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Martin T. Stein, Robert D. Clemons and D. Jeffrey Newport

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# CHALLENGING CASE: BEHAVIORAL CHANGES

## Temper Tantrums, Impulsivity, and Aggression in a Preschool-Aged Boy\*

### CASE

James is a 4-year-old boy whose parents had become increasingly concerned about his behavior since age 12 months. His mother described him as a "high-needs" infant since birth. He was "colicky" as a young infant, cried frequently, and was difficult to settle. His mother brought him to play groups after his first birthday; she had to leave early approximately half of the time because of James' aggressive behavior. James would often walk up to another child and "punch him in the face for no reason." He also had problems with pulling hair and exhibiting long temper tantrums.

James was the first child of parents with a stable marriage and a loving home. His mother chose to stay at home to care for James and was well informed about parenting techniques. At age 2 years, she discussed James' behavior problems with his pediatrician who suggested that she enroll in a parenting course. She complied with this suggestion, although she had already read many parenting books in her attempt to better understand and manage James' behaviors. At age 2.5 years, James' mother brought him to the pediatrician again, described his behaviors, and stated that she could not understand the reasons for his patterns of behavior. She told the pediatrician that there was no modeling for aggressive or abusive behavior in the home and that neither parent ever used physical punishment. She described periods of intense anger over relatively minor events during which James would clench his fists or shake with rage and strike out at those around him. Once again, she was encouraged to read parenting books, attend parenting classes, and use "time outs" for disciplining his behaviors.

Between ages 2 and 3 years, James developed an attachment to dolls and carried a favorite Barbie doll with him everywhere he went. During this time, he also began frequent masturbation. He continued to strike out at playmates, often with no provocation. In frustration, James' parents took him to a child psychologist, who suggested that he had psychological "issues" that needed to be addressed and recommended three-times-weekly psychotherapy. The parents did not follow his recommendation.

At age 3 years, he began preschool and was sent home nine times during the year for hitting others or for other unacceptable behaviors. His parents consulted a new pediatrician, who also suggested a par-

enting class. The family took James to another psychologist who performed a battery of psychometric tests. She told the parents that James had above-average intelligence, but problems with fine motor control, socialization, and sensory-motor integration, which would likely require treatment by an occupational therapist. She also recommended a structured behavior modification system, which the family found to be helpful. James was enrolled in a new preschool, where he was placed in a class with older children. A "strict-but-loving" teacher and consistent use of behavior modification techniques helped to decrease his unacceptable behaviors. James' behavior at home continued to be characterized by excessive crying and sudden outbursts of anger.

James was taken back to his pediatrician at age 4 years and was tested for allergies at the mother's request to find a medical explanation for his behavior. His physical examination was described as normal. His height was plotted at the 95th percentile for his age, and his weight was plotted at the 75th percentile. His relatively tall stature was attributed to the fact that his father is 6 feet, 3 inches tall. His allergy evaluation was normal. The pediatrician asked the parents to complete a questionnaire to screen James for attention-deficit/hyperactivity disorder (ADHD) as a possible explanation for his impulsive behavior. After reviewing the results of the questionnaire, neither James' mother nor his pediatrician believed that James met the criteria for the diagnosis of ADHD. James' parents were instructed to continue using behavior modification and were told that his behavior might improve as he aged and as his impulse control improved.

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Index terms: *temper tantrums, aggression, precocious puberty, congenital adrenal hyperplasia.*

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### Dr. Martin T. Stein

It is the experience of most pediatricians who see young children with severe and persistent tantrums that parent education about normal developmental expectations, coupled with an opportunity to learn about behavior management, usually provides a therapeutic benefit. As parents learn to reward positive behaviors and manage the negative behaviors with strategies targeted to specific symptoms, a gradual improvement occurs. Attention to the child's temperament, home environment, and parenting styles is an important component of the diagnostic process that may suggest additional interventions.

The case of James is particularly challenging in that even after efforts were made to assist the parents in learning behavior management skills and after a

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psychological assessment was performed, he continued to demonstrate a disruptive behavior pattern at home. In addition, as outlined in the first commentary, James received a diagnosis of an organic disorder of endocrine metabolism that suggests an etiology for his externalizing behaviors.

Two clinicians were asked to write a commentary about this case. **Dr. Robert B. Clemons** is a pediatric endocrinologist at the Kaiser Permanente Medical Group in San Diego, California. **Dr. D. Jeffrey Newport** is a psychiatrist at Emory University School of Medicine in Atlanta, Georgia. His research is in the area of psychoneuroendocrinology.

