

ADRENAL HYPERPLASIA V

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HORMONE – PRODUCING CELLS OF THE ADRENAL GLAND

- adrenal glands located atop the kidneys
- adrenal glands include two major regions: medulla and cortex
- cortex contains 3 zones each producing a primary hormone endproduct:
 - glomerulosa – produces aldosterone, a mineralocorticoid
 - promote reuptake of sodium (salt retention) by the kidney into the blood (see Fig. 4)
 - more blood sodium leads to water retention to maintain blood pressure
 - fasciculata – produces cortisol, a glucocorticoid (Fig. 1)
 - helps the body respond to chronic stress and food deprivation overnight
 - increases availability of glucose (blood sugar) and fats in blood as fuels for body functions
 - reticularis – produces androstenedione, an androgen (Fig.2)
 - associated with male sex characteristics
 - males: most androgen (testosterone) produced in testes with much lesser amounts in adrenals
 - females: adrenal is the only tissue in which androgens are produced

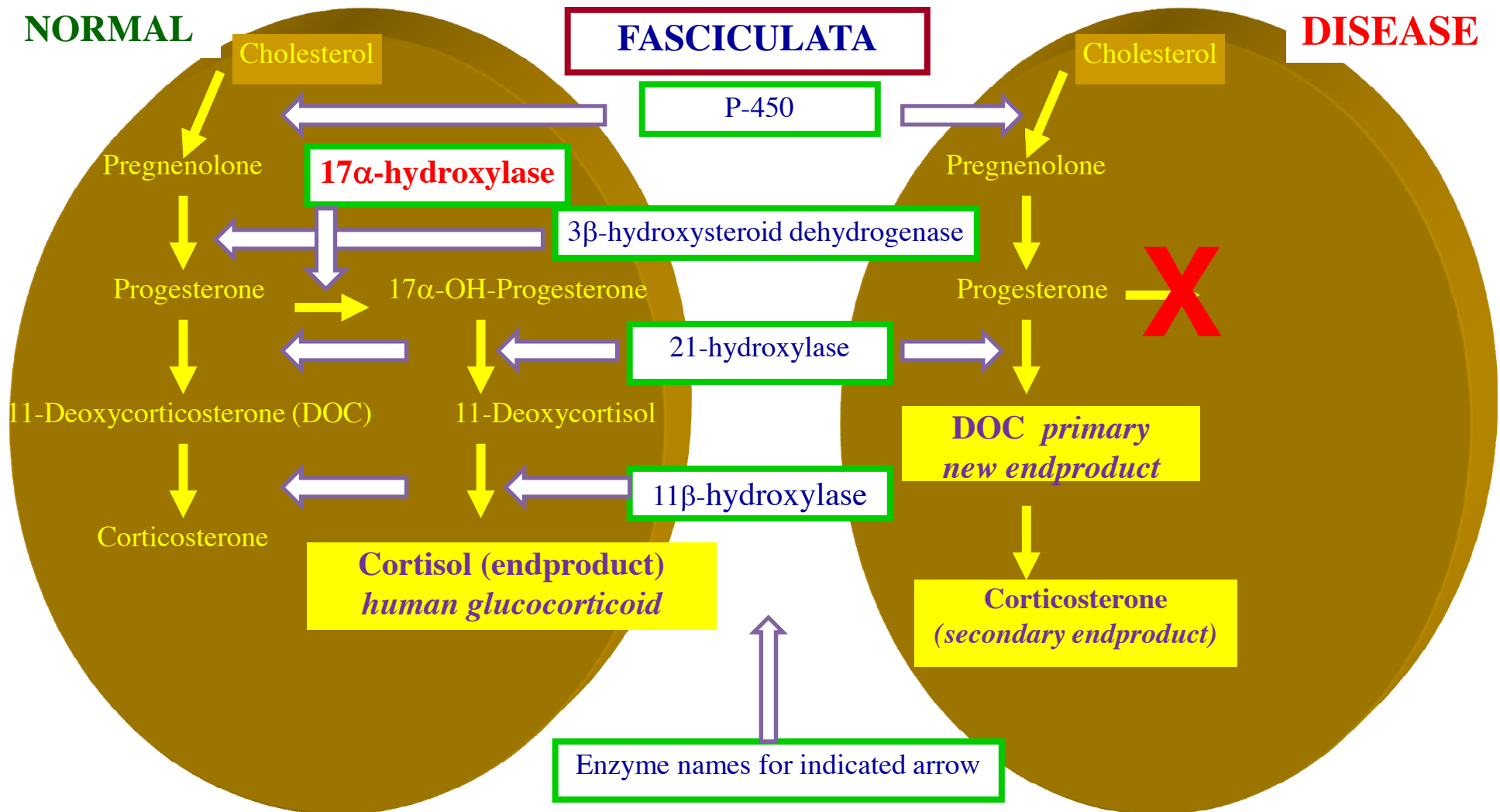


Figure 1. Cholesterol in fasciculata normally (left) yields **cortisol as endproduct** following **ACTH signal**. In adrenal hyperplasia V (right), defective **17α-OHase** prevents cortisol formation (**X**). DOC becomes the primary endproduct. **DOC acts like aldosterone (mineralocorticoid) causing excessive salt retention and hypertension without normal control** (see Figures 3 and 4). Without **cortisol** (glucocorticoid), the patient **responds poorly to chronic stress and food deprivation** overnight. Patients must be treated with replacement glucocorticoid.

