23 - 18 Gender Dysphoria

01 - 18 Gender Dysphoria

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18

Gender Dysphoria The term gender dysphoria appears as a diagnosis for the first time in the fifth edition of the Diagnostic and Statistical Manual of Mental Disorders (DSM-5) to refer to those persons with a marked incongruence between their experienced or expressed gender and the one they were assigned at birth. It was known as gender identity disorder in the previous edition of DSM. The term gender identity refers to the sense one has of being male or female, which corresponds most often to the person’s anatomical sex. Persons with gender dysphoria express their discontent with their assigned sex as a desire to have the body of the other sex or to be regarded socially as a person of the other sex. The term transgender is a general term used to refer to those who identify with a gender different from the one they were born with (sometimes referred to as their assigned gender). Transgender people are a diverse group: There are those who want to have the body of another sex known as transsexuals; those who feel they are between genders, of both genders, or of neither gender known as genderqueer; and those who wear clothing traditionally associated with another gender, but who maintain a gender identity that is the same as their birth-assigned gender known as crossdressers. Contrary to popular belief, most transgender people do not have genital surgery. Some do not desire it and others who do may be unable to afford it. Transgender people may be of any sexual orientation. For example, a transgender man, assigned female at birth, may identify as gay (attracted to other men), straight (attracted to women), or bisexual (attracted to both men and women). In DSM-5, no distinction is made for the overriding diagnostic term gender dysphoria as a function of age. However, criteria for diagnosis in children or adolescents are somewhat different. In children, gender dysphoria can manifest as statements of wanting to be the other sex and as a broad range of sex-typed behaviors conventionally shown by children of the other sex. Gender identity crystallizes in most persons by age 2 or 3 years. A specifier is noted if the gender dysphoria is associated with a disorder of sex development. EPIDEMIOLOGY Children Most children with gender dysphoria are referred for clinical evaluation in early grade school years. Parents, however, typically report that the cross-gender behaviors were apparent before 3 years of age. Among a sample of boys younger than age 12 who were referred for a range of clinical problems, the reported desire to be the other sex was 10

percent. For clinically referred girls younger than age 12, the reported desire to be the other sex was 5 percent. The sex ratio of children referred for gender dysphoria is 4 to 5 boys for each girl, which is hypothesized to be due in part to societal stigma directed toward feminine boys. The sex ratio is equal in adolescents referred for gender dysphoria. Researchers have observed that many children considered to have shown gender nonconforming behavior do not grow up to be transgender adults; conversely many people who later come out as transgender adults report that

they were not identified as gender nonconforming during childhood. Adults The estimates of gender dysphoria in adults emanate from European hormonal/surgical clinics with a prevalence of 1 in 11,000 male-assigned and 1 in 30,000 female-assigned people. DSM-5 reports a prevalence rate ranging from 0.005 to 0.014 percent for maleassigned and 0.002 to 0.003 percent for female-assigned people. Most clinical centers report a sex ratio of three to five male patients for each female patient. Most adults with gender dysphoria report having felt different from other children of their same sex, although, in retrospect, many could not identify the source of that difference. Many report feeling extensively cross-gender identified from the earliest years, with the crossgender identification becoming more profound in adolescence and young adulthood. Overall the prevalence of male to female dysphoria is higher than female to male dysphoria. An important factor in diagnosis is that there is greater social acceptance of birth-assigned females dressing and behaving as boys (so-called tomboys) than there is of birth-assigned males acting as females (so-called sissies). Some researchers speculate that one in 500 adults may fall somewhere on a transgender spectrum, based on population data rather than clinical data. ETIOLOGY Biological Factors For mammals, the resting state of tissue is initially female; as the fetus develops, a male is produced only if androgen (set off by the Y chromosome, which is responsible for testicular development) is introduced. Without testes and androgen, female external genitalia develop. Thus, maleness and masculinity depend on fetal and perinatal androgens. Sexual behavior in lower animals is governed by sex steroids, but this effect diminishes as the evolutionary tree is scaled. Sex steroids influence the expression of sexual behavior in mature men or women; that is, testosterone can increase libido and aggressiveness in women, and estrogen can decrease libido and aggressiveness in men. But masculinity, femininity, and gender identity may result more from postnatal life events than from prenatal hormonal organization. Brain organization theory refers to masculinization or feminization of the brain in utero. Testosterone affects brain neurons that contribute to the masculinization of the brain in such areas as the hypothalamus. Whether testosterone contributes to so-called