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#### **Dr. Robert D. Clemons**

James' behaviors continued unchanged until age 5 years, at which point his mother noted the onset of pubic hair growth and brought him back to the pediatrician for evaluation. James was referred for an endocrinology evaluation, and a diagnosis of the simple virilizing form of congenital adrenal hyperplasia (CAH) due to 21-hydroxylase deficiency<sup>1-3</sup> was quickly established.

Pertinent physical findings during James' initial examination at age 5 years, 1 month included tall stature with a height age of 7.5 years, sparse Tanner Stage III pubic hair, and a stretched penile length of 9 cm, well above normal for a prepubertal child. Testicular volume was small (<1 mL), consistent with the suppression of pituitary gonadotropin secretion from an abnormal (adrenal) source of androgen secretion. Laboratory studies revealed a 17-hydroxyprogesterone level of 8858 ng/dL (normal range, 3-90 ng/dL), androstenedione level of 728 ng/dL (8-50 ng/dL), and testosterone level of 92 ng/dL (<10 ng/dL). Both luteinizing hormone and follicle-stimulating hormone levels were suppressed to <1.0 mIU/mL. A normal 11-deoxycortisol value eliminated the possibility of CAH resulting from 11-hydroxylase deficiency. James' bone age was 13 years, and his predicted final adult height was estimated to be 5 feet or shorter.

Treatment with oral hydrocortisone was initiated, and his abnormal laboratory values gradually corrected. James' parents and teacher noted a definite improvement in his behavior within the first weeks of treatment. As anticipated in a child with significantly advanced skeletal maturation, James' testes increased to an early pubertal volume 6 months after beginning treatment, indicating the activation of the hypothalamic-pituitary-gonadal axis. At age 5 years, 7 months, a pubertal gonadotropin response was noted during a gonadotropin-releasing hormone (GnRH) stimulation test. Treatment with a combination of long-acting GnRH analog, to prevent the progression of central precocious puberty, and growth hormone, to attempt to maximize final adult height, was begun.<sup>4</sup>

This case typifies an all-too-common sequence of events that occurs before the diagnosis of simple

virilizing CAH in male patients. The majority of female infants with CAH receive the diagnosis at birth because of the presence of ambiguous genitalia. The diagnosis of CAH in male infants without salt-wasting is usually delayed because their physical findings are subtle and easily overlooked in the newborn nursery and during well-baby examinations. In states that have not implemented newborn screening for CAH, the average age at diagnosis for boys is 62 months.<sup>5</sup> A retrospective review of newborn examination records reveals an occasional comment that the phallus seemed large. Other physical findings that may be present in the newborn include small testicular size, due to the suppression of gonadotropin secretion, and hyperpigmentation of the genital skin, resulting from excessive adrenocorticotrophic hormone secretion. As the child grows, the first clinical manifestation of androgen excess is often accelerated growth with increasing height percentile for age. Increased penis size also occurs relatively early. The other common physical findings of increased androgen production include the presence of adult apocrine odor, acne, or increased sebaceous gland activity and the presence of pubic hair. Increased muscle strength and muscle definition tend to occur later in the child's clinical course.

The behavioral manifestations of excessive androgen production in children include aggressive behaviors such as pushing, hitting, and biting (often for no apparent reason), emotional lability, and unprovoked outbursts of anger. An increased frequency of penile erections is common, as is frequent masturbation. Other sexual behaviors including an inappropriate interest in dolls, attempts to kiss "girlfriends," inappropriate sexual comments, and even ineffectual attempts at mimicking intercourse with inanimate objects or female playmates may also occur. These behaviors are very disturbing to parents and are frequently brought to the attention of the child's physician. All too often, parents state that their concerns were brushed aside or were attributed to "behavior problems," with subsequent referral to a child psychologist. Parental complaints about aggressive or impulsive behaviors may be interpreted as indicative of attention-deficit/hyperactivity disorder (ADHD) by the busy pediatrician. Sexual behaviors may raise concern about the possibility of sexual abuse and may lead to investigation by or referral to Child Protective Services.

James' diagnosis was delayed because his presenting symptoms were interpreted as reflective of a behavior disorder. Although it is true that some of James' behaviors are common in young children, the combination of tall stature with severe, persistent aggressive behavior associated with inappropriate sexual behavior should have suggested the possibility of androgen excess. It is also often true that during well-child visits, the genital examination is either omitted or performed in a cursory fashion. A careful physical examination, including examination of the genitalia, may have led to an earlier diagnosis. James' mother later commented that she had thought his penis seemed large for his age for some time, but

had not previously cared for a male infant and did not express her concern to the pediatrician.