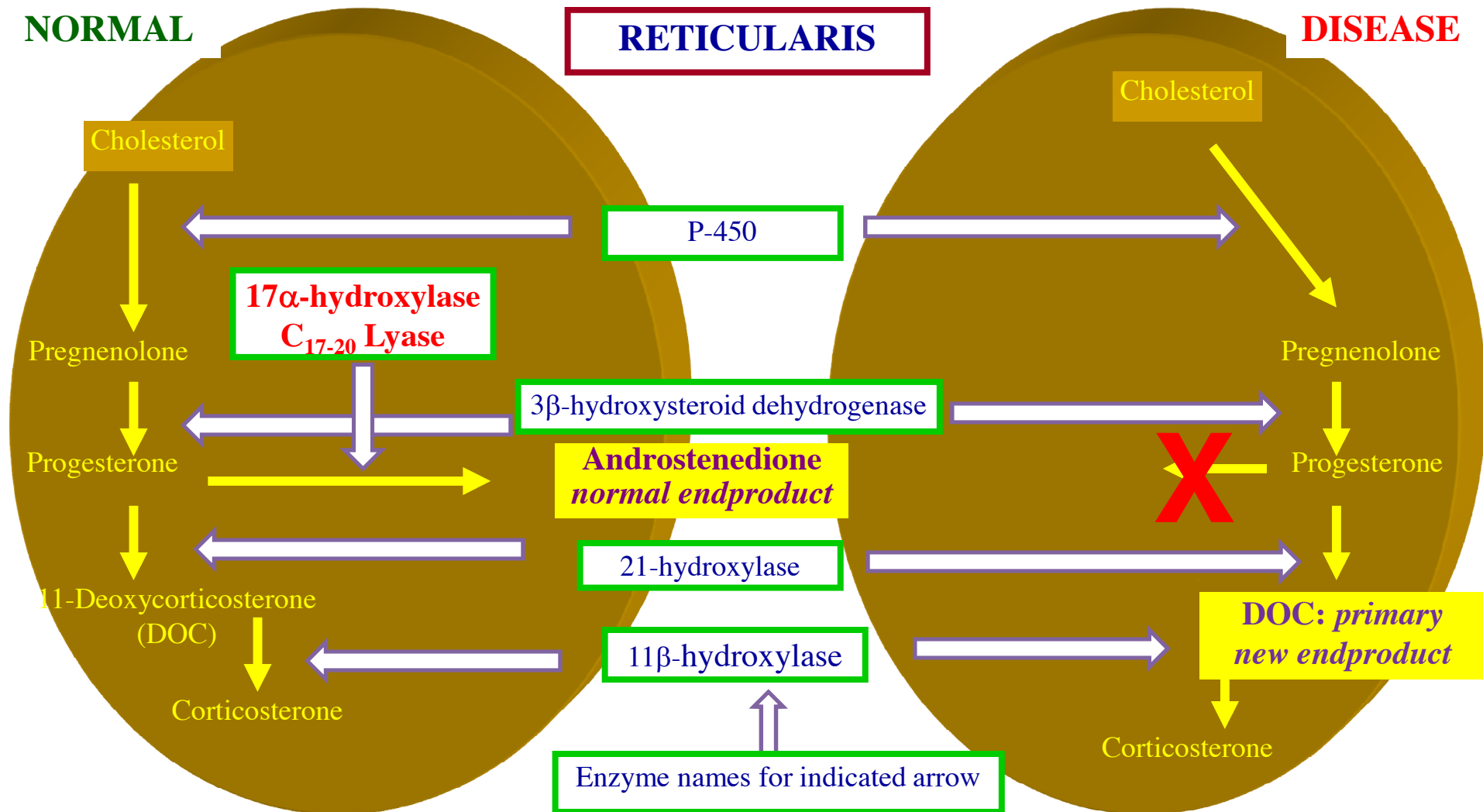
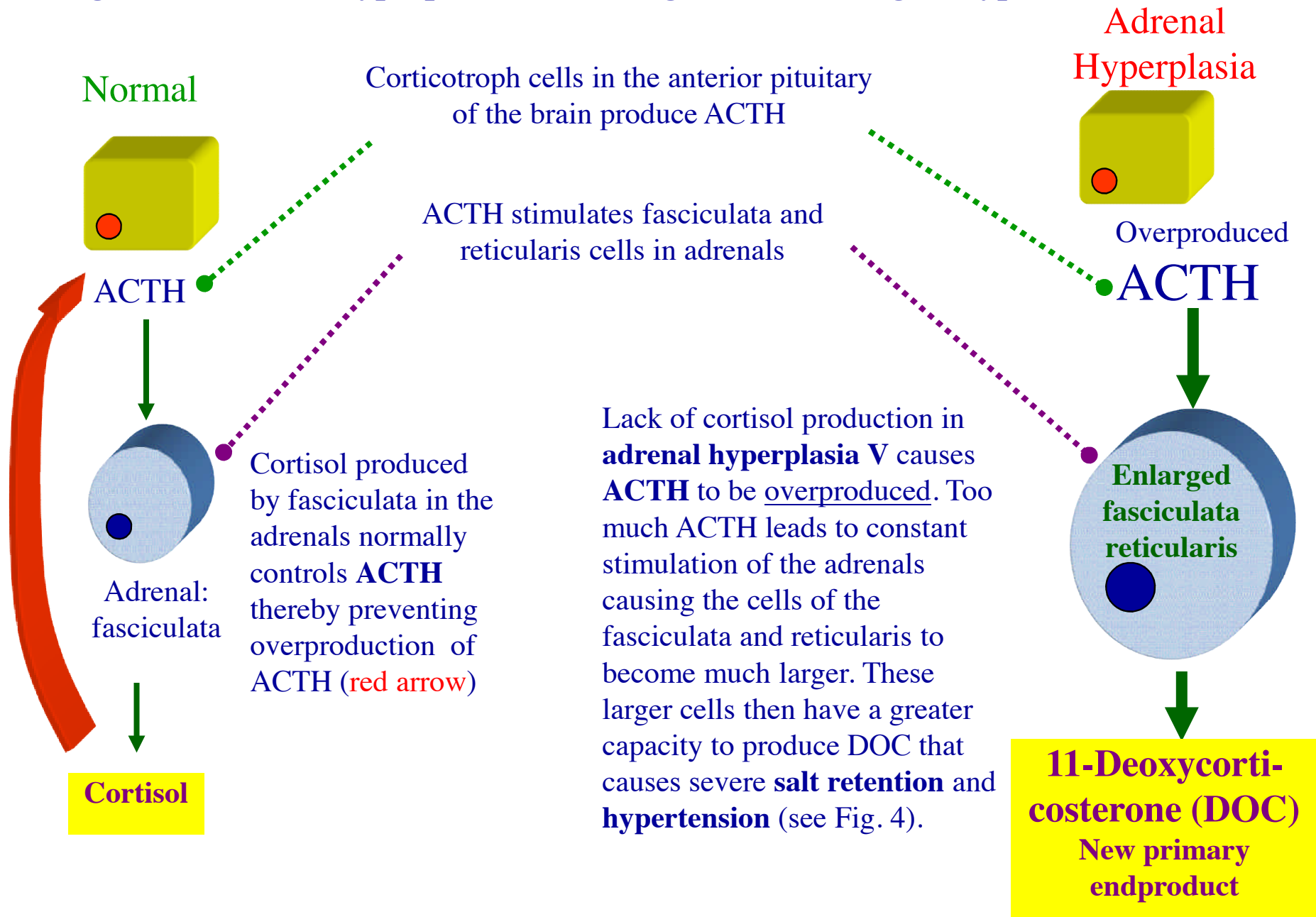


Figure 2. Cholesterol in reticularis normally (left) yields **androstenedione as endproduct** following **ACTH signal**. In adrenal hyperplasia IV (right), defective **17 α -OHase/C_{17,20}Lyase** prevents formation of androstenedione (**X**) and DOC becomes a primary endproduct. **DOC acts like mineralocorticoid causing excessive salt retention and hypertension without normal control** (see Figures 3 and 4).

