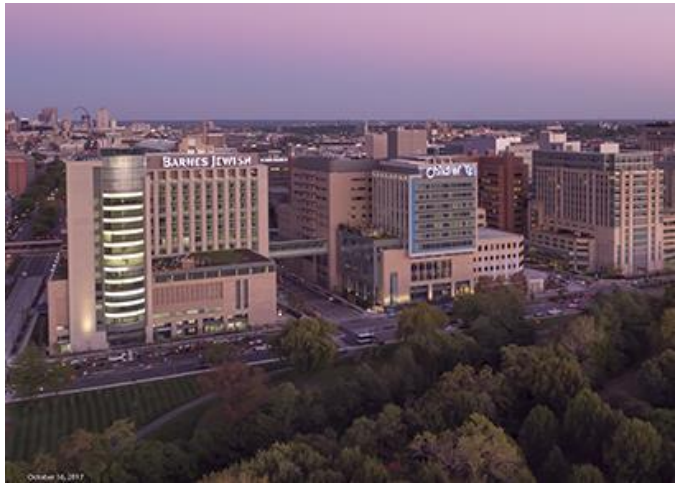

CP without AP

CEL MODY or UPR

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Disclosures

Royalties for UpToDate

CEL-MODY

CEL

- Expressed in pancreas and lactating mammary gland
- Critical for fat digestion in newborns

CEL-MODY

- Deletion, frameshift variant in the VNTR associates with MODY and chronic pancreatitis

Clinical Features of CEL-MODY

- Rare autosomal dominant disease
- Pancreatic lipomatosis evident in children
- Develop diabetes as young adults
  - No episodes of ketoacidosis
  - Autoantibodies not present
  - Diabetics require insulin
- Exocrine pancreatic insufficiency is evident in childhood
- Pancreatic cysts common in adults
  - Calcifications not seen
- Episodes of acute pancreatitis not documented
  - Most had history of mild, recurrent abdominal pain starting in the second decade life