It is imperative that physicians listen to parental concerns carefully and evaluate them thoughtfully. This case reminds us of the constant vigilance that busy physicians must maintain to avoid overlooking or quickly dismissing important clues to a diagnosis. Although medical conditions do not often present with purely behavioral symptoms, it is the physician's responsibility to consider medical problems before referring to a behavioral pediatrician or a mental health specialist.

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#### Dr. D. Jeffrey Newport

The case of 5-year-old James, who ultimately received a diagnosis of CAH when the physical sequelae of androgen excess became unmistakable, illustrates several issues pertinent to psychoneuroendocrinology. First, there are clinical questions directly applicable to James' condition. Could the diagnosis have been made sooner? Now that the diagnosis has been made, what is the appropriate course of treatment? However, there are also matters of more global relevance arising from James' case. How does one define normal endocrine function? How do gonadal steroids (in this case, androgens) impact normal development, psychosexual differentiation, and the pathophysiology of disorders such as CAH?

Although there is a growing understanding of the underlying neurobiology of many psychiatric illnesses, psychiatric nosology remains predicated upon clinical phenomenology. Thus, the psychiatric diagnostic evaluation must focus on the observed symptoms. In James' case, the early symptoms included aggression and impulsivity, a preference for dolls as toys, frequent masturbation, and a tall stature. In addition, psychometric testing indicated deficiencies in fine motor skills and sensory/motor integration.

Taken in isolation, these symptoms are nonspecific. For example, although James' masturbation can now be recognized as a harbinger of an impending precocious puberty, there are other plausible explanations. Sexual abuse must be considered, but even

more likely is normal nonpurposive masturbation not uncommon among toddlers as they begin to explore their bodies and discover the pleasurable sensations associated with touching the genitals. Likewise, James' preference for dolls does not seem to be particularly helpful. Although CAH seems to masculinize the toy preferences of girls, there is no clear impact on toy preferences in boys.<sup>1</sup> However, if James' play with the dolls was frequently sexualized in content, this would have been a more immediate cause for concern. As noted in the case presentation, James' stature could be explained by his father's height. James' specific psychomotor abnormalities are more common in boys than in girls and by implication may be a consequence of androgen excess,<sup>2</sup> but they are certainly too nonspecific to be indicative of an endocrinopathy when taken in isolation.

Of particular concern to James' family and physician were his predilection for aggression and his impulsive outbursts of anger. This is, again, a rather nonspecific finding. Childhood aggression may be a normal variant (although James' case does seem quite extreme and, hence, pathological), a consequence of child abuse, or symptomatic of ADHD. With regard to CAH, the evidence that androgen excess in humans causes aggression is inconsistent at best.<sup>3,4</sup> The only study specifically investigating aggression in children with CAH demonstrated significant increases in aggression among girls with CAH, but found no differences in boys with the disorder.<sup>5</sup> In summary, no single symptom presented in James' case is especially helpful. However, the complex of symptoms including aggression/impulsivity, tall stature, and precocious sexual activity certainly suggests a syndrome of androgen excess. Given that triad, a neuroendocrine screening is warranted, although it is certainly understandable how this possibility was initially missed.

The mainstay of treatment for James' condition should be hormonal therapy, including glucocorticoid supplementation and possibly androgen suppression. However, this may not be sufficient. It is important to understand that gonadal steroids have both acute/activational effects and developmental/organizational effects.<sup>3</sup> The acute effects of androgen excess were quickly ablated when hormonal therapy was started. Consequently, James' parents and teacher noted improvement within a few weeks of initiating hormonal therapy. However, gonadal steroids also act on the brain during discrete developmental windows and thereby induce permanent changes in brain structure and function. Therefore, the impact of perinatal androgen excess may, to some extent, persist despite corrected endocrine function beginning at age 5 years.

The case presentation demonstrates that James will benefit from other treatment modalities. Behavioral techniques implemented by his teacher helped to modulate his aggression and impulsivity. Behavior management will no doubt remain helpful in managing any residual psychiatric symptoms of James' illness. Hopefully, the combination of psychosocial treatment with hormonal therapy will be adequate. If not, traditional psychopharmacological

treatments for the management of aggression and impulsivity may be indicated (e.g., serotonergic antidepressants, anticonvulsants, clonidine).

