Editor’s Note

I want to wish all of you Happy Holidays and a Happy New Year! I recently returned from a trip to Pakistan where I spoke at the Biennial Meeting of the Pakistan Pediatric Association in Islamabad. As is often the case I also got an opportunity to speak with the faculty of my alma mater, King Edwards Medical University in Lahore, at a symposium on medical writing and editing. While these professional interactions are so important, even better were my visits to two centers for poor children where they are provided free education and some medical care. While here in the United States we often are concerned, and rightly so, about the lack of resources for our children, visiting these centers was an eye opener.

I was energized after attending the American Academy of Pediatrics’ National Conference & Exhibition in Orlando and hearing so many motivating stories about the wonderful work people are doing for children all over the world. I wanted to see for myself what some of the activities happening for children occurring in Pakistan were. There are so many that I will not be able to cover them here, but I wanted to share two that I thought were innovative and have tremendous potential.

First, I visited the “ILM Center”. In Urdu, the national language of Pakistan, Ilm is the word for “knowledge”. This center is a collaboration between PAHCHAAN (https://www.facebook.com/Pahchaan/) and local Non-Governmental Organization (NGO; another term for a 501c3 charitable organization) and the University of Lahore. It is headed by my medical school classmate, pediatrician and a child rights advocate Dr. Naeem Zafar. They have 90 students enrolled in two, four-hour sessions of 45 students each. During these sessions, students are given basic education and one meal all for less than $0.50/day. The students are all ages from 5 years to 13 years, and the majority of them are girls. The older students often work and earn for their families the other half day as apprentices learning to become tailors or automobile mechanics. This is by no means a tacit approval of child labor but rather a statement of facts. The kids were so full of life, enthusiastic, and most important of all, wanting to learn. The teachers and operators of the center told me that these students almost never miss a day of school. The center has engaged the local community called “mohalla”, loosely translated as neighborhood, and also brings in mothers for a few hours to teach them life skills, good nutrition, child development, and child rearing. There are even plans to add a primary care center to provide basic healthcare needs for the children.

I also visited another school cum primary healthcare center funded by the Shahid Afridi Foundation (https://ShahidAfridifoundation.org/), another NGO. Shahid Afridi is a retired cricketer (a very popular sport in Pakistan – Google it!) who is devoting his life to the welfare of children. This project is led by a Dr. Muhammad Moaz, a friend and a committed child rights advocate. This center has free schooling for 200 kids up to grade 8, with plans to increase to grade 10. These children will also receive their primary and acute care right there at the center. This center is located in a poor, working class village in the outskirts of Lahore, where most of the villagers work. The idea is to replicate this model in other parts of the country so that children not only receive education but also healthcare. Most remarkable is again the low cost per student at this center.

What also impressed me in both of these centers was the commitment of the employees and the volunteers, and also how a small amount of funds can have a significant impact in the life of these children. As pediatricians and advocates for children, we should be advocating for children all over the world. If we want to make the world a better place, let’s make sure that all children have good education and are healthy. The global village is shrinking and if we don’t take care of today’s children, tomorrow’s adults will for sure be a problem.

I know that all of you are working hard for children of America. I also know that many pediatricians are working for better life for children all over the world. Perhaps you can share those activities with us. Tell us some of the things you are doing in the U.S. and internationally and we can report them in your Journal.

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Florida Chapter ———
American Academy of Pediatrics
INTRODUCTION

According to a 2011 joint consensus statement from the American Academy of Pediatrics (AAP), the American Academy of Family Physicians and the American College of Physicians – American Society of Internal Medicine, health care transition (HCT) is “a dynamic lifelong process that seeks to meet the individual needs of youth as they move from childhood to adulthood.” A transition is therefore not a discrete event, but a series of actions taking place over a number of patient, family and provider interactions. The process must provide a comprehensive plan of action over time to address both medical and non-medical needs.

