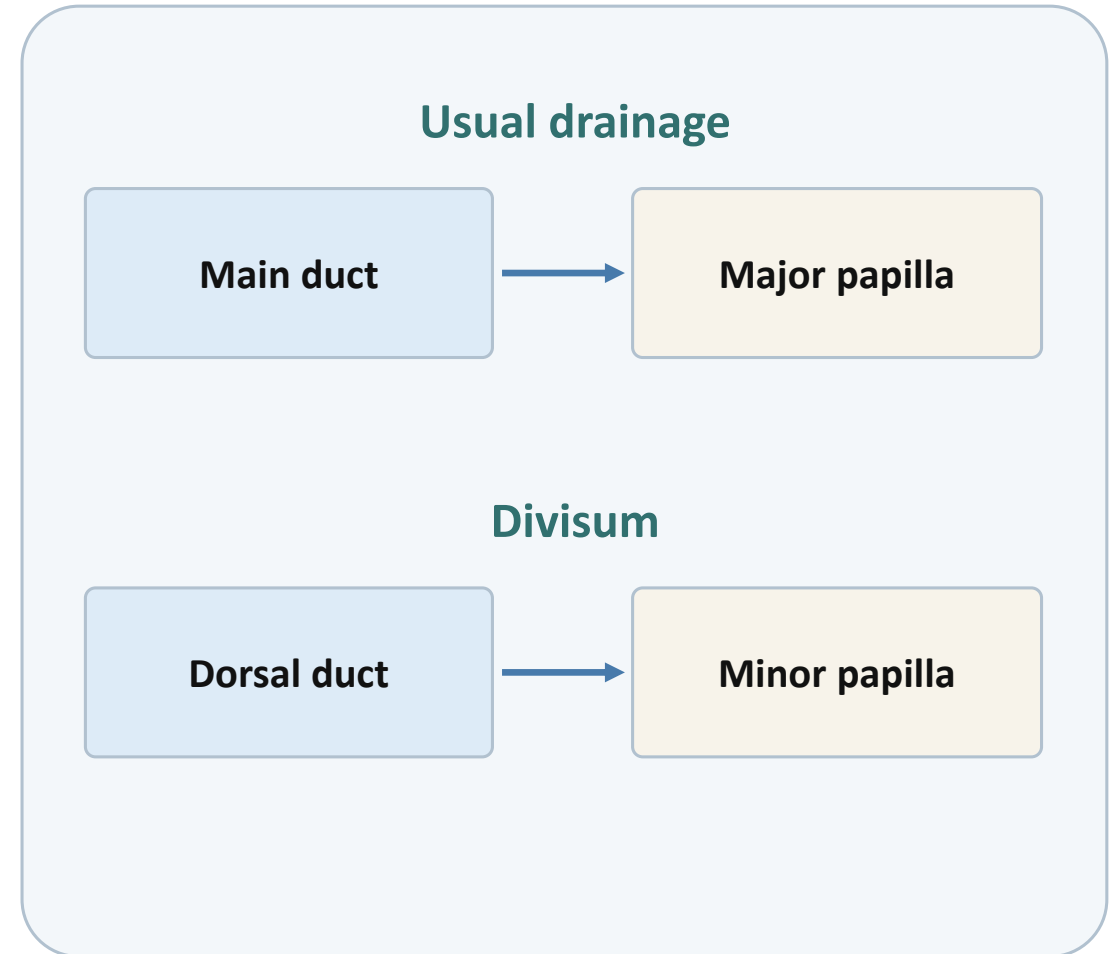
Normal versus divisum: The crucial distinction

Normal: Most pancreatic secretion follows the main-duct route to the major papilla.

Divisum: Secretion from the dorsal bulk exits through the minor papilla.

The principal difference is which outlet carries the largest drainage load.

Schematic — simplified drainage only



Divisum is a relatively frequent variant

Reported by Robbins

An incidence of 3%–10%.

The reported range describes occurrence of the anatomical variant, not pancreatitis.

Frequency of anatomy must be separated from frequency of clinical complications.

3%–10%

Pancreas divisum frequency

Denominator and clinical context matter

Most individuals with divisum are asymptomatic

Reported by Robbins

Most individuals are asymptomatic.

Only about 5% develop pancreatitis in the cited discussion.

An incidental diagnosis cannot automatically explain abdominal symptoms.

≈5%

Develop pancreatitis

Denominator and clinical context matter

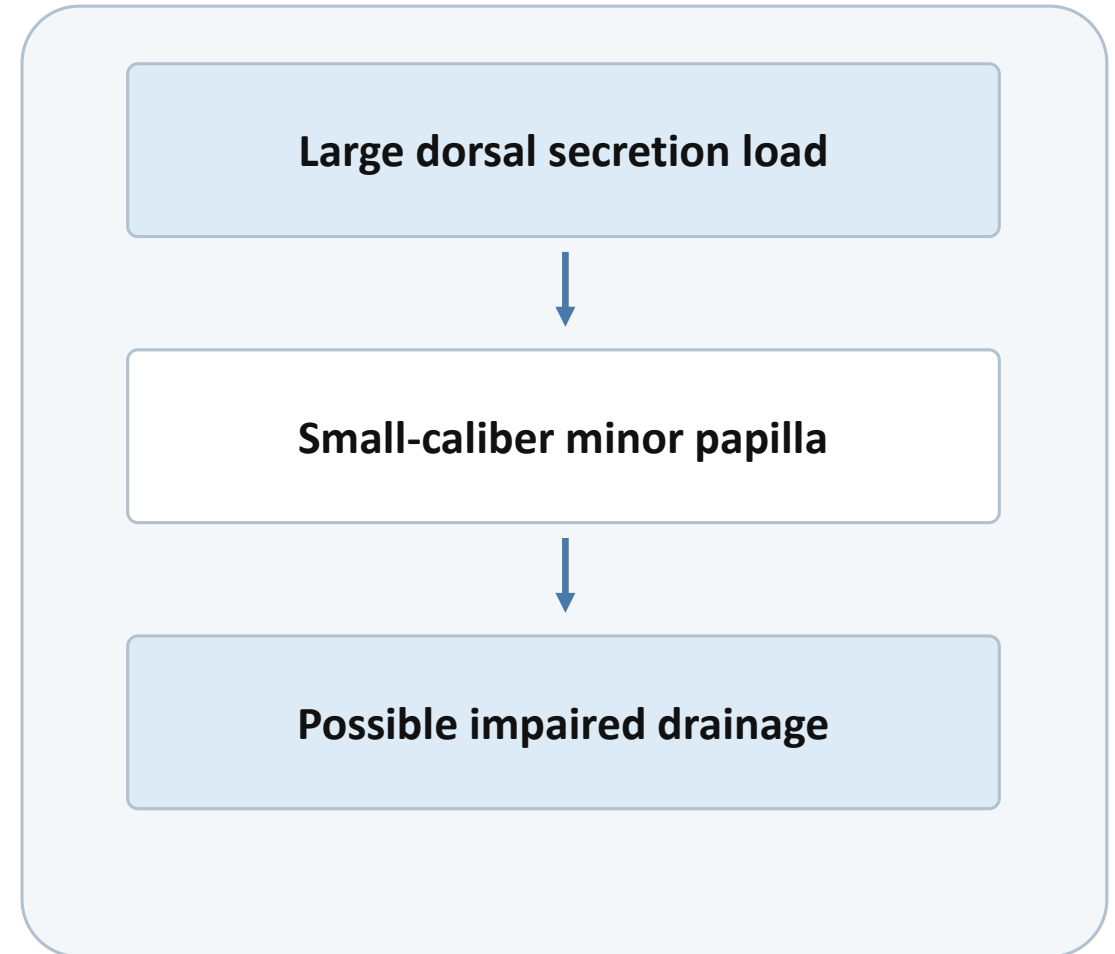
The minor-papilla bottleneck hypothesis

The minor papilla is small-caliber yet drains most of the pancreas in divisum.

Insufficient drainage has been proposed as a cause of increased ductal stress.

This proposed mechanism is not a universal consequence of the anomaly.

Schematic — causal or drainage sequence



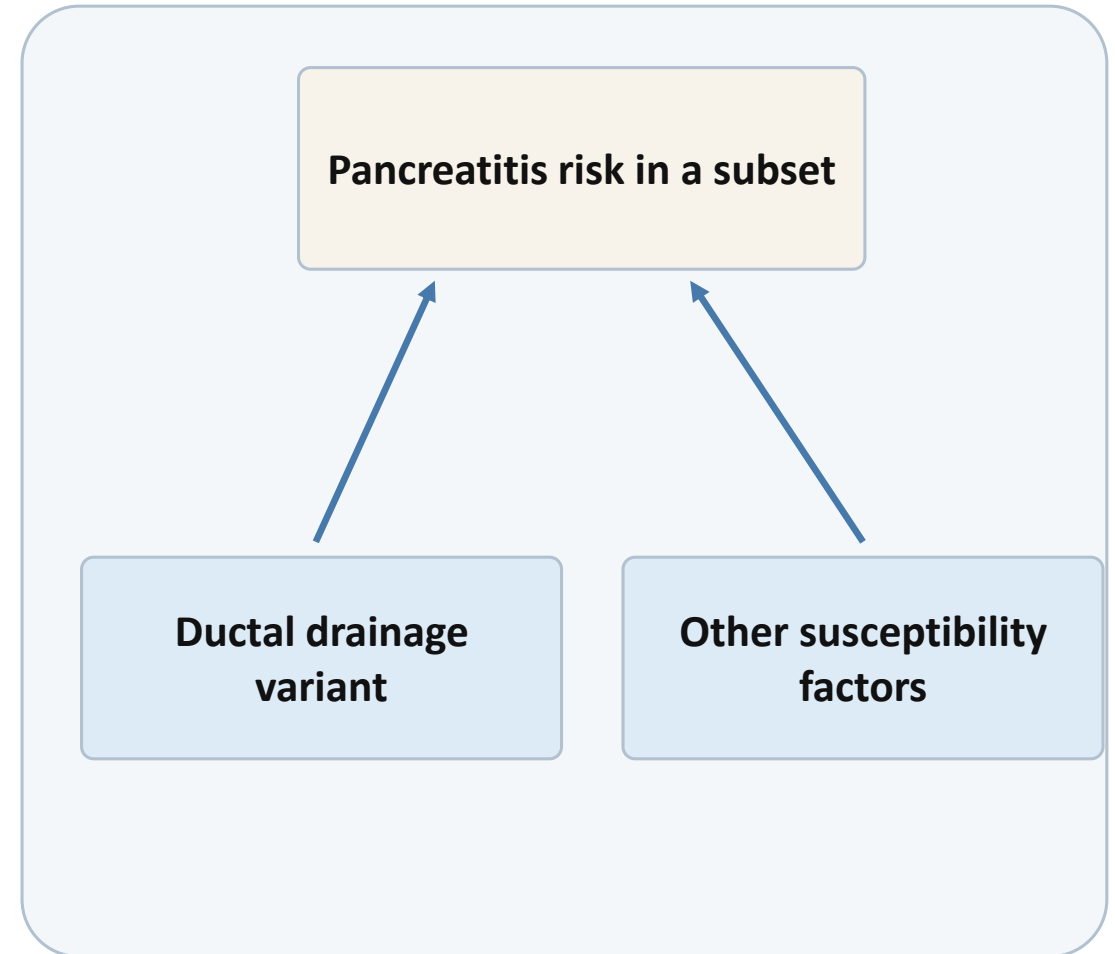
Genetic susceptibility may modify divisum risk

Genetic defects increasing pancreatitis susceptibility may coexist with divisum.

A combined anatomic and genetic predisposition is suggested, not established for every carrier.

The anomaly should therefore be interpreted within the larger susceptibility context.

Schematic — developmental relationships



Pancreatitis association: Avoid causal overreach

An association between divisum and pancreatitis does not prove that divisum caused an attack.

Most carriers do not develop pancreatitis.

The textbook frames drainage limitation as a proposed predisposing mechanism.

