CLINICAL PAYMENT AND CODING POLICY

If a conflict arises between a Clinical Payment and Coding Policy (CPCP) and any plan document under which a member is entitled to Covered Services, the plan document will govern. If a conflict arises between a CPCP and any provider contract pursuant to which a provider participates in and/or provides Covered Services to eligible member(s) and/or plans, the provider contract will govern. "Plan documents" include, but are not limited to, Certificates of Health Care Benefits, benefit booklets, Summary Plan Descriptions, and other coverage documents. BCBSTX may use reasonable discretion interpreting and applying this policy to services being delivered in a particular case. BCBSTX has full and final discretionary authority for their interpretation and application to the extent provided under any applicable plan documents.

Providers are responsible for submission of accurate documentation of services performed. Providers are expected to submit claims for services rendered using valid code combinations from Health Insurance Portability and Accountability Act (HIPAA) approved code sets. Claims should be coded appropriately according to industry standard coding guidelines including, but not limited to: Uniform Billing (UB) Editor, American Medical Association (AMA), Current Procedural Terminology (CPT®), CPT® Assistant, Healthcare Common Procedure Coding System (HCPCS), ICD-10 CM and PCS, National Drug Codes (NDC), Diagnosis Related Group (DRG) guidelines, Centers for Medicare and Medicaid Services (CMS) National Correct Coding Initiative (NCCI) Policy Manual, CCI table edits and other CMS guidelines.

Claims are subject to the code edit protocols for services/procedures billed. Claim submissions are subject to claim review including but not limited to, any terms of benefit coverage, provider contract language, medical policies, clinical payment and coding policies as well as coding software logic. Upon request, the provider is urged to submit any additional documentation.

Testing for Alpha-1 Antitrypsin Deficiency

Policy Number: CPCPLAB061

Version 1.0

Enterprise Medical Policy Committee Approval Date: 1/25/2022

Plan Effective Date: May 1, 2022

Description

BCBSTX has implemented certain lab management reimbursement criteria. Not all requirements apply to each product. Providers are urged to review Plan documents for eligible coverage for services rendered.

Reimbursement Information:

Serum Testing

1. Serum quantification of alpha-1 antitrypsin (A1AT) protein and/or A1AT phenotyping by isoelectric focusing or A1AT proteotyping (Pi-typing or protease inhibitor typing) for Z and S alleles by liquid chromatography-tandem mass spectrometry **may be reimbursable** in the following situations:
    a. Symptomatic adults with emphysema, COPD or asthma
    b. Individuals with unexplained liver disease
    c. Individuals with persistent obstruction on pulmonary function tests without identifiable risk factors (e.g., cigarette smoking, occupational exposure)
    d. Adults with necrotizing panniculitis
    e. Siblings of an individual with known alpha-1 antitrypsin (AAT) deficiency
    f. Individuals with anti-proteinase three-positive vasculitis (C-ANCA [anti-neutrophil cytoplasmic antibody]-positive vasculitis)
    g. Individuals with bronchiectasis without evident etiology

2. Isoelectric focusing/phenotyping **may be reimbursable** when there is strong suspicion of the disease based on laboratory testing and symptoms and individual has a negative genotype testing for common variants or discordant results between A1AT serum levels and proteotype.

3. Testing for alpha-1 antitrypsin deficiency **is not reimbursable** in all other situations.

Note:
In 2003, the American Thoracic Society published recommendations on the diagnosis and management of individuals with AAT deficiency.

Recommendations were classified as follows:
Type A: Genetic testing is recommended
Type B: Genetic testing should be discussed and could be accepted or declined
Type C: Genetic testing is not recommended, i.e., should not be encouraged
Type D: Recommend against genetic testing, i.e., should be discouraged

Type A recommendations for diagnostic testing in the following situations:

1. Symptomatic adults with emphysema, COPD or asthma with airflow obstruction that is not completely reversible with aggressive treatment with bronchodilators
2. Individuals with unexplained liver disease
3. Asymptomatic individuals with persistent obstruction on pulmonary function tests with identifiable risk factors (e.g., cigarette smoking, occupational exposure)
4. Adults with necrotizing panniculitis
5. Siblings of an individual with known alpha-1 antitrypsin (AAT) deficiency

Type B recommendations for diagnostic testing in the following situations:

1. Adults with bronchiectasis without evidence etiology
2. Adolescents with persistent airflow obstruction
3. Asymptomatic individuals with persistent airflow obstruction and no risk factors
4. Adults with C-ANCA positive (anti-proteinase 3-positive) vasculitis

5. Individuals with a family history of COPD or liver disease not known to be attributed to AAT deficiency
6. Distant relatives of an individual who is homozygous for AAT deficiency
7. Offspring or parents of an individual with homozygous AAT deficiency
8. Siblings, offspring, parents, or distant relatives of an individual who is heterozygous for AAT deficiency
9. Individuals at high risk of having AAT deficiency-related diseases
10. Individuals who are not at risk themselves of having AAT deficiency but who are partners of individuals who are homozygous or heterozygous for AAT deficiency

Type C recommendations for diagnostic testing in the following situations:

1. Adults with asthma in whom airflow obstruction is completely reversible
2. Pre-dispositional testing
3. Population screening of smokers with normal spirometry

Type D recommendations for diagnostic testing in the following situations:

1. Pre-dispositional fetal testing
2. Population screening of either neonates, adolescents, or adults*

* Population screening is not recommended currently. However, a possible exception (type B recommendation) may apply in countries satisfying all three of the following conditions:

(1) the prevalence of AAT deficiency is high (about 1/1,500, or more);
(2) smoking is prevalent; and
(3) adequate counseling services are available.

According to the 2003 joint statement on diagnosis and management of alpha-1 antitrypsin deficiency by the American Thoracic Society/European Respiratory Society:

The following features should prompt suspicion by physicians that their patient may be more likely to have AAT deficiency:

Clinical Factors

1. Early-onset emphysema (age of 45 years or less)
2. Emphysema in the absence of a recognized risk factor (smoking, occupational dust exposure, etc.)
3. Emphysema with prominent basilar hyperlucency
4. Otherwise, unexplained liver disease
5. Necrotizing panniculitis
6. Anti-proteinase three-positive vasculitis (C-ANCA [anti-neutrophil cytoplasmic antibody]-positive vasculitis)
7. Bronchiectasis without evident etiology

Procedure Codes

Codes
82103, 82104, 82542, 83789

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Policy Update History:

5/1/2022	New policy