As pediatricians, we are well versed in the variations of physiologic, cognitive and behavioral development that present in the youth we care for. As such, the experience of becoming an adult will be different for every patient, depending on the developmental circumstances. It is our role as care providers to identify the unique needs and challenges of both our patients and their families to facilitate this transition to adulthood. This is especially relevant to youth and young adults with special health care needs (YYASHCN), who are especially vulnerable to poor outcomes if adequate resources are not in place, as they become adults. While there is an ongoing debate about setting age limits in pediatrics and whether certain youth with special needs should remain in pediatric care, the fact remains that all youth and families will need counseling about health care transition. This will need to occur irrespective of whether or not a discrete transfer of services from pediatrics to internal or family medicine occurs.

According to the 2009/10, National Survey of Children with Special Health Care Needs, one in four households in the United States (US) have at least one child with a special health care need. Special health care need is defined as having a chronic medical, behavioral, or developmental condition and the need for prescription medications and/or specialized therapies. In the US, more than 90% of children with a chronic or disabling condition will live beyond 20 years and each year approximately 500,000 YYASHCN will reach the age of 18 years. The 2016 National Survey of Children’s Health found that nationwide only 16.5% of families with YYASHCN receive services related to HCT and only 7.5% in Florida. Lack of support during HCT may lead to poor outcomes related to fragmented care, lack of care coordination, poor preventive services and loss of health care coverage and access. These risks are especially high in young adults with chronic conditions where gaps in care translate into declines in health and quality of life.

It is critical that pediatric practices provide YYASHCN guidance in HCT to ensure optimal health and well-being. Longitudinal transition activities typically occur in the outpatient subspecialty or primary care setting. It is, however, important to note that discussions about HCT may occur in the emergency department or inpatient setting and may often be the initial trigger for a patient or family to begin the process of preparing for the future. This paper will specifically focus on HCT in the outpatient setting. Supporting a patient and family through the transition process is a team effort and often involves multiple players from both the medical and non-medical setting.

MAKING YOUR PRACTICE “HCT READY”/SETTING THE STAGE FOR TRANSITION

Transition services occur in a variety of ways during clinic encounters. Some examples include integration of transition questions and counseling during a routine visit, scheduling time outside of office visits specific to transition goal setting and planning, or offering a dedicated HCT day when patients needing such services are preferentially booked and staff knowledgeable in HCT are available for consultation. Delivery of such services will require a clear and organized approach.

The “Got Transition” website describes the “Six Core Elements of Transition” which includes; development of a transition policy, transition tracking and monitoring, transition readiness assessment, transition planning, transfer of care and transfer completion. Practices may utilize tools from “Got Transition” to measure the level of transition planning that is taking place and develop goals for future improvements. Implementation of transition-related activities may be more effective if quality improvement (QI) techniques are followed by repeated “Plan Do Study Act” (PDSA) cycles. Using this framework allows for identification of gaps in practice, planning, implementation and measurement of change and further refinement of actions to promote continuous QI of transition services.

In addition to considering the “Six Core Elements of Transition”, it is also crucial for practices to consider the needs of the population they serve and the priority transition needs of their patients. Other team members such as medical assistants, nurses, social workers or case managers may play a role in HCT activities. It also helps to create a clinic flow diagram (Figure 1) to identify points in the clinic visit where transition counseling or services may occur. Incorporating transition tools or questions (Figure 2) such as a transition care plan into clinic encounters may promote continuous QI of transition services.

Figure 1: Clinic Flow Diagram. Consider points in a patient visit where Health Care Transition screening and counseling may occur. Determine who would be responsible for these tasks.
Advocacy efforts to better fund transition initiatives are currently ongoing with a specific focus on improved reimbursement rates. The AAP and “Got Transition” recently published a coding and billing tip sheet that provides guidance on appropriate billing codes to use for transition-related services. In order for practices to provide HCT services, ongoing advocacy is needed to ensure reimbursement for such services.

**PATIENT AND FAMILY APPROACH:**

One may define adulthood by legal age, socio-cultural or developmental factors. Youth move into adult roles through a number of processes that involve complex physical and psychological changes which are shaped by their social and cultural environments. Being an adult often involves both a celebration and fear of independence as youth cut ties with family, move out on their own, become financially independent and potentially start a family of their own. For those with developmental or cognitive limitations, adulthood will look quite different from those without such impairments and preparation should be in line with expected evolving capacities. With this in mind, the first step in transition counseling is a discussion with the family and patient about how they define youth and adulthood and how this relates to their cultural/personal, psychosocial, developmental/cognitive, physical/environmental, educational and economic needs and goals (Table 1).

