

Disorders of sexual development health and social care essay



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Providing Holistic Care of Infants with Disorders of Sexual Development Amy

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10032. Contact: acr2151@columbia.edu Abstract Studies estimate that the

incidence of genital anomalies could be as high as 1 in 300 births. While it is

rare for an infant to present with truly ambiguous genitalia, it is plausible

that the primary care nurse practitioner will encounter a patient with some

form of disorders of sexual development (DSD) in his or her career. Cases of

DSD can be challenging due to complexities of diagnosis, gender

assignment, uncertain outcomes, treatment options, and psychosocial

stressors. This article discusses the management of infants with DSD and the

nurse practitioners' role in providing integrated primary care and

management of the child. The nurse practitioner is the ideal patient and

family advocate, providing education and anticipatory guidance about

diagnosis, complications, and treatment options; acting as a liaison between

specialists and translating specialty findings; helping the patient and family

through psychological and social stresses; and ensuring comprehensive,

holistic, and continuous primary care in childhood for the patient with DSD

and his or her family. Keywords: disorders of sexual development, DSD,

intersex, primary care What's Going on Down There: Providing Holistic Care

of Infants with Disorders of Sexual Development A new mother arrives at your

practice with her 9-day-old infant for a newborn visit. When reviewing the

chart, you notice that the parents were told that their baby would be female

based on prenatal ultrasound results. However, amniocentesis revealed a

karyotype of 46, XY. Therefore the parents had expected a son, but the

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delivery room genital exam revealed a micropenis with penoscrotal hypospadias and partially fused labia majora. No labia minora are present, no testes are palpable, and the anus is placed normally and is anatomically correct. How will you educate this parent about their child's disorder? How will you help them decide on what treatment, if any, to pursue? How will you answer their questions about their child's future gender identity, sexuality, and fertility?

Overview

The term disorder of sexual development (DSD) encompasses "congenital conditions in which development of chromosomal, gonadal, or anatomic sex is atypical" (Lee, Houk, Ahmed, & Hughes, 2006). Studies estimate that the incidence of true genital ambiguity in DSD is 1 in 5000 to 1 in 4500 births (Thyen, Lanz, Holterhus, & Hiort, 2006)(Ahmed & Rodie, 2009). It is estimated that the presence of genital anomalies, however, could be as high as 1 in 300 births (Ahmed & Rodie, 2009). While it is rare for an infant to present with truly ambiguous genitalia, it is plausible that the primary care nurse practitioner will encounter a patient with some form of DSD in his or her career. Cases of DSD can be challenging due to complexities of diagnosis, gender assignment, treatment options, and psychosocial stressors. This article will discuss the management of infants with DSD to inform the nurse practitioner's integrated primary care and management of the child with DSD. It is the nurse practitioner's role to provide education and anticipatory guidance about diagnosis, complications, and treatment options, act as a liaison between specialists, translate specialty findings, help the patient and family through psychological and social stresses, and ensure

holistic primary care in childhood for the patient with DSD and his or her family.

Etiology

It is important for the nurse practitioner to have an understanding of the cause and pathophysiology of the various DSDs in order to educate the family and patient in a developmentally and intellectually appropriate way. Many parents will feel guilt in that the origin of their child's DSD may have been their own doing somehow. It is important to continuously emphasize that DSDs organically and spontaneously arise, and that they are not implicated in any way. DSD were categorized into three distinct groups (See Table A1) by the Lawson Wilkins Pediatric Endocrine Society (LWPES) in 2006: sex chromosome DSD, 46, XY DSD, and 46, XX DSD, with some overlap between these groups (Lee et al., 2006). A revised nomenclature was also proposed by this group (See Table A2). Normal sexual development follows a complex pathway of genetics and gonadal, ductal and genital development (Murphy, Allen, & Jamieson, 2011). Therefore there are multiple steps in which interference or interruption of normal physiology will result in atypical genitalia at birth (See Figure B1).

Chromosomal DSD

Chromosomal DSD includes gonadal dysgenesis and ovotesticular DSD, caused by sex chromosome abnormality, and there is some overlap with 46, XX and 46, XY DSD. Infants with ovotesticular DSD have both ovarian and testicular tissues, and external genitalia are most often ambiguous.

