Table 36-2 Causes of Hyperkalemia

Spurious laboratory value	Acquired Addison disease
Hemolysis	21-Hydroxylase deficiency
Tissue ischemia during blood drawing	3β-Hydroxysteroid-dehydrogenase deficiency
Thrombocytosis	Lipoid congenital adrenal hyperplasia
Leukocytosis	Adrenal hypoplasia congenita
Increased intake	Aldosterone synthase deficiency
IV or PO	Adrenoleukodystrophy
Blood transfusions	Hyporeninemic hypoaldosteronism
Transcellular shifts	Urinary tract obstruction
Acidemia	Sickle cell disease
Rhabdomyolysis	Kidney transplant
Tumor lysis syndrome	Lupus nephritis
Tissue necrosis	Renal tubular disease
Hemolysis/hematomas/gastrointestinal bleeding	Pseudohypoaldosteronism type 1
Succinylcholine	Pseudohypoaldosteronism type 2
Digitalis intoxication	Urinary tract obstruction
Fluoride intoxication	Sickle cell disease
β-Adrenergic blockers	Kidney transplant
Exercise	Medications
Hyperosmolality	ACE inhibitors
Insulin deficiency	Angiotensin II blockers
Malignant hyperthermia	Potassium-sparing diuretics
Hyperkalemic periodic paralysis	Cyclosporine
Decreased excretion	NSAIDs
Renal failure	Trimethoprim
Primary adrenal disease	

ACE, Angiotensin-converting enzyme; *IV*, intravenous; *NSAIDs*, nonsteroidal anti-inflammatory drugs; *PO*, oral.

potassium content of stored blood. Increased intake may precipitate hyperkalemia if there is an underlying defect in potassium excretion.

The intracellular space has a high potassium concentration, so a **shift of potassium from the intracellular space** to the extracellular space can have a significant impact on the plasma potassium. This shift occurs with acidosis, cell destruction (rhabdomyolysis or tumor lysis syndrome), insulin deficiency, medications (succinylcholine, β-blockers), malignant hyperthermia, and hyperkalemic periodic paralysis.

Hyperkalemia secondary to decreased excretion occurs with renal insufficiency. Aldosterone deficiency or unresponsiveness to aldosterone causes hyperkalemia, often with associated metabolic acidosis (see Chapter 37) and hyponatremia. A form of congenital adrenal hyperplasia, **21-hydroxylase deficiency,** is the most frequent cause of aldosterone deficiency in children. Male infants typically present with hyperkalemia, metabolic acidosis, hyponatremia, and volume depletion. Female infants with this disorder usually are diagnosed as newborns because of ambiguous genitalia.

Renin, via angiotensin II, stimulates aldosterone production. A deficiency in renin, resulting from kidney damage, can lead to decreased aldosterone production. These patients typically have hyperkalemia and a metabolic acidosis, without hyponatremia. Some patients have impaired renal function, partially accounting for the hyperkalemia, but the impairment in potassium excretion is more extreme than expected for the degree of renal insufficiency.

Children with **pseudohypoaldosteronism type 1** have hyperkalemia, metabolic acidosis, and salt wasting, leading to hyponatremia and volume depletion; aldosterone levels are elevated. In the autosomal recessive variant, there is a defect in the renal sodium channel that is normally activated by aldosterone. In the autosomal dominant form, patients have a defect in the aldosterone receptor, and the disease is milder, often remitting in adulthood. Pseudohypoaldosteronism type 2, also called **Gordon syndrome,** is an autosomal dominant disorder characterized by hypertension secondary to salt retention and impaired excretion of potassium and acid leading to hyperkalemia and metabolic acidosis. The risk of hyperkalemia secondary to medications that decrease renal potassium excretion is greatest in patients with underlying renal insufficiency.

Clinical Manifestations

The most import effects of hyperkalemia are due to the role of potassium in membrane polarization. The cardiac conduction