Figure 3. Adrenal Hyperplasia (cell enlargement) Leading to Hypertension



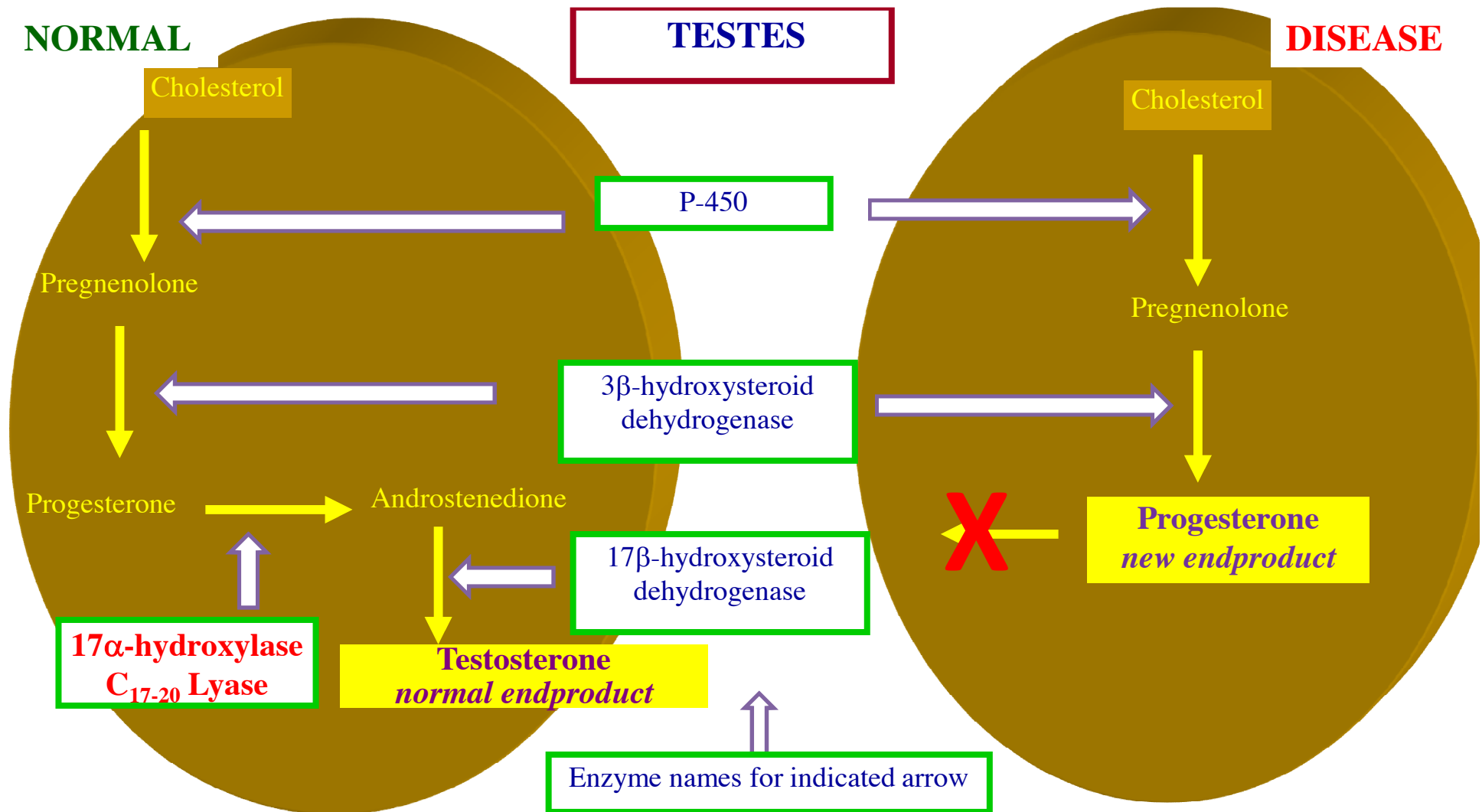


Figure 5. Cholesterol in testes normally (left) yields **testosterone as endproduct** following **luteinizing hormone signal**. In adrenal hyperplasia II (right), defective **17 α -OHase/C_{17,20}Lyase** prevents formation of testosterone (**X**). Progesterone becomes the new endproduct. This lack of production of testosterone in the testes leads to ambiguous genitalia because of the lack of dihydrotestosterone, which is normally made from testosterone and is responsible for external male genitalia (Fig. 6). The overproduction of progesterone may cause development of female secondary sexual characteristics.

