



Genetic Steroid Disorders: Chapter 2. Adrenal Development

Yewei Xing, John C. Achermann, Gary D. Hammer

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The adrenal glands comprise two distinct endocrine organs: the inner medulla and the outer cortex. The inner medulla is made up of neuroectodermal cells derived from the neural crest and produces the catecholamine hormones norepinephrine and epinephrine, which are crucial for stress responses. The outer cortex is derived from the mesoderm and synthesizes steroid hormones that are essential to maintain fluid and electrolyte balance, modulate intermediary metabolism and regulate inflammatory processes. Steroidogenesis in the adrenal cortex is mainly regulated by trophic hormones controlled by the hypothalamus–pituitary endocrine axes. Adrenal organogenesis and development of adult steroidogenesis are carefully orchestrated by action of a number of gene products. Although the pattern of development differs somewhat in diverse primates, the same genes appear to regulate the basic developmental program in all mammalian species. Most basic laboratory research is done in mice, in which prenatal development occurs within a compressed period of approximately 19 days and in which adrenals at birth are considerably less developed than in their human counterparts. This chapter describes the contributions of genes responsible for the proper development of the adrenal cortex, as well as how an understanding of adrenal gland disease provides novel fundamental insights into the regulation of adrenal development and steroidogenesis.

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