

# Management of Childhood Congenital Adrenal Hyperplasia—An Integrative Review of the Literature

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## ABSTRACT

Congenital adrenal hyperplasia (CAH) is an endocrine condition that requires parents to administer steroids up to three times daily, supplementing maintenance doses with oral or injectable doses during times of illness. The purpose of this integrative review was to explore the management, care, and associated health issues for children with CAH and the family response to the condition. Four literature indexes

were searched, with 39 articles included. Four themes emerged: (a) *Health- and Development-Related Issues*, (a) *Effects of Excess Androgens*, (c) *Life Experience of CAH*, and (d) *Managing and Averting Adrenal Crisis*. Families having a child with CAH face complex concerns related their child's growth, the fear of adrenal crisis events, and the consequences of atypical genitalia for affected girls. Future studies should focus on interventions that provide guidelines to increase parental preparedness in managing adrenal crises and creating support systems for affected girls. *J Pediatr Health Care*. (2017) ■, ■-■.

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## KEY WORDS

Congenital adrenal hyperplasia, atypical genitalia, adrenal crisis

## INTRODUCTION

Classic congenital adrenal hyperplasia (CAH) is a rare, life-threatening endocrine disorder that affects boys and girls equally. CAH has an incidence in the United States of approximately 1 in 15,000 live births annually (Speiser et al., 2010). Management of CAH requires parents to administer oral steroids, typically hydrocortisone, up to three times daily. If a child is prescribed too much hydrocortisone, adverse effects can include growth suppression, obesity, and other Cushingoid features. If the dose of hydrocortisone is not sufficient, children with CAH are at a high risk for precocious puberty, which can also lead to stunted growth. Determining the proper dosage of steroids is typically achieved by obtaining routine laboratory work and measuring height, weight,

and bone age every 3 to 6 months in a growing child (Merke & Bornstein, 2005; Speiser et al., 2010). Additionally, stress dosing (doubling or tripling the oral steroid dose) is required when the child is experiencing an acute illness, such as fever or a broken bone. An emergency intramuscular injection of hydrocortisone is required when a child is unable to tolerate oral medications and/or if signs of adrenal crisis are present (Merke & Bornstein, 2005; Speiser et al., 2010; Witchel & Azziz, 2011). The need for stress dosing, either orally or by injection, related to simple viral and bacterial childhood illnesses is frequent and unpredictable, often requiring parents to make complex treatment decisions (Merke & Bornstein, 2005).

Girls born with CAH often experience virilization, which results in atypical genitalia at birth, because of elevated testosterone related to adrenal dysfunction. Boys born with CAH have typically appearing male genitalia (Witchel & Azziz, 2011). Families having a girl with CAH may face multiple surgeries into adolescence if parents, in consultation with their daughter's physicians, make the determination that reconstructive feminizing genitoplasty is warranted (Witchel & Azziz, 2011).

There are multiple challenges for practitioners and families regarding the treatment and monitoring of children with CAH, including achieving optimal glucocorticoid replacement, height and weight difficulties, excess testosterone exposure, and the need for stress dosing during times of illness (Kim, Ryabets-Lienhard & Geffner, 2012; Merke & Bornstein, 2005; Speiser et al., 2010). In addition to these physiological concerns, children with CAH and their families must also manage the psychosocial and behavioral aspects of chronic illness, such as dealing with school personnel who are unfamiliar with the condition and the possible stigmas associated with chronic illness and, for girls, atypical genitalia (Schaeffer 2011; Speiser et al., 2010). The purpose of this integrative review was to explore the management and care, associated health issues, and growth and developmental consequences for children with CAH, as well as the family response to the condition.

## METHODS

Based on established guidelines for conducting integrative reviews (Whitemore & Knafl, 2005) and in consultation with a university health sciences librarian with expertise in Web-based searches, four literature indexes were searched for articles published in peer-reviewed journals from January 1, 2000 through June

1, 2015. Databases searched included PubMed, CINAHL, PsycINFO, and Family and Society Studies Worldwide using the following search terms: *congenital adrenal hyperplasia* and *family* (or *parents* or *siblings*) as well as *adrenal crisis* and *parents*. The words *family*, *parents*, and *siblings* were truncated for maximum results.

Empirical articles meeting the following criteria were included in this review: (a) focus on CAH management, experience, and health outcomes for children under 18 years; (b) written in the English language and published between January 1, 2000 and June 1, 2015; and (c) peer reviewed. Articles were excluded if they (a) focused exclusively on pathology, (b) addressed other adrenal disorders or other disorders that may result in atypical genitalia, and (c) focused on nonclassical or rare types of CAH. The initial search yielded 470 articles (see Figure). After further screening of the article titles, 389 were excluded, leaving 81 articles from the initial search. Additional review of the abstracts and full text excluded an additional 55 articles, mainly because of a focus on pathology, nonempirical studies, and studies that included adults, leaving 26 articles. Thirteen additional articles that met the criteria were added based on review of the reference lists of relevant research articles, resulting in a total of 39 research reports (encompassing 39 separate studies). A structured template was used to extract relevant information about the research (author/title, research purpose, location of study, participant information, data collection methods, and findings) and study results pertinent to the aims of the review. Data analysis was completed using a systematic approach consisting of sorting, categorizing, and summarizing data in an effort to create meaningful conclusions about the state of the knowledge (Whitemore & Knafl, 2005).

## RESULTS

Thirty-nine empirical research studies met the criteria and were included in this review. Thirty-four studies used a quantitative design, four used a mixed-methods design, and one used a qualitative design. These studies had global representation. Of the 39, 15 were conducted in the United States, 13 in Europe (with most European studies conducted in the United Kingdom), eight in Asia (Jordan, Turkey, India, and Malaysia), three in Brazil, and one in Australia. One study was conducted jointly in the United Kingdom and the United States.

Across the 39 studies, four focus areas were identified: (a) *Health and development issues associated with a diagnosis of CAH* ( $n = 10$  studies), (b) *Physical and psychosocial consequences for the child resulting from prenatal exposure to excess androgens* ( $n = 18$  studies), (c) *Life experience of having CAH for those affected and their families* ( $n = 7$  studies), and (d) *Times of acute illness and managing and averting adrenal crisis* ( $n = 4$  studies).

There are multiple challenges for practitioners and families regarding the treatment and monitoring of children with CAH.

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