Schematic — key structures or categories

Common variant

Usually asymptomatic

Proposed causal modifier

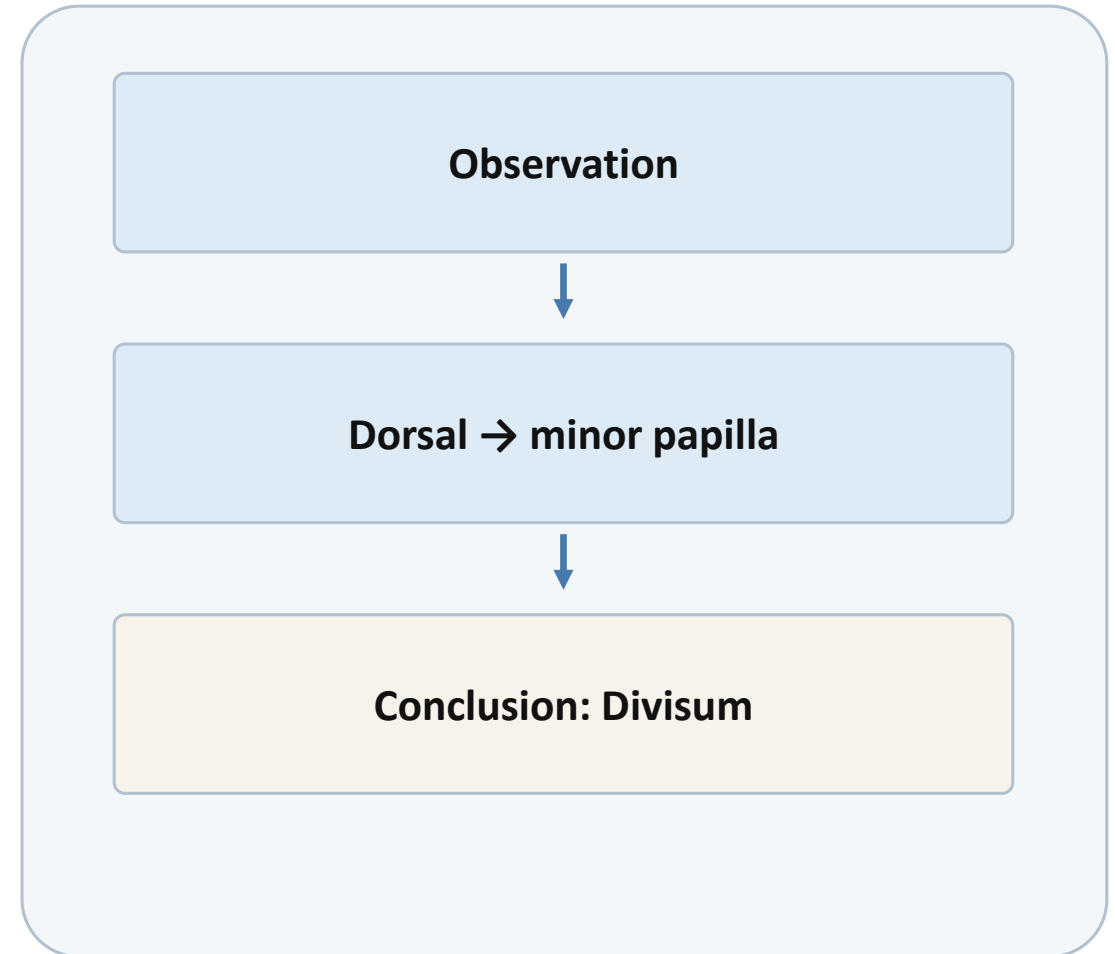
Divisum: Diagnostic interpretation exercise

Teaching exercise — hypothetical example

An asymptomatic individual is found to have separate dorsal and ventral drainage.

The dorsal territory drains predominantly through the minor papilla.

Interpretation: A ductal variant is present;
Symptoms or complications are not implied.



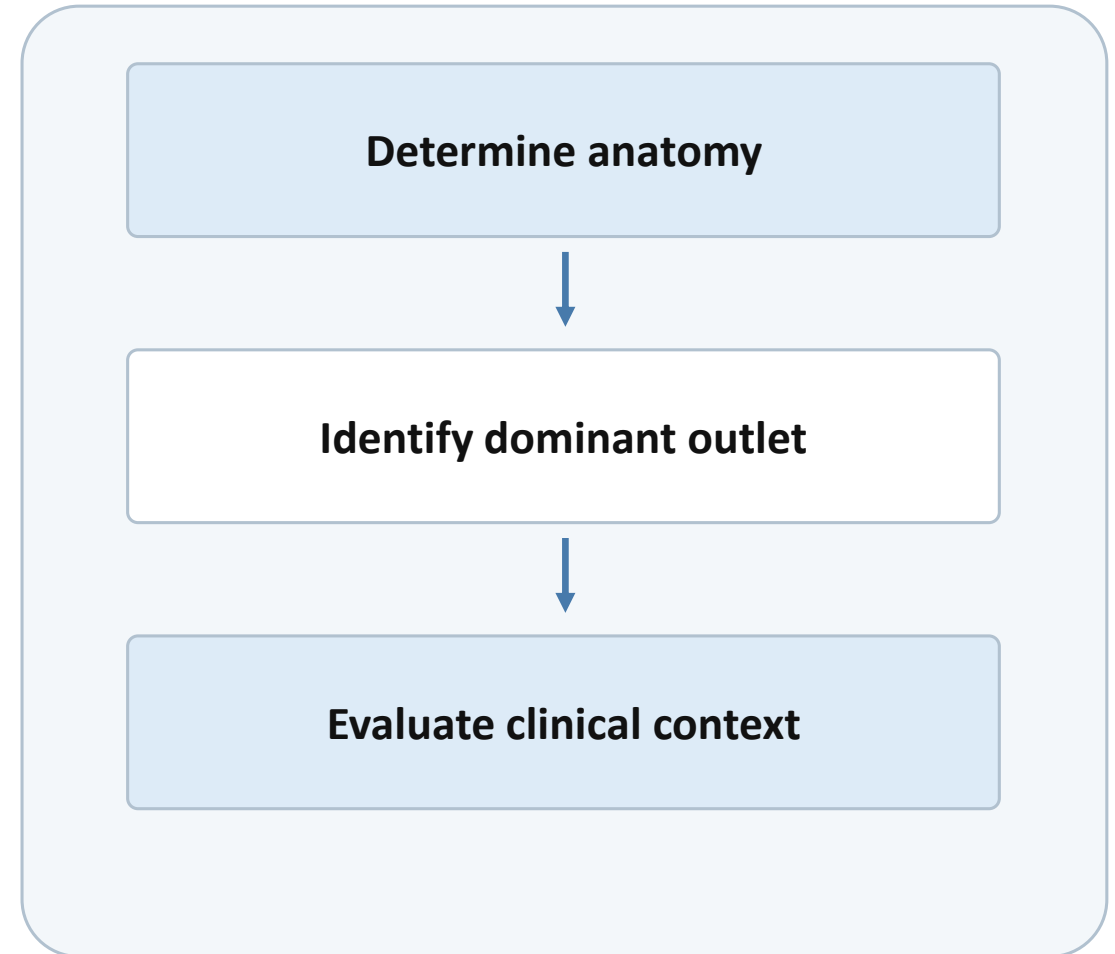
Divisum: Why anatomy matters clinically

Identify which duct collects secretions from the larger pancreatic territory.

Determine whether drainage proceeds to the minor or major papilla.

Assess any pancreatitis association cautiously, rather than treating the variant as causal proof.

Schematic — causal or drainage sequence



Ductal comparison at a glance

Feature	Usual anatomy	Pancreas divisum
Dorsal duct	Communicates with main drainage	Drains bulk through minor papilla
Wirsung	Main outflow to major papilla	Only small ventral head portion
Minor papilla	Accessory outlet	Dominant dorsal outlet
Clinical course	Usual anatomy	Most with divisum asymptomatic

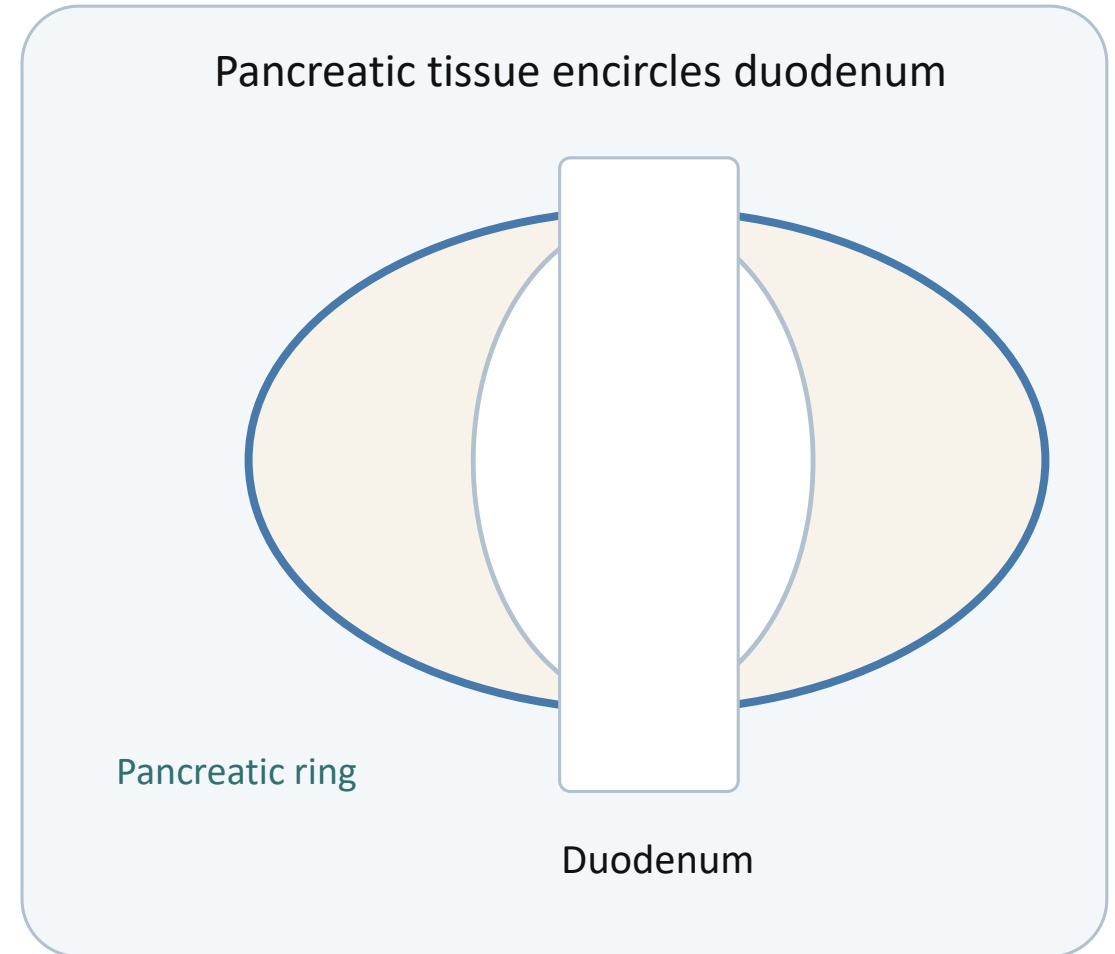
Annular pancreas: A rotation anomaly

Schematic — not a patient image

Annular pancreas results from incomplete rotation of the ventral pancreatic bud.

Normal pancreatic tissue forms a band-like ring.

The ring completely encircles the second portion of the duodenum.