Ovotesticular DSD has varied presentations, such as one ovary and one

testis, 2 ovotestes, or one ovotestis partnered with either an ovary or a testis
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(Murphy et al., 2011). One third of patients have chimeric (46, XY/46, XX) or mosaic (46, XY/47, XXY or 45, X/46, XY) karyotypes. The most common genotype, 46, XX, is exhibited by over half of patients with ovotesticular DSD, while only a small minority are genetically 46, XY (Holm, 2005). Virilization of the genitalia is related to the whether or not Müllerian ducts have developed into female structures or if the testis is able to secrete testosterone. The internal genitalia often coincide with the presentation of the external genitalia on that side; for example, a testis on the left side externally will coincide with Wolffian structures on the left side internally (Murphy et al., 2011). Gonadal dysgenesis refers to incomplete or partial gonads. If a testis is not formed completely, it is known as a dysgenetic testis, while an ovary that is incompletely formed is called a streak gonad (Ocal, 2011). Infants with complete gonadal dysgenesis may have a 46, XX, 46, XY, or Turner's syndrome karyotype (45, XO, or 45, XO/46, XX). These infants will have streak gonads, female external genitalia, and will often exhibit delayed puberty (Murphy et al., 2011). A rare form of complete gonadal dysgenesis is Sawyer syndrome, in which the genotype is 46, XY, but the internal and external genitalia are female. It is proposed that Sawyer syndrome is caused by a mutation in the SRY gene or other possible genes involved in testis determination, but the true cause remains unknown (Hyun & Kolon, 2004). Partial gonadal dysgenesis is marked by partial testicular development, and includes mixed gonadal dysgenesis (MGD) and testicular or ovarian regression (Hyun et al, 2004). The most common presentation of MGD is asymmetric development of the testes (Ocal, 2011). Patients with MGD show a streak or absent gonad on one side and a testis (often dysgenetic) on the other side. The most common genotype are 45, XO/46, <https://assignbuster.com/disorders-of-sexual-development-health-and-social-care-essay/>

XY, or 46, XY. The virilization of the external genitalia is dependent upon the androgen synthesis capability of the often dysgenetic testis, and when two dysgenetic testes are present, ambiguous genitalia often results from low levels of testosterone (Holm, 2005). Klinefelter's syndrome is sometimes included in partial gonadal dysgenesis, and is characterized by small digenetic testes due to low levels of testosterone.

46, XY DSD

Patients with 46, XY DSD have one of three underlying etiologies: defects in androgen synthesis and metabolism, resistance to androgens, or malformation syndromes. Infants with defects in androgen synthesis exhibit normal testes, but adrenal androgen production is compromised or the conversion of testosterone to dihydrotestosterone is inhibited, resulting in atypical genitalia. Common causes for undervirilization include CAH and 5 α -reductase deficiency (Murphy et al., 2011). Infants with androgen resistance are described as having either partial (PAIS) or complete androgen insensitivity syndrome (CAIS). Both conditions are caused by genetic defects located on the single X chromosome that interferes with androgen receptor function (Sultan et al., 2002). Infants with CAIS will have female external genitalia and two undescended testes which appear to be bilateral inguinal hernias. The testicles are still able to secrete Müllerian inhibiting substance, and so no fallopian tubes or uterus form (Murphy et al., 2011). PAIS is a more heterogeneous condition, comprising phenotypes that resemble CAIS to infertile males with male genitalia. Malformation syndromes of 46, XY DSD include somatic chromosomal abnormalities and single gene mutations, such as Trisomy 13 and Beckwith-Wiedemann syndrome.

46, XX DSD

Patients with 46, XX DSD typically exhibit female internal genitalia, including structures derived from the Müllerian duct and ovaries. However, external genitalia are virilized due to exposure to fetal or maternal androgens in utero. The most common cause of 46, XX DSD is congenital adrenal hyperplasia (CAH), although it does not always result in ambiguous genitalia (Demirici & Witchell, 2008). Other etiologies of 46, XX DSD include maternal exposure to androgens, such as androgen-secreting adrenal and ovarian tumors, exogenous ingestion, or rarely, PCOS (Hyun & Kolon, 2004). Ovarian development disorders are not well understood, but are proposed to be an imbalance of male- and female-promoting factors, tipping the balance in favor of testicular development. The theory that other genes are involved in testicular development is supported by a finding that 10% of males lacking an SRY gene are able to form testes (Hughes, 2008) (Blaschko, Cunha, & Baskin, 2012). A more in-depth understanding of the pathogenesis of ovarian development disorders will evolve as the nuance of the human genome unravels in future research.

