# Idiopathic Pancreatitis PLUS

**Key Points:**
- Most people who have one episode of unexplained acute pancreatitis will not have another.
- In patients with recurrent unexplained acute pancreatitis, workup should include a CT of the abdomen, endoscopic ultrasound, pancreatic function tests, and ERCP. Empiric cholecystectomy should be considered if all of the above are negative.
- Groove pancreatitis is a localized, segmental chronic pancreatitis that may be difficult to distinguish clinically from pancreatic cancer.
- Charcot-Marie-Tooth disease is a hereditary motor sensory neuropathy characterized by motor weakness, calf/foot deformities, and distal sensory neuropathy.
- Based on one small study, patchy or nodular ground-glass on HRCT is 100% sensitive and 89% specific for PCP in AIDS patients with normal, equivocal, or non-specific xrays.

## 1. What is the standard of care for working up “idiopathic” pancreatitis?

- The usual investigations (history and physical, basic labs including Ca<sup>2+</sup> and TG, and ultrasound) will not reveal a cause for pancreatitis in 30% of patients with their first episode of acute pancreatitis. In one retrospective study, only 1/31 patients with an unexplained episode of acute pancreatitis went on to suffer another episode over the next 36 months.
- A prospective study examined 126 patients with recurrent (>1 episode) idiopathic pancreatitis with ERCP, sphincter of Oddi manometry, and bile analysis. A presumed cause was identified in 79% of the patients (papillary stenosis 21%, pancreas divisum 7%, choledocholithiasis 5%; SOD dysfunction 21% of patients with a gallbladder, 47% of those without). 75% of cases were “endoscopically treatable” per the authors, and freedom from pancreatitis rates ranged from 67-100% (depending on cause) over 29 months of follow-up. Of note, sphincter of Oddi manometry is not without complications (up to 22%) and is therefore not recommended by all authorities.
- Some authorities recommend the following path for recurrent idiopathic pancreatitis:

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From *UptoDate 2002, “Etiology of Acute Pancreatitis”*
2. **What is groove pancreatitis?**
Groove pancreatitis is a form of chronic segmental pancreatitis affecting the head of the pancreas. The groove referred to lies between the head of the pancreas, the duodenum, and the common bile duct. Some authorities have speculated that Santorini’s duct (minor) is more frequently causative than Wirsung’s duct (major). The etiologies of groove pancreatitis are not different from those of other types of pancreatitis; however, it may be difficult to differentiate between groove pancreatitis and pancreatic cancer on radiologic grounds.

3. **What is Charcot-Marie-Tooth (CMT) disease?**
CMT disease is a broad name for a heterogeneous group of disorders also known as hereditary motor sensory neuropathies (HMSN). CMT is the most common hereditary peripheral neuropathy. All of the disorders are caused by specific mutations in myelin genes and can be autosomal dominant, autosomal recessive, or X-linked. Pathologically, there is demyelination of peripheral motor and sensory nerves. Clinically, patients typically present in the 2nd or 3rd decade with distal leg weakness (sprained ankles, gait difficulty), foot deformity (hammer toes, pes cavus (high-arching foot)), hand/foot/calf atrophy (“stork leg deformity” or “champagne glass calves”), and distal proprioception and vibration loss. Treatment is supportive with stretching, orthotics, and surgery for deformities. Life expectancy is not affected.

4. **How good is HRCT in the diagnosis of PCP?**
The easy answer – it’s OK with a normal, equivocal, or non-specific xray. Hard to believe, but there is one good prospective trial from SFGH of 51 patients (yes, 51). Sensitivity was 100%, specificity 89%. Given this, it is now part of the SFGH algorithm for PCP and absence of ground-glass rules out PCP with normal, equivocal, or non-specific xray findings.

5. **How does MRCP work and what is it good for?**
MRCP uses T2-weighted images which make stationary or slow-flowing fluid within the bile and pancreatic ducts appear very bright relative to the low signal intensity solid tissue and to the flowing blood which has little or no signal. It is best used for bile duct obstruction, choledocholithiasis, pancreatitis, pancreas divisum, and pancreatic cancer. It has limitations in small stones, PSC, strictures, and the inability to do an intervention.

**References**