

Membranous Nephropathy (MN) ICD-10 CM Code Recommendation

Joseph Vassalotti, MD
Chief Medical Officer
National Kidney Foundation
Clinical Professor of Medicine
Icahn School of Medicine

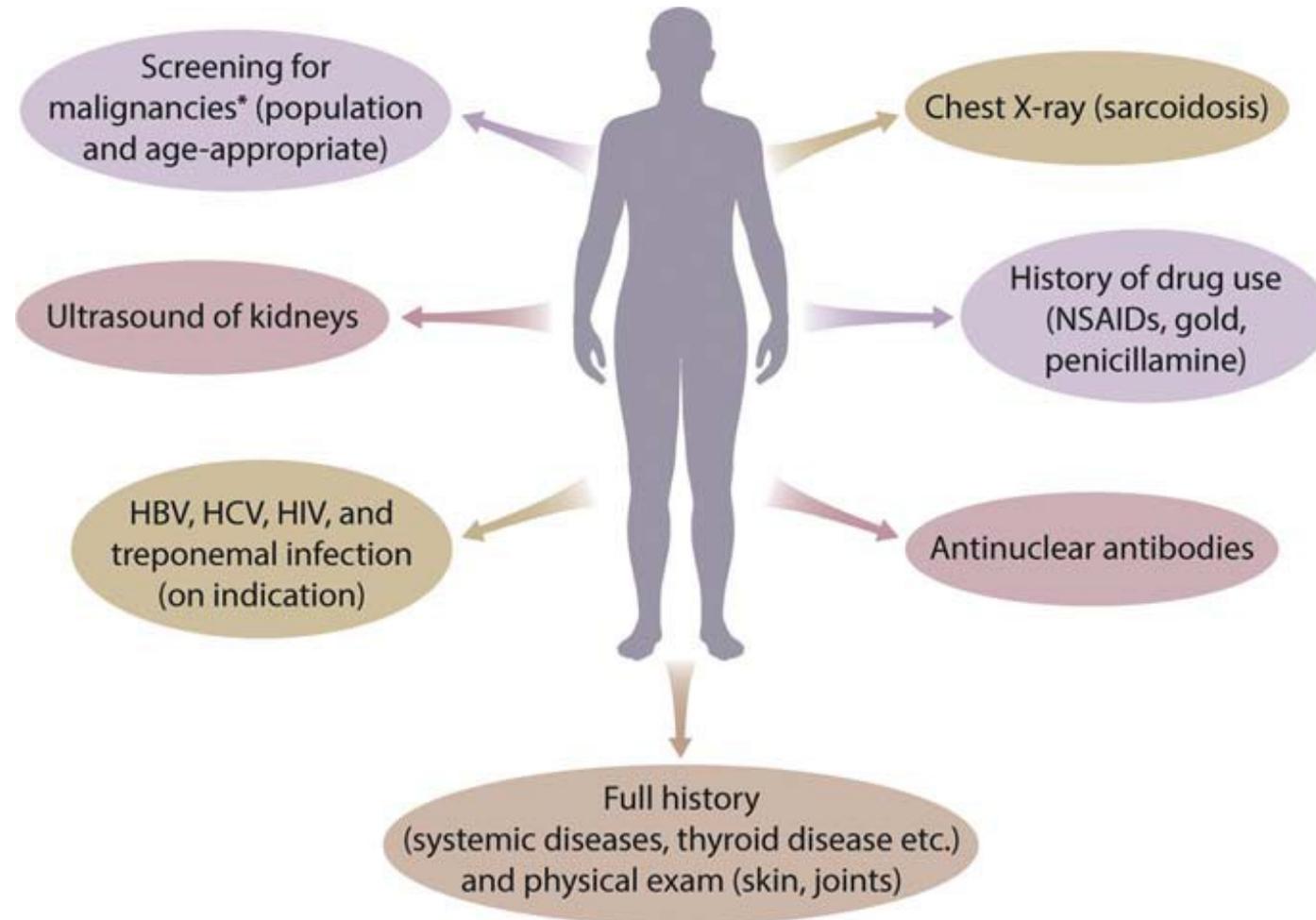
DEFINITION OF MEMBRANOUS NEPHROPATHY

- MN is an immune-mediated disease of the glomeruli
- It is one of the most common causes of nephrotic syndrome in Caucasian non-diabetic adults
- The term “membranous” reflects the hallmark histologic feature of the disease: thickening of the glomerular wall without significant cellular infiltration or proliferation

