

FIGURE 3-30

Mechanism of hyperkalemia in pseudohypoaldosteronism type I (PHA I). This rare autosomally transmitted disease is characterized by neonatal dehydration, failure to thrive, hyponatremia, hyperkalemia, and metabolic acidosis. Kidney and adrenal function are normal, and patients do not respond to exogenous mineralocorticoids. Genetic mutations responsible for PHA I occur in the  $\alpha$  and  $\beta$  subunits of the amiloride-sensitive sodium channel of the collecting tubule. Frameshift or premature stop codon mutations in the cytoplasmic amino terminal or extracellular loop of either subunit disrupt the integrity of the sodium channel and result in loss of channel activity. Failure to reabsorb sodium results in volume depletion and activation of the renin-aldosterone axis. Furthermore, since sodium reabsorption is indirectly coupled to potassium and hydrogen ion secretion, hyperkalemia and metabolic acidosis ensue. Interestingly, when mutations are introduced into the cytoplasmic carboxyl terminal, sodium channel activity is increased and Liddle's syndrome is observed [4].

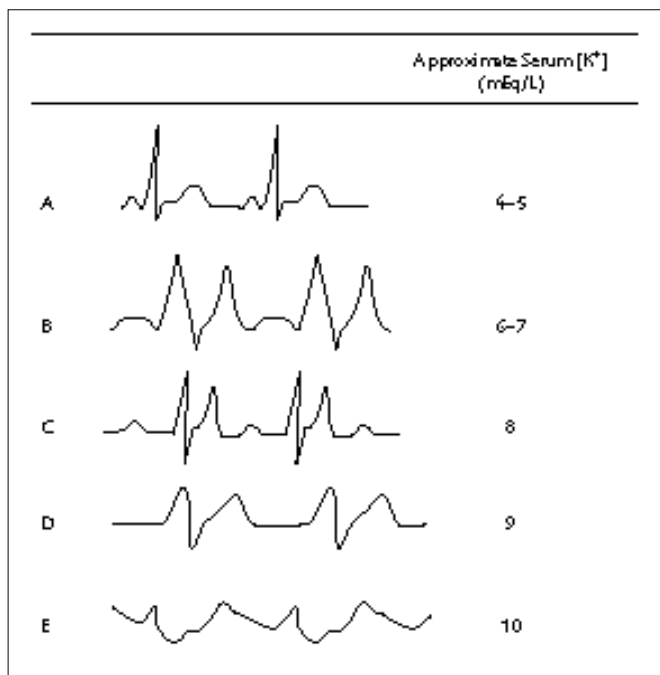
## Hyperkalemia: Clinical Manifestations

### CLINICAL MANIFESTATIONS OF HYPERKALEMIA

Cardiac	Renal electrolyte
Abnormal electrocardiogram	Decreased renal $NH_4^+$ production
Atrial/ventricular arrhythmias	Natriuresis
Pacemaker dysfunction	Endocrine
Neuromuscular	Increased aldosterone secretion
Paresthesias	Increased insulin secretion
Weakness	
Paralysis	

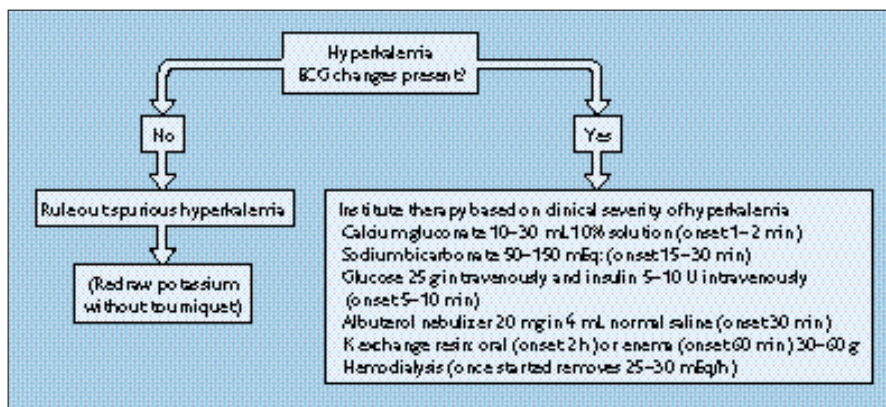
FIGURE 3-31

Clinical manifestations of hyperkalemia.

**FIGURE 3-32**

Electrocardiographic (ECG) changes associated with hyperkalemia. **A**, Normal ECG pattern. **B**, Peaked, narrow-based T waves are the earliest sign of hyperkalemia. **C**, The P wave broadens and the QRS complex widens when the plasma potassium level is above 7 mEq/L. **D**, With higher elevations in potassium, the P wave becomes difficult to identify. **E**, Eventually, an undulating sinusoidal pattern is evident. Although the ECG changes are depicted here as correlating to the severity of hyperkalemia, patients with even mild ECG changes may abruptly progress to terminal rhythm disturbances. Thus, hyperkalemia with any ECG changes should be treated as an emergency.

## Hyperkalemia: Treatment

**FIGURE 3-33**

Treatment of hyperkalemia.

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