Diagnosis

Diagnosis of the infant with DSD is often a complicated and multi-step process involving multidisciplinary input. While the nurse practitioner may not be directly involved in the process of developing a working diagnosis, it is important that he or she understand the medical conclusions that inform the diagnosis. An understanding of how the diagnosis was reached is crucial to planning treatment guidelines and reducing parental uncertainty through education of functionality and fertility possibilities. It is also important that

the nurse practitioner understands the psychological trauma that may be associated with multiple physical exams, blood draws, and imaging. The first step toward a correct diagnosis is a thorough family and prenatal history, including consanguinity, infertility, gonadal and urogenital malformations, pregnancy history, antenatal drug use, and maternal symptoms of androgen excess. A general physical exam should then be conducted with an eye for dysmorphic features in addition to an assessment of genital anatomy. Close attention should be paid to extragenital components such as hydration status, blood pressure, which may be altered in patients with CAH, as well as jaundice, and hyperpigmentation of areola. (Murphy et al., 2011). Common findings consistent with presentation of DSD are listed in Table 3.

Laboratory

The gold standard for testing infants with DSD includes karyotyping, imaging, hormone levels, serum electrolytes, and urinalysis (Lee et al., 2006). Karyotype with X- and Y- specific probe detection is necessary even when prenatal karyotype is available. Genetic evaluations of specific molecular studies to screen for the presence of mutations or gene imbalance has recently become available (such as SRY, SOX9, 17 β hydroxysteroid dehydrogenase, 5 α -reductase-2, and others), however specific testing is limited by cost and accessibility (Ocal, 2011). While significant progress has been made in determining the genetic etiology of DSD, molecular and genetic diagnosis is determined in only 20% of DSD cases (Lee et al., 2006). Only fifty percent of patients with 46, XY DSD will receive a definitive diagnosis (Ahmed & Rodie, 2010). Hormone levels of 17-hydroxyprogesterone, testosterone, gonadotropins, and anti-Müllerian

hormone, as well as serum electrolytes and urinalysis should be conducted and interpreted for normal values for chronological and gestational age (Lee et al., 2006). The results of most of these tests are available within 48 hours and can facilitate a working diagnosis while further investigations are planned.

Imaging

Imaging is a useful tool in visualizing internal structures, and an abdominopelvic ultrasound should be done to visualize internal anatomy. This can be extremely helpful in diagnosis by confirming the presence of a uterus, identifying gonads, and tracing the renal and genital tracts. However, Steven et al. reports that in a study of 18 patients with complex DSD from birth to age 14 years that pelvic ultrasonography was unable to identify Müllerian structures in 40% of patients (Steven, O'Toole, Lam, MacKinlay & Cascio, 2012). When necessary, a sinogram may be preformed, which entails injecting contrast into the urinary and/or genital tract to trace its course. When possible, this should be avoided to prevent unnecessary irradiation of the child. While diagnostic algorithms exist (Ogilvy-Stuart & Brain, 2004), there is such a varied spectrum of findings that no algorithm can be recommended for the diagnosis of all presentations of DSD.

Gender Assignment

Recent recommendations suggest gender assignment should be done in the first weeks of life, not in the immediate perinatal period, as was historically done, to give the multidisciplinary team time for a full evaluation and consultation before arriving at a working diagnosis and treatment plan.

Gender assignment of infants with DSD is based on genital appearance, <https://assignbuster.com/disorders-of-sexual-development-health-and-social-care-essay/>