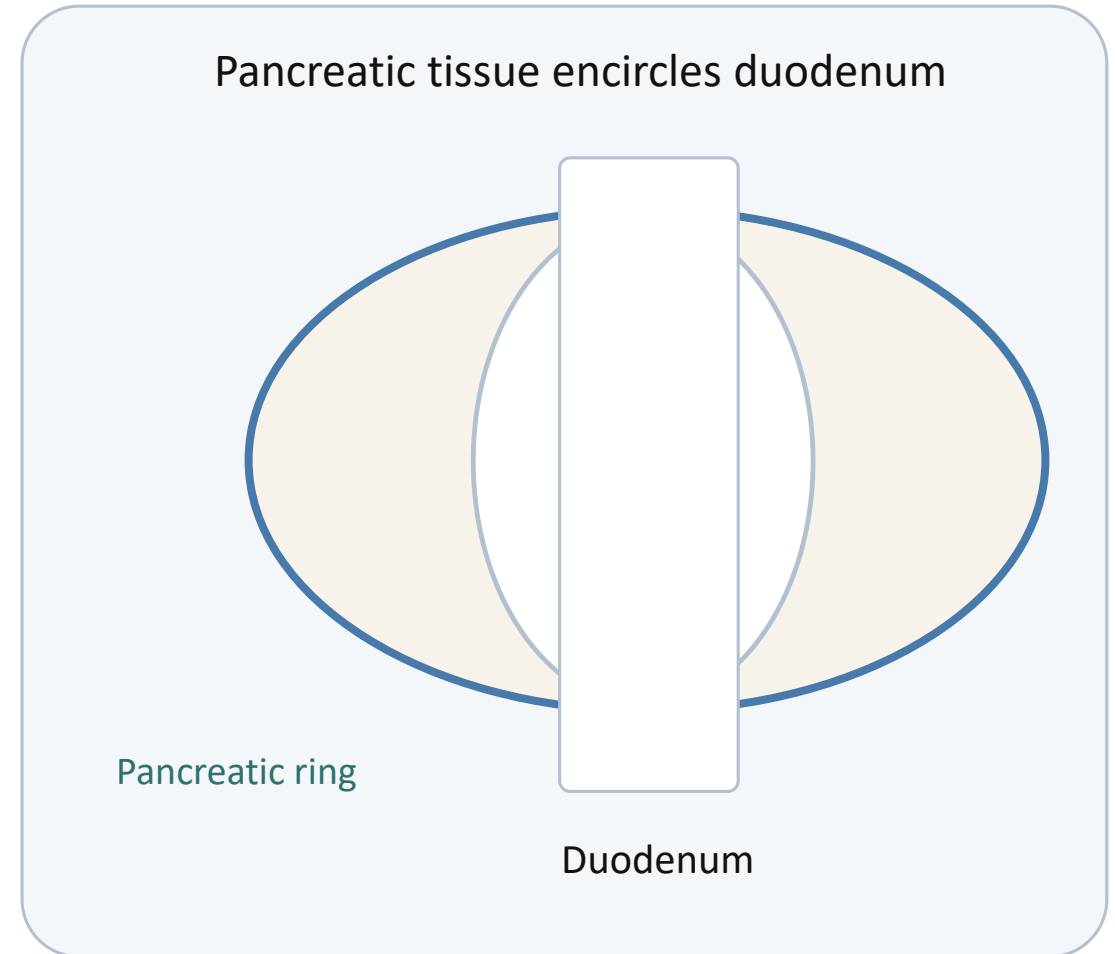
A ring of pancreatic tissue surrounds the duodenum

Schematic — not a patient image

The encircling tissue is histologically pancreatic, not a novel tissue type.

The defining topographic relationship is pancreatic tissue around the duodenal lumen.

The second duodenal portion is the specified anatomic site.



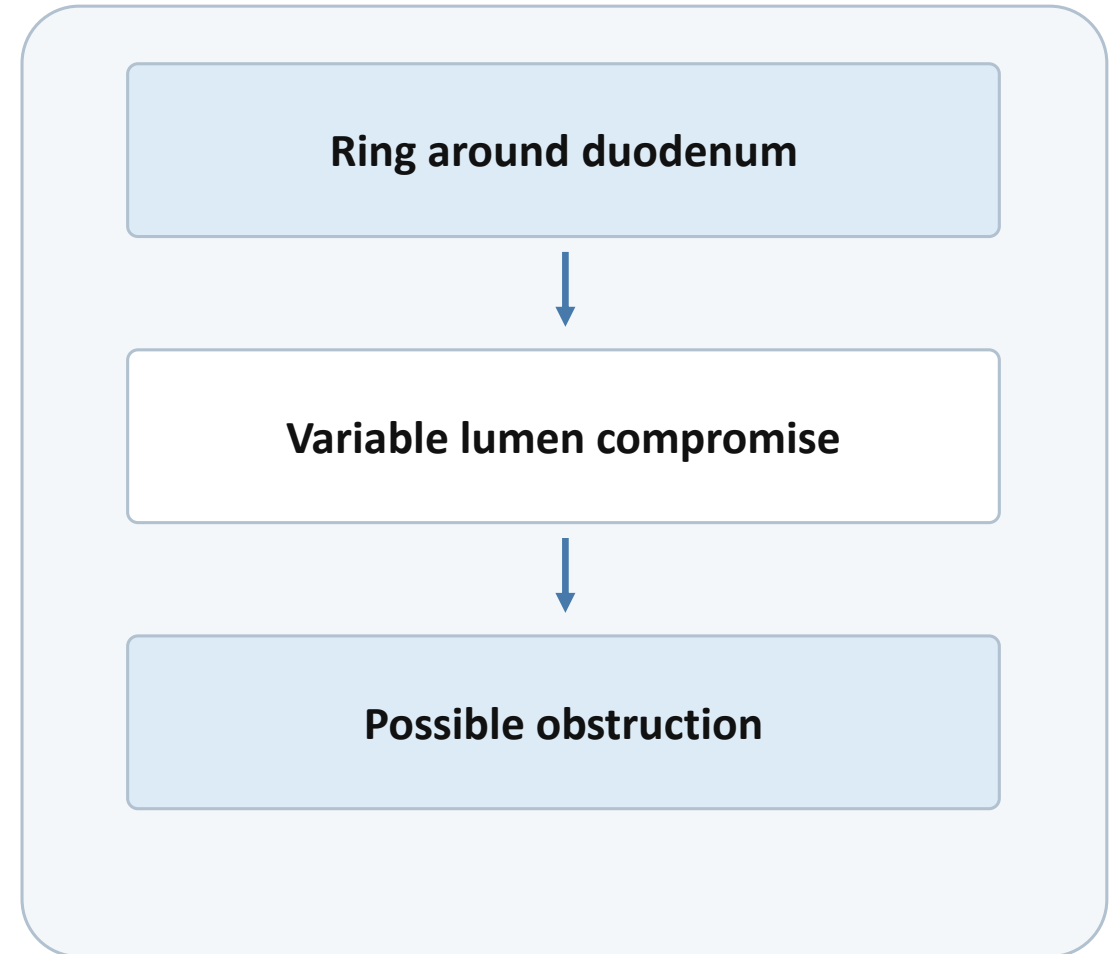
Annular pancreas: Anomaly and effect differ

A circumferential pancreatic ring may exist without symptoms.

The same structural configuration can narrow or obstruct duodenal transit in some patients.

The presence of the ring and the presence of obstruction are separate observations.

Schematic — causal or drainage sequence



Annular pancreas may be found incidentally

Reported by Robbins

Annular pancreas can be an incidental imaging finding.

Approximately one-third of cases are asymptomatic.

Incidental detection should not be equated with clinically important obstruction.

~1/3

Asymptomatic annular pancreas

Denominator and clinical context matter

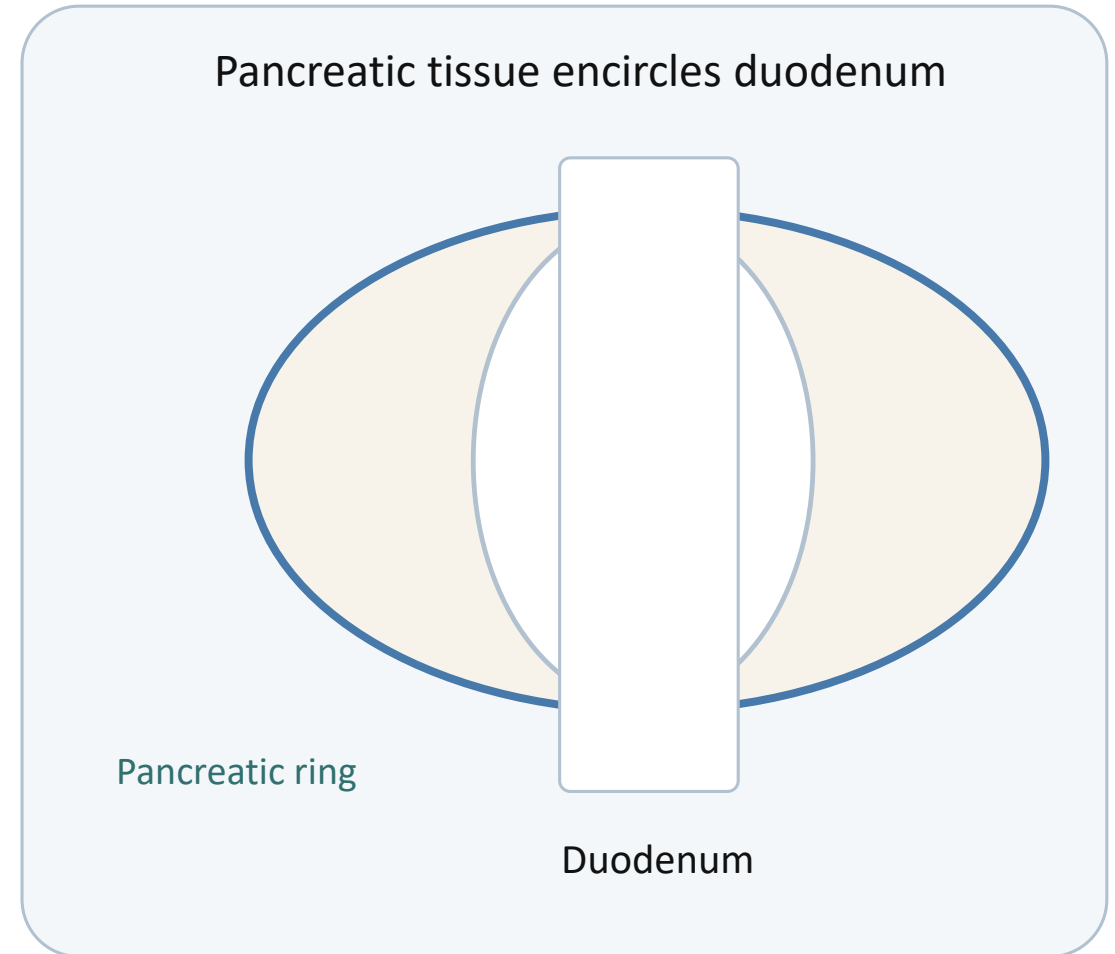
Duodenal obstruction is the principal complication

Circumferential pancreatic tissue surrounds the second duodenal portion.

In a subset of cases it creates duodenal obstruction.

The complication is a mechanical consequence of anatomy rather than abnormal tissue histology.

Schematic — not a patient image



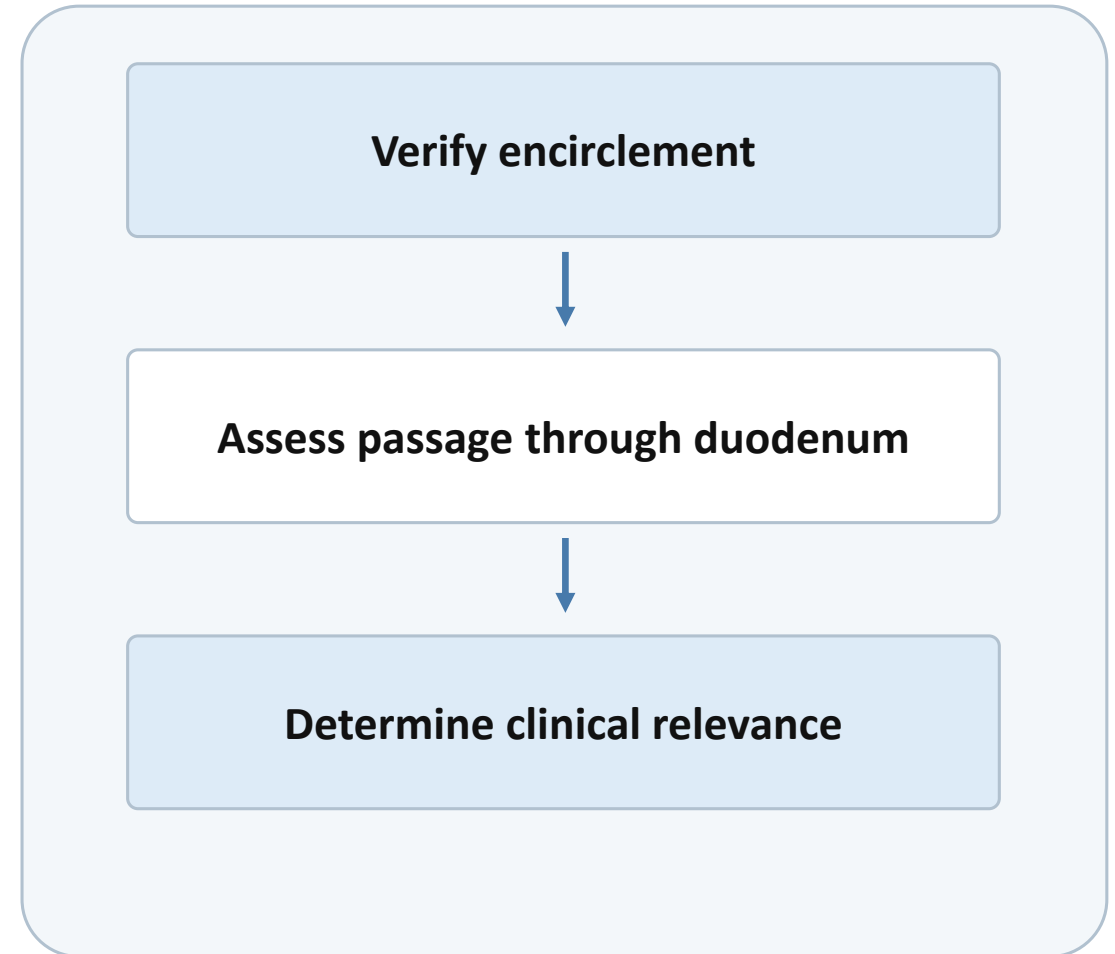
Annular pancreas: Structural reasoning

First identify a complete ring around the duodenum.

