Review Article

Challenges and Opportunities in Adolescent Gynecology Patients with Surgically-Treated Congenital and Acquired Anomalies: Transition of Care from Pediatric to Adult Surgery

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A B S T R A C T

The transition from adolescence to young adulthood in patients with reproductive health care needs such as disorders of sexual development and congenital anomalies is a complex process that occurs over several years. The transition process for these patients is still poorly understood. The patients with disorders of sexual development and reproductive issues have specific and unique issues regarding timing of disclosure of diagnosis, genital examinations, gonadectomy, vaginal treatments, surgical procedures, hormone replacement therapy, use of long-term medication, and potential cancer screening. The purpose of this review is to briefly describe complex genital malformations and their associated anomalies with long-term concerns and then provide an overview of what has been published at this time regarding the transition of care to provide some guidance for providers who care for those patients.
Key Words: Adolescents, Disorders of sexual development, DSD, Intersex, Congenital adrenal hyperplasia, Gynecologic anomalies, Uterovaginal agenesis, MRKH syndrome, Ovarian conditions, Transition of care

Introduction

The transition from adolescence to young adulthood even in healthy young women is challenging, but there is an added complexity when those patients have undergone reproductive surgical procedures during their childhood. Adolescents with such issues will benefit from communication and long-term planning about the transition from pediatric to adult care.[1] This need for provision of age-appropriate care and continuity between pediatric and adult care has been recognized in other conditions such as cardiac anomalies, cerebral palsy, rheumatologic disorders, cystic fibrosis, and diabetes.[2–7]

Transition guidelines have been developed by the American Academy of Pediatrics, the American Academy of Family Physicians, and the American College of Physicians but none are specific for gynecology and reproductive issues.[8] Patients with disorders of sexual development (DSD), previous reproductive neoplasms (ovarian tumors), and/or other gynecologic malignancies might respond better to the transition if this has been discussed by the pediatric providers caring for the adolescent/young adult and her parents.[9,10]

Improvements in the care of these patients resulting in favorable outcomes have shifted expectations from mere survival to that of an increasing emphasis on wanting to be normal in genital appearance and function, including sexual and reproductive health.[11] This has created new challenges for health care providers and patients and it is increasingly recognized that clinicians managing these patients need specific knowledge and expertise to ensure optimal long-term outcomes. A key to that outcome is the successful transition of the patients into adult care. There is little in the way of evidence-based research on which to rely for managing the transition of these patients as they approach adulthood. Most of the available literature centers around initial studies on transition of care with patients with DSD conditions.[12] The purpose of this review is to summarize available data on transition from pediatric to adult surgery in general and more specifically as it relates to patients with complex gynecologic malformations and associated anomalies. We include a brief summary of these malformations and make recommendations on transition timing, content, and structure.

Transition of Care

Transition of health care has been defined as the purposeful planned shift from receiving pediatric health care to receiving primary and specialty adult-oriented care that addresses the medical, psychosocial, and educational/vocational needs of adolescents and young adults with chronic physical and medical conditions.[13,14] Originally described as it related to pediatric patients with complex chronic medical conditions such as cerebral palsy, diabetes, and renal failure, transition of care has more recently comprised a far wider spectrum of disease processes. In the

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past 5 years, there has been a marked increase in publications and discussions centered on transition of care from pediatric to adult surgery, addressing conditions such as anorectal malformations, Hirschsprung disease, congenital heart disease, solid organ transplantation, and myriad others.[15]

The goal of successful health care transition is the optimization of health outcomes and quality of life for adolescents with chronic conditions by providing personalized care in which the patient is given a sense of control.[11] In adolescence, young people with particular needs in medical care gradually transfer from child-centered to adult-centered medicine. It is important to recognize that transition of care is a process that takes years and progresses as the patient passes from early adolescence into young adulthood and involves a fundamental philosophical shift from family-centered to individually focused care.[5,15]

Creighton and Wood[11] wrote that transition is not simply a move from one healthcare team to another but is an important phase in adolescent life when there exists a shift in the doctor-patient relationship. Whereas in pediatric care, the physician's principle interaction is with the parents with reference to the child, in adolescence, the aim is to create a reversal with the primary relationship being the one with the doctor and the patient with the support from the parents.

The value and clinical importance for transition from pediatric to adult care has been shown in studies of patients with various other chronic medical conditions[2,4,6,7,15,16] and should be the same for patients with reproductive issues including DSDs.