masculine or feminine behavioral patterns remains a controversial issue. Genetic causes of gender dysphoria are under study but no candidate genes have been identified, and chromosomal variations are uncommon in transgender populations. Case reports of identical twins have shown some pairs that are concordant for transgender issues and others not so affected. A variety of other approaches to understanding gender dysphoria are underway. These include imaging studies that have shown changes in white matter tracts, cerebral blood flow, and cerebral activation patterns in patients with gender dysphoria; but such studies have not been replicated. An incidental finding is that transgender persons are likely to be left handed, the significance of which in unknown. Psychosocial Factors Children usually develop a gender identity consonant with their assigned sex. The formation of gender identity is influenced by the interaction of children’s temperament and parents’ qualities and attitudes. Culturally acceptable gender roles exist: Boys are not expected to be effeminate, and girls are not expected to be masculine. There are boys’ games (e.g., cops and robbers) and girls’ toys (e.g., dolls and dollhouses). These roles are learned, although some investigators believe that some boys are temperamentally delicate and sensitive and that some girls are aggressive and energized —traits that are stereotypically known in today’s culture as feminine and masculine, respectively. However, greater tolerance for mild cross-gender activity in children has developed in the last few decades. Sigmund Freud believed that gender identity problems resulted from conflicts experienced by children within the Oedipal triangle. In his view, these conflicts are fueled by both real family events and children’s fantasies. Whatever

interferes with a child's loving the opposite-sex parent and identifying with the same-sex parent interferes with normal gender identity development. Since Freud, psychoanalysts have postulated that the quality of the mother–child relationship in the first years of life is paramount in establishing gender identity. During this period, mothers normally facilitate their children's awareness of, and pride in, their gender: Children are valued as little boys and girls. Analysts argue that devaluing, hostile mothering can result in gender problems. At the same time, the separation–individuation process is unfolding. When gender problems become associated with separation–individuation problems, the result can be the use of sexuality to remain in relationships characterized by shifts between a desperate infantile closeness and a hostile, devaluing distance. Some children are given the message that they would be more valued if they adopted the gender identity of the opposite sex. Rejected or abused children may act on such a belief. Gender identity problems can also be triggered by a mother's death, extended absence, or depression, to which a young boy may react by totally identifying with her—that is, by becoming a mother to replace her. The father's role is also important in the early years, and his presence normally helps the separation–individuation process. Without a father, mother and child may remain overly close. For a girl, the father is normally the prototype of future love objects; for a boy, the father is a model for male identification. Learning theory postulates that children may be rewarded or punished by parents and teachers on the basis of gendered behavior, thus influencing the way children express their gender identities. Children also learn how to label people

according to gender and eventually learn that gender is not dictated by surface appearance such as clothing or hairstyle. DIAGNOSIS AND CLINICAL FEATURES Children The DSM-5 defines gender dysphoria in children as incongruence between expressed and assigned gender, with the most important criterion being a desire to be another gender or insistence that one is another gender (Table 18-1). By emphasizing the importance of the child's self-perception, the creators of the diagnosis attempt to limit its use to those children who clearly state their wishes to be another gender, rather than encompassing a broader group of children who might be considered by adults to be gender nonconforming. However a child's behavior may also lead to this diagnosis. Table 18-1 DSM-5 Diagnostic Criteria for Gender Dysphoria

