ADRENAL HYPERPLASIA IV

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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 (Fig. 1)
        - promote reuptake of sodium (salt retention) by the kidney into the blood (see Fig. 5)
        - more blood sodium leads to water retention to maintain blood pressure
    - fasciculata – produces cortisol, a glucocorticoid (Fig. 2)
        - 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.3)
        - 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

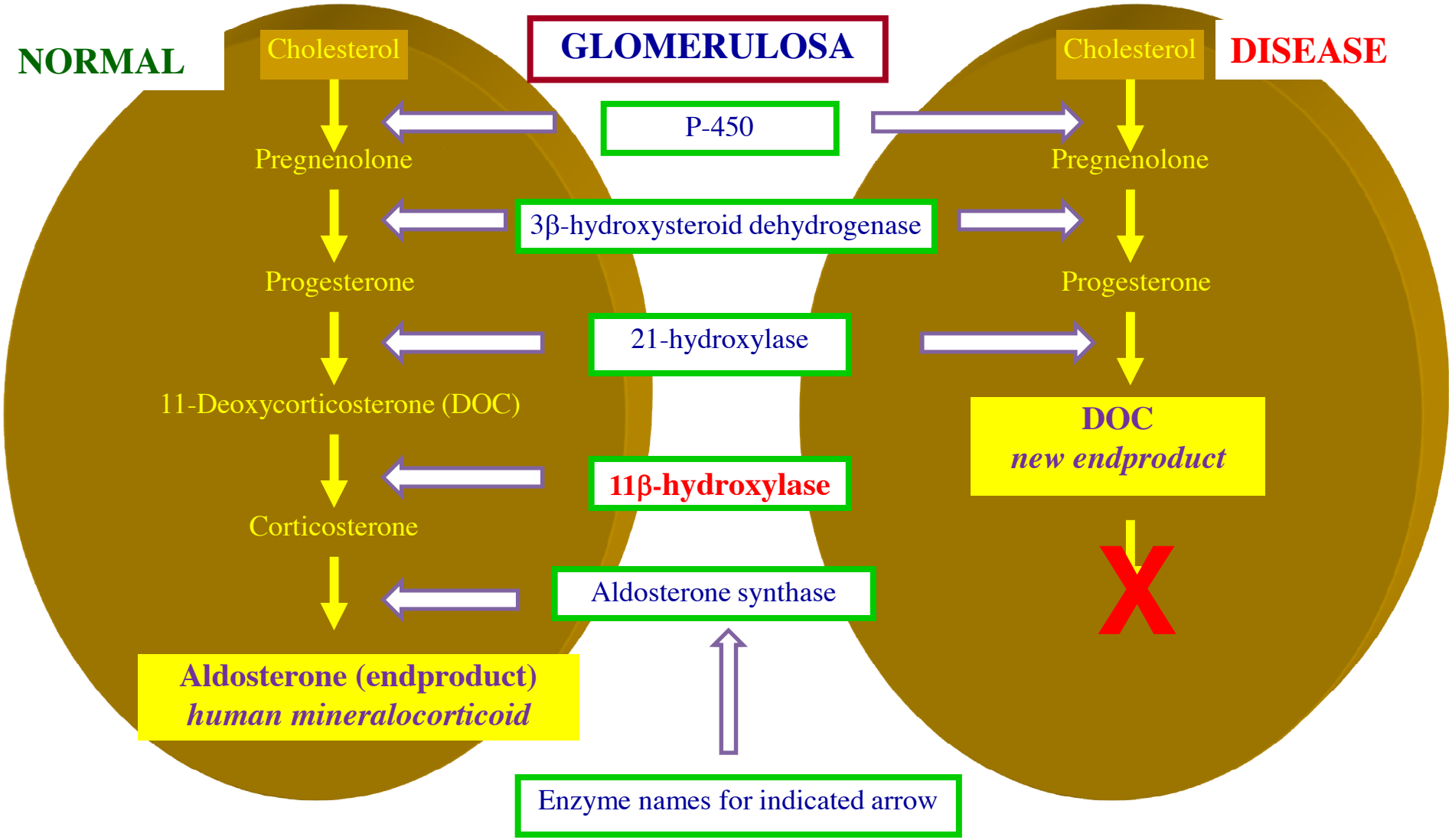


Figure 1. Cholesterol in glomerulosa normally (left) yields **aldosterone as endproduct** following **angiotensin signal**. In adrenal hyperplasia IV (right), defective **11β-OHase** prevents aldosterone formation (**X**). DOC becomes the new endproduct. DOC overproduction in fasiculata and reticularis accounts for consequences of the disease (see Figures 2 and 3, respectively).

