

## Opis choroby \*

### Definicja

A very rare, malignant, epithelial tumor of the pancreas characterized, macroscopically, by a usually large, well-circumscribed, fully or partially encapsulated, solid mass, often with hemorrhage, necrosis and cystic changes, in any portion of the pancreas and, histologically, by neoplastic cells with variable degrees of differentiation and morphology, ranging from acinar structures similar to normal pancreatic acini to large sheets of poorly differentiated neoplastic cells. Presenting symptoms are typically non-specific and include abdominal pain, weight loss, vomiting, nausea, and/or, less commonly, jaundice. Immunohistochemical evidence of acinar-specific products is observed. Association with Lynch syndrome, familial adenomatous polyposis, and pancreatic panniculitis has been reported.

### Dane

#### Klasyfikacja

Choroba

#### Synonimy

Pancreatic acinar cell carcinoma

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#### Kod ORPHA

424046

#### Kod OMIM

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#### Kod ICD10

C25.2

#### Kod ICD11

2C10.0

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#### \*Źródło

orphanet