Many children with gender dysphoria prefer clothing typical of another gender, preferentially choose playmates of another gender, enjoy games and toys associated with another gender, and take on the roles of another gender during play. For a diagnosis to be made, these social characteristics must be accompanied by other traits less likely to be socially influenced, such as a strong desire to be the other gender, dislike of one's sexual anatomy, or desire for primary or secondary sexual characteristics of the desired gender. Children may express a desire to have different genitals, state that their genitals are going to change, or urinate in the position (standing or sitting) typical of another gender. It is notable that characteristics used to diagnose children with gender dysphoria must be accompanied by clinically significant distress or impairment on the part of the child, and not simply on the part of the adult caregivers, who may be uncomfortable with gender nonconformity. Differential Diagnosis of Children

Children diagnosed with gender dysphoria, predicted to be more likely than others to identify as transgender as adults, are differentiated from other gender nonconforming children by statements about desired anatomical changes, as well as persistence of the diagnosis over time. Children whose gender dysphoria persists over time may make repeated statements about a desire to be or

belief that they are another gender. Other gender nonconforming children may make these statements for short periods but not repeatedly, or may not make these types of statements, and may instead prefer clothing and behaviors associated with another gender, but show contentment with their birthassigned gender. The diagnosis of gender dysphoria no longer excludes intersex people, and instead is coded with a specifier in the cases where intersex people are gender dysphoric in relation to their birth-assigned gender. A medical history is important to distinguish between those children with intersex conditions and those without. The standards of care for intersex children have changed dramatically over the last few decades due to activism by intersex adults and supportive medical and mental health professionals. Historically, intersex babies were often subjected to early surgical procedures to create more standard male or female appearances. These procedures had the potential to cause sexual dysfunction, such as inability to orgasm, and permanent sterility. Recently, these practices have changed considerably so that more intersex people are given the chance to make decisions about their bodies later in life. Adolescents and Adults Adolescents and adults diagnosed with gender dysphoria must also show an incongruence between expressed and assigned gender. In addition, they must meet at least two of six criteria, half of which are related to their current (or in the cases of early adolescents, future) secondary sex characteristics or desired secondary sex characteristics. Other criteria include a strong desire to be another gender, be treated as another gender, or the belief that one has the typical feelings and reactions of another gender (see Table 18-1). In practice, most adults who present to mental health practitioners with reports of gender-related concerns are aware of the concept of transgender identity. They may be interested in therapy to explore gender issues, or may be making contact in order to request a letter recommending hormone treatment or surgery. The cultural trope of being “trapped in the wrong body” does not apply to all, or even most, people who identify as transgender, so clinicians should be aware to use open and affirming approaches, taking language cues from their patients. The DSM-5 criteria are noticeably open to the idea that some people do not fit into the traditional gender binary, and may desire to be alternative genders, such as genderqueer. Like the diagnosis in childhood, the adolescent and adult diagnosis also requires that those diagnosed be personally distressed or impaired by their feelings, rather than their behaviors or identities being pathologized by others while not upsetting to the people themselves. The adolescent and adult criteria also contain a

post-transition specifier, which can be used for those people who live in their affirmed genders. They are required, however, to have undergone or be preparing to undergo at least one medical or surgical procedure in order to qualify for this specifier. Differential Diagnosis of Adolescents and Adults Those who meet the criteria for a diagnosis of gender dysphoria must experience clinical distress or impairment related to their gender identity. This excludes from the diagnosis those transgender or gender nonconforming people who are not clinically distressed by their gender identities. There are certain mental illnesses in which transgender identity may be a component of delusional thinking, such as in schizophrenia. However, this is extremely rare and can be differentiated from transgender identity or gender dysphoria through the diminishment of transgender feelings with the successful treatment of psychosis versus the persistence of these feelings in periods that are psychosis free. Body dysmorphic disorder may be a differential diagnosis for some patients who present with a desire to change gendered body parts. However, those with body dysmorphic disorder generally focus on a body part because of a belief that it is abnormal, rather than due to a desire to change their assigned gender. The Paraphilic Disorders chapter of the DSM-5 contains the diagnosis transvestic disorder, which is defined as recurrent and