Literature Review and Models of Transition of Care

A 2006 international consensus recommends the approach to DSD patients be multidisciplinary, holistic, and patient-centered,[17] and ideally, the transition of care would represent a multidimensional and active process that targets the physiological, medical, psychosocial, and educational needs of young people. The current problem exists that although models of best practice have been established in the pediatric care services for those with DSD, the same type of care has not been matched by similar efforts in the adult services.[18]

Additional factors that have hampered development of transitional care is that although the understanding of the molecular underpinnings of DSD has undergone considerable development over the past 20 years, that progress has not been matched by research on the long-term effects on the physical, psychological, social, educational, sexual, and vocational outcomes for these patients.

This lack of data is somewhat due to a legacy of decades of nondisclosure such that individuals with DSD were unaware of their underlying diagnosis into adulthood despite the parents and clinicians having that information. The effect of nondisclosure limited the availability of clinical information about DSD patients and fueled the stigma and shame associated with the diagnosis of DSD and might have reduced the likelihood of these affected individuals seeking care.[18]

Although this lack of data on long-term outcomes of individuals with DSD is a documented challenge over the past decade multiple articles have been published related to transition of care of adolescents with sexual developmental disorders.[10,11,18–23]

In an attempt to define the initial requirements of a transition service for young women with DSD, Liao et al prospectively assessed 50 adolescent girls with DSD at University College Hospital (London) during a 6-month period (October 2009 to March 2010) and the main clinical needs of each individual were categorized as endocrine, gynecological and/or urological, or psychological. Clinicians then further categorized the urgency of each need. Twenty-eight percent of the patients were considered to have an urgent clinical need, which, in most was a psychological issue.[24]

The need for psychological support during transition has been additionally underscored by research that assessed 60 adolescents aged 13-16 years who had been diagnosed with DSD. Although the general psychological well-being of the group with DSD did not show substantial detriment compared with that of their unaffected peers, individuals who required hormonal therapy to induce puberty showed impaired well-being compared with those who underwent spontaneous puberty. Additionally, adolescents with DSD showed reduced sexual activity compared with their unaffected peers.[25]

Little else is known with respect to transition of patients with DSDs or reproductive concerns. However, one might be able to extrapolate from findings from a research group that identified 5 factors that were likely to contribute to successful transition outcomes when transitioning diabetic patients.[26] They included: (1) initiation of the transition process 1 year before the intended date of transfer; (2) assessment of the individual for readiness for transition; (3) provision of guidance and education to the patient and the family; (4) using available online resources; and (5) maintenance of continual communication between pediatric and adult services.

Although this set of 5 recommendations from the diabetologists might be generalizable to the gynecologic population, it is important to note that the American Academy of Pediatrics suggest that the transition process begin in the early teen years. Therefore, starting the conversation earlier than 1 year might be beneficial.

This same research team recommended that the transition process be managed and coordinated by a specific clinician, who would be able to provide continuity of care for the individual and ensure that communication is maintained between the patient and their family as well as with the pediatric and adult teams.

Researchers who conducted a series of semistructured interviews with young adults with various long-term conditions, all of whom had experienced transfer of care from pediatric to adult services, interviewed parents and patients and determined that the transition was complicated by methodological and cultural gaps between the two services. There was a lack of multidisciplinary team approach, reduced attention paid to psychological issues, and a need for the individual to assume responsibility for all practical arrangements such as clinic visits and appointments in adult services.

They therefore identified 2 central issues in their recommendations for improving transition: (1) better preparation for parents and patients; and (2) better organization and communication between pediatric and adult services. Of note in this research, parents and patients expressed a preference for meeting the new adult health care providers before the transfer took place.[27]

This need for improvement in communication behavior, information management, and for psychological support that is designed and specific to the target group was also noted in a systematic literature review of the care experience of girls and young women with Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome. Their review identified the care requirements and recommendations for structuring health care of girls and young women with MRKH and well as studies of medical care during the adolescent transition period for various other diseases. They noted 9 publications on MRKH care and concerns and 10 publications on the transition from adolescence into adulthood for other chronic diseases.[28] Medical care requirements and measures were identified for the following areas: diagnosis during adolescence and organization of medical care, reactions to the diagnosis, functional infertility, psychological stress and threat to self-image, contact with others, and dealing with MRKH coping strategies. The investigators noted that there were only a few studies that addressed care requirements and that provided recommendations for transition management. The primary need for improvement was found particularly for communication behavior and transmission of information for patients with MRKH. The authors mention that specific educational courses might be needed to train medical staff on how to interact with these girls and young women. There are no specific studies identified in this review that evaluated transition for MRKH patients and as such there is a need for research in transition of patients with reproductive issues and DSDs.