It is critical to determine if the focus of transition activities is on preparing the patient, the caregiver or both, as this may vary in different circumstances, depending on the age and maturity of the patient. For example, at younger ages, patient-related activities may include diagnosis naming and education while parents focus more on issues of health insurance and coverage as the child ages. Assessment of family relationships and dynamics is also helpful in transition planning; consider enabling and hindering factors when developing the transition plan. While some caregivers are ready to let go, others are not. Similarly, some youth are ready to be independent and others are quite fearful of the process. Many families and patients are also reluctant to see new doctors and work with new therapists, nurses, pharmacies and medical supply companies, all of which may change when they become an adult. Gathering this information will give insight into possible barriers to the transition plan. While some caregivers are ready to let go, others are not. Similarly, some youth are ready to be independent and others are quite fearful of the process. Many families and patients are also reluctant to see new doctors and work with new therapists, nurses, pharmacies and medical supply companies, all of which may change when they become an adult. Gathering this information will give insight into possible barriers to the transition process.

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Transition to adulthood involves transitions in the non-medical domains of education/vocation, public benefits/insurance coverage, living arrangements/home supports and decision-making capacity/guardianship.1 Offices should be prepared to screen for needs and place patient-centered referrals as necessary to appropriate agencies. Collaboration with community partners is critical to close gaps in care and to ensure service provision. The healthcare team should also have up-to-date handouts and/or brochures of the suggested referral service. Fragmented transition to adult-based health care results from a lack of focus on non-medical transition needs and this is, therefore, a critical part of the transition visit.

Transition readiness questionnaires and transition planning tools are available through a number of online sources for self-directed learning about screening for HCT needs.12,13 It is helpful to create a flexible goal oriented timeline according to age and developmental stage. Allow flexibility in the beginning stages of transition as, despite expected benchmarks based on development, the emotional stress of transition may slow progress. It is necessary to identify any barriers or challenges that may arise in the different transition domains and work toward resolution. The healthcare team should offer continual guidance to coordinate efforts with any other involved agencies to avoid duplication of efforts.

Education/Vocation: YYASHCN often receive Exceptional Student Education (ESE), or accommodations, which provide specialized services for students living with disabilities. After evaluation to determine eligibility, students will either receive ESE services, including an Individualized Educational Plan (IEP) or ineligibility for ESE, a 504 plan. The IEP and 504 plan implement differently. An IEP addresses special instructions and goals for specific disabilities affecting one’s education, while the 504 plan allows for special accommodations in the face of physical or mental impairments. The Rehabilitation Act of 1973, Section 504 and Individuals with Disabilities Education Act (IDEA) are educational policies structured by the US Department of Education to protect the rights and educational choices of YYASHCN.11 In addition; the IEP is a valuable resource for the HCT team as it provides information about psychosocial, learning and developmental capacities to guide transition goals. The IEP also provides information for post-graduation planning (college, career, vocational and life skills).

If considering college, YYASHCN should research federal, state and local scholarship and financial aid opportunities and seek the assistance of a guidance counselor in this process. Information about federal financial aid and scholarship opportunities is available online (https://studentaid.ed.gov/sa/eligibility).13 Students should also learn about campus disability services and register with the office of disability to receive continuity of special accommodations. The Division of Vocational Rehabilitation Florida Department of Education offers eligible individuals sixteen years or older employment evaluation services to predetermine job readiness skills to prepare for the workforce.10 Other post-graduation options include, adult day training programs, life skills focused programs and group social activities. With proper planning, graduation from school should lead to new experiences and opportunities that promote engagement in community life.