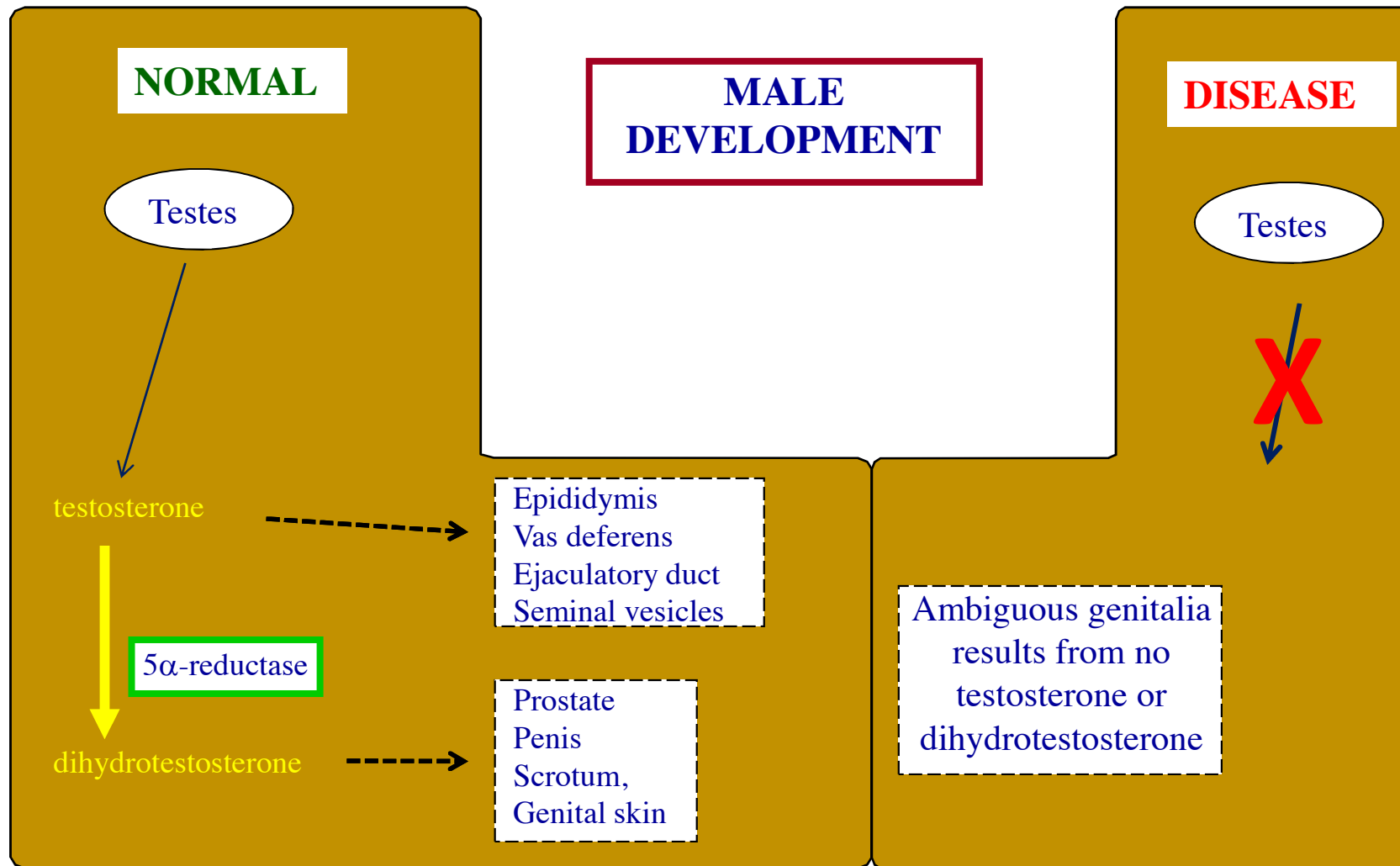


Figure 6. Normally during fetal development production of testosterone by the testes leads to formation of internal male tissues. In some tissues, testosterone is converted to dihydrotestosterone by the enzyme 5 α -reductase and causes formation of external male tissues and the prostate. In adrenal hyperplasia II (right), the lack of testosterone (X) and hence of dihydrotestosterone leads to ambiguous genitalia

