

ENTITLEMENT ELIGIBILITY GUIDELINES

CHRONIC PANCREATITIS

MPC 00951
ICD-9 577.1

DEFINITION

Chronic Pancreatitis is a chronic inflammatory disease of the pancreas characterized by progressive fibrosis, and destruction of exocrine tissue and endocrine tissue.

Please note: Entitlement should be granted for a chronic condition only. For VAC purposes, “chronic” means that the condition has existed for at least 6 months. Signs and symptoms are generally expected to persist despite medical attention, although they may wax and wane over the 6 month period and thereafter.

DIAGNOSTIC STANDARD

A diagnosis from a qualified medical practitioner is required, supported by test results of serum amylase.

The following investigations are normally conducted but are not mandatory:

- plain film x-rays of abdomen and chest
- CT scan or MRI of abdomen
- blood glucose
- serum lipase

Endoscopic retrograde cholangio-pancreatography (ERCP) is conducted on occasion.

Evidence of duration of a disability for at least 6 months should be provided.

ANATOMY AND PHYSIOLOGY

The pancreas is an elongated organ located in the retroperitoneal space with its head within the curve of the duodenum and its tail extending towards the spleen. The retroperitoneal location and the absence of a capsule surrounding the pancreas are important factors in understanding how pancreatitis evolves. Pancreatic inflammation and fibrosis may spread unimpeded by anatomic barriers to involve the spleen, the splenic artery and vein, the duodenum and distal common bile duct, the mesocolon, the diaphragm and pararenal spaces, the lesser omental sac, and the celiac and superior mesenteric ganglia.

The acinar cells of the exocrine pancreas synthesize approximately 20 digestive enzymes including colipase, a secretory trypsin inhibitor, and lithostathine S₂₋₅, a protein that inhibits the precipitation of calcium carbonate from pancreatic juice. These secretory proteins are sorted into condensing vacuoles, which then become zymogen granules.

Premature activation of zymogens within the pancreas is the key event in the pathogenesis of acute pancreatitis. Protein and calcium carbonate precipitate within the pancreatic duct system play a major role in the development of chronic pancreatitis.

Acute pancreatitis is an acute inflammatory condition due to auto-digestion of pancreatic tissue by its own enzymes, typically presenting with abdominal pain. It is usually associated with raised pancreatic enzymes in blood or urine. Only rarely does acute pancreatitis lead to chronic pancreatitis. Chronic pancreatitis may follow acute pancreatitis when there have been disturbances of the duct system, most frequently produced when trauma is the inciting cause.

Degrees of severity vary, with severe representing multisystem failure and/or development of a complication.

There are many accepted causes of acute pancreatitis. There must be many pathophysiological mechanisms involved but the basic abnormality appears to be release from the pancreatic acinar cells of activated enzymes into the pancreatic and surrounding tissue due to disturbances in the plasma membranes of the pancreatic acinar cells.

Chronic pancreatitis is a chronic inflammatory condition typically presenting with chronic abdominal pain and a progressive fibrosis of the pancreas with loss of exocrine (steatorrhea) and endocrine (diabetes mellitus) function. Acute relapsing pancreatitis (recurrent attacks of acute pancreatitis) are often superimposed. The disease may be focal, segmental or diffuse. Clinically and pathologically there may be episodes of acute pancreatic inflammation during the course of the disease, leading to nearly all the possible complications of acute pancreatitis. Chronic pancreatitis may be caused by pancreatic duct obstruction. Relief of the obstruction cannot only arrest the progress of the disease but can lead to some restitution of morphology and function. It may be at a stage in which fibrosis, the end result of chronic inflammation, may replace the gland completely without any evidence of active inflammation or pancreatic acinar tissue when examined.

Specific diseases involving fibrosis and acinar atrophy in the pancreas, e.g. cystic fibrosis of the pancreas and hemochromatosis, are not considered under the name of chronic pancreatitis.

Chronic pancreatitis can occur, but not necessarily with the same clinical course as in alcoholic or idiopathic chronic pancreatitis, in the following conditions:

- chronic renal failure
- some congenital abnormalities of the pancreas
- hereditary pancreatitis
- after renal, hepatic and cardiac transplantation

Pancreatic pseudocysts are a circumscribed local accumulation of leaking pancreatic secretions that occur as the result of pancreatic inflammation. Pseudocysts can develop a fibrous pseudo capsule and may contain bacteria and inflammatory cells but no liquefaction necrosis of the pancreas. They do not have an epithelial lining. Acute and chronic pseudocysts occur and a significant number of acute pseudocysts resolve slowly spontaneously over weeks. Unresolved pseudocysts can lead to complications which can be fatal.

CLINICAL FEATURES

Chronic inflammatory disease of the pancreas may present as episodes of acute inflammation in a previously injured pancreas or as chronic damage with persistent pain and malabsorption.

The classic presentation of acute pancreatitis is development of steady, dull or boring pain in the mid-epigastric area. It is often associated with nausea and vomiting. Pain may radiate to the back or chest. Persons may go on to develop low grade fever, hypotension and tachycardia. One to two weeks later ecchymoses may appear in the flanks (Grey Turner's sign) or umbilicus (Cullen's sign).

The clinical manifestations of chronic pancreatitis comprise abdominal pain, followed by the results of progressive loss of exocrine and endocrine function (leading to maldigestion and malabsorption of nutrients and glucose intolerance). A proportion of cases of chronic pancreatitis are pain-free. Loss of 80% of exocrine function can occur without demonstrable abnormality in digestion or absorption.

Diabetes mellitus is not likely to occur unless more than 80% of the gland has been destroyed. Because of coexisting loss of glucagon from islet cells, the diabetic status in chronic pancreatitis is frequently brittle.

