



Figure 1. Pathways of adrenal steroid synthesis. Enzymes and encoding genes are indicated at arrows. When an enzymatic defect occurs, steroids proximal to the defect increase and are frequently shunted to other products, and steroids distal to the defect decrease (eg, a deficiency of 11 $\beta$ -hydroxylase will cause increased levels of deoxycorticosterone, progesterone, 11-deoxycortisol, and 17-hydroxyprogesterone and decreased levels of corticosterone, aldosterone, and cortisol). 17 $\alpha$ , 17 $\alpha$ -hydroxylase; 17,20, 17,20-lyase; 3 $\beta$ , 3 $\beta$ -hydroxysteroid dehydrogenase; 21, 21-hydroxylase; 11 $\beta$ , 11 $\beta$ -hydroxylase; A, two-step process of aldosterone synthesis: 1) hydroxylation of corticosterone to form 18-hydroxycorticosterone, 2) oxidation of 18-hydroxycorticosterone to form aldosterone; DHEA, dihydroepiandrosterone; DHEAS, dihydroepiandrosterone sulfate.

<sup>a</sup> StAR, steroid acute regulatory protein: transports cholesterol from the outer to inner mitochondrial membrane.