CATEGORIES OF MEMBRANOUS NEPHROPATHY

- MN is considered as either primary or secondary
- Primary MN (PMN) comprises about 80% of cases and is caused by circulating autoantibodies
- In secondary MN (about 20% of MN cases), an underlying cause is identified, such as autoimmune disease (e.g., lupus) malignancies, hepatitis B and C antigenemia, and certain drugs
 - No autoantibodies have been identified as causative agents in secondary MN

DISTINGUISHING PRIMARY FROM SECONDARY MEMBRANOUS NEPHROPATHY: EVALUATION FOR PATHOGENIC ANTIBODIES AND CLINICAL ASSESSMENT



EPIDEMIOLOGY

- Estimated incidence of PMN in U.S.:
 - 12/million/year
- The average age of onset of PMN:
 - Between 50 – 60 years
- Males are affected twice as frequently as females
 - Children rarely develop PMN
- U.S. incidence of end stage kidney disease (ESKD) due to PMN:
 - About 1.9 million/year

CLINICAL COURSE AND PROGNOSIS OF PRIMARY MN

- PMN usually progresses slowly but it is associated with high morbidity and mortality
- Progressive decline in the glomerular filtration rate (GFR) occurs in about 60% of patients
- About one-third of patients with PMN:
 - Experience spontaneous remission without treatment
 - Reach ESKD
 - Experience an indolent disease progression
- Without remission, nephrotic patients will likely progress to ESKD
- Conversely, PMN patients who achieve remission have an excellent prognosis

Overview of treatment of MN

Primary and secondary forms of MN have distinct treatment pathways

- Initial treatment for PMN involves antibody-targeted therapies, immunosuppressive agents, and supportive care for blood pressure control and proteinuria reduction
- Initial treatment for secondary MN follows the treatment pathways of the underlying condition
- Since treatment is often targeted at the etiologic cause in secondary MN, differential diagnosis is critical to distinguish between the two forms
- To reflect the process and time it takes to achieve a specific diagnosis in MN, a code for unspecified MN is also necessary

TREATMENT FOR PRIMARY MN

Initial, supportive therapy

Typical initial treatment starts with a conservative, non-specific, supportive approach

- ACE inhibitors and ARBs
 - Targeting reduction in albuminuria/proteinuria and controlling blood pressure,
- Statins
 - Cardiovascular risk reduction
- Anticoagulants
 - In selected patients at risk for thromboembolic events

TREATMENT FOR PRIMARY MN (cont'd)

First line therapy

For patients who do not respond to supportive therapy, first-line therapies for PMN aim to reduce the immune-related causes of the disease:

- Alkylating agents (cyclophosphamide, chlorambucil)
- B cell depletion (rituximab)
- Calcineurin inhibitors (cyclosporine, tacrolimus)
- Glucocorticoids
- Patients who cannot tolerate or are not responsive to these therapies may be given mycophenolate mofetil, ACTH, or plasma exchange

Rationale for ICD-10 -CM Code Revision

- Current ICD-10-CM coding for MN does not adequately capture the current clinical understanding of MN
- An advisory panel of expert clinicians, researchers, and patient educator/advocates indicated that:
 - Current codes do not capture MN as its own diagnosis and only represent it as an add-on with other conditions
 - The term used in the current codes; “diffuse membranous glomerulonephritis” is not used in clinical guidelines to describe MN, nor in typical clinical practice
- The revisions proposed here would update the existing code descriptions to be more clinically accurate, useful, and up-to-date with scientific advances
- This proposal builds on a proposal submitted in December 2021 and accounts for feedback received from the NCHS