PUBLIC BENEFITS/INSURANCE:

A review of public benefits needs is necessary as eligibility determinations for continuity of services may change during transition. Patients need to be aware of designated dates and times of (re)evaluation of services, as this may result in the loss of benefits for health care, food, and cash assistance.10,15 Gaps in health insurance, during HCT, lead to a decline in health status and poor outcomes for YYASHCN. According to the 2016 US Census report, young adults in the US continue to be uninsured at the highest rates when compared with all other age groups.17 Pre-planning to determine what options exist, if any, for those who will lose coverage is crucial. Those screening positive for public benefits assistance need referral to a social worker or public benefits attorney to review the circumstances and provide assistance as indicated. Medical complexity involves costly services and care, which are often not available through adult health plans. Patients may therefore require long-term care (LTC) waiver services to ensure ongoing medically necessary services. Initiating the intake for an LTC waiver may be as early as age eighteen and requires attention at least six months prior to one’s 21st birthday in order to avoid a gap in services. Just the Facts: The 411 on Health Insurance for Young Adults Ages 18-30 in Florida is a resource-filled guide for pre-planning and navigating health insurance.16

LIVING ARRANGEMENTS/HOME SUPPORTS/TRANSPORTATION

Transition to adulthood also involves consideration of future living arrangements. If patients are considering living away from caregivers, the supports and needs of the patient are imperative to housing arrangements. Housing should be accessible and have modifications in place according to state and federal regulations that will optimize safety, function, and independence. If assisted living is an option, the family and patient must determine which daily tasks will require some or full support. Transportation needs such as obtaining a driver’s permit/license, free Medicaid transportation services, and public transportation for special needs individuals, and/or newer options such as application-based transportation services are all options. In the case where a caregiver with a full care dependent child is no longer able to provide care, group homes are an option and financial support for this may be available through long-term care waiver programs or other government supports.

DECISION MAKING CAPACITY/GUARDIANSHIP

Prior to turning eighteen years of age, families should determine decision-making capacity and seek legal counsel on this issue. Options will depend on the YYASHCN’s cognitive and developmental capacity. Decision-making may range from less restrictive capacity, such as a self-supported decision to very limiting, full guardianship with no decision-making rights.22 Caregivers not filing legal documentation before the patient 18th birthday may experience delay or denial to access information and services. Florida Statue, Chapter 765, identifies those permissible decision makers for an incapacitated person without legal documentation.23 Guardianship is also an area where medical-legal services are useful.

MEDICAL HCT NEEDS

Part of the transition plan will likely include the need to establish care with adult physicians, allied health professionals, durable medical equipment companies and other special skilled services. Identification of providers and benefits of service in the patient’s insurance network and early referral to these providers will eliminate harmful gaps in services and access. Patients should request a medical transfer note or complete a portable medical summary, obtain relevant medical records or request providers do a warm handoff call to ensure medical information is not lost during transfer. Providers that share electronic medical records systems facilitates easy sharing of information. Communication between pediatric and adult providers during the transition period allows the transfer to be smoother once a young adult completely moves to adult providers.2

SELF-MANAGEMENT

Developmentally appropriate self-management skills are an important goal of HCT. The goal is to optimize skills to the best of one’s developmental capacity and promote independence that is in line with an individual’s expected skill set. When appropriate to do so, patients see the doctor alone in the visit to allow for discussion of private matters such as sexual and reproductive health issues, mental health, substance use, trauma, and abuse. Patients and caregivers should be aware that seeing the doctor alone promotes autonomy and is part of the shift to adulthood.1 Caregivers are asked to leave during the appointment as early as 12 years of age in the pediatric setting and mark the beginning of the shifting roles between the caregiver and the patient with respect to healthcare. Children may participate in conversations about their health even earlier if providers make a dedicated effort to do so. For example, the use of medication pictures or diagrams of body parts may be useful in explaining management plans and disease states to younger children. Engagement in office visits from an early age will make self-management skills easier as the child ages.