Next ask whether the duodenal lumen is compromised.

Do not infer severity solely from discovery of the ring.

Schematic — causal or drainage sequence



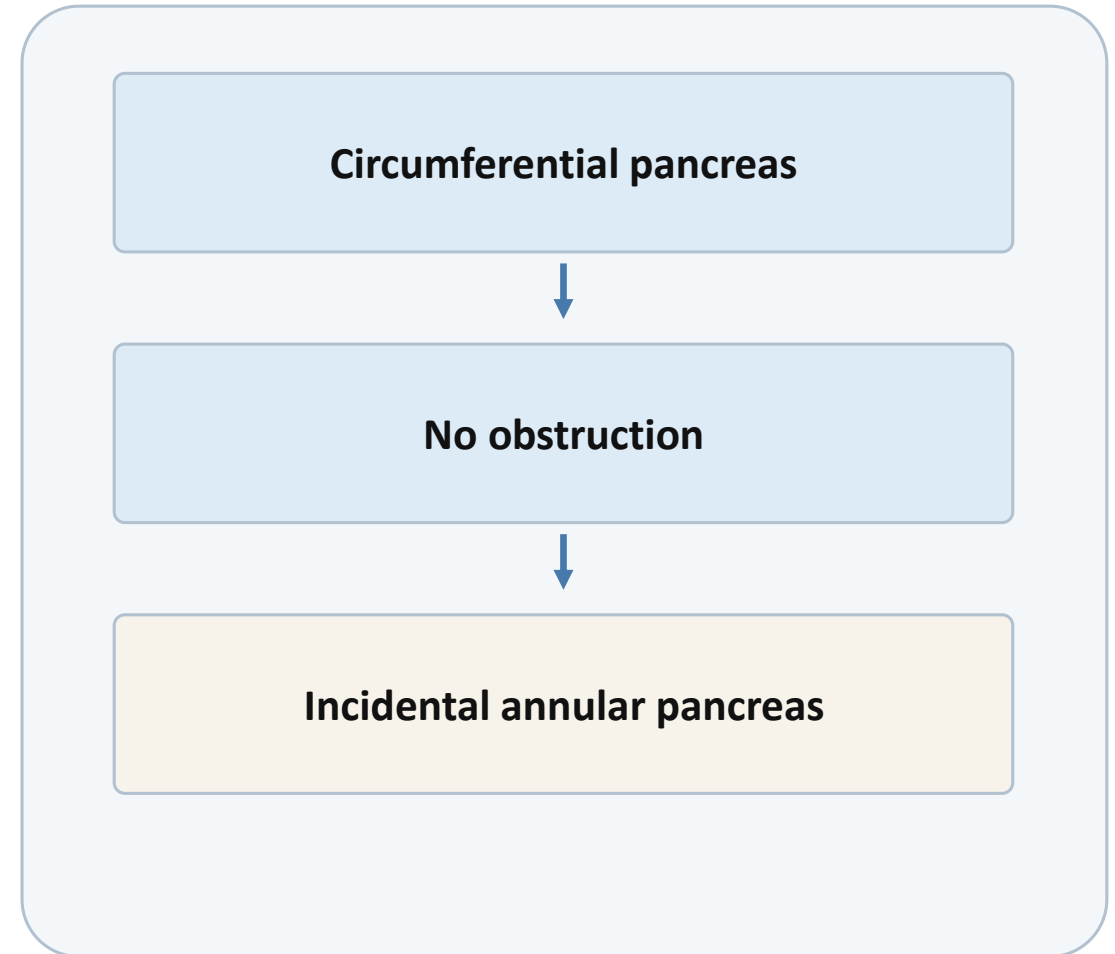
Annular pancreas: Teaching vignette

Teaching exercise — hypothetical example

Imaging demonstrates pancreatic tissue completely surrounding the second duodenal portion.

There is no evident mechanical obstruction.

Interpretation: Annular pancreas may be incidental despite striking anatomy.



Annular and divisum reflect different defects

Dimension	Pancreas divisum	Annular pancreas
Developmental defect	Failed duct fusion	Incomplete ventral rotation
Principal feature	Separate drainage routes	Tissue ring around duodenum
Potential consequence	Pancreatitis in minority	Duodenal obstruction in subset
Often incidental?	Yes	Yes

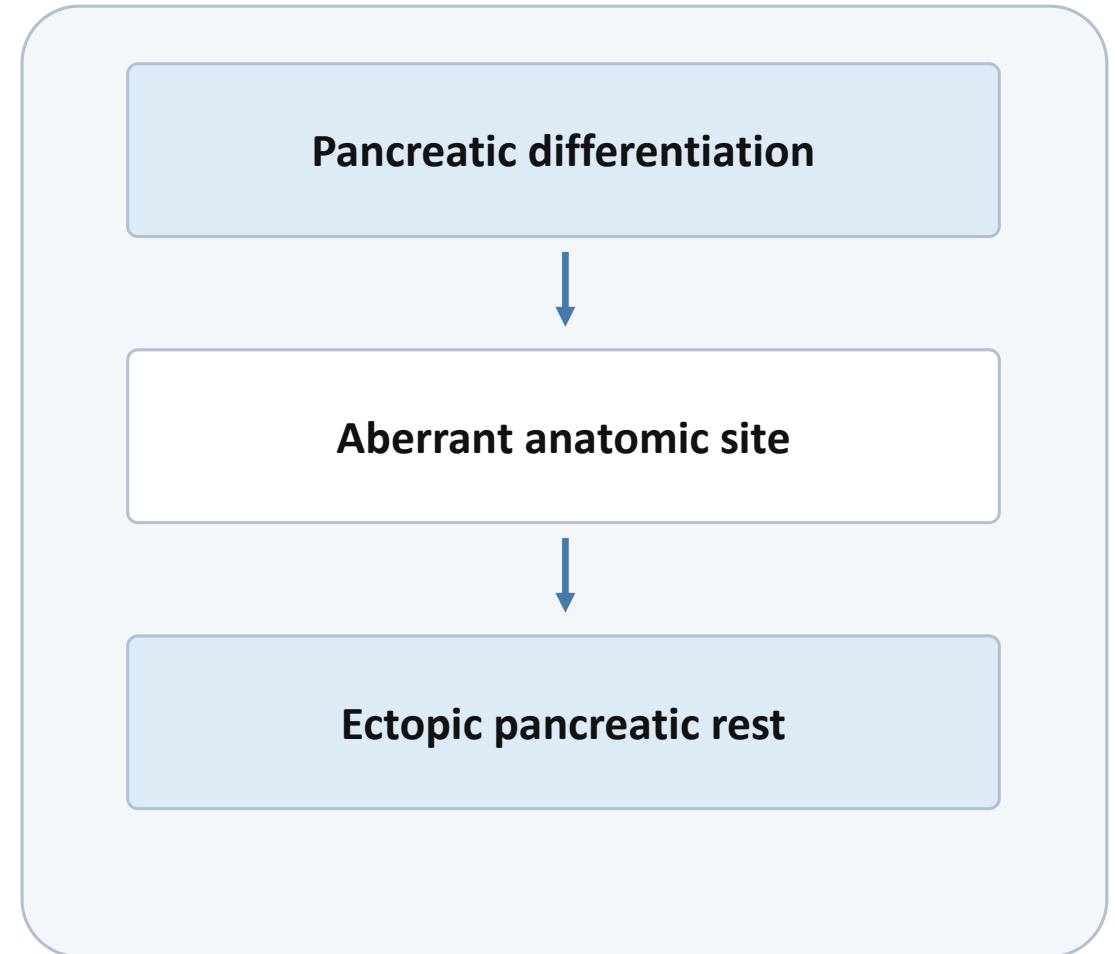
Ectopic pancreas: Pancreatic tissue out of place

Ectopic pancreas is pancreatic tissue located in an aberrant site.

It represents an embryologic pancreatic rest.

It is usually an incidental finding but may produce localized complications.

Schematic — causal or drainage sequence



Ectopia is primarily a location abnormality

The ectopic tissue is described as normal-appearing pancreatic tissue.

The key abnormality is location, not necessarily cytologic atypia.

Symptoms arise when the local rest becomes inflamed or causes bleeding.

Schematic — key structures or categories

Normal-appearing tissue

Abnormal location

Potential local effects

Most favored ectopic sites: Stomach and duodenum

Schematic — key structures or categories

The stomach and duodenum are favored sites for pancreatic ectopia.

These upper gastrointestinal sites should be considered when interpreting an ectopic pancreatic rest.

The site itself does not imply symptoms.

Stomach

Duodenum

Additional ectopic sites extend into the intestine

Schematic — key structures or categories

The jejunum, Meckel diverticula, and ileum.

The distribution emphasizes that ectopia is not confined to the duodenum.

A rest is classified by pancreatic tissue identity despite its intestinal location.

Jejunum

Meckel diverticulum

Ileum

Ectopic pancreas: Frequency depends on ascertainment

Reported by Robbins

Ectopic tissue in about 2% of careful routine postmortem examinations.

This is specifically a postmortem-observation figure.

It should not be relabeled as the prevalence of symptomatic ectopic pancreas.

≈2%

Careful postmortem
examinations

Denominator and clinical context matter

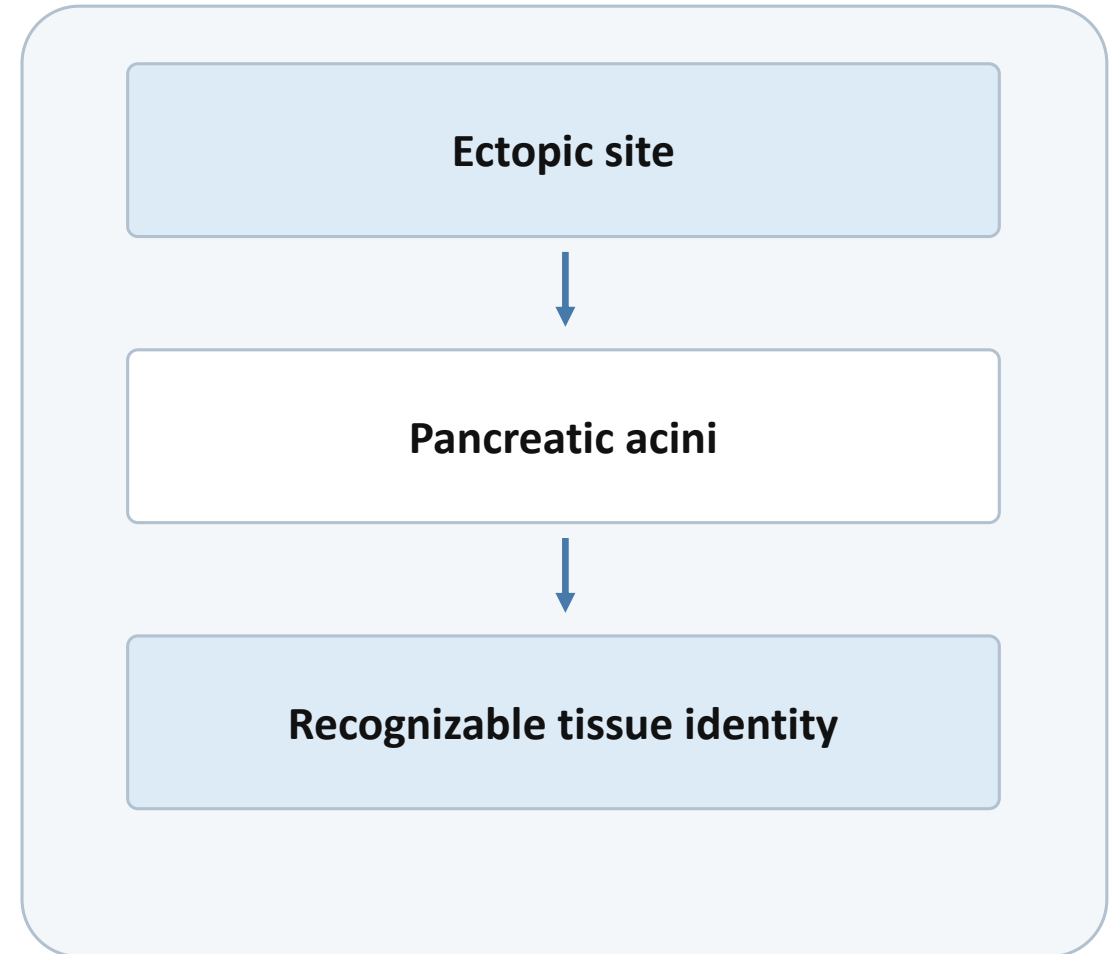
Ectopic pancreatic rests can form acini

Ectopic pancreatic tissue can contain normal-appearing acini.

