

If a conflict arises between a Clinical Payment and Coding Policy 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. Blue Cross and Blue Shield of Texas 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 approved code sets. Claims should be coded appropriately according to industry standard coding guidelines including, but not limited to: Uniform Billing Editor, American Medical Association, Current Procedural Terminology, CPT® Assistant, Healthcare Common Procedure Coding System, ICD-10 CM and PCS, National Drug Codes, Diagnosis Related Group guidelines, Centers for Medicare and Medicaid Services National Correct Coding Initiative 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

**Approval Date:** February 5, 2025

**Plan Effective Date:** May 15, 2025

## Description

The plan 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:

1. For individuals who are suspected of having alpha-1 antitrypsin (AAT) deficiency, serum quantification of alpha-1 antitrypsin (AAT) protein **and** AAT phenotyping **or** AAT proteotyping (see **NOTE 1**) **may be reimbursable** once per lifetime in **any** of the following situations:
  - a. For symptomatic individuals 18 years of age or older with emphysema, COPD, or asthma
  - b. For individuals with unexplained liver disease (e.g., chronic hepatitis with or without cirrhosis, chronically elevated aminotransferase levels, portal hypertension, primary liver cancer)
  - c. For individuals with persistent obstruction on pulmonary function tests without identifiable risk factors (e.g., cigarette smoking, occupational exposure)
  - d. For individuals 18 years of age or older with necrotizing panniculitis
  - e. For the siblings of an individual with known alpha-1 antitrypsin (AAT) deficiency
  - f. For individuals with anti-proteinase three-positive vasculitis (C-ANCA [anti-neutrophil cytoplasmic antibody]-positive vasculitis)
  - g. For individuals with bronchiectasis without evident etiology
  - h. For individuals with neonatal cholestasis
2. For individuals who have negative genotype results for common variants or who have discordant results between AAT serum levels and proteotyping, but for whom a clinical suspicion of AAT deficiency remains, isoelectric focusing/phenotyping **may be reimbursable**.
3. For all other situations not described above, testing for alpha-1 antitrypsin (AAT) deficiency **is not reimbursable**.

### **Note 1:**

AAT phenotyping should be performed using isoelectric focusing. AAT proteotyping (Pi-typing or protease inhibitor typing) for Z and S alleles should be performed using liquid chromatography-tandem mass spectrometry.

## Procedure Codes

The following is not an all-encompassing code list. The inclusion of a code does not guarantee it is a covered service or eligible for reimbursement.

Codes
82103, 82104, 82542, 83789

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### Policy Update History:

Approval Date	Effective Date; Summary of Changes
02/05/2025	05/15/2025; Document updated with literature review. The following changes were made to Reimbursement Information: Added “once per lifetime” to #1. Now reads: “1) For individuals who are suspected of having alpha-1 antitrypsin (AAT) deficiency, serum quantification of alpha-1 antitrypsin (AAT) protein and AAT phenotyping or AAT proteotyping (see Note 1) may be reimbursable once per lifetime in any of the following situations:” Edited #1.b. to include examples of unexplained liver disease. Now reads: “b) For individuals with unexplained liver disease (e.g., chronic hepatitis with or without cirrhosis, chronically elevated aminotransferase levels, portal hypertension, primary liver cancer).” New #1.h.: “h) For individuals with neonatal cholestasis. References revised.
09/13/2024	01/01/2025: New policy.