The psychiatric manifestations of James' illness may have implications beyond CAH itself. One common practice has been to work backward from an endocrine disorder with well-documented psychiatric symptoms to understand the pathophysiology of similar "functional" psychiatric syndromes. For example, the frequent occurrence of depression in patients with Cushing's disease or hypothyroidism has led psychiatric researchers to study the function of the adrenal and thyroid neuroendocrine axes in depressed patients without these endocrine disorders. The result is that subtle endocrine changes that fall short of an endocrinologist's definition of abnormal contribute to the pathophysiology of depression. Similar research is beginning into the potential role of subtle aberrations in androgen function in disorders marked by impulsivity and/or aggression, such as ADHD and conduct disorder.<sup>6</sup>

This, of course, raises the question of just what constitutes normal endocrine function. These definitions have historically and understandably been defined from the perspective of the endocrinologist. Thus, normal ranges for hormonal assays in most clinical laboratories were established with a view to maximizing the sensitivity and specificity of the test for diagnosing the underlying cause of a clinically overt medical syndrome. Clinical endocrinologists are therefore prone to remark that purely psychiatric presentations of endocrine disorders are rare. Evidence from psychoneuroendocrine research, however, suggests otherwise. Certain psychiatric disorders may not only be associated with but may also be a consequence of subtle endocrinopathies that do not meet the thresholds established by endocrinologists. For example, an astute psychoneuroendocrinologist often interprets a thyroid profile differently than an endocrinologist does. Insights gleaned from the behavioral manifestations of CAH and other disorders of androgen excess may lead to similar redefinitions of normal gonadal function.

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#### Web Site Discussion

The case summary for this Challenging Case was posted on the Developmental and Behavioral Pediatrics web site\* and the *Journal's* web site\* (<http://www.lww.com/DBP>). Dr. Hank Shapiro (University of South Florida, All Children's Hospital) wrote: "There are studies on the effect of androgen excess on psychosexual development, and behavior in general, particularly aggression and sexual orientation.<sup>1-5</sup> My sense of these studies is that there is increased aggression in females compared to controls, and male controls compared to female controls. It is not clear that male aggression is increased, or that there are measurable effects on male behavior. This assumes that the individual did not experience a salt-wasting crisis and severe illness. Lower IQ scores have been recorded in children with CAH who had a salt-losing syndrome.<sup>5</sup>

"In the absence of clear evidence that androgen excess itself is responsible for this child's behavioral problems, we have to consider the full range of family, environmental, and temperamental issues regardless of the medical treatment. There is a substantial literature on learning, behavioral, social, and emotional problems with short stature, but not much on tall stature, which is found in untreated children with CAH prior to treatment. I have often wondered about this. A cross-sectional study of behavior and emotional function analyzed with respect for height percentile would be interesting. I wonder whether tall-for-age children are seen as more "immature" and as having more behavioral problems. Curiously, there is a good chance that the child in the challenging case might end up with short stature in the long run due to accelerated bone maturation!"

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Dr. Shapiro echoed the earlier comment of Dr. Newport, who wrote, "... the evidence that androgen excess in humans causes aggression is inconsistent at best." A recent study among adolescent boys and girls with delayed onset of puberty (a hypogonadal condition) approached the association between androgens and behavior in a novel way.<sup>1</sup> The sub-

\*A bimonthly discussion of an upcoming challenging case takes place at the Developmental and Behavioral Pediatrics web site. This web site is sponsored by the Maternal and Child Health Bureau and the American Academy of Pediatrics section on Developmental and Behavioral Pediatrics. Henry L. Shapiro, M.D., is the editor of the web site. Martin Stein, M.D., the Challenging Case editor, incorporates comments from the Web discussion into the published Challenging Case. To become part of the discussion at the Developmental and Behavioral Pediatrics home page, go to <http://www.dbpeds.org>.

jects were given three different doses of testosterone for 3 months each, alternating with a placebo control pill. The amount of testosterone at the three dose levels approximated early, middle, and late adolescence. A self-reported, standardized test for aggression demonstrated a significant hormone effect on physical, but not verbal, aggressive behavior. Interestingly, conjugated estrogen administered to hypogonadal girls had a similar but less robust effect on aggression. The adolescents in the study did not have CAH, so the results may not be directly applicable to patients such as James.

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**Dr. Ed Christophersen** (Children's Mercy Hospital, Kansas City, MO) discussed James' behaviors in the context of a child who presents to a pediatric clinician without an organic diagnosis. He offers an approach to behavioral diagnosis and management that makes use of tools different from those described in the case summary: "This case really must be dealt with as almost two separate cases. The information obtained around the time of the child's fifth birthday, when his mother noted the onset of pubic hair growth, was not available when he was 2, 3, and 4 years old. Thus, the practitioner is faced with deciding how to deal with these questions when they are asked at the earlier ages, prior to any knowledge of biological findings. And, in the vast majority of cases, there would not be the biological findings.