The acinar component supports pancreatic tissue identity.

Anatomic misplacement can occur despite preserved microscopic differentiation.

Schematic — causal or drainage sequence



Ductal structures may be present in ectopia

The source describes glands in ectopic pancreatic rests.

Acinar and duct/glandular elements contribute to the pancreatic appearance.

A rest need not resemble the entire adult pancreas in size or location.

Schematic — key structures or categories

Pancreatic acini

Glandular/ductal elements

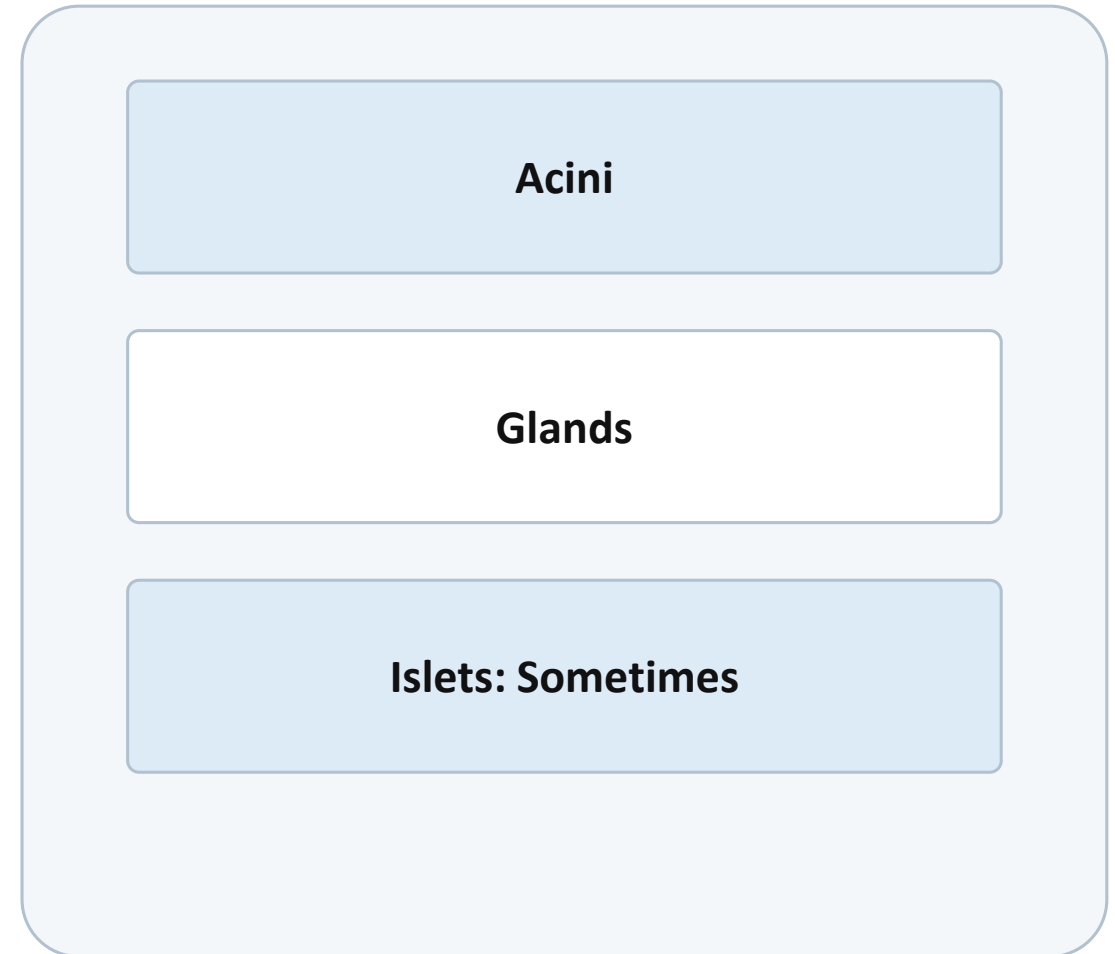
Some rests contain endocrine islets

Islets of Langerhans are sometimes present in ectopic tissue.

Islets are not required by the wording of the textbook description.

Therefore, microscopic composition may vary among pancreatic rests.

Schematic — key structures or categories



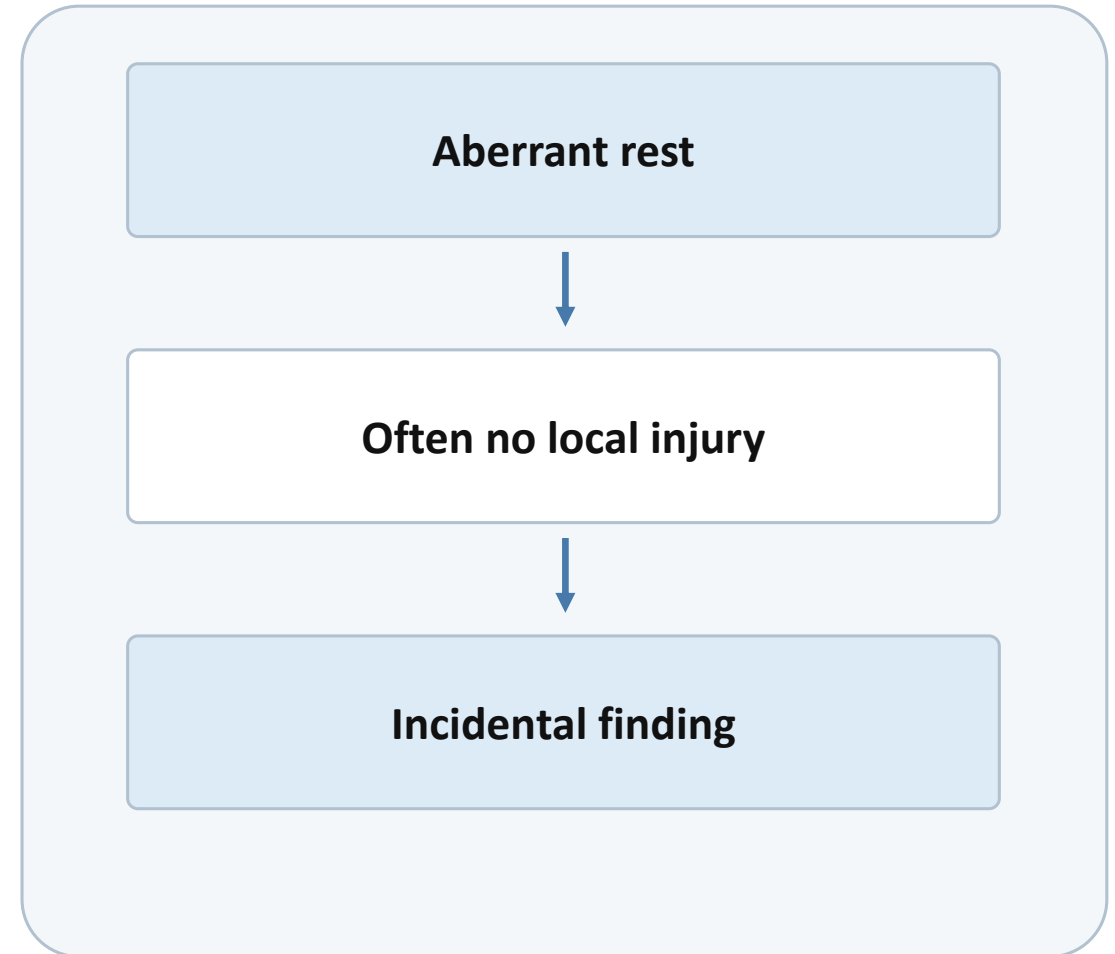
Why ectopic pancreas is often silent

Most pancreatic rests as incidental findings.

Merely finding pancreatic tissue at an abnormal site does not establish a functional disorder.

Clinical importance increases when local inflammation or bleeding occurs.

Schematic — causal or drainage sequence



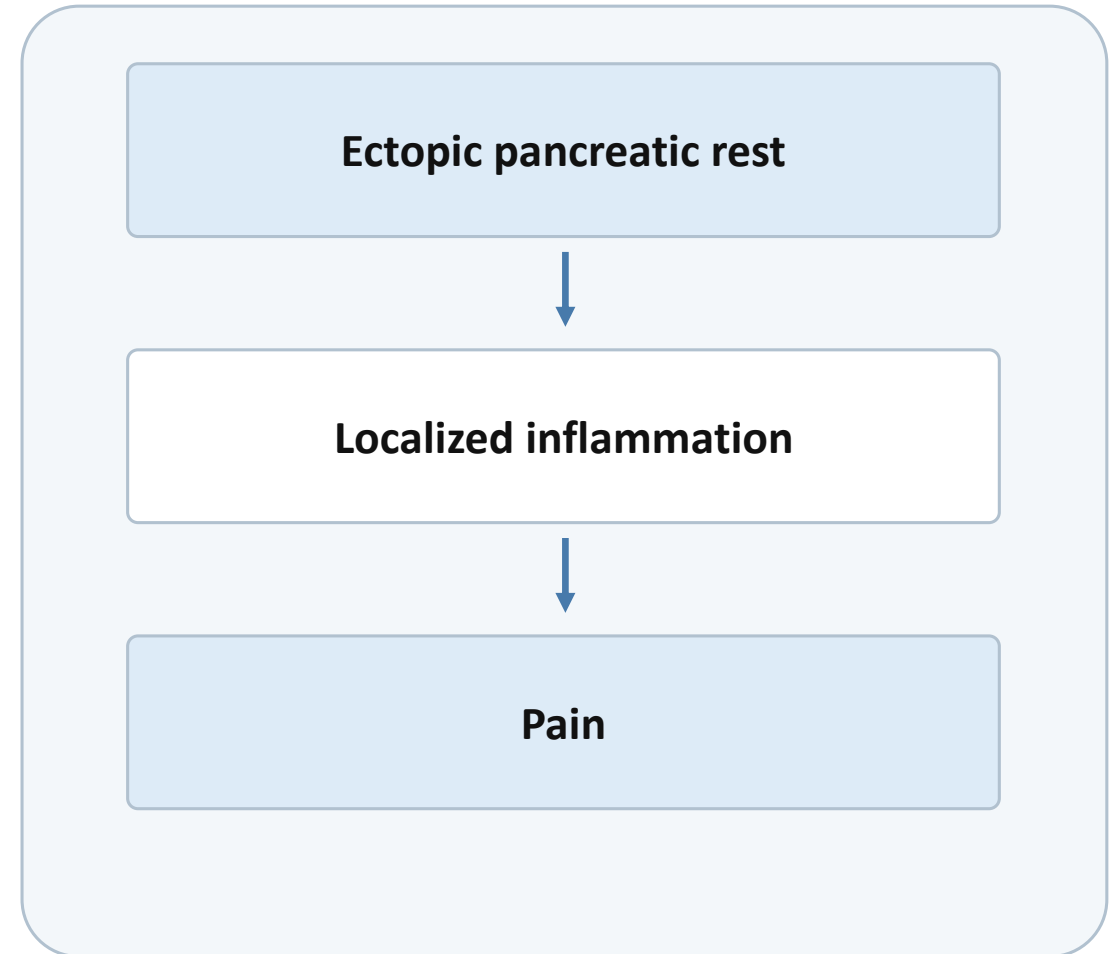
Localized inflammation can cause pain

A pancreatic rest can develop localized inflammation.