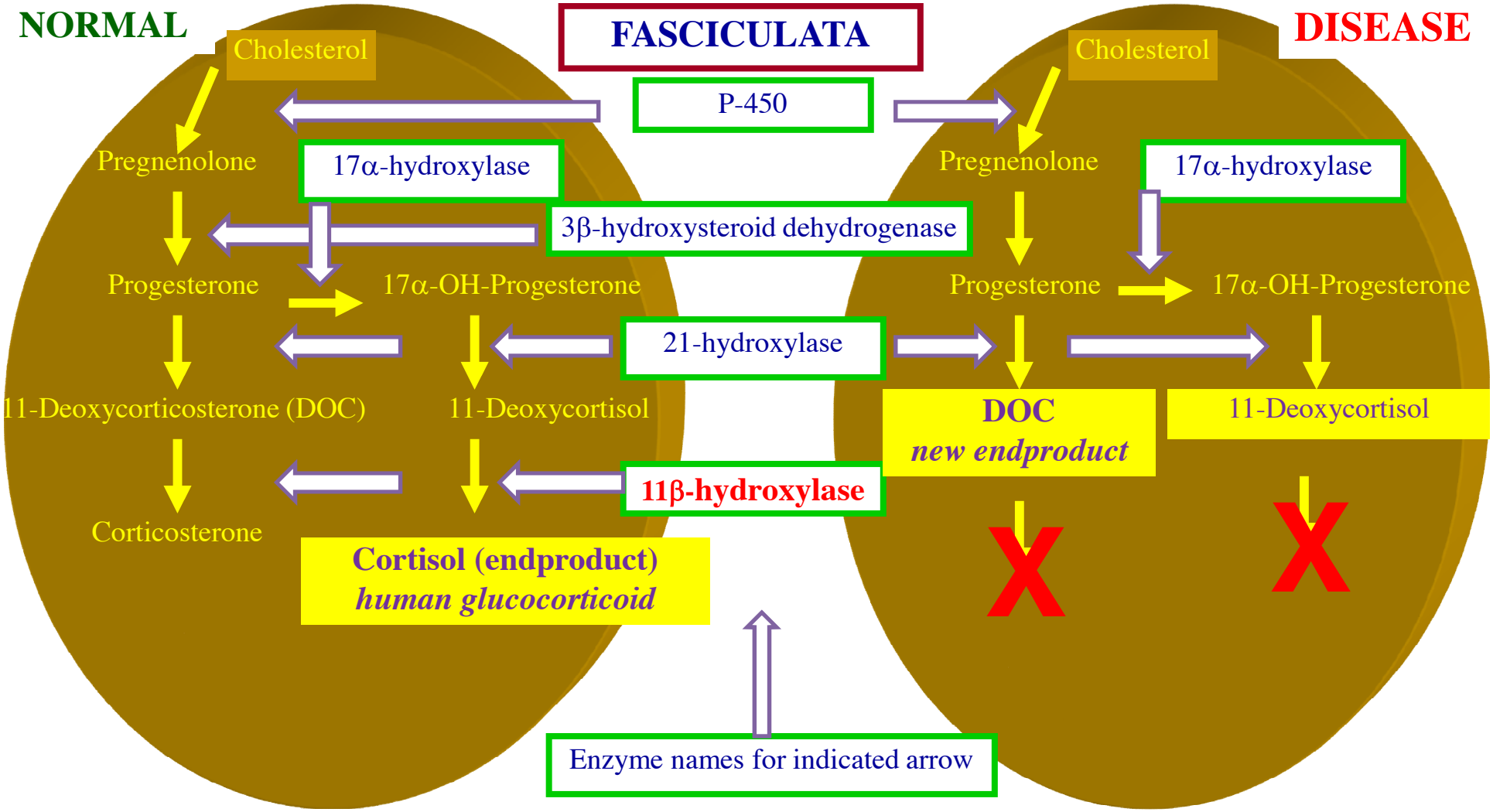


Figure 2. Cholesterol in fasciculata normally (left) yields **cortisol as endproduct** following **ACTH signal**. In adrenal hyperplasia IV (right), defective **11β-OHase** prevents cortisol formation (**X**). DOC becomes the primary endproduct. DOC **acts like aldosterone (mineralocorticoid) causing excessive salt retention and hypertension without normal control** (see Figures 4 and 5). Without **cortisol** (glucocorticoid), the patient **responds poorly to chronic stress and food deprivation** overnight. Patients must be treated with replacement glucocorticoid.