Several self-management tools in the form of transition readiness questionnaires exist.1 Transition tools may be generalizable to all conditions or more disease-specific depending on patient needs.22 They include questions related to managing medications, appointment keeping, tracking of health issues, talking with providers and managing daily activities. EMR or mobile application portals are another effective and favorable self-management tool for YYASHCN. EMR portals allow patient medical information sharing from the healthcare facility to the patient’s secured assigned account and are accessible by most mobile electronic devices. YYASHCN may utilize the messaging feature, which helps to build patient-provider rapport.1 Patients should track, recognize and reward themselves for the progress of self-management skills while accepting unpredictable fluctuations in health status, which may hinder efforts. The use of motivational interviewing has also been shown to be highly effective in transition planning with YYASHCN and when setting goals.25

SELF-ADVOCACY

Empowering young adults to communicate their comfort level in self-management is important to set realistic care goals and involve caregivers as needed. Offices should create a safe, judgment-free zone for patients to learn and practice health care self-management skills over time. With ongoing health care reform, health care teams should be well informed of policy changes affecting this population and effectively educate patients and families on this topic. Health is impacted by all policy and therefore advocacy around medical, educational, vocational and social platforms is necessary. Patients may not fully understand their health care rights and new role as a young adult in the adult care system; therefore, self-advocacy information and guidance is available through organizations such as Disability Rights Florida.24

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CONCLUSION
HCT counseling is critical to ensure that as youth emerge into adulthood, in particular, those with special health care needs, there is adequate preparation and support for success in the adult world. Preparation begins in the pediatric office setting and then flows into a number of other settings. It takes a village to close the health care disparity gap for YYASHCN; therefore, community resources and collaborations are the foundation to provide HCT services.

Creating a system of care with community partners is critical to ensure YYASHCN are not lost during the critical time of transition into adulthood. The development of adequate screening processes and a robust collaborative referral network will provide the infrastructure for successful HCT. In addition, these organizations must work together to advocate for system and policy changes that promote the health and well-being of YYASHCN in our communities.

REFERENCES:
The Care of Transgender Youth

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ABSTRACT

The general pediatric office may be the first resource a family turns to when seeking care for a gender nonconforming child or adolescent. Thus, it is important that pediatricians are familiar with certain terms and definitions relating to children with gender dysphoria, and understand the psychological and medical needs of this population. This review article focuses on providing the general pediatrician with an overview of the care of transgender youth, including diagnostic criteria, natural history, psychological considerations, and medical management.

DEFINITIONS AND PREVALENCE

At birth, children are assigned a sex based on the outward appearance of their genitalia, determined by chromosomes and hormones. However, “gender identity” refers to a person’s inner sense of maleness or femaleness, and this can only be expressed through a behavior that is consistent with that gender identity. Gender identity can be male, female, or somewhere along the continuum. When one’s biologic sex and gender identity does not align, the distress and anguish that is felt is referred to as “gender dysphoria” (GD). The Diagnostic and Statistical Manual, 5th Edition specifies that in order to meet the criteria for gender dysphoria, one must have a marked incongruence between one’s birth sex and expressed gender for at least 6 months duration, manifested by at least 2 of the following: 1. Incongruence of expressed gender and one’s or anticipated secondary sexual characteristics. 2. Desire to be of (or prevent) one’s secondary sexual characteristics. 3. Desire for the development of female gender identity. 4. Desire to be the other gender. 5. Desire to be treated as the other gender. 6. Conviction that one has the typical feelings/reactions of a different gender. 3

Due to a feeling of incongruency between natal sex and gender identity, a transgender or gender nonconforming person expresses their gender identity based on their true, deep inner sense of their gender. People express their gender identity in many ways, including their preferred name and pronouns, clothing, haircut, body characteristics, voice, and mannerisms.

A transgender male refers to a person assigned a female sex at birth, but who identifies as male. A transgender female refers to a person assigned a male sex at birth, but who identifies as female. Sexual orientation should not be confused with gender identity. Sexual orientation refers to a person’s physical and emotional attraction to another person. A transgender person may be attracted to men, women, both (bisexual), or neither (asexual). 3

The actual prevalence of gender dysphoria in children and adolescents is unknown. Shields, et al sampled 2730 6-8th graders in 2011, and found that 1.3% self-identified as transgender. 10 In a 2012 random sample of 8166 high school students in New Zealand, 1.2% self-identified as transgender, and 2.5% reported as not being sure about their gender. 11 The literature has supported a recent shift in the number of natal males vs females who present with gender dysphoria. Aitken, et al reported that prior to 2006, more natal males than females were referred for gender dysphoria with a ratio of 2.11 to 1; however between 2006-2013, that ratio changed to 1 to 1.76, favoring girls. 11 However, a recent report from the Netherlands estimating the prevalence in 2015 to be 1.3,800 for natal men (transwomen) and 1.5,200 for natal women (transmen). 12