Pain as a possible clinical effect.

In a symptomatic case, the local lesion and surrounding tissue effects matter.

Schematic — causal or drainage sequence



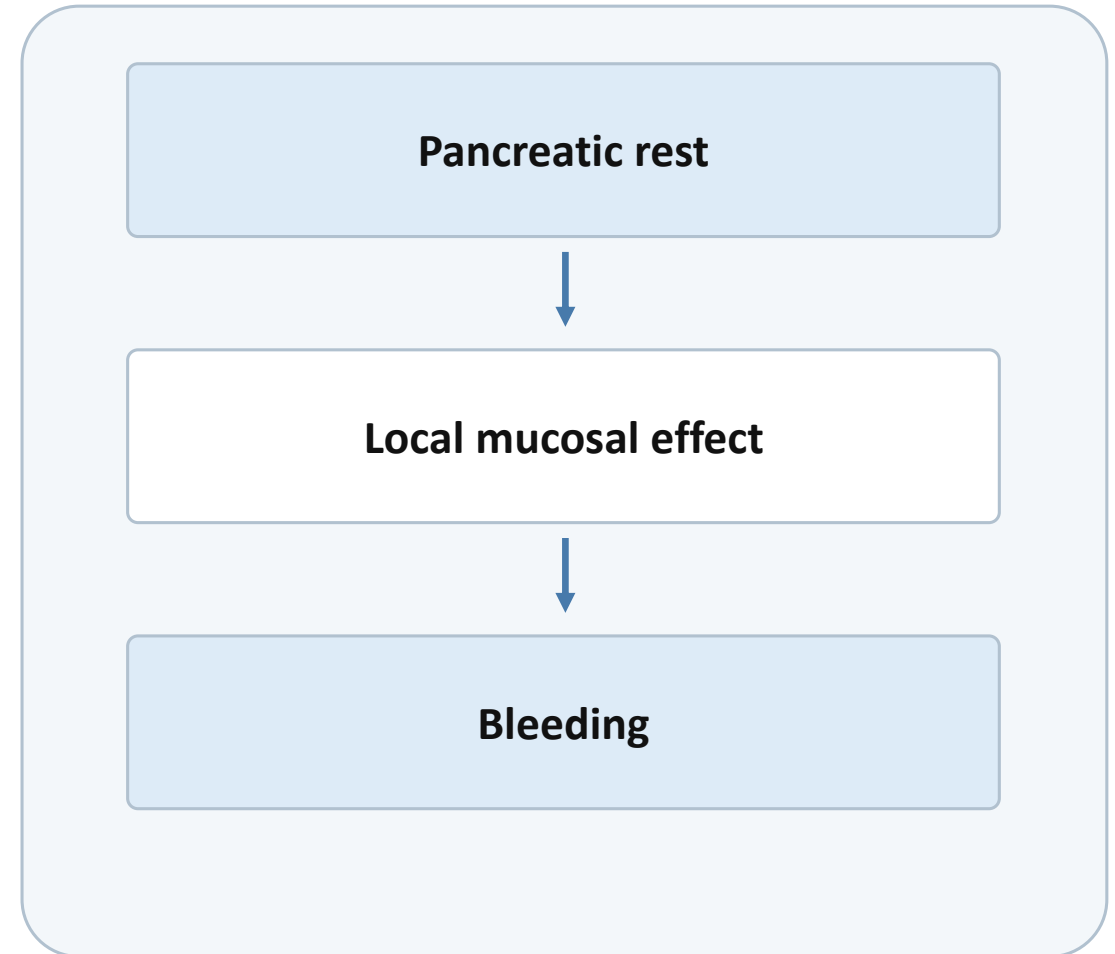
Mucosal bleeding is another possible effect

Ectopic pancreas may incite mucosal bleeding.

The relevant mechanism is a local effect of an aberrant tissue rest.

Bleeding is a possible consequence, not an inevitable feature of ectopia.

Schematic — causal or drainage sequence



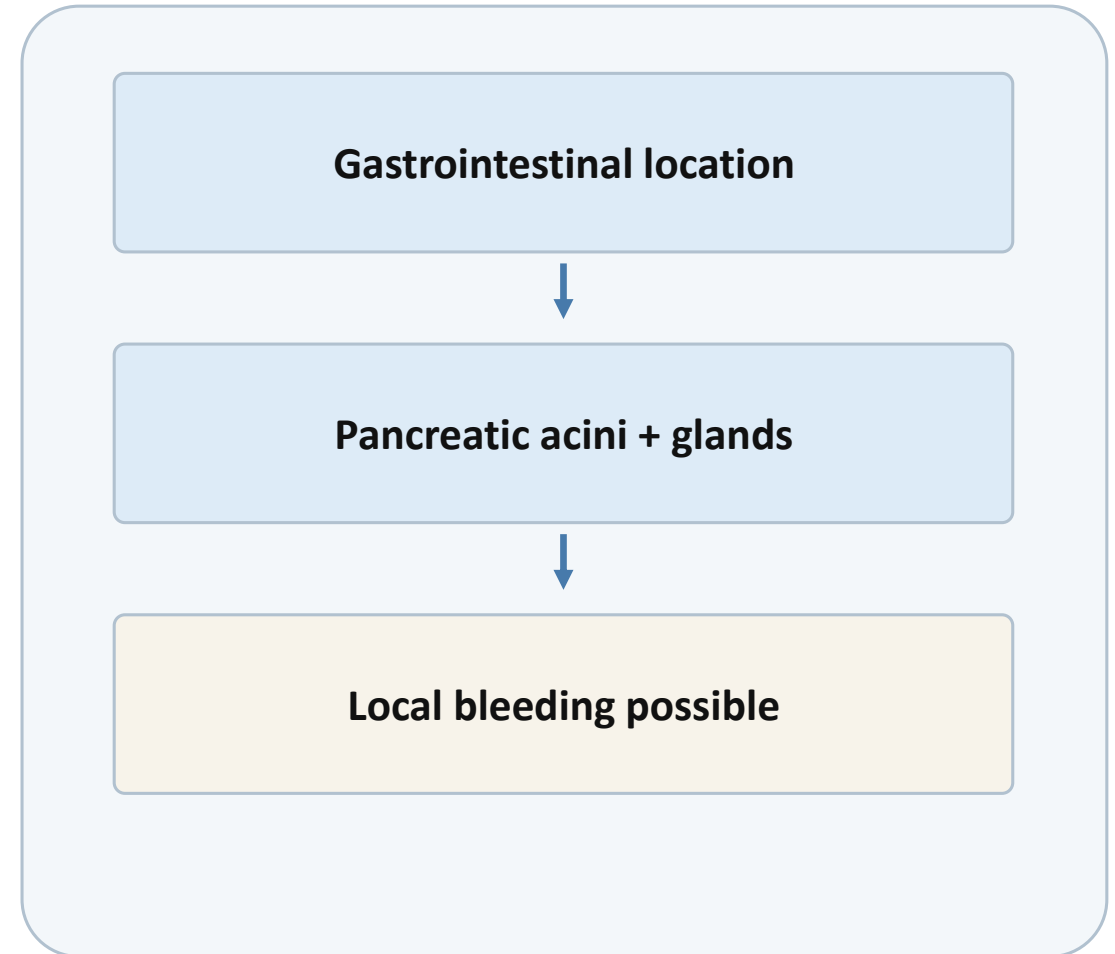
Ectopic pancreas: Clinicopathologic exercise

Teaching exercise — hypothetical example

A gastrointestinal lesion contains pancreatic acini and glands outside the normal pancreas.

The overlying mucosa shows a local bleeding complication.

Interpretation: Ectopic pancreas with a symptomatic local effect.



Ectopia and annular pancreas are not equivalents

Feature	Ectopic pancreas	Annular pancreas
Anatomic issue	Pancreatic tissue at separate site	Ring around duodenum
Typical sites	Stomach/duodenum; Other bowel	Second portion of duodenum
Microscopy	Acini, glands $\pm$ islets	Normal pancreatic tissue
Potential problem	Local pain or mucosal bleeding	Duodenal obstruction

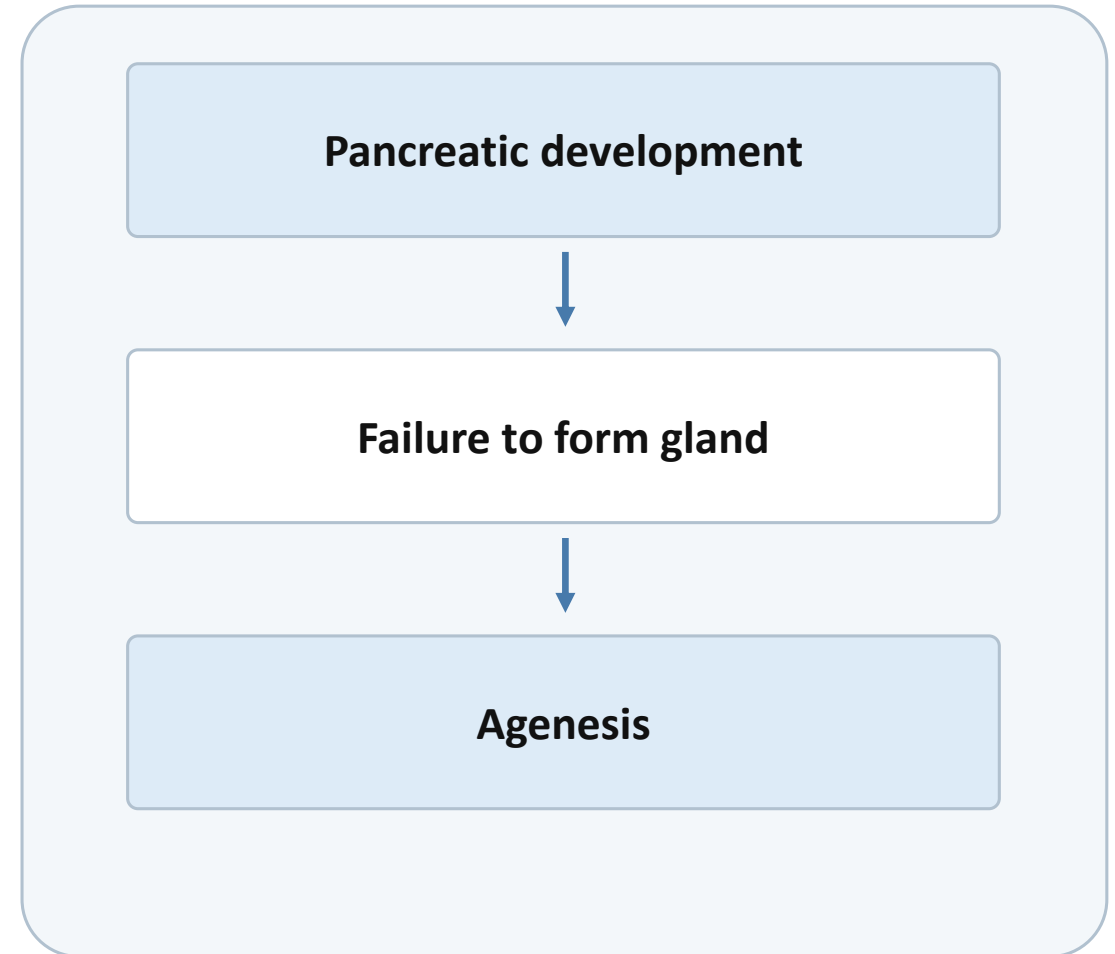
Pancreatic agenesis: Absence of development

Pancreatic agenesis means the pancreas fails to develop.

The condition is very rare description.

It differs fundamentally from abnormal rotation or duct drainage of an otherwise formed gland.