Simoes et al reported data from a German-funded TransCareO project that examined in a mixed method design health care provisional deficits, preferences, and barriers in health care access as perceived by female adolescents affected by MRKH.[20] To our knowledge, this is the first study to provide a comprehensive assessment of the provisional situation and in addition provide a full examination of the sociocultural findings, values, beliefs and attitudes, and competing demands that influence adolescents with MRKH in transition. In their initial interviews with MRKH patients they identified critical elements of MRKH-related care and then completed in a communicative validation workshop with MRKH patients ages 10-19 years at time of their diagnosis. They noted that the MRKH patients experienced deficits predominately in the availability of information in their present health care services and that they perceived those deficits as crucial barriers to obtaining appropriate health care at the time of diagnosis and in transition of care. The TransCareO project comprised the first phase of a complex intervention to improve health care for female patients with rare genital malformation by initiating evidence-based transitional care with prospective follow-up.

Personal stories from patients with DSD reflect the lack of informed health care providers as well as the desire for accurate information regarding their diagnoses. DSD patients who are members of the Inter-Act Youth blog (http://inter-actyouth.tumblr.com) post personal stories regarding their experiences. What is often reflected is that many adult physicians who treat adolescent and adult patients with DSD typically receive little to no education about the various types of DSD and the related medical, surgical, and psychological management.[29,30] As a result, affected patients are often unable to find satisfactory medical care and might result in them exiting the medical system as they age out of childhood.[31] This loss of access to care for patients with chronic illness or special health care needs has been associated with a deterioration in health care status.[32]

What is continued to be noted in the available literature regarding MRKH and might be extrapolated to other reproductive conditions in young women, is that patients prefer decision-making on health issues to be completed in personal communication with medical professionals. This includes validation of information in relation to the patient's personal situation as well as in open discussion of fears, disease-related burdens, and health concerns, thus empowering the patient for informed consent. Because information ranks among the highest concerns from this research and access to knowledgeable clinicians might require long distance travel, the adolescents experiencing this barrier suggest use of e-health facilities.[20]

Reproductive Conditions with Need for Transitional Care

The congenital and acquired gynecologic anomalies and other reproductive surgical issues that might benefit from planned transition of care are described briefly in the following sections.

Gynecologic Anomalies

These anomalies occur embryologically with the development of Müllerian ducts and predominantly present in the adolescent at the time of menarche with either absent or obstructed menstruation.

Uterovaginal Agenesis (MRKH Syndrome)

Uterovaginal or Müllerian agenesis results in failure of the uterus and the vagina and occurs in approximately 1 in 5000 female live births. Associated congenital anomalies of the upper urinary tract are seen in 30%-40% of cases. In addition to the associated renal anomalies, skeletal, and auditory abnormalities have also been documented and evaluation for such is indicated.[17]

These patients commonly present in adolescence with primary amenorrhea in the presence of otherwise normal pubertal development. Young women with this condition will not menstruate or carry a pregnancy because they do not have uterine function, however they might have their own genetic children using oocyte retrieval, assisted contraception, and use of a surrogate. As such, this diagnosis is often devastating to the adolescent and family.

The initial therapy is that of education and psychological support. For those who choose vaginal creation, vaginal dilation is the most risk-free and effective treatment to

improve vaginal length and as such is considered the first-line treatment.[33] There are a variety of surgical reconstructive procedures described for use when dilation is not successful. Infertility is a consequence of MRKH that is difficult for affected patients to accept. In 2014, the first clinical uterine transplant was reported[34] and in 2015 a successful live birth was reported after uterine transplant.[35] There have been multiple reports of live donor uterine transplants since that time but this option is still considered experimental.[36]

Obstructed Menstruation

Menstrual obstruction might result from an anatomic obstruction at any level from the hymen to the uterus. The true incidence of these anomalies is not known and varies depending on whether a general population is evaluated at the time of obstetric delivery or one with a history of infertility or habitual miscarriage.[37–39]

Patients often present with abdominal pain associated with amenorrhea or persistent dysmenorrhea with or without an abdominal mass. These anomalies will require surgical correction but the type of procedure will vary on the basis of the type and location of the anomaly present. Magnetic resonance imaging is the gold standard to evaluate the anomaly and to assist in planning surgical treatment.