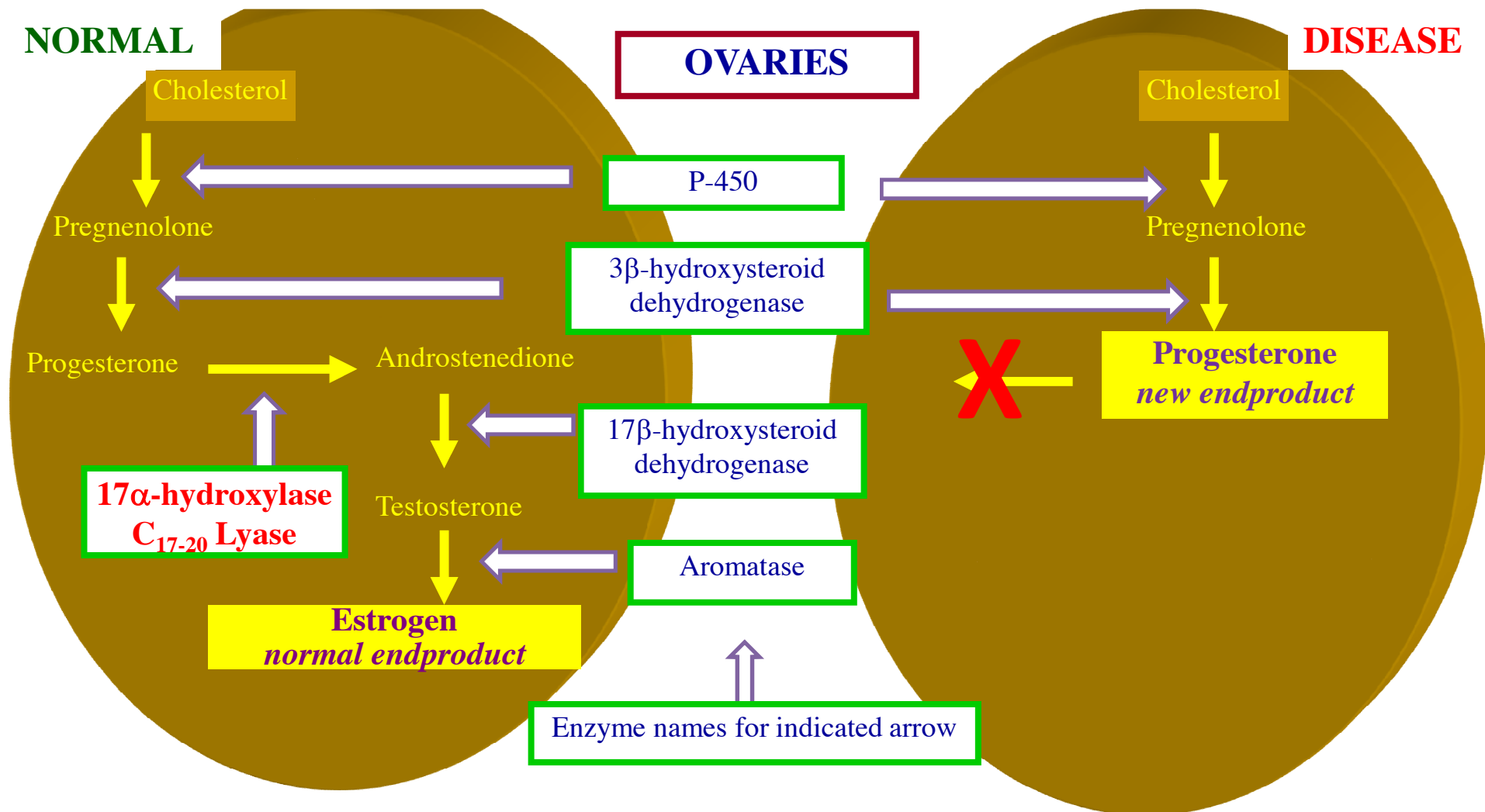


Figure 7. Cholesterol in ovaries normally (left) yields **estrogen as endproduct** following **luteinizing hormone and follicle stimulating hormone signals**. In adrenal hyperplasia II (right), defective **17 α -OHase/C_{17,20}Lyase** prevents formation of testosterone (X). Progesterone becomes the new endproduct. This lack of production of estrogen leads to lack of menstruation and infertility.