Schematic — causal or drainage sequence



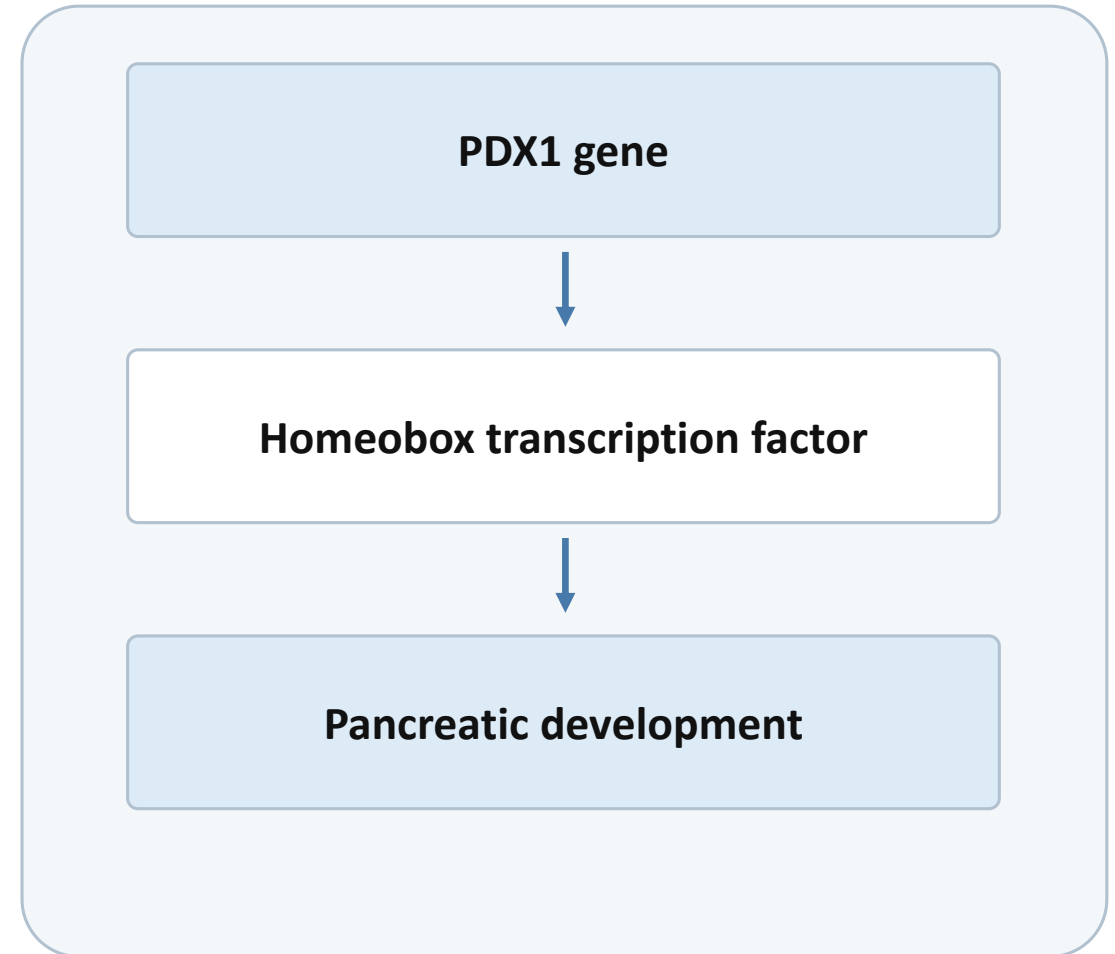
PDX1 is required for pancreatic development

PDX1 encodes a homeobox transcription factor.

The factor has a critical developmental role in the pancreas.

Pathogenic changes in this developmental regulator can cause pancreatic agenesis.

Schematic — causal or drainage sequence



Agenesis: Inherited mutations explain some cases

Homozygous germline PDX1 mutations in some cases.

The source does not assign PDX1 mutations to every case of agenesis.

This example illustrates how a developmental regulator can affect organ formation.

Schematic — key structures or categories

Homozygous germline variants

PDX1 in a subset

Failure of organ development

Agensis contrasts with ductal variants

Characteristic	Agensis	Pancreas divisum
Underlying process	Gland fails to form	Fetal duct systems fail to unite
Anatomic phenotype	Absent organ development	Gland with separate drainage
Molecular association	PDX1 in some cases	No single gene specified here
Main concept	Developmental formation	Ductal architecture

The mechanism–phenotype map

Embryologic disturbance	Anatomic outcome	Possible significance
Duct-system fusion failure	Pancreas divisum	Pancreatitis in minority
Incomplete ventral-bud rotation	Annular ring	Duodenal obstruction
Aberrant location of pancreatic rest	Ectopic tissue	Pain or mucosal bleeding
Failure of pancreatic development	Agenesis	Absent gland

Which findings predict an incidental variant?

Divisum is usually asymptomatic despite its ductal configuration.

Annular pancreas is asymptomatic in approximately one-third of cases.

Ectopic pancreatic rests are usually incidental.

Morphology alone is insufficient to claim a symptom is caused by the finding.

Schematic — key structures or categories

Divisum: Mostly silent

Annular: ~1/3 silent

Ectopia: Often incidental

Match the complication to the anomaly

Anomaly	Structural emphasis	Clinical correlation
Divisum	Minor-papilla-dominant drainage	Proposed pancreatitis predisposition
Annular pancreas	Duodenal encirclement	Obstruction in a subset
Ectopic pancreas	Local pancreatic tissue rest	Pain or mucosal bleeding
Agenesis	Failure of organ formation	Genetic developmental basis in some

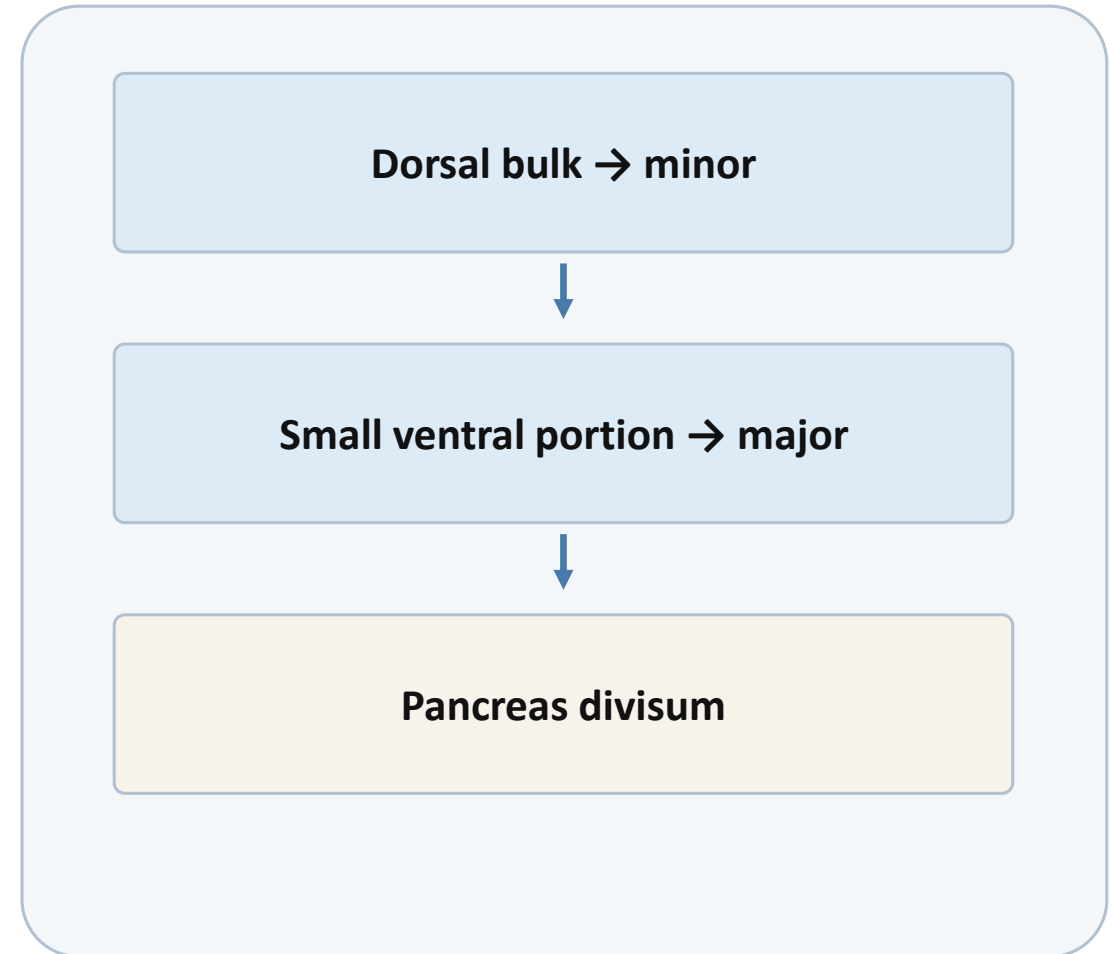
Integrated case: Accessory outlet dominates

Anatomic tracing shows the bulk of the gland draining into the duodenum through the minor papilla.

Only a small ventral head portion drains through Wirsung and the major papilla.

Best explanation: Pancreas divisum following failed ductal fusion.

Teaching exercise — hypothetical example



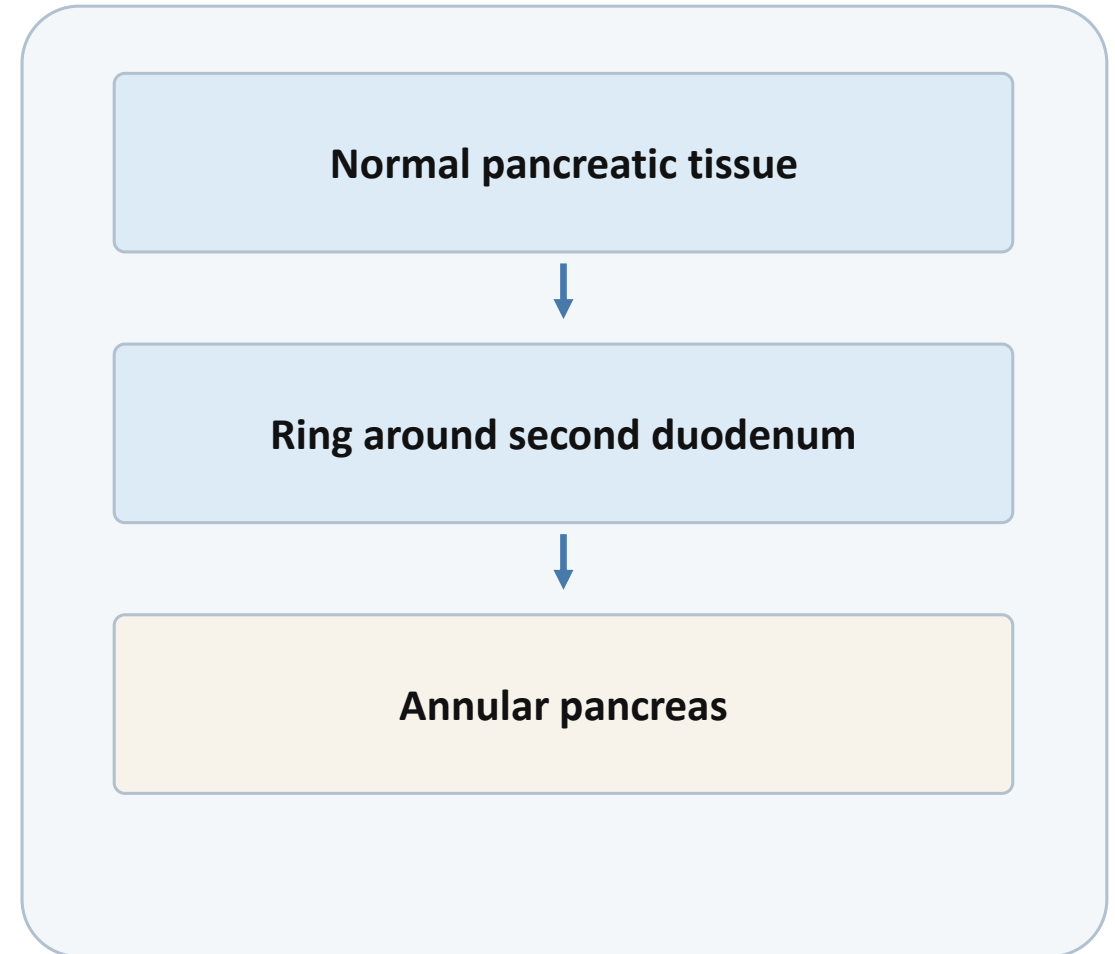
Integrated case: Duodenal encirclement

Teaching exercise — hypothetical example

A ring of normal pancreatic tissue completely surrounds the second duodenal portion.