Typically, for low obstructions such as imperforate hymen or low transverse vaginal septa, primary excision via a vaginal procedure will result in normal menstrual egress and allow normal sexual function. Higher obstructive lesions as seen with high transverse vaginal septa or cervical agenesis will require complex surgical treatment and is best completed with a multidisciplinary team of surgeons.[11] On occasion, the definite treatment for the obstructed vagina will require interposition vaginoplasty, replacing the obstructed vagina with a segment of bowel.

Currently obstetric outcomes after obstructive anomalies are only reported in small case series and case reports. This makes decision-making for more complex disorders to be best completed within multidisciplinary team with experience with these anatomic and reproductive anomalies.[11]

DSD

DSD are relatively rare congenital conditions in which the development of the chromosomal, gonadal, or anatomic sex is atypical. Data on the prevalence and incidence of DSD are limited, although it is thought that the incidence is increasing.[40] By strict definition of "ambiguous genitalia at birth," the incidence of DSD is approximately 1:2000,[41] but an incidence comprehensive of all abnormalities is much more difficult to define. These patients might present at birth with ambiguous genitalia or later in childhood with virilization or at puberty or in adolescence with primary amenorrhea.

These DSDs are categorized into 3 main types: (1) 46, XX DSD: This includes disorders of gonadal development such as ovotesticular DSD and gonadal dysgenesis as well as androgen excess conditions as in congenital adrenal hyperplasia and vaginal agenesis, also known as MRKH syndrome or Müllerian aplasia. (2) 46, XY DSD: This includes disorders of gonadal development such as Swyer's syndrome and disorders of androgen synthesis or action such as androgen insensitivity syndrome. (3) Sex chromosome DSD: These are conditions of gonadal ambiguities or absence as a result from chromosomal abnormalities or syndromes. Included in this category are Turner syndrome 45 X as well as 46, XY mixed gonadal dysgenesis.

Table 1
An Adapted Checklist for Assessing Whether an Adolescent or Young Adult with 46, XY DSD or 46, XX DSD s Prepared to Transition Their Health Care to Adult Medical Specialists

List Item
Health Care Skills
I have decided who will be my DSD physician for adult care.
I can explain my DSD to unfamiliar physicians.
I can find information about my condition online, and know how to connect with DSD advocacy and support groups.
I can schedule and keep a calendar of my own medical appointments.
I wear medic alert jewelry to alert others of my life-threatening allergies or conditions (when needed).
I prepare and ask questions of my health care team members.
I can explain the side effects of my medications.
I can explain complications associated with my DSD and how to avoid them, including the importance of healthy eating and exercise.
I know which physical and mental health symptoms of my DSD require urgent care and where to go for that care.
I can describe how my DSD affects pubertal development, sexual functioning, and fertility.
I keep records of my menstrual periods (when needed).
I understand safe sexual practices.
I am informed about family planning and have access to contraceptives (when needed).
I know how to connect with genetic counselors to discuss my condition.
I perform breast self exams (when needed).
I perform testicular exams (when needed).
Health History Knowledge
What is DSD? What type of DSD do you have?
What medications do you currently take? How much and how often?
Why do you take these medications? What happens if you take too much or too little?
When and how was your DSD diagnosed?
Have you had any surgeries for your DSD? If yes, what procedures and when?
Do you have a copy of your medical records? If not, do you know how to get this?
What is the natural history of your DSD (if known)?
Are you fertile?
Do you know what kinds of mental health and health care specialists are available to help you as you get older (eg, psychiatrist, reproductive endocrinologist, couples counselor)?

DSD, disorders of sexual development.Adapted from Gleeson and Wisniewski[29, pp 130] and Hullmann et al.[10, pp 157]

Ovarian Conditions

Ovarian neoplasms are more commonly seen conditions in the pediatric and adolescent patient that might result in reproductive surgical intervention. Such interventions might include ovarian cystectomy, untwisting of an adnexal or ovarian torsion, with or without oophoropexy, or might result in oophorectomy. Although there is minimal consensus on the appropriate monitoring for such patients with benign ovarian lesions, when transition of care for these patients is completed, some coordination should be undertaken as with any other reproductive anomaly.

