

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/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

Includes defects of catalase and peroxidase

E80.0	Hereditary erythropoietic porphyria	E80.3	Defects of catalase and peroxidase
E80.1	Porphyria cutanea tarda	E80.4	Gilbert syndrome
E80.2	Other and unspecified porphyria	E80.5	Crigler-Najjar syndrome
E80.20	Unspecified porphyria	E80.6	Other disorders of bilirubin metabolism
E80.21	Acute intermittent (hepatic) porphyria	E80.7	Disorder of bilirubin metabolism, unspecified
E80.29	Other porphyria		

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Overview 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/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

Excludes2 Alzheimer's disease (G30.0-)

E85.0	Non-neuropathic hereditary amyloidosis <i>Code also associated disorders, such as: autoinflammatory syndromes (M04.-) Excludes2 transthyretin-related (ATTR) familial amyloid cardiomyopathy (E85.4)</i>	E85.4	Organ-limited amyloidosis
E85.1	Neuropathic hereditary amyloidosis	E85.8	Other amyloidosis
E85.2	Hereditary amyloidosis, unspecified	E85.81	Light chain (AL) amyloidosis
E85.3	Secondary systemic 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/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 E88

Use additional code for associated conditions

Excludes1 histiocytosis X (chronic) (C96.6)

E88.8 Other specified metabolic disorders

E88.81 Metabolic syndrome and other insulin Resistance

Use additional codes for associated manifestations, such as: obesity (E66.-)

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

Excludes1 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

Excludes1 cachexia NOS (R64) nutritional marasmus (E41)

Excludes2 failure to thrive (R62.51, R62.7)

References

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