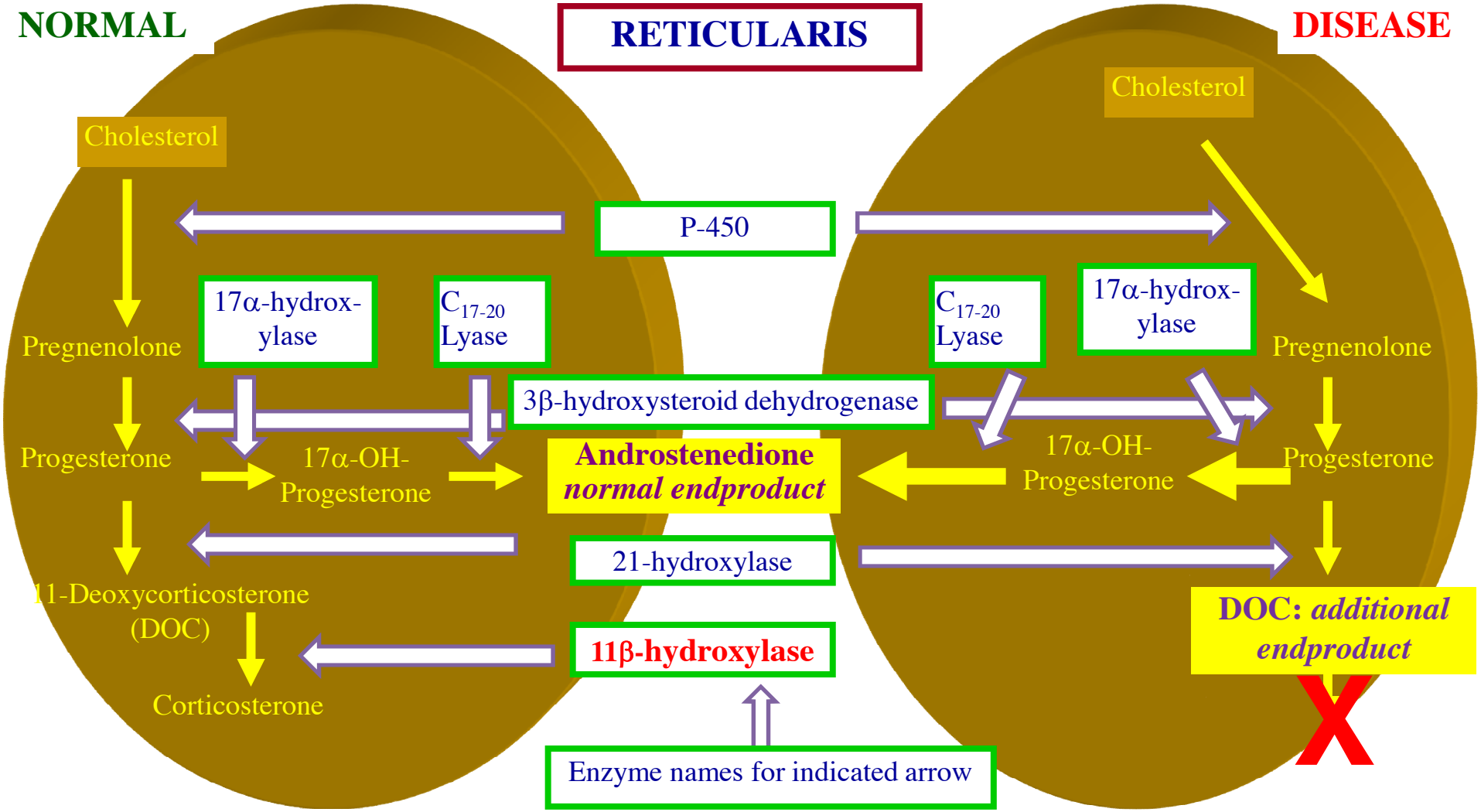