Preparation for Transition

The transition of care of these patients will encompass a multistep process. One of the first steps in preparation for

transition of care includes collation of the patient's medical record and should include operative reports, pertinent genetic and other laboratory testing results, pathology reports, radiologic imaging of sex organs, and bone density as well as any psychological evaluations.[19,22]

There is no evidenced-based information regarding how to transition patients with reproductive issues. However, on the basis of the American Academy of Pediatrics general recommendations for transition of care in the context of chronic illness,[13,14] Hullman et al[10] suggests a strategy for transitioning cognitively normal DSD patients from pediatric to adult providers that involves a sequential process beginning at ages 12-13 years and concluding between the ages of 18-21 years. The initial step includes the introduction of the topic of transition and education regarding how health care services and treatment goals change as DSD patients mature. Over the next 2 years, a transition plan should be developed that includes the anticipated age of transition, recommended adult providers, and outlines the responsibilities of the pediatric providers, the patient, and the caregivers to ensure that the transition occurs. This transition plan is reviewed as the patient ages, and copies are given to the patient. Finally, when the patient is 18-21 years of age, the transition plan is implemented. This plan should include a summary of the patient's previous care, including medical and surgical, that is provided to the patient and all adult providers who the patient will be seeing. The summary should include all operative reports with intraoperative findings, a description of all procedures performed, relevant pre-/intra-/postoperative pictures, a summary of any postoperative complications and their resolutions, and any planned or discussed future procedures.[23]

The goals of these discussions are to identify and discuss with the patient and her parent/caregiver their needs and goals in self-care.[8]

A sample transition readiness assessment tool is shown in Table 1. A similar assessment tool might also be completed by the parent. Both of these will help to assess the patient's awareness of their own medical condition, needs, concerning symptoms, medications, allergies, and their ability to explain their medical needs to others.

It will be also be helpful to provide resources for families for additional information (Table 2).

Table 2
Resources for Health Care Providers, Patients, and Families

Sexual development/overview of sexual differentiation
- https://pie.med.utoronto.ca/htbw/SexDevelopmentAnOverview/SexualDifferentiation

Transition resources from adolescent to adult care
- GotTransition.org
- NASPAG.org

DSD
- DSD translational research
- Network (http://dsdtrn.org)
- Accord Alliance (accordalliance.org)
- DSD families (dsdfamilies.org; http://interactyouth.tumblr.com)
- AIS DSD support group (AISDSD.org)

Müllerian agenesis
- Beautifulyoumrkh.org
- MRKH.org

Androgen insensitivity
- Aisdsd.org

Turner syndrome
- Turnersyndrome.org

Congenital adrenal hyperplasia
- Caresfoundation.org

Legal issues
- Advocates for informed choice (aiclegal.org)

AIS, androgen insensitivity syndrome; DSD, disorders of sexual development.

Table 3
Physician and Staff Providers and Roles in the DSD Multidisciplinary Team

Provider	Role
Primary care provider	Well care and direct specialty referral
Neonatologist	Newborn care/workup of infant with DSD
Geneticist	Consultant evaluation, testing, and counseling
Endocrinologist	Directs hormonal therapy as needed at all stages of development
Psychologist/psychiatrist	Provides counseling as needed to the patient and family
Surgical specialist team • Pediatric surgery/urology • Adult gen surgery/urology • Pediatric and adolescent gynecology	Surgical consultation and treatment as indicated during all stages of development
Pediatric and adolescent gynecology	Provides female pubertal reproductive counseling and hormonal recommendations during adolescence; works with endocrinology to provide continuity of care
Pediatric and adult urology	Provides male pubertal reproductive counseling and hormonal recommendations during adolescence; works with endocrinology to provide continuity of care
Reproductive endocrinologist	Female and male fertility counseling and ART if indicated
Andrologist	Male fertility counseling and ART if indicated; works with reproductive endocrinologist
High-risk obstetrician/MFM	Women with previous reproductive tract surgery including UGS or Müllerian anomalies
Child life/expressive therapy	Assist with emotional and psychological needs of the patient/family
Social services	Assist with social needs, home health, and community/school integration
Chaplain	Assist with spiritual needs of the patient and family

ART, assisted reproductive techniques; DSD, disorders of sexual development; MFM, maternal-fetal medicine; UGS, urogenital sinus.Adapted from McCracken and Fallat.[23, pp 90]

Communication

Current consensus recommendations suggest that the multidisciplinary team supporting young people with DSD and their families should include pediatric subspecialists in endocrinology, surgery and/or urology, psychology/psychiatry, gynecology, genetics, social work, and nursing (Table 3).