intense sexual arousal from cross-dressing that causes clinically significant distress or impairment. This diagnosis is differentiated from gender dysphoria by the patient's gender identity being consistent with their gender assigned at birth, and by sexual excitement linked to cross-dressing coming to interfere with the person's life. COURSE AND PROGNOSIS Children Children typically begin to develop a sense of their gender identity around age 3. At this point they may develop gendered behaviors and interests, and some may begin to express a desire to be another gender. It is often around school age that children are first brought for clinical consultations, as this is when they begin to interact heavily with classmates and to be scrutinized by adults other than their caregivers. Some children who will later identify as transgender as adults do not show behaviors consistent with another gender at this age. Some say later that they worked hard to appear stereotypical to their assigned gender, whereas others deny being able to recall gender identity concerns. Approaching puberty, many children diagnosed with gender dysphoria begin to show increased levels of anxiety related to anticipated changes to their bodies. Children diagnosed with gender dysphoria do not necessarily grow up to identify as transgender adults. A number of studies have demonstrated that more than half of those diagnosed with gender identity disorder, based on the DSM-IV, later identify with their birth-assigned gender once they reach adulthood. Those children who do identify as transgender as adults have been shown to have more extreme gender dysphoria as

children. Many studies show increased rates of gay and bisexual identity among those who were gender nonconforming as children. Comorbidity in Children Children diagnosed with gender dysphoria show higher rates than other children of depressive disorders, anxiety disorders, and impulse-control disorders. This is likely related to the stigma faced by these children related to their gendered behaviors and identities. There are also reports that those diagnosed with gender dysphoria are more likely than others to fall on the autism spectrum. Some researchers posit that this may be related to intrauterine hormone exposure. Adults Some people diagnosed with gender dysphoria as adults recall the continuous development of transgender identity since childhood. In these cases, some have periods of hiding their gender identity, many entering into stereotypic activities and employment in order to convince themselves and others that they do not have gender nonconforming identities. Others do not recall gender identity issues during childhood. Lesbian and gay communities are often havens for gender nonconforming people, and some people identify as gay, lesbian, or bisexual before coming out as transgender. Comorbidity in Adults Adults diagnosed with gender dysphoria show higher rates than other adults of depressive disorders, anxiety disorders, suicidality and self-harming behaviors, and substance abuse. The lifetime rate of suicidal thoughts in transgender people is thought to be about 40 percent. The minority stress model predicts increases in mental illness in groups that are stigmatized, discriminated against, harassed, and abused at higher rates than others. DSM-5 reports that persons with late-onset gender dysphoria may have greater fluctuations in the extent of their distress and more ambivalence about and less satisfaction after sex reassignment surgery. TREATMENT Children Treatment of gender identity issues in children typically consists of individual, family, and group therapy that guides children in exploring their gendered interests and identities. There are some providers who practice reparative, or conversion therapy, which attempts to change a person's gender identity or sexual orientation. This type of therapy is contrary to position statements by the American Psychiatric Association and practice guidelines of the American Academy of Child and Adolescent Psychiatry. Adolescents

