

cases. A plasma aldosterone:PRA ratio of >50, due to suppression of circulating renin and an elevation of circulating aldosterone, is suggestive of hyperaldosteronism. Patients with hyperaldosteronism or apparent mineralocorticoid excess may require further testing, for example, adrenal vein sampling ([Chap. 386](#)) or the clinically available testing for specific genetic causes (e.g., FH-I, SAME, Liddle's syndrome). Patients with primary aldosteronism should thus be tested for the chimeric FH-I/GRA gene (see above) if they are younger than 20 years of age or have a family history of primary aldosteronism or stroke at a young age (<40 years). Preliminary differentiation of Liddle's syndrome due to mutant ENaC channels from SAME due to mutant 11 β HSD-2 (see above), both of which cause hypokalemia and hypertension with aldosterone suppression, can be made on a clinical basis and then confirmed by genetic analysis; patients with Liddle's syndrome should respond to amiloride (ENaC inhibition) but not spironolactone, whereas patients with SAME will respond to spironolactone.

TREATMENT

Hypokalemia

The goals of therapy in hypokalemia are to prevent life-threatening and/or serious chronic consequences, to replace the associated K⁺ deficit, and to correct the underlying cause and/or mitigate future hypokalemia. The urgency of therapy depends on the severity of hypokalemia, associated clinical factors (e.g., cardiac disease, digoxin therapy), and the rate of decline in serum K⁺. Patients with a prolonged QT interval and/or other risk factors for arrhythmia should be monitored by continuous cardiac telemetry during repletion.

Urgent but cautious K⁺ replacement should be considered in patients with severe redistributive hypokalemia (plasma K⁺ concentration <2.5 mM) and/or when serious complications ensue; however, this approach has a risk of rebound hyperkalemia following acute resolution of the underlying cause. When excessive activity of the sympathetic nervous system is thought to play a dominant role in redistributive hypokalemia, as in TPP, theophylline