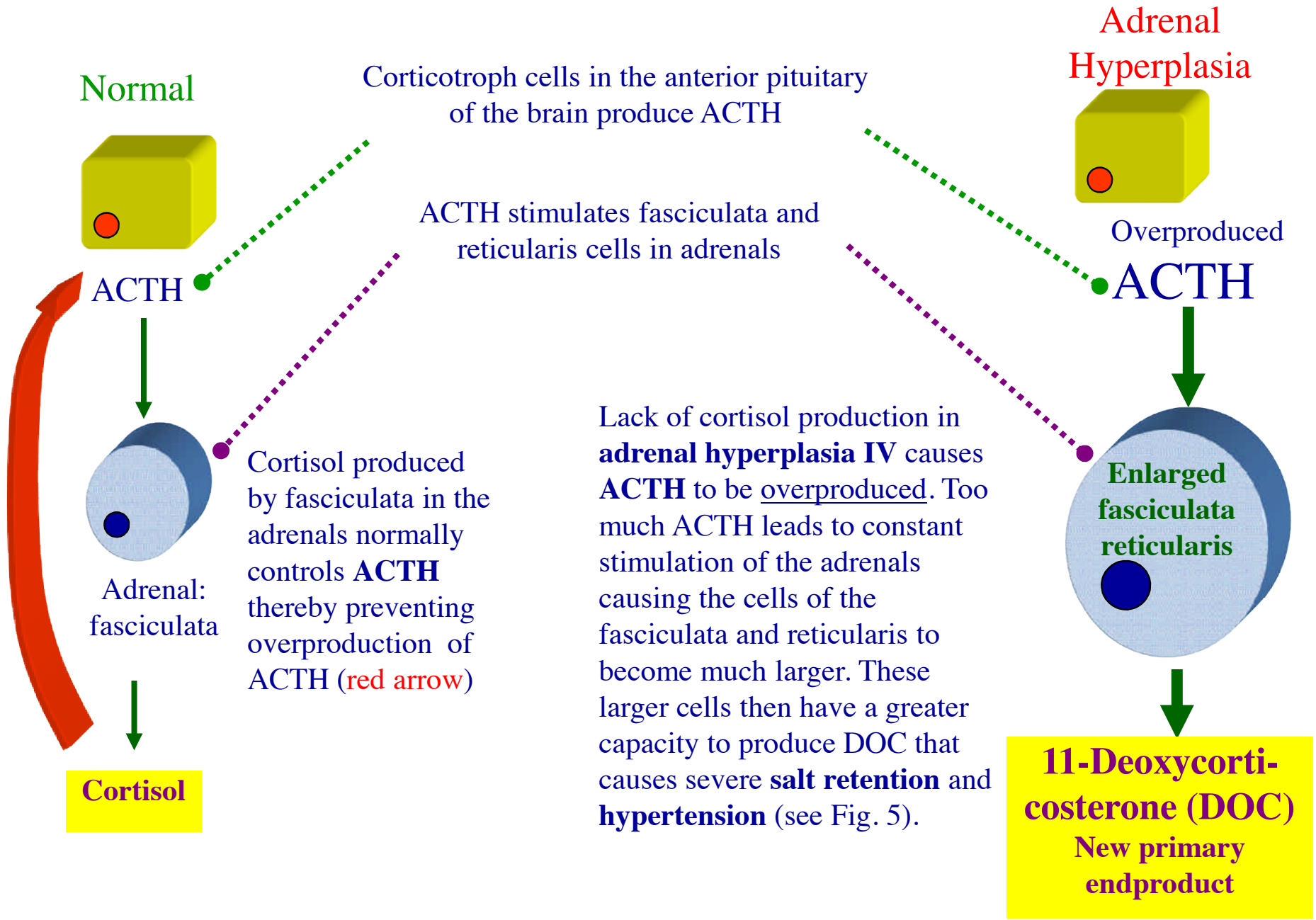