As gender-nonconforming children approach puberty, some show intense fear and preoccupation related to the physical changes they anticipate or are beginning to experience. In addition to providing psychotherapy, many clinicians use these adolescents' reactions to the first signs of puberty as a compass to determine if pubertyblocking medications should be a consideration. Puberty-blocking medications are gonadotropin-releasing hormone (GnRH) agonists that can be used to temporarily block the release of hormones that lead to secondary sex characteristics, giving adolescents and their families time to reflect on the best options moving forward. GnRH agonists have been used for many years in other populations (e.g., children with precocious puberty) and are felt to be safe. However, such steps should be considered carefully. Adults Treatment of adults who identify as transgender may include psychotherapy to explore gender issues, hormonal treatment, and surgical treatment. Hormonal and surgical interventions may decrease depression and improve quality of life for such persons. Mental Health Treatment The history of poor treatment and medicalization of transgender people by mental health providers has led to a decreased interest on the part of trans-identified people in engaging in mental health care. Many surgeons, and some physicians who prescribe transition-related hormones, require a letter from a mental health provider, so many transgender people are engaged with mental health in a gatekeeping model. Many community clinics are now using informed consent models for hormone treatment, thereby decreasing the need for mental health providers to play the role of gatekeepers. The World Professional Association for Transgender Health (WPATH) Standards of Care (SOC) for the health of transsexual, transgender, and gender-nonconforming people have recently become more flexible and open to informed consent models. Some mental health providers are specializing in working with transgender populations, and this is increasing the rate at which transgender people engage in psychotherapy. Hormones Hormone treatment of transgender men is primarily accomplished with testosterone, usually taken by injection every week or every other week. Initial changes with testosterone therapy include increased acne, muscle mass, and libido, as well as cessation of menses, usually within the first few months. Subsequent, and more permanent, changes include deepening of the voice, increased body hair, and enlargement of the clitoris. Monitoring includes hemoglobin/hematocrit levels, as testosterone can rarely cause an increase in red blood cell counts that can lead to stroke. Like all steroid hormones, testosterone is processed in the liver, so routine liver function tests should be obtained. Clinicians also want to monitor cholesterol and screen for diabetes, as testosterone treatment may increase the likelihood of lipid abnormalities

and diabetes. Those beginning hormone treatments are routinely counseled on fertility, as future fertility may be affected on testosterone. Transgender women may take estrogen, testosterone-blockers, or progesterone, often in combination. These hormones can cause softening of the skin and redistribution of fat, as well as breast growth. Breast development varies between people, but does not generally exceed bra cup size B. It is generally recommended to be on hormones for 18 to 24 months before having breast augmentation, allowing the breasts to develop to their final size. Sex drive can decrease, as well as erections and ejaculation. Body hair can decrease somewhat, but often not as much as desired, prompting many women to obtain electrolysis. There is no change in voice, as testosterone has permanently altered the vocal cords, and many women seek out voice coaching. Those on estrogen should avoid cigarette smoking, as the combination can lead to increased risk of blood clots. Blood pressure should be monitored, as well as liver function and cholesterol. In addition, providers routinely test prolactin as this hormone can increase on estrogen therapy, and in rare cases transgender women may develop prolactinomas. Reproductive

counseling is very important before beginning estrogen treatment because permanent sterility is almost always the outcome. Surgery Many fewer people undergo gender-related surgeries than take hormones. Some people do not desire gender-related surgeries. Others cannot afford them, or are not convinced that they will be satisfied with currently available results. The most common type of surgery for both trans-men and trans-women is “top surgery,” or chest surgery. Transgender men may have surgery to construct a malecontoured chest. Trans-women may have breast augmentation. “Bottom surgery” is less common. Transgender men may have a metoidioplasty, in which the clitoris is freed from the ligament attaching it to the body, and tissue is added, increasing its length and girth. Scrotoplasty, the placement of testicular implants, is another way to create male-appearing genitalia. Phalloplasty, the creation of a penis, is less commonly performed because it is expensive, involves multiple procedures, requires donor skin from another part of the body, and has limited functionality. Bottom surgery for women is typically vaginoplasty, also commonly known as Sex Reassignment Surgery (SRS). In this procedure, the testicles are removed, the penis is reconstructed to form a clitoris, and a vagina is created. Techniques for vaginoplasty are becoming very good, but the procedure remains expensive. Because of this, some women, especially those with less money, may have orchiectomies, where the testes alone are removed. These can be in-office procedures with local anesthetic, and are effective in substantially decreasing the body’s production of androgens like testosterone. Less widely discussed, but important to many women, are facial feminization surgeries that alter the cheeks, forehead, nose, and lips to create a more feminine facial appearance. The face is often used by persons to recognize gender in another person and having facial features that match one’s affirmed gender can facilitate social interaction and provide safety from