This team approach would be appropriate for all young women with reproductive or pediatric adolescent gynecologic concerns for which transition of care is needed.

As patients transition into adulthood, several members of the original team, such as the neonatologist will no longer be needed and additional members will need to be added such as reproductive endocrinology and/or andrology for fertility concerns.

Success of Transition

Successful health care transition of adolescents and young adults with chronic illnesses has been associated with their ability to function autonomously, keep medical appointments without a parent or sibling, and be compliant with adherence to medical regimens.[42] As such, clinicians caring for young adult patients in transition with reproductive health issues should be assessed for successful transition using this information. This might include ensuring patients are able to attend their medical appointments, adhere to medication prescribed, and have knowledge of safe sexual practices with appropriate screening and use of contraception as needed.

Ongoing Care

Before transition it is important to ensure that the adolescent understands her diagnosis, what treatments she has had, and an understanding of her adult care needs. In many cases, reproductive and DSD diagnoses are complex and might require the clinician to help disclose age-appropriate information to the patient regarding the pathophysiology of her reproductive condition avoiding medical jargon and explaining interaction of chromosomes, hormones, and anatomic structures. For example, when providing this information, it is important to avoid confusing terminology such as “blind vagina” in the cases of vaginal agenesis, and “phallic structure” when describing DSD conditions.

Patients with atypical genitalia often have psychosocial concerns and assessing the patient's psychosocial support, gender identity, and sexual attraction is necessary. Many of these patients have experienced anxiety and distress related to shame, stigma, infertility, and identity concerns. As such they experience higher rates of suicide attempt than adults with other medical conditions and should be screened for depression. Peer support and advocacy groups (Table 2) might be helpful for the patient and her family to feel that they are not alone and might enable them to gain tools to navigate life, relationships, and health care with reproductive conditions.

It is crucial to obtain an understanding of the patient's gender expression, identity, sexual attraction, and orientation as preparation is made for transition of care. Although most individuals identify with their assigned gender of rearing, higher rates of gender dysphoria are noted in the DSD population.[43] It is important to remember to allow the patient to consider gender in his/her own terms and decide for him/herself how he/she wants to identify. It is equally important for the medical team to recognize that gender identity (the gender the patient perceives him/herself to be) is independent of sexual orientation (the sex to whom one is sexually and romantically attracted). There are increased bisexual and homosexual attractions noted in some DSD conditions[44] and an awareness of where on this spectrum of identity/attraction that your patient falls will allow the medical provider to provide optimal transition of care.

In patients with reproductive health care issues, transition of care might include the need for surveillance for future reproductive neoplasms, assessment and continuation of hormonal replacement, ongoing assessment of sexual function and satisfaction, postsurgical evaluation of any previous surgically created vagina, and potential genetics evaluation.

Future Research

Research on transition of care in patients with reproductive concerns is needed to inform future practice. At this time, there are very few data on the process of transition of these patients. Information is needed on assessment for transition readiness in these patients to identify factors that will predict successful transition and help the medical team to identify when an individual is ready to transition to adult-oriented care. The proposed biological and behavioral indicators identified from other disease processes and transition of care should be empirically evaluated. In addition, these biologic and psychologic factors should be monitored at regular intervals throughout the transition process as should evaluation to identify barriers to successful transition to identify targets for intervention. Finally, long-term outcome data regarding transition of this patient population is needed to assess the entire transition process.[10]

There is a need for long-term evaluation of individuals receiving hormone replacement therapy and relevant biological markers that include bone density measurements and similarly for patients receiving steroid replacement therapy evaluation of long-term endogenous adrenal hormone levels.

Continued monitoring and research regarding expected timing of gonadectomy, vaginal dilation, or reconstructive surgery and the associated anatomical and psychological outcomes of those interventions should be measured and reported.

