AAVBC
AMERICAN ACADEMY OF VALUE BASED CARE

Addison's Disease

Quick Reference Guide

2026

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1 CLINICAL SNAPSHOT

Definition: Addison's disease (primary adrenal insufficiency, PAI) is a chronic, life-limiting **endocrine disorder** in which destruction of the adrenal cortex produces deficient cortisol and, in most primary cases, deficient aldosterone.[1] In North America and Europe roughly **90%** of cases are autoimmune (autoimmune adrenalitis); the remainder follow **infection** (historically tuberculosis, now also HIV and fungal disease), **hemorrhage, infiltrative disease, or bilateral adrenalectomy**.[2] Because the adrenal glands lose most of their reserve **before** overt symptoms appear, the presentation is **insidious** and the **stress-response axis fails silently** — any physiologic stressor can then precipitate a **life-threatening adrenal crisis.**[3,4] Addison's must be conveyed as **dual-hormone** loss: it is not simply low cortisol, and most patients require both glucocorticoid and mineralocorticoid replacement.[5]

CLINICAL PEARL: PRIMARY VS. SECONDARY ADRENAL INSUFFICIENCY

In **primary** adrenal insufficiency (Addison's disease), the adrenal gland itself is destroyed, so both **cortisol and aldosterone** are deficient, and **ACTH is elevated** due to loss of negative feedback - often producing **hyperpigmentation**.

In **secondary** adrenal insufficiency, the defect lies in the pituitary or hypothalamus (deficient ACTH or CRH), so **aldosterone production is preserved** (regulated independently by the renin-angiotensin system), **ACTH is low**, and hyperpigmentation is absent. This distinction matters clinically: only primary adrenal insufficiency requires mineralocorticoid replacement.

ICD-10 Codes:

Confirmed Addison's disease is reported as **E27.1** (primary adrenocortical insufficiency), the standard code for autoimmune or other primary adrenal-gland failure. Acute decompensation is **E27.2** (Addisonian/adrenal crisis)and, when a crisis is the reason for the encounter, **E27.2** is sequenced as the principal diagnosis. Insufficiency from exogenous steroid withdrawal is **E27.3** (drug-induced), with an added code for the causative drug. **E27.40** (unspecified) and **E27.49** (other) should be reserved for genuinely undetermined cases. Secondary (pituitary) adrenal insufficiency is a distinct disorder coded **E23.0** — pituitary ACTH failure, not adrenal-gland failure — and does not require mineralocorticoid replacement. Autoimmune polyglandular failure is **E31.0**, and **A18.7** (tuberculosis of the adrenal gland) is coded in addition to **E27.1** when relevant. Long-term steroid replacement is documented with **Z79.52**.[6]

Prevalence and Burden: Addison's disease occurs equally in males and females, affecting an estimated 1 in 100,000 people in the United States. Overall prevalence is estimated at **40-60 cases per million** in the general population, though underdiagnosis makes true prevalence difficult to determine. While Addison's disease can occur at any age, it most commonly presents between **30-50 years**.[7] The Under-Recognition Problem: Diagnostic delay is the central clinical failure. Up to **50% of patients first present in adrenal crisis**, about **20% report nonspecific symptoms for more than 5 years** before diagnosis, and **40% are diagnosed more than 6 months** after symptom onset — with fewer than 30% of women diagnosed within the first 6 months.[8]