harassment and violence. Transgender men rarely undergo facial surgeries, as testosterone typically causes the face to appear more masculine. Because surgery is inaccessible to many, there are rare cases of self-surgery and some people have surgeries performed under unsafe conditions. Women may inject industrial grade silicone to produce body curves. Silicone injection that is not done under the supervision of a medical professional can result in body mutilation, infection, and even silicone blood clots that can lead to embolism and death. Other Specified The category other specified gender dysphoria can be used in cases where the presentation causes clinically significant distress or impairment but does not meet the full criteria for gender dysphoria. If this diagnosis is used, the clinician records the specific reason that the full criteria were not met. Unspecified The category unspecified gender dysphoria can be applied when full criteria are not met and the clinician chooses not to specify why they are not met. ICD-10/11 In the current iteration of the International Statistical Classification of Diseases and Related Health Problems (ICD-10), gender identity issues appear under Disorders of Adult Behavior and Personality in the category Gender Identity Disorders (F64), and include five diagnoses: transsexualism (F64.0), dual-role transvestism (F64.1), gender identity disorder of childhood (F64.2), other gender identity disorders (F64.3), and gender identity disorder, unspecified (F64.4). The ICD Working Group on the Classification of Sexual Disorders and Sexual Health is recommending that for ICD-11, gender identity concerns be moved from the psychological sections and is considering options that would list these concerns in their own separate chapter, as medical diagnoses, or as part of a new chapter on sexual health and sexual disorders. A 27-year-old assigned female at birth was referred to a gender identity clinic reporting having felt different as a child from other girls, although unable then to identify the source. As a young girl, she enjoyed playing sports with girls and boys, but generally preferred the companionship of boys. She preferred wearing unisex or boyish clothes and

resisted wearing a skirt or dress. Everyone referred to her as a tomboy. She tried to hide her breast development by wearing loose fitting tops and stooping forward. Menses were embarrassing and poignantly reminded her of her femaleness, which was becoming increasingly alienating. As sexual attractions evolved, they were directed exclusively to female partners. In her late teens, she had

one sexual experience with a man, and it was aversive. She began socializing in lesbian circles, but did not feel comfortable there and did not consider herself lesbian, but more a man. For sexual partners, she wanted heterosexual women and wanted to be considered by the partner as a man. As gender dysphoric feelings became increasingly pronounced, she consulted transsexual sites on the Internet and contacted a female-to-male transsexual community support group. She then set into motion the process of clinical referral. She transitioned to living as a man, had a name change, and was administered androgen injections. The patient's voice deepened, facial and body hair grew, menses stopped, and sex drive increased, along with clitoral hypertrophy. After 2 years, the patient underwent bilateral mastectomy and is on the wait list for phalloplasty and hysterectomy-oopherectomy. Employment as a man continues, as does a 3-year relationship with a female partner. The partner has a child from a previous marriage. (Adapted from case of Richard Green, M.D.) Intersex Conditions Intersex conditions include a variety of syndromes in which persons are born with anatomies that do not correspond with typical male or female bodies. Congenital Adrenal Hyperplasia. Congenital adrenal hyperplasia is a condition in which an enzymatic defect in the production of adrenal cortisol, beginning prenatally, leads to overproduction of adrenal androgens and, when the chromosomes are XX, virilization of the female fetus. Postnatally, excessive adrenal androgen can be controlled by steroid administration. The androgenization can range from mild clitoral enlargement to external genitals that look like a normal scrotal sac, testes, and a penis, but behind these external genitals are a vagina and a uterus. Other parts of the body remain feminized (i.e. there is breast development at puberty). Most people with congenital adrenal hyperplasia are raised female, except in cases of extreme virilization. If the parents are uncertain about the sex of their child, sometimes an intersex identity results. Gender identity usually reflects the rearing practices, but hormones may help determine behavior. Studies showed that sex-disordered children raised as girls had a more intense tomboy quality than that found in a control group. The girls most often had a heterosexual orientation, but higher rates of bisexual or homosexual behavior were reported. In those brought up female, about 5% show severe gender dysphoria, whereas about 12% of those assigned male are gender dysphoric. Androgen Insensitivity Syndrome. Androgen insensitivity syndrome was formerly called testicular feminization. In persons with complete androgen insensitivity and the XY karyotype, tissue cells are unable to use testosterone or other androgens. Therefore, the person appears to be a normal female at birth and is raised as a girl. She is later found to have cryptorchid testes, which produce the testosterone to which the

