ICD-10-CM Documentation
and Coding Best Practices

Guidance on Documentation and Coding for Porphyria, Amyloidosis and Metabolic Syndrome

Overview of disorders of porphyrin and bilirubin metabolism (E80)

Porphyria refers to a group of disorders that result from a buildup of natural chemicals that produce porphyrin in your body. Porphyrins are essential for the function of hemoglobin — a protein in your red blood cells that links to porphyrin, binds iron, and carries oxygen to your organs and tissues. High levels of porphyrins can cause significant problems and typically affects the nervous system and skin. Porphyria is usually inherited and cannot be cured.

Types of porphyria

- Acute – mainly affecting nervous system.
- Cutaneous – mainly affecting the skin.

Signs and symptoms

- Acute type – severe abdominal pain, pain in the chest, legs or back, constipation, diarrhea, vomiting, nausea, urination problems, mental changes, palpitations, high blood pressure and seizures.
- Cutaneous – skin redness, edema, sensitivity to the sun and blisters on exposed skin.

Documentation guidance for porphyria

Medical record documentation should include symptoms, diagnosis tests/labs ordered as well as interpretation of results when received, referrals to specialist, and treatment plan.

ICD-10-CM Code information

ICD-10 Category E80

E80.0 Hereditary erythropoietic porphyria

E80.1 Porphyria cutanea tarda

E80.20 Unspecified porphyria

E80.21 Acute intermittent (hepatic) porphyria

E80.29 Other porphyria

E80.3 Defects of catalase and peroxidase

E80.4 Gilbert syndrome

E80.5 Crigler-Najjar syndrome

E80.6 Other disorders of bilirubin metabolism

- E80.7 Disorder of bilirubin metabolism, unspecified

(continued)

Overview of disorders of Amyloidosis (E85)

Amyloidosis is a rare condition that occurs when the abnormal amyloid protein builds up in body organs and interferes with their normal function. Some varieties are hereditary while others are caused by outside factors, such as inflammatory diseases or long-term dialysis.

Subtypes of amyloidosis:

- AL amyloidosis – also called primary amyloidosis. It usually affects the heart, kidneys, liver, and nerves.
- AA amyloidosis – also known as secondary amyloidosis. This type is usually triggered by an inflammatory disease, such as rheumatoid arthritis.
- Hereditary amyloidosis – inherited type often affecting the nerves, heart, and kidneys.
- Wild-type amyloidosis – this type occurs when the TTR protein produces amyloid for unknown reasons. This type tends to affect men over age 70 and typically targets the heart.
- Localized amyloidosis – typical sites affected include the bladder, skin, throat, or lungs.

Signs and symptoms

May include swelling of ankles and legs, severe fatigue, shortness of breath, numbness and tingling of hands or feet, diarrhea, irregular heartbeat, enlarged tongue and skin changes.

Documentation guidance for amyloidosis

- Medical record documentation should include symptoms, diagnosis tests/labs ordered as well as interpretation of results when received, referrals to specialist, and treatment plan.
- Document complications and the organ it affects.
- Document underlying conditions that occur in association with certain varieties of amyloidosis.

ICD-10-CM Code information

ICD-10 Category E85

E85.0 Non-neuropathic heredofamilial amyloidosis
Code also associated disorders, such as:
- autoinflammatory syndromes (M04.-)
- Excludes transthyretin-related (ATTR)
  - familial amyloid cardiomyopathy (E85.4)

E85.1 Neuropathic heredofamilial amyloidosis

E85.2 Heredofamilial amyloidosis, unspecified

E85.3 Secondary systemic amyloidosis

E85.4 Organ-limited amyloidosis

E85.81 Light chain (AL) amyloidosis

E85.82 Wild-type transthyretin-related (ATTR) Amyloidosis

E85.89 Other amyloidosis

E85.9 Amyloidosis, unspecified

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Overview of Metabolic Syndrome (E88)

Metabolic Syndrome: Group of health risks that increase the likelihood of developing heart disease, stroke, and diabetes. These risks include certain parameters for blood pressure, cholesterol, and glucose levels.

Signs and symptoms

- Increased blood pressure
- High blood sugar
- Excess body fat around the waist
- Abnormal cholesterol or triglyceride levels

Causes

- Linked to overweight/obesity and inactivity
- Linked to insulin resistance

Documentation guidance for metabolic syndrome

Medical record documentation should include symptoms, diagnosis tests/labs ordered as well as interpretation of results when received, and referrals to specialist.

Document a treatment plan including counseling for prevention when patients are at risk of developing chronic diseases.

- Exercise.
- No smoking.
- Eating plenty of vegetables, fruits, lean proteins, and whole grains.
- Maintain a healthy weight.
- Restrict saturated fats and salt from diet.

ICD-10-CM Code information

ICD-10 Category E85

E88.810 Metabolic syndrome

E88.811 Insulin resistance syndrome, Type A

E88.818 Other insulin resistance

E88.819 Insulin resistance, unspecified

E88.89 Other specified metabolic disorders

- Excludes adult pulmonary Langerhans cell histiocytosis (J84.82)

E88.9 Metabolic disorder, unspecified

E88.A Wasting disease (syndrome) due to underlying condition

- Code first underlying condition
- Excludes cachexia NOS (R64) nutritional
- Marasmus (E41)
- Excludes failure to thrive (R62.51, R62.7)

References

- "ICD-10." Centers for Medicare & Medicaid Services, CMS.gov. http://www.cms.gov/medicare/coding/icd10