surgical options, diagnosis, potential for fertility, views of the family, cultural practices, and the need for lifelong therapy. Determining the appropriate gender of rearing in infants with DSD is difficult, and no clinical data can reliably predict gender identity. It is tantamount, therefore, that the parents, as the primary care giver and legal guardian of the child, be involved in the gender assignment process. It is the role of the nurse practitioner to make sure they are adequately educated and supported in order to make an informed decision. The majority of patients with DSD are assigned a female gender during initial management. More than 90% of patients with 46, XX CAH (Dessens, Slijper & Drop, 2005) and 46, XY CAIS are assigned as female in infancy (Mazur, 2005). Genetically female patients with CAH have a high potential for fertility and studies have shown that most have a female post-pubertal gender orientation (Dessens, et al., 2005). Genetic males with complete gonadal dysgenesis typically have entirely female external genitalia and are traditionally raised female (Barthold, 2011). It is suggested that all 46, XY patients with micropenis should be raised male due to the possibility of further virilization during puberty and maintaining sexual function (Ocal, 2011). Male or female gender may be assigned in patients with PAIS, 5- α -reductase deficiency, 46, XY partial gonadal dysgenesis, mixed gonadal dysgenesis and ovotesticular DSD. Studies have documented favorable fertility potential in patients with 5- α -reductase deficiency assigned as male (Lee et al., 2006) but more comprehensive studies are needed to provide empirical evidence. When deciding on the gender of rearing for a patient with ovotesticular DSD, the potential fertility based on the gonadal differentiation and genital development should be considered, as well as an assessment of whether genitalia can be consistent with the chosen sex (Lee <https://assignbuster.com/disorders-of-sexual-development-health-and-social-care-essay/>)

et al., 2006). In mixed gonadal dysgenesis, prenatal androgen exposure, testicular function, phallic development and gonadal location should be taken into account. Infants with 45, X/46, XY mixed gonadal dysgenesis are usually assigned the gender most congruent with testicular function and external masculinization (Lee et al., 2006). More than ever, families are considering a decision to raise children with DSD as gender neutral in an effort to avoid influencing their child's gender identity. Gender assignment in patients with severe DSD is complicated and there is no way to ensure that gender assignment will be congruent with the child's gender identity as he or she matures. The role of the nurse practitioner should be to educate parents about the status of the working diagnosis, the possibility of fertility and sexual function, and the consensus of specialty opinions so that they may make an informed decision. Parents of children with DSDs should be counseled that children with DSDs are more likely than the general population to feel that their gender assignment at birth was incorrect. It may be helpful to describe the process of sex determination and adult implications with the family, including not just physical outcomes of fertility and puberty, but the psychological factors involved. Definitions of gender identity, sex-role behavior, and sexual orientation may help describe such psychosocial affects. Every effort should be made to facilitate shared decision making between health care providers and the family. With the exception of a conclusion that will cause obvious harm to the child, the gender of rearing should ultimately be the parental decision, as they are the legal guardians and primary care givers for the child.

Treatment

As the primary health care provider, the nurse practitioner provides patient- and family centered comprehensive care in a setting that fosters shared decision making. The nurse practitioner may be the first outpatient health care provider that patients with DSD and their families see in an outpatient setting. It is therefore important to understand the etiology behind the child's DSD diagnosis and the various testing that the child and the family have been put through in order to create a trusting patient-provider relationship. The nurse practitioner should provide an open, non-judgmental caring atmosphere where the patient and family can be comfortable to share feelings, admit fears, and ask questions. Good communication skills are critical to successful treatment and management of the patient with DSD.

Medical Treatment

Many DSDs are associated with underlying medical problems that require management. It is the role of the nurse practitioner to educate patients and families about their treatment plan and its medical basis. For predictable problems, care can be organized as a routine (Schober et al., 2012).

However, because of lack of outcomes data in DSD, providers must keep an open mind, as medical advances and further research may provide answers to what may now seem unpredictable. Hormone Therapy. Underlying endocrine disturbances are present in many cases of DSD and therefore require long-term hormone therapy. In patients with androgen synthesis or action disorders who are raised as males, testosterone replacement therapy may help to increase penile size (Moshiri et al., 2012). Patients with gonadal dysgenesis may not reach puberty due to a lack of functioning gonadal

tissue, and may require hormone therapy to induce puberty. In males, intramuscular testosterone injections are administered, while females receive estrogen supplements and added progesterone to induce menses in those patients with a uterus (Lee et al., 2006). Patients with 5- α -reductase deficiency are given dihydrotestosterone. Steroid supplementation when necessary is vital for the management of patients with CAH, as adrenal insufficiency can result if not carefully monitored. A study of 11 parents of children with CAH suggested that the most predominant worry was about the management of medications (Kogan et al., 2012). Therefore it is important for the nurse practitioner to provide a written treatment plan and offer strategies to help facilitate medicine adherence. Surgery. Surgery in the first year of life in patients with DSD is controversial. Many patient advocate groups have pushed for deferment of all genital surgeries until the affected individual is capable of comprehending the lifelong irreversible consequences and consenting or assenting to surgery. However, it is impossible to know at what age a patient with DSD will be ready to make such a decision, and the psychosexual affects of raising a child with ambiguous genitalia has not been studied. Therefore, the LWPES recommends that parents should make the ultimate decision of whether to defer surgery or not. Emphasis should be placed on maintaining innervation and sexual function of genitalia rather than cosmetic reconstruction. Some studies report that surgery in the first year of life relieves parental stress and facilitates parent-child bonding and attachment, but there is a need for studies providing empirical evidence of this association (Baskin, 2004). In cases of clitoromegaly, surgery should only be considered in cases of severe virilization (Prader stages 3-5, see Figure B2); patients with mild or moderate