tissues do not respond, and minimal or absent internal sexual organs. Secondary sex characteristics at puberty are female because of the small, but sufficient, amount of estrogens, which results from the conversion of testosterone into estradiol. The patients usually sense themselves as females and are feminine. However, some experience gender conflicts and distress. In partial androgen insensitivity, persons may have a range of anatomical structures and gender identities. TURNER'S SYNDROME. In Turner's syndrome, one sex chromosome is missing, such that the sex karyotype is simply X. Persons with Turner's syndrome have female genitalia, are short, and sometimes have anomalies such as a shield-shaped chest and a webbed neck. As a

consequence of dysfunctional ovaries, they require exogenous estrogen to develop female secondary sex characteristics. Gender identity is typically female (Fig. 18-1). FIGURE 18-1Turner's syndrome in a patient aged 23. Note webbed neck, increased carrying angle, failure of breast development, and lack of pubic hair. (From Douthwaite AH, ed. French's Index of Differential Diagnosis. 7th ed. Baltimore: Williams & Wilkins; 234.) KLINEFELTER'S SYNDROME. An extra X chromosome is present in Klinefelter's syndrome, such that the karyotype is XXY. At birth, persons with Klinefelter's appear to be normal

males. Excessive gynecomastia may occur in adolescence. Testes are small, usually without sperm production. They are tall, and body habitus is eunuchoid. Reports suggest a higher rate of gender dysphoria. 5-α-Reductase Deficiency. In 5-α-reductase deficiency, an enzymatic defect prevents the conversion of testosterone to dihydrotestosterone, which is required for prenatal virilization of the genitalia. At birth, the affected person appears to be female, although some variance is visible. In earlier generations, before childhood identification of the disorder was common, these persons, raised as girls, virilized at puberty and usually changed their gender identity to male. Later generations were expected to virilize and, thus, may have been raised with ambiguous gender. Over half of those with 5-α-reductase deficiency identify as male as adults. There are reports of a small number of patients for whom early removal of the testes and socialization as girls have resulted in a female gender identity. Treatment. Because intersex conditions are present at birth, treatment must be timely. The appearance of the genitalia in diverse conditions is often ambiguous, and a decision must be made about the assigned sex (boy or girl) and how the child should be reared. Intersex conditions should be addressed as early as possible, so that the entire family can regard the child in a consistent, relaxed manner. This is particularly important because intersex patients may have gender identity problems because of complicated biological influences and familial confusion about their actual sex. When intersex conditions are discovered, a panel of pediatric, urological, and psychiatric experts works with the family to determine the sex of rearing on the basis of clinical examination, urological studies, buccal smears, chromosomal analyses, and assessment of the parental wishes. Education of parents and presentation of the range of options open to them is essential, because parents respond to the infant's genitalia in ways that promote the formation of gender identity. Although the label of boy or girl may be assigned to the infant on the basis of chromosomal and urological examination, the parents can then react to the child according to sex role assignment with leeway to adjust the sex assignment should the child act definitively as a member of the sex different from the one designated. Some studies have shown that an equal number of persons assigned to be female at birth choose to become male as adults as do those assigned to be male at birth who choose to become female. In general, the sex of rearing is the best predictor of later gender identity. In the past, many intersex infants underwent surgical procedures at an early age in order to normalize genital appearance. It is easier to surgically assign a child to be female than to assign one to be male, because male-to-female genital surgical procedures are far more advanced than female-to-male procedures. That is an insufficient reason, however, to assign a chromosomal male to be female. The standards of care related to intersex infants have changed considerably due to

work by intersex people and their allies, so that it is no longer recommended that infants have immediate surgical procedures performed. Instead, families are encouraged to choose a sex of rearing that is flexible, and to wait for the intersex person to decide on their own later whether to have surgery. Early surgeries are typically avoided now because they may interfere with later