Symptoms, if present, may reflect obstruction at that segment.

Best explanation: Annular pancreas resulting from incomplete ventral rotation.



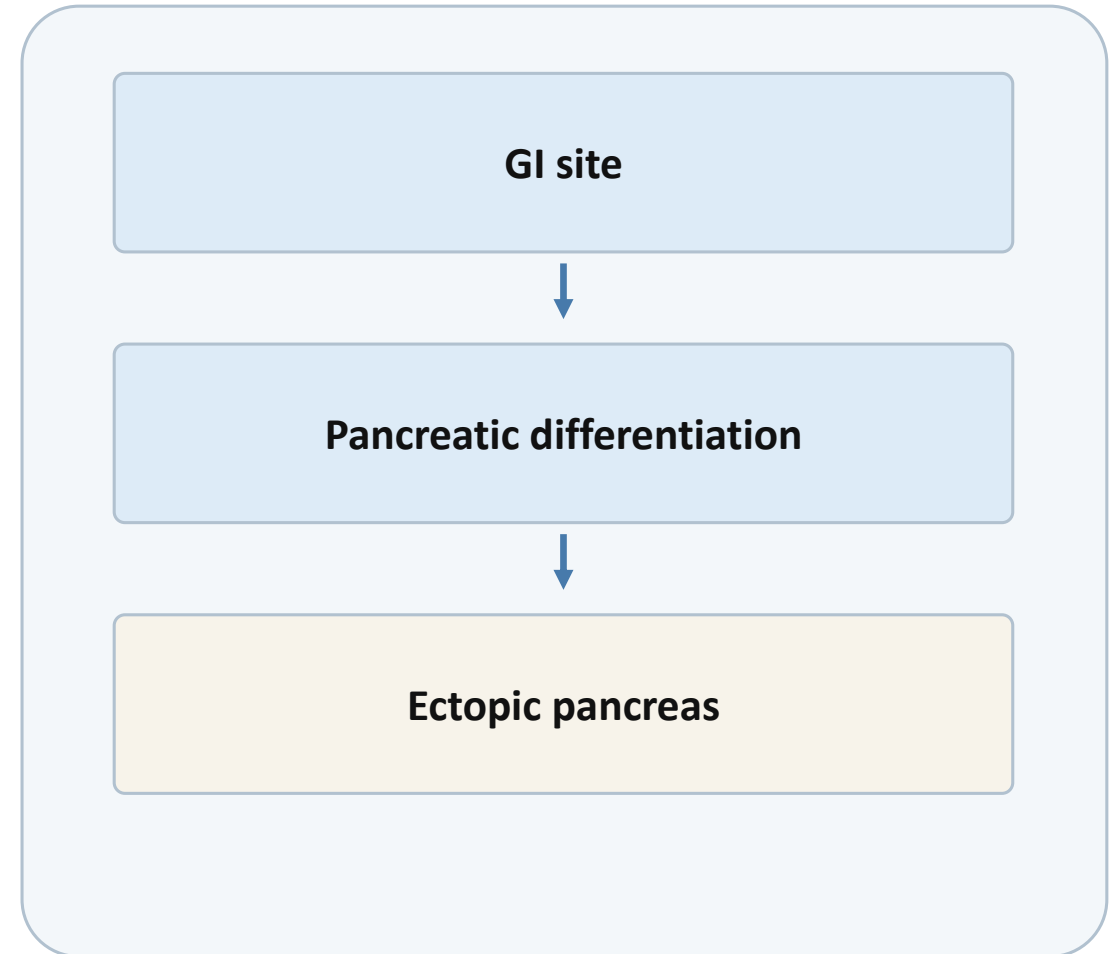
Integrated case: Normal pancreas at an unusual site

Teaching exercise — hypothetical example

A localized gastrointestinal focus contains pancreatic acini and glands.

It is distinct from the normal organ location.

Best explanation: Ectopic pancreatic tissue, often incidental unless locally complicated.



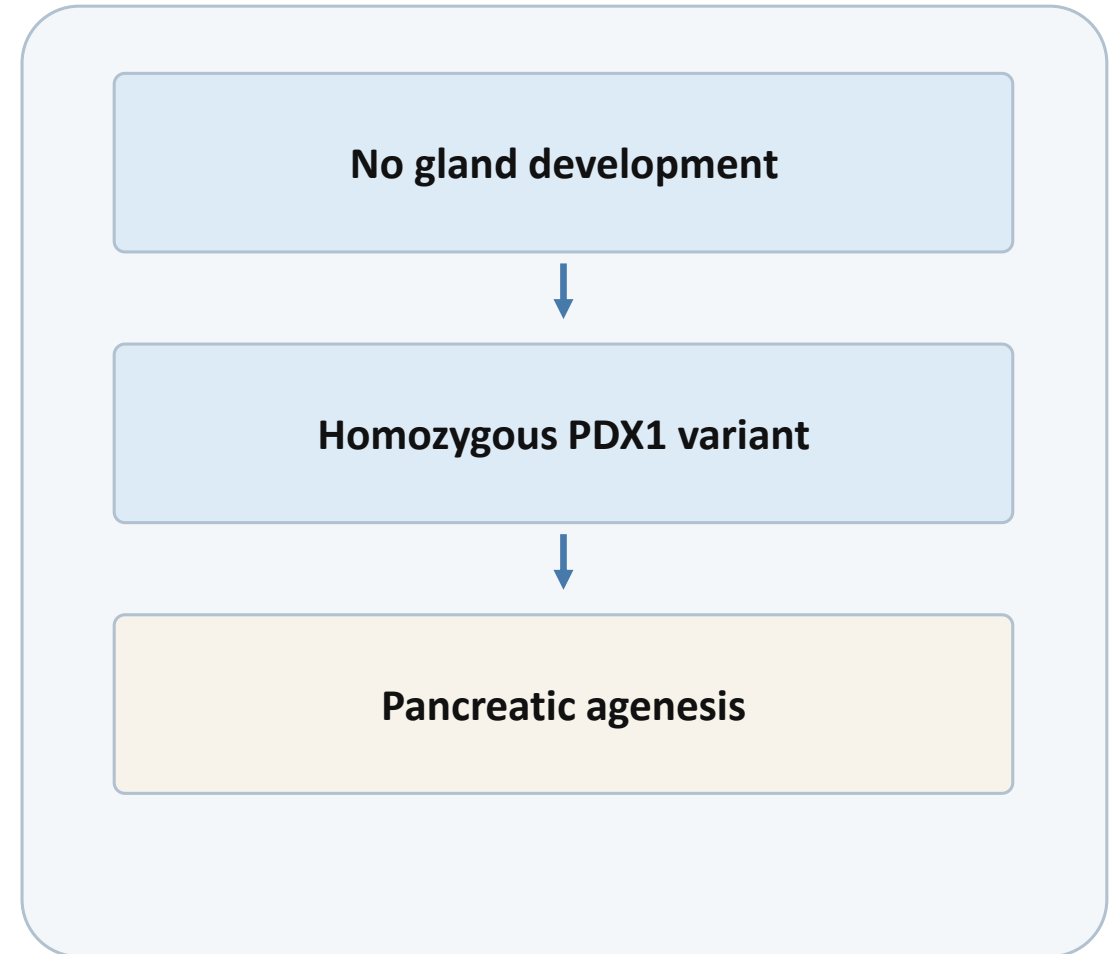
Integrated case: Loss of organ development

Teaching exercise — hypothetical example

The pancreas has failed to develop.

A homozygous germline PDX1 alteration is identified in the hypothetical case.

This supports the molecularly associated subset of pancreatic agenesis.



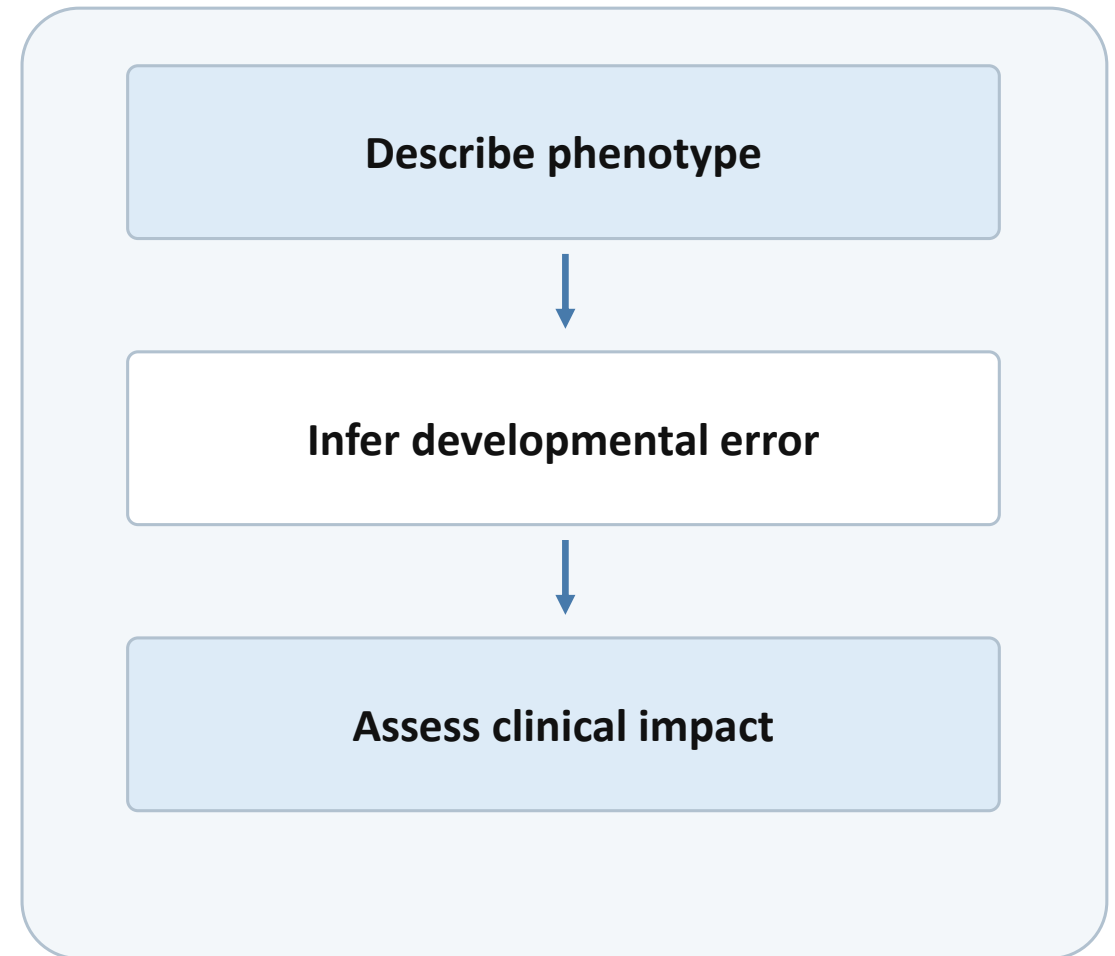
Interpret congenital pancreatic findings in three steps

Define the anatomical phenotype: Duct division, duodenal ring, ectopic rest, or absent pancreas.

Connect the phenotype to its developmental mechanism.

Only then consider whether there is a supported clinical consequence.

Schematic — causal or drainage sequence



Essential distinctions to retain

Fusion failure changes ductal routing;
Incomplete rotation can encircle the
duodenum.

Ectopic tissue is misplaced pancreatic
parenchyma; Agenesis is failure of gland
formation.

Divisum, annular pancreas, and ectopia are
commonly incidental or asymptomatic.

Report a complication only when the clinical
and anatomic context supports it.

Schematic — key structures or categories

Fusion

Rotation

Location

Organ formation

Reference and source boundaries

Robbins, Cotran & Kumar. Pathologic Basis of Disease. 11th ed. (2026), Chapter 19, pp. 800–801.

Developmental anatomy, duct physiology, and congenital variants are drawn from the supplied chapter text.

All mechanism drawings are original editable schematics, not textbook figures or patient images.