Creighton and Wood[11] pose that current research questions also include:

- What are the long-term outcomes of reproductive surgery in childhood?
- Would outcomes be different without surgery in childhood when safe to avoid it?
- Will intervention in utero alter outcomes later in life?
- Is there a predictable genetic basis for conditions that have not been previously identified?

Conclusions

Transition of patients with reproductive health care needs such as congenital anomalies and DSD is a complex process that occurs over multiple years. Patients report that psychological support is an essential component to support the process. Although transition models used in the care of adolescent patients with other chronic diseases can provide valuable protocols to follow, transition for these patients with reproductive needs is still poorly understood. The patients with reproductive issues have specific unique issues regarding timing of disclosure of diagnosis, genital examinations, gonadectomy, vaginal treatments, hormone

replacement therapy, use of long-term medication, and potential cancer screening recommendations.[18] Fortunately, at least 1 center in Germany is working on a long-term transition of DSD patients but at the time of this publication, evidence-based care is not available for this patient population and research on transition of care is needed to inform future practice.

References

1. Zuckerman AL: Transition of care to an adult provider. Curr Opin Obstet Gynecol 2017; 29:295
2. Kovacs AH, McCrindle BW: So hard to say goodbye: transition from pediatric to adult cardiology care. Nat Rev Cardiol 2014; 11:51
3. Tuchman LK, Schwartz LA, Sawicki GS, et al: Cystic fibrosis and transition to adult medical care. Pediatrics 2010; 125:566
4. Feitell S, Hankins SR, Eisen HJ: Adjunctive therapy and management of the transition of care in patients with heart failure. Cardiol Clin 2014; 32:163
5. Gleeson H, Turner G: Transition to adult services. Arch Dis Child Educ Pract Ed 2012; 97:86
6. Blackman JA, Conaway MR: Adolescents with cerebral palsy transitioning to adult health care services. Clin Pediatr 2014; 53:356
7. McDonagh JE: Transition of care from pediatric to adult rheumatology. Arch Dis Child 2007; 92:802
8. Got Transition. Available. http://www.gottransition.org. Accessed August 12, 2018.
9. Rosen DS, Blum RW, Britto M, et al: Transition to adult health care for adolescents and young adults with chronic conditions: position paper of the Society for Adolescent Medicine. J Adolesc Health 2003; 33:309
10. Hullmann SE, Chalmers LJ, Wisniewski AB: Transition from pediatric to adult care for adolescents and young adults with a disorder of sex development. J Pediatr Adolesc Gynecol 2012; 25:155
11. Creighton SM, Wood D: Complex gynaecological and urological problems in adolescents: challenges and transition. Postgrad Med J 2013; 89:348
12. Campbell F, Biggs K, Aldiss SK, et al: Transition of care for adolescents from pediatric services to adult health services. Cochrane Database Syst Rev 2016; 4:CD009794
13. American Academy of Pediatrics, American Academy of Family Physicians: American College of Physicians-American Society for Internal Medicine: A consensus statement on health care transitions for young adults with special health care needs. Pediatrics 2002; 110:1304
14. American Academy of Pediatrics, American Academy of Family Physicians, American College of Physicians, et al: Supporting the health care transition from adolescence to adulthood in the medical home. Pediatrics 2011; 128:182
15. Rothstein DH, Dasgupta R: Delivery of Surgical Care Committee of the American Academy of Pediatrics Section on Surgery: Transition of care from pediatric to adult surgery. Pediatrics 2016; 138:e20161303
16. McDonagh JE, Viner RM: Lost in transition? Between paediatric and adult services. BMJ 2006; 332:435
17. Lee PA, Houk CP, Ahmed SF, et al: Consensus statement on management of intersex disorders. International Consensus Conference on Intersex. Pediatrics 2006; 118:e448
18. Crouch NS, Creighton SM: Transition of care for adolescents with disorders of sex development. Nat Rev Endocrinol 2014; 10:436
19. Oelschlager AM, Muscarella M, Gomez-Lobo V: Transition to adult care in persons with disorders of sexual development: the role of the gynecologist. Obstet Gynecol 2015; 126:845