reproductive capacity and sexual functioning. Transvestic Disorder Transvestic disorder appears in the DSM-5 section on Paraphilic Disorders, and is defined as a period of at least 6 months of recurrent and intense sexual arousal from crossdressing that causes clinically significant distress or impairment. Those who cross-dress are diverse, and many use cross-dressing as a form of entertainment or pleasure that does not cause distress, and therefore do not meet the criteria for this diagnosis. Crossdressing does not imply gender dysphoria—many people who cross-dress do so while retaining a gender identity that matches their assigned gender. Cross-dressers do not necessarily have a preoccupation with getting rid of their primary and secondary sex characteristics and acquiring the sex characteristics of the other sex. However, there are those who may be diagnosed with both gender dysphoria and transvestic disorder. The prevalence of transvestic disorder is unknown. It is more common in males and extremely rarely diagnosed in females, most likely due to comparable societal acceptance of women dressing in male-typical clothing. Those diagnosed with transvestic disorder often remember a fascination with female clothing in childhood. They may have periods of stress-related cross-dressing that produces sexual excitement, but also reduces tension and anxiety. There may be periods where the person buys a number of articles of clothing, wears them for sexual excitement, and then becomes distressed by their behavior and throws them out. Transvestic disorder can coexist with other paraphilic disorders, most commonly sexual masochism disorder and fetishistic disorder. Treatment. A combined approach, using psychotherapy and pharmacotherapy, is often useful in the treatment of transvestic disorder. The stress factors that precipitate the behavior are identified in therapy. The goal is to help patients cope with the stressors appropriately and, if possible, eliminate them. Intrapsychic dynamics about attitudes toward men and women are examined, and unconscious conflicts are identified. Medication, such as antianxiety and antidepressant agents, is used to treat the symptoms. Because cross-dressing can occur impulsively, medications that reinforce impulse control may be helpful, such as fluoxetine (Prozac). Behavior therapy and hypnosis are alternative methods that may be of use in selected patients. Preoccupation with Castration Preoccupation with castration does not appear in the DSM-5 but can be serious and lifethreatening if castration is carried out without medical supervision. This preoccupation occurs at times in people who do not have a desire to acquire the sex characteristics of

the other sex, but may be uncomfortable with their assigned sex, and their lives are driven by the fantasy of what it would be like to be a different gender. They may be asexual and lack sexual interest in either men or women. REFERENCES Adelson SL; American Academy of Child and Adolescent Psychiatry (AACAP) Committee on Quality Issues (CQI). Practice parameter on gay, lesbian, or bisexual sexual orientation, gender nonconformity, and gender discordance in children and adolescents. J Am Acad Child Adolesc Psychiatry. 2011;51(9):957–974. Carmel T, Hopwood R, Dickey L. Mental health concerns. In: Erickson-Schroth L, ed. Trans Bodies, Trans Selves. New York: Oxford University Press; 2014. Devor AH. Witnessing and mirroring: A fourteen stage model of transsexual identity formation. Journal of Gay and Lesbian Psychotherapy. 2004;8(1/2): 41–67. Drescher J. Queer diagnoses: Parallels and contrasts in the history of homosexuality, gender variance, and the Diagnostic and Statistical Manual. Arch Sex Behav. 2009;39:427–460. Drescher J, Cohen-Kettenis P, Winter S. Minding the body: Situating gender identity diagnoses in the ICD-11. Int Rev Psychiatry, 2012;24(6): 568–577. Erickson-Schroth L. Update on the biology of transgender identity. Journal of Gay & Lesbian Mental Health. 2013;17(2):150–174. Erickson-Schroth L, Gilbert MA, Smith TE. Sex and gender development. In: Erickson-Schroth L, ed. Trans Bodies, Trans Selves. New York: Oxford University Press. Grant JM, Mottet LA, Tanis J, Harrison J, Herman JL, Keisling M. Injustice at every turn: A report of the national transgender discrimination survey, Washington, DC:

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