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clitoromegaly should defer surgical reconstruction until adolescence (Houk & Lee, 2008). Often cosmetic surgeries are now done in conjunction with repair of the urogenital sinus. Part of surgical therapy is anticipated revision and additional reconstruction at the time of puberty, and vaginal dilation does not occur until after pubarche. Patients with an absent or inadequate vagina typically require a vaginoplasty which is usually done during puberty, when the patient can be fully invested and participate in vaginal dilation. In patients with DSDs associated with hypospadias, various surgical techniques including chordee release and urethral reconstruction can be used in conjunction with testosterone supplementation therapy. When parents are deciding on the gender assignment, care must be taken to provide realistic outcomes and discuss the complexity of phalloplasty in adulthood.

Neophalloplasties can be successful in adulthood, but have little erectile function. An erectile prosthesis can be inserted for coital intercourse, but is associated with high morbidity (Lee et al., 2006). Parents of children with CAIS or PAIS who will be raised female may choose to remove the testes at the time of diagnosis to facilitate estrogen replacement therapy. Early removal of dysgenetic testes has been recommended due to their potential for malignancy; however, the earliest report of testicular malignancy in CAIS is 14 years, and therefore parents may decide to defer surgery until adolescence (Lee et al., 2006). MGD patients with streak gonads should have testes removed in early childhood. Patients with androgen synthesis defects who will be raised female should receive gonadectomy before puberty.

Patients with bilateral ovotestes have the potential for fertility due to ovarian tissue. Separation of ovarian and testicular tissue can be difficult, and so must be accomplished early in life (Lee et al., 2006). The nurse practitioner <https://assignbuster.com/disorders-of-sexual-development-health-and-social-care-essay/>

must work with the family closely, and address the uncertainties of DSD management with the parents. Because sex assignment is not inextricably linked to surgical revision of the genitals, there is an opportunity to defer surgery until the affected child may become involved in management plans that have lifelong consequences. Gonadal Malignancy. Patients with DSD who bear Y chromosome material in their karyotype are at increased risk for gonadal malignancy, specifically type II germ cell tumors. Type II germ cell tumors arise from immature germ cells, which are often found in gonadal tissue of patients with DSD. Some markers of these germ cells seem to provide them with the ability to proliferate and suppress apoptosis (Pleskacova et al., 2010). An additional factor in malignant potential is the TSPY protein, which may cause gonadoblastomas, is over-expressed in germ cells of DSD gonads. The risk of a germ cell tumor has been estimated at 15% in patients with PAIS, up to 30% in patients with gonadal dysgenesis, 0.8% in patients with CAIS, and 2.6% of patients with ovotesticular DSD (Cools, Drop, Wolffenbuttel, Oosterhuis & Looijenga, 2006). 46, XY or 45, X/46, XY patients with gonadal dysgenesis seem to be at the greatest risk. Early identification and careful monitoring of patients at increased risk of gonadal malignancy is important to clinical management, especially patients who have deferred gonadectomy and have retained gonads.

Psychosocial Care

Care of patients with DSD and their families should inherently include psychological support. Families and patients require ongoing counseling by experienced health care providers to deal with the challenges that accompany a diagnosis of DSD. Family support. The birth of a child with DSD