20. Simoes E, Sokolov AN, Kronenthaler A, et al: Information ranks highest: expectations of female adolescents with rare genital malformation towards healthcare services. PLoS One 2017; 12:e0174031
21. Koyle M, Bowlin P: Issues in the long-term management of adolescents and adults with DSD: management of gonads, genital reconstruction, and late presentation of the undiagnosed DSD. In: Wood HM, Wood D, editors. Transition and Lifelong Care in Congenital Urology. Switzerland, Springer International Publishing, 2015, pp 65–75
22. Gomez-Lobo V, Oelschlager AM: Disorders of sexual development in adult women. Obstet Gynecol 2016; 128:1162
23. McCracken KA, Fallat ME: Transition from pediatric to adult surgery care for patients with disorders of sexual development. Semin Pediatr Surg 2015; 24:88
24. Liao LM, Tacconelli E, Wood D, et al: Adolescent girls with disorders of sex development: a needs analysis of transitional care. J Pediatr Urol 2010; 6: 609
25. Kleinemeier E, Jurgensen M, Lux A, et al: Psychological adjustment and sexual development of adolescents with disorders of sex development. J Adolesc Health 2010; 47:463
26. Jones SE, Hamilton S: The missing link: paediatric to adult transitioning diabetes services. Br J Nurs 2008; 17:842
27. van Staa AL, Jedeloo S, van Meeteren J, et al: Crossing the transition chasm: experiences and recommendations for improving transitional care of young adults, parents and providers. Child Care Health Dev 2011; 37:821
28. Wagner A, Brucker SY, Ueding E, et al: Treatment management during the adolescent transition period of girls and young women with Mayer-Rokitansky-Küster-Hauser syndrome (MRKH): a systematic literature review. Orphanet J Rare Dis 2016; 11:152
29. Gleeson H, Wisniewski A: Working with adolescents and young adults to support transition. In: O. Hiort and S.F. Ahmed (eds) Understanding Differences and Disorders of Sex Development (DSD). Endocr Dev Basel Karger 2014; 27:128
30. Auchus RJ, Arlt W: Approach to the patient: the adult with congenital adrenal hyperplasia. J Clin Endocrinol Metab 2013; 98:2645
31. Gleeson H, Davis J, Jones J, et al: The challenge of delivering endocrine care and successful transition to adult services in adolescents with congenital adrenal hyperplasia: experience in a single centre over 18 years. Clin Endocrinol 2013; 28:23
32. Okumura MJ, Hersh A, Hilton JF, et al: Change in health status and access to care in young adults with special health care needs: results from the 2007 national survey of adult transition and health. J Adolesc Health 2013; 52:413
33. American College of Obstetricians and Gynecologists Committee on Adolescent Health: Müllerian agenesis: diagnosis, management and treatment. Committee opinion 728. Obstet Gynecol 2018; 131:196
34. Brannstrom M, Johannesson L, Dahm-Kahler P, et al: First clinical uterus transplantation trial: a six-month report. Fertil Steril 2014; 10:1228
35. Brannstrom M, Johannesson L, Bokstrom H, et al: Livebirth after uterus transplantation. Lancet 2015; 385:607
36. Flyckt R, Davis A, Farrell R, et al: Uterine transplantation: surgical innovation in the treatment of uterine factor infertility. J Obstet Gynaecol Can 2018; 40:86
37. Stray-Petersen B, Stray-Petersen S: Etiologic factors and subsequent reproductive performance in 195 couples with a prior history of habitual abortion. Am J Obstet Gynecol 1984; 148:140
38. Simon C, Tortajada M, Martinez L, et al: Müllerian defects in women with normal reproductive outcome. Fertil Steril 1991; 56:1192
39. Lin P, Bhatnagar KP, Nettleton GS, et al: Female genital anomalies. Fertil Steril 2002; 78:899
40. Nabhan Z, Lee P: Disorders of sex development. Curr Opin Obstet Gynecol 2007; 19:440
41. Maharaj N, Dhai A, Wiersma R, et al: Intersex conditions in children and adolescents: surgical, ethical and legal considerations. J Pediatr Adolesc Gynecol 2005; 18:399
42. Fair CD, Sullivan K, Gatto A: Indicators of transition success for youth living with HIV: perspectives of pediatric and adult infectious disease care providers. AIDS Care 2011; 23:965
43. Yang JH, Baskin LS, DiSandro M: Gender identity in disorders of sex development: review article. Urology 2010; 75:153
44. Frisen L, Nordenstrom A, Falhammar H, et al: Gender role behavior, sexuality, and psychosocial adaptation in women with congenital adrenal hyperplasia due to CYP21A2 deficiency. J Clin Endocrinol Metab 2009; 94:3432