"Faced with the mother of a 2–3 year old who strikes out at playmates, often without provocation, the pediatrician should start with multi-informant, objective input. Preference should be given to standardized rating scales that have been normed with this age population. The two most popular are the Child Behavior Checklist (both the Parent and the Teacher forms) and the Behavior Assessment System for Children (both the Parent and the Teacher forms).<sup>1,2</sup> Also, since both systems have computerized scoring available, they require relatively little commitment on the part of office staff.

"An office interview of the parent(s), with the child present, should be adequate to obtain a history of any prior problems in either biological family, as well as a brief assessment of parenting practices, family stresses (such as marital problems, depression, recent moves, job changes, and the like), and attachment issues. A written form that is completed by the parents, prior to the interview, typically results in the acquisition of more information in a shorter period of time, with less risk of leaving out important information.<sup>3</sup> Many pediatric practitioners will already have asked the parents to complete a standardized form for assessment of the child's overall development such as the Child Development Inventory or the Denver II. Further developmental assessment appears to be unwarranted at that time.

"Referral to a mental health practitioner should be based upon the prior history with that practitioner,

as well as their training and qualifications. Some communities have well-established parent training groups that are predicated upon empirically supported intervention strategies (such as the programs developed by Patterson, Webster-Stratton and Hammond, or Barkley and Benton).<sup>4–6</sup> Other communities have well-established parent training programs that serve one family at a time. The choice between such programs should be predicated upon prior outcomes.

"A clinician's approach to behavioral issues in a primary care setting is similar to dealing with biological issues in that it starts with comprehensive assessment including objective findings, interviews, and observation. As the child's story unfolds, further biological assessments can rather easily be included as new information is obtained."

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## Dr. Martin T. Stein

When I initially heard about James at a weekly departmental pediatric case conference, I was immediately struck by the apparent delay in the diagnosis CAH. There seemed to be a pattern of behaviors that preceded, by several years, the discovery of the somatic effects of increased androgen during a physical examination. Why was James' condition not correctly diagnosed as hyperandrogenemia a few years before the diagnosis when his mother brought him to the pediatrician? A complete physical examination, I reasoned, would have detected signs of increased circulating androgens. In a short time, I came to recognize my error.

The incidence of CAH in the United States is 1 in 12,000 (1 in 680 to 15,000 worldwide). As Dr. Clemens observed, CAH is not diagnosed in the first few years of life in male infants. In the most common form, a deficiency of the 21-hydroxylase enzyme, ambiguous external genitalia in a female newborn usually leads to the correct diagnosis. In neonates with CAH who develop a salt-losing syndrome resulting from decreased mineralocorticoids, severe dehydration, electrolyte imbalance, and shock may suggest the diagnosis in both genders in the first 2 weeks of life. In the boys without salt-losing syndrome, normal male external genitalia at birth and the absence of symptoms preclude an early diagnosis. James was in this group.

Secondary sexual characteristics were apparent when James' mother brought him to a pediatrician when he was 5 years old after she observed pubic

hair. A physical examination revealed the effects of increased androgens—an enlarged phallic length, sparse Tanner Stage III pubic hair, small testes, and tall stature. As Dr. Clemons observed, the mean age of diagnosis for the non-salt-losing male patients with CAH is 5 years, 2 months. As with all forms of inborn errors of metabolism, the degree of deficiency of the putative enzyme is variable. In CAH, there is a variability in the onset of the somatic effect of increased androgens. James may not have revealed secondary sexual characteristics during a physical examination at 2 to 4 years of age. Even at the onset of puberty in normally developing boys, the initial physical signs—a slight increase in testicular volume, followed by reddening and thinning of the scrotum—may be difficult to detect. Nevertheless, an axiom of good clinical practice is that all children with problems of behavior and development must undergo a thorough physical examination as part of a complete evaluation.

The standardized screening tests recommended by Dr. Christophersen are reliable indicators of behavioral conditions. However, in current pediatric pri-

mary care practice, their use may be limited. Traditionally, pediatricians rely on a medical interview with the parents and child to assess problems of behavior or development. This model is consistent with all other areas of diagnosis in medical practice. From that perspective, it is not surprising that standardized tests of behavior have not found their way into most primary care office practices. Perhaps with the increased training time for developmental and behavioral pediatrics in residency programs, there will be greater opportunity to demonstrate the value of standardized tests in primary care practice. However, if these tests are to become useful to pediatricians, residents must be shown the way an office practice can be organized in a manner that efficiently incorporates standardized testing. At the same time, teaching the skills required for an effective and efficient medical interview should not be given short shrift. Checklists cannot substitute for an opportunity to listen to a patient's story!

#### ACKNOWLEDGMENT

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