Figure 3. Cholesterol in reticularis normally (left) yields **androstenedione as endproduct** following **ACTH signal**. In adrenal hyperplasia IV (right), defective **11β-OHase** prevents metabolism of DOC (**X**) that becomes an additional endproduct. DOC **acts like mineralocorticoid causing excessive salt retention and hypertension without normal control** (see Figures 4 and 5). Androstenedione is overproduced due to hyperplasia (enlargement) of reticularis. In females this powerful androgen leads to severe virilization (see Figure 6). In males the problem is minor with excess androgens before puberty.

Figure 4. Adrenal Hyperplasia (cell enlargement) Leading to Hypertension



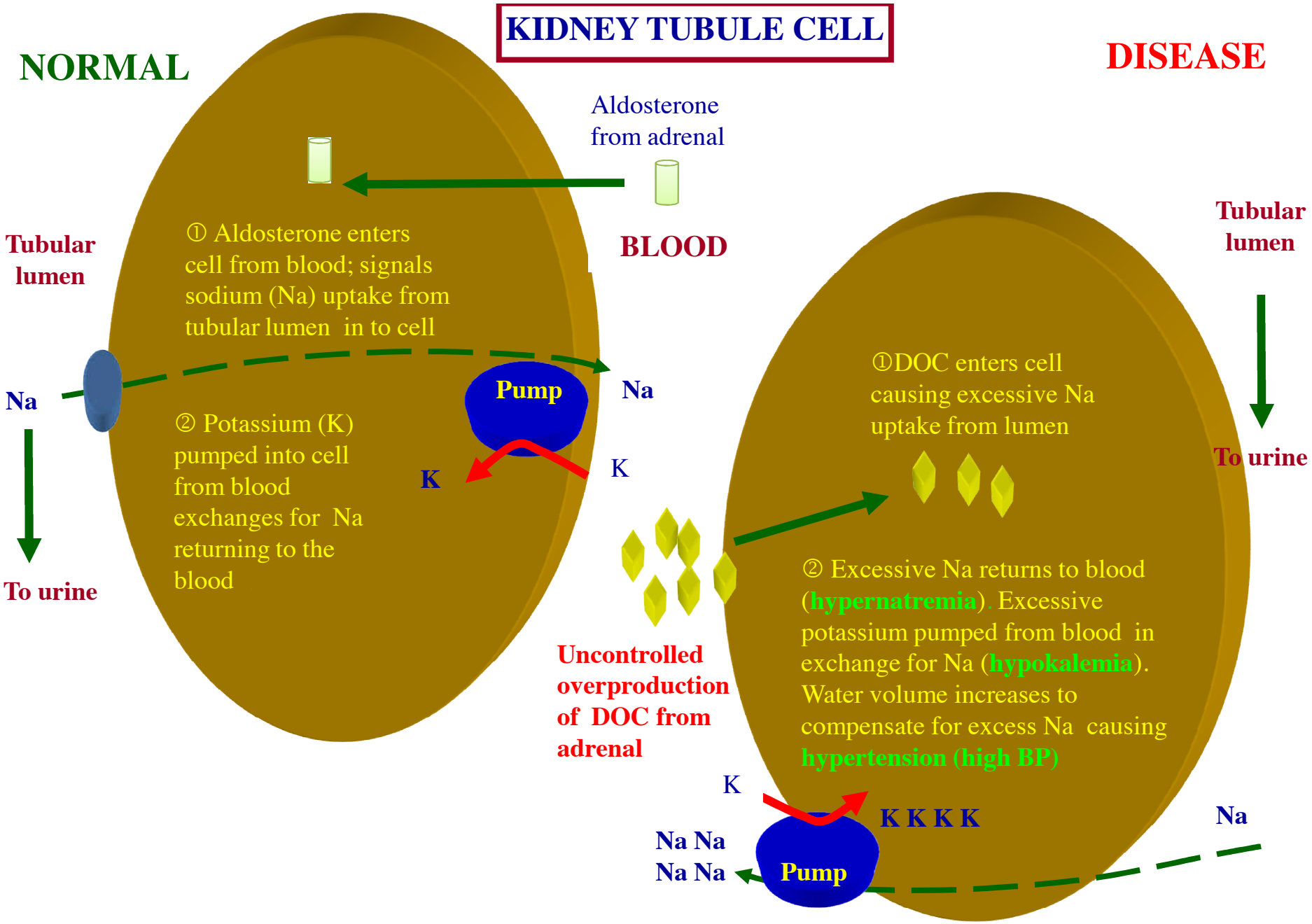


Figure 5. Action of mineralocorticoid

Figure 6. Adrenal Hyperplasia (cell enlargement) Leading to Excessive Androgen Production