

The adrenal cortex produces three classes of corticosteroid hormones: glucocorticoids (e.g., cortisol), mineralocorticoids (e.g., aldosterone), and adrenal androgen precursors (e.g., dehydroepiandrosterone [DHEA]) (Fig. 406-1). Glucocorticoids and mineralocorticoids act through specific nuclear receptors, regulating aspects of the physiologic stress response as well as blood pressure and electrolyte homeostasis. Adrenal androgen precursors are converted in the gonads and peripheral target cells to sex steroids that act via nuclear androgen and estrogen receptors.

Disorders of the adrenal cortex are characterized by deficiency or excess of one or several of the three major corticosteroid classes. Hormone deficiency can be caused by inherited glandular or enzymatic disorders or by destruction of the pituitary or adrenal gland by autoimmune disorders, infection, infarction, or iatrogenic events such as surgery or hormonal suppression. Hormone excess is usually

the result of neoplasia, leading to increased production of adrenocorticotropic hormone (ACTH) by the pituitary or neuroendocrine cells (ectopic ACTH) or increased production of glucocorticoids, mineralocorticoids, or adrenal androgen precursors by adrenal nodules. Adrenal nodules are increasingly identified incidentally during abdominal imaging performed for other reasons.

ADRENAL ANATOMY AND DEVELOPMENT

The normal adrenal glands weigh 6–11 g each. They are located above the kidneys and have their own blood supply. Arterial blood flows initially to the subcapsular region and then meanders from the outer cortical zona glomerulosa through the intermediate zona fasciculata to the inner zona reticularis and eventually to the adrenal medulla. The right suprarenal vein drains directly into the vena cava, while the left suprarenal vein drains into the left renal vein.

During early embryonic development, the adrenals originate from the urogenital ridge and then separate from gonads and kidneys at about the sixth week of gestation. Concordant with the time of sexual differentiation (seventh to ninth week of gestation, Chap. 410), the adrenal cortex starts to produce cortisol and the adrenal sex steroid

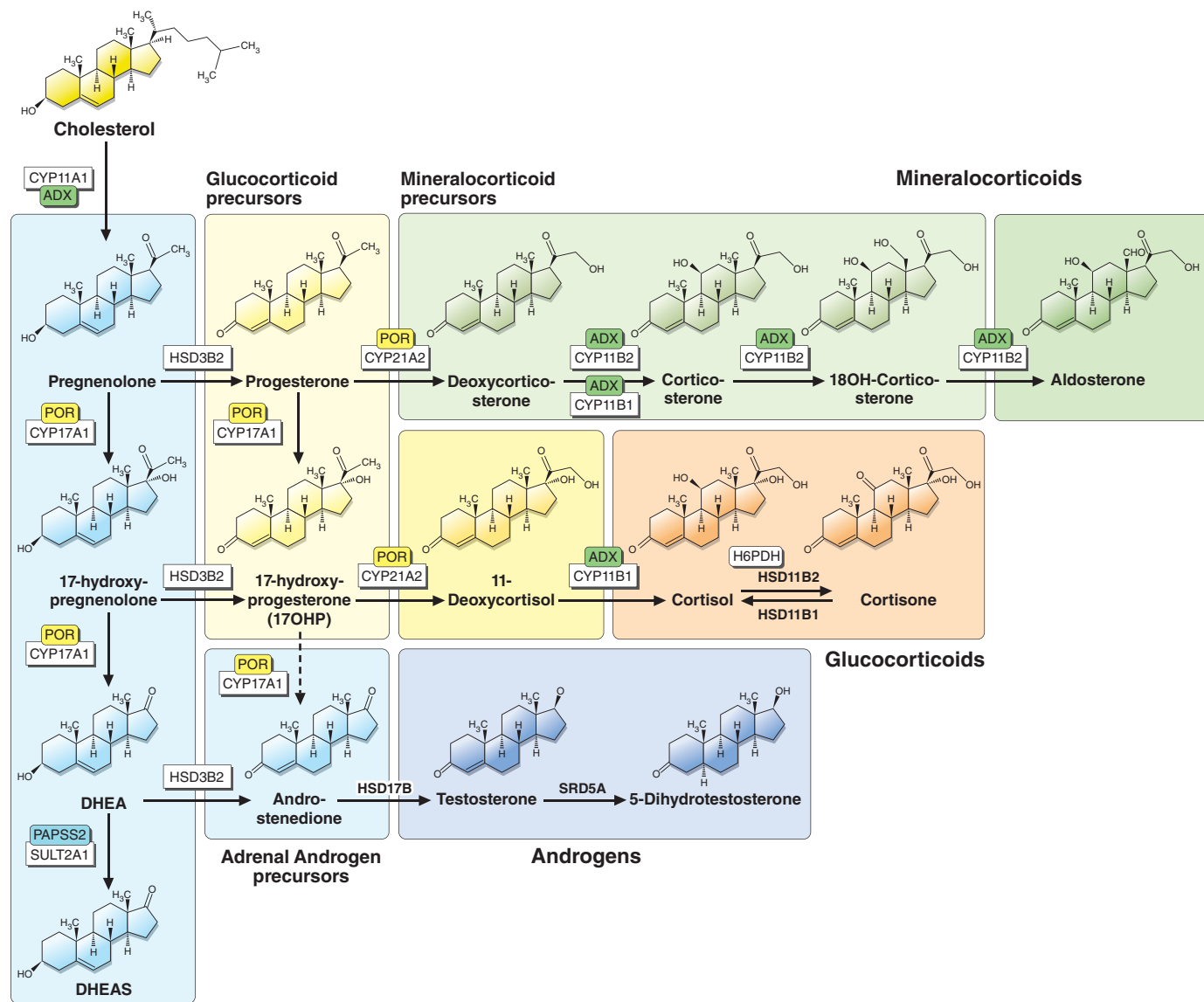


FIGURE 406-1 Adrenal steroidogenesis. ADX, adrenodoxin; CYP11A1, side chain cleavage enzyme; CYP11B1, 11 β -hydroxylase; CYP11B2, aldosterone synthase; CYP17A1, 17 α -hydroxylase/17,20 lyase; CYP21A2, 21-hydroxylase; DHEA, dehydroepiandrosterone; DHEAS, dehydroepiandrosterone sulfate; H6PDH, hexose-6-phosphate dehydrogenase; HSD11B1, 11 β -hydroxysteroid dehydrogenase type 1; HSD11B2, 11 β -hydroxysteroid dehydrogenase type 2; HSD17B, 17 β -hydroxysteroid dehydrogenase; HSD3B2, 3 β -hydroxysteroid dehydrogenase type 2; PAPSS2, PAPS synthase type 2; POR, P450 oxidoreductase; SRD5A, 5 α -reductase; SULT2A1, DHEA sulfotransferase.