Steatorrhea does not occur unless the secretion of pancreatic lipase is reduced to less than 10% of normal. Thus, steatorrhea is not common in chronic pancreatitis unless there is substantial damage to the pancreas or complete blockage of the pancreatic duct in the head of the gland.

It is not possible to say when or how chronic pancreatitis (of any cause) begins.

There is no pancreatic function test that has been shown to be sensitive in recognizing early chronic pancreatitis. This often results in a delay in establishing a diagnosis of chronic pancreatitis. ERCP and computerized scanning may show apparent minor abnormalities, but as yet there has been incomplete correlation with morbid anatomy. Only major changes from normal can be interpreted with confidence in well-advanced disease.

PENSION CONSIDERATIONS

A. CAUSES AND/OR AGGRAVATION

THE TIMELINES CITED BELOW ARE NOT BINDING. EACH CASE SHOULD BE ADJUDICATED ON THE EVIDENCE PROVIDED AND ITS OWN MERITS.

1. Prolonged and heavy alcohol consumption prior to clinical onset or aggravation

Chronic alcohol abusers represent 70% to 80% of persons with chronic pancreatitis. No particular level of alcohol consumption beyond that of a "heavy drinker" can be stated to give rise to chronic pancreatitis in those who are susceptible, for clearly there are large numbers of persons who consume alcohol in equivalent quantities without developing chronic pancreatitis.

Alcoholic liver disease develops in 40% to 50% of persons afflicted and frequently becomes manifest 5 to 10 years after the onset of pancreatitis. It is not possible to say in every person who has an attack of acute pancreatitis whether it is an episode in the course of already established chronic pancreatitis, unless a long story of characteristic pain is present as in the case of chronic pancreatitis. Calcification takes a further 8 years on average to appear as does exocrine insufficiency with detectable maldigestion/malabsorption.

Abstention from alcohol will most frequently abolish the attacks of painful acute pancreatitis which punctuate the progress of chronic alcoholic pancreatitis. It does not prevent pain in those who already have it continuously, nor is it believed to arrest the progress of the disease. It could not be expected to alter the effects of loss of active pancreatic tissue (maldigestion/malabsorption), diabetes, or calcification.

2. Idiopathic

This category comprises 10% to 20% of affected persons. The features, pathologically and clinically, resemble those of alcohol-produced chronic pancreatitis, except that the course is slower, and duct changes and calculi are

less frequent.

3. Disturbance of anatomical structure of the pancreatic ductal system **through disease** at time of clinical onset or aggravation

This involves a demonstrated alteration to the pancreatic ductal system with consequent disturbed pancreatic outflow. Diseases which may lead to such disturbance include, but are not limited to:

- pancreatic pseudocysts (e.g. following acute pancreatitis)
- periampullary tumors
- diseases which lead to mechanical or structural changes of the pancreatic duct sphincter, e.g. surgery, stenosis

4. Disturbance of anatomical structure of the pancreatic ductal system **through trauma** at time of clinical onset or aggravation

Only severe trauma to the abdomen would result in trauma to the pancreas. Penetration of the abdominal cavity is not required, as abdominal bleeding may result from a severe blow to the abdomen. The trauma would include, but is not limited to:

- Physical injury, e.g. a hit, blow, knock, or twisting, bending, crushing injury (as may result from shell and bomb explosions, vehicle and machinery accidents)
- Penetrating injury, e.g. from projectiles such as bullets and shrapnel
- Surgery, i.e. post-operative pancreatitis

5. Tropical pancreatitis prior to clinical onset or aggravation

This occurs frequently in malnourished persons in southern India, Africa and Indonesia. It is a disease of the teens and young adult life. Although abdominal pain is common, the diagnosis is frequently made on the basis of newly discovered diabetes or pancreatic calcifications. While malnutrition is suspected to play a role, this form of chronic pancreatitis is not found in other areas where malnutrition is equally common. There is no relationship to alcohol consumption. Tropical pancreatitis is alleged to predispose to adenocarcinoma of the pancreas.

6. Hereditary pancreatitis prior to clinical onset or aggravation

Pancreatitis is inherited as an autosomal dominant trait, with 40% to 80% penetrance accounting for approximately 2% of persons with chronic pancreatitis. Episodes of abdominal pain usually start at ages 5 to 12. The etiology is amino acid substitutions in the cationic trypsin molecule.

7. Senile/pancreatic atrophy prior to clinical onset or aggravation

Of persons with chronic pancreatitis, 10% to 20% are older than 60 at initial presentation. Smoking and obesity, but not alcohol abuse, have been implicated. Senile atrophy and lipomatosis of the pancreas are thought likely to represent the same poorly understood entity.

8. Post-renal transplant prior to clinical onset or aggravation9. Endocrine disorder, including hyperparathyroidism, prior to clinical onset or aggravation

Hyperparathyroidism can result in hypercalcemia.

10. Hyperlipidemia prior to clinical onset or aggravation11. One or more courses of therapeutic radiation to the region of the pancreas, where the first exposure occurred at least one year before clinical onset or aggravation

This may occur with periaortic node radiation and radiation for other conditions, including cancer of the stomach and/or Hodgkin's Lymphoma.

12. Inability to obtain appropriate clinical management**B. MEDICAL CONDITIONS WHICH ARE TO BE INCLUDED IN ENTITLEMENT/ASSESSMENT**

- Pancreatic pseudocyst
- Pancreatic abscess
- Pancreatic ascites
- Secondary malabsorption

C. COMMON MEDICAL CONDITIONS WHICH MAY RESULT IN WHOLE OR IN PART FROM CHRONIC PANCREATITIS AND/OR ITS TREATMENT

- Diabetes mellitus
- Adenocarcinoma

REFERENCES FOR CHRONIC PANCREATITIS

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