can be psychologically stressful for any family. Some of these stressors include uncertainty about gender of upbringing, genital appearance, inadequate coping mechanisms, ambiguous gender role, behavior, or identity, and altered genital function (Creighton, Chernausek, Romao, Ransley, & Salle, 2012). Studies also have identified that parents may feel they cannot disclose information about their child's DSD to family and friends, removing established social support systems (Brinkmann, Schuetzmann & Richter-Appelt, 2007). Many of these negative outcomes could be avoided, or at least minimized, by active disclosure and sensitive supportive interactions with families. While it is the role of the nurse practitioner to advocate for the child with DSD, important decisions are often made on their behalf early in life by family members in collaboration with the medical team. It is therefore tantamount that holistic care encompasses the family unit, allowing them to address fears, uncertainties, and reflect upon their experience with the birth and early management of their child. Families may require substantial support in accepting the uncertainty of gender assignment and sex of rearing, especially in cases where diagnostic tests do not provide a foreseeable solution (Crissman et al, 2011). Considerable psychological attention should be placed on parents' reactions to a traumatic birth experience, unkind words from careless health care professionals, incorrect information from the internet, and unresolved fears which may resurface later in the child's life (Brain et al., 2010). Of particular importance is discussing how to inform family and friends while gender assignment is pending, which should focus on honesty in order to avoid creating a sense of shame (Consortium on the Management of Disorders of Sexual Development, 2006). Individual support. First and foremost, the nurse practitioner is an <https://assignbuster.com/disorders-of-sexual-development-health-and-social-care-essay/>

advocate for the child. He or she must constantly consider the long term effects of decisions made by the medical team and parents. Studies suggest that there is a greater risk of depression, social and sexual avoidance, and dissatisfaction with self-image in individuals with DSD, but quality of life data is limited, and may vary with type of DSD (Barthold, 2011). Nonetheless, quality of life of patients with DSD can be significantly improved by positive and adaptive coping mechanisms fostered by psychosocial support. Like all other children, children with DSDs are given a gender assignment at birth, but it is impossible to predict which gender any child will identify with in the future. Little definitive evidence has been uncovered about the role gender identity and the effects of androgens on the brain (Cohen-Kettenis, 2010). An estimated 8.5-20% of patients with DSDs experience gender dysphoria, with higher rates of gender dysphoria in patients with severe DSD, such as cloacal exstrophy or penile agenesis (Furtado et al., 2012). Genetically female patients with CAH have a high potential for fertility and studies have shown that most have a female post-pubertal gender orientation (Dessens, et al., 2005). As much as 25% of patients with PAIS, biosynthetic androgen defects and incomplete gonadal dysgenesis expressed dissatisfaction with the gender they were assigned at birth, no matter what gender that was (Murphy et al., 2011). Sixty percent of patients with 5-a-reductase deficiency assigned as females and most assigned as males who virilize at puberty live their adult lives as males (Murphy et al., 2011). Individuals with cloacal exstrophy reared female show variability in gender identity outcome but more than 65% live as female (Lee et al., 2006). Psychological support should be considered an ongoing process, and not a routine treatment, in

which the child and family can initiate treatment when they determine it is needed.

Patient Centered Care

Patient-centered care should focus clearly on the well-being of the child and his or her family. First, medical and surgical care should be provided to the child to prevent complications and maintain physical health. Surgical and hormonal treatments should be delayed, if possible, until the patient can actively participate in decision making. The nurse practitioner should consider whether irreversible interventions are truly necessary for the health of the child or are simply being offered as a way to assuage parental fears and uncertainty. Mental health care providers should be involved in the advising the decision-making process and fostering effective coping skills. Child and family psychosocial stress should be explicitly addressed with the support of mental health care and support groups. The nurse practitioner should respect parents concerns honestly, empathically, and explicitly and foster a trusting environment where patients and families can feel comfortable expressing such concerns. Additionally, the nurse practitioner must minimize feelings of shame by avoiding stigmatized terminology, excessive genital examinations or photography, and involving unnecessary examiners for the purpose of teaching or speculation. It is also important that the nurse practitioner not impose a notion of physical or social norms on the patient or family, as no one definition (such as genital size/shape, gender-specific behaviors) is suitable for all patients. Finally, the nurse practitioner must always explain findings and answer questions truthfully to

the patient and family. When possible, questions should be answered promptly and honestly.

Conclusion

The nurse practitioner, as the primary care provider for a child with DSD, must support the family by providing care, protection, and participation in decisions. As the primary care provider, the nurse practitioner must understand the etiology and medical basis behind the child's DSD diagnosis. He or she must provide patient- and family-centered care in medical and psychosocial treatment of DSD. Cases of DSD can be challenging due to complexities of diagnosis, gender assignment, treatment options, and psychosocial stressors, but the nurse practitioner is suited to provide holistic care that can minimize negative outcomes for these children and their families.