Nursing Alert

The parents should be advised that there is no physical harm in treating for suspected adrenal insufficiency that is not present, but the consequence of not treating acute adrenal insufficiency can be fatal.

Pheochromocytoma

Pheochromocytoma is a rare tumor characterized by secretion of catecholamines. The tumor most commonly arises from the chromaffin cells of the adrenal medulla but may occur wherever these cells are found, such as along the paraganglia of the aorta or thoracolumbar sympathetic chain. In children, they are frequently bilateral or multiple and are generally benign. Often there is a familial transmission of the condition as an autosomal dominant trait (White, 2016b).

The clinical manifestations of pheochromocytoma are caused by an increased production of catecholamines, producing hypertension, tachycardia, headache, decreased gastrointestinal activity and resulting constipation, increased metabolism with anorexia, weight loss, hyperglycemia, polyuria, polydipsia, hyperventilation, nervousness, heat intolerance, and diaphoresis. In severe cases, signs of congestive heart failure are evident.

Diagnostic Evaluation

The clinical manifestations mimic those of other disorders, such as hyperthyroidism or DM. Usually the tumor is identified by computed tomography (CT) scan or MRI. Definitive tests include 24-hour measurement of urinary levels of the catecholamine metabolites, histamine stimulation, and α-adrenergic blocking agents.

Therapeutic Management

Definitive treatment consists of surgical removal of the tumor. In children, the tumors may be bilateral, requiring a bilateral adrenalectomy and lifelong glucocorticoid and mineralocorticoid therapy. The major complications that can occur during surgery are