Biological Determinants of Sex and Biology of Gender Dysphoria

In the 6th week of pregnancy, the testes or ovaries begin to develop. If the sex determining gene on the Y chromosome (SRY) is present, testes will form, which secrete testosterone, and this hormone is converted to dihydrotestosterone, a potent androgen. Without the SRY present, ovaries will form. Between 6 and 12 weeks gestation, the male sexual organs form under the influence of androgens, or female sexual organs form in the absence of androgens. Sexual differentiation of the brain then occurs predominantly due to the presence or absence of androgens. For example, in complete androgen insensitivity syndrome (CAIS), a 46,XY person is born with normal testosterone production, but due to a receptor defect, is completely insensitive to that hormone. Those with CAIS develop phenotypically into females, and despite the presence of male chromosomes, they form a female gender identity. 13 Alternatively, a 46,XX female with congenital adrenal hyperplasia (CAH) who has been exposed to high levels of androgens in utero, has a 300-1000 greater likelihood of developing transsexuality, but still the risk is only 1-3%. 13

Twin studies suggest that there is an inheritability component to gender identity disorder. 14 Further studies need to clarify the role of genetics in gender identity disorder. Several sex hormone related candidate genes have been identified in the development of gender identity. For example, an increased prevalence of a polymorphisms in the CYP17 gene (involved in steroidogenic pathway) has been reported in female to male transsexuals. There are conflicting reports regarding polymorphisms associated with a higher prevalence of transsexuality in the androgen receptor gene, estrogen receptor beta gene (ERB), and the aromatase gene (CYP19A1). 15

NATURAL HISTORY OF TRANSGENDER YOUTH

Long term follow-up studies have demonstrated that the majority of prepubertal children who present with gender dysphoria early in childhood will not continue to have gender dysphoria into adolescence and adulthood. A 2008 follow up study of 25 girls with gender identity disorder showed that while 60% of the girls met the Diagnostic and Statistical Manual of Mental Health Disorders (DSM) criteria for gender identity disorder at an average age of 8.8 years, only 12% met this criteria by an average age of 23.2 years. 16 In another long term follow up study of 77 children with gender dysphoria who presented at an average age of 8 years, only 27% had persistence of the gender dysphoria by a mean age of 18 years. 17 Steensma, et al identified certain factors that predict persistence of gender dysphoria from childhood to adolescence, including older age at presentation, lower socioeconomic status, female natal sex, and early social transition. 18 However, though there is limited data on the persistence of gender dysphoria from adolescence to adulthood, one study demonstrated that 100% of Dutch adolescents who presented to a gender clinic for hormone transition continued to be transgender young adults. 19

PSYCHOLOGICAL CONSIDERATIONS

The first step in management of a child or adolescent who expresses gender dysphoria is referral to a qualified mental health professional who has expertise in transgender youth. The psychologic evaluation consists of obtaining information from the child and parent regarding the child’s psychosocial development, general functioning, and mental health status. There is also an assessment of the child’s support structure and family’s ability to endure stress and deal with the child’s complex situation. After evaluation, the mental health professional decides if the patient meets the criteria for gender dysphoria and evaluates for other co-morbid mental health conditions. The provider determines if the child truly has gender dysphoria, or another mental health disorder with similar features, such as body dysmorphic disorder or body identity integrity disorder. Also, when clinically appropriate, the mental health professional plays a role in evaluating psychosocial readiness for hormonal or surgical intervention. 20

DEFINITIONS AND PREVALENCE
The psychologist counsels the parent on how to deal with the gender dysphoric youth, and assists the family regarding social transition. Gender variant behavior should not be discouraged or punished, however some mental health professionals argue against young children making a complete social transition since most gender dysphoric children will not continue to have gender dysphoria during adolescence. (1) The aim of this recommendation is to prevent having to do a complex change back to the role of their natal gender. (1)

Co-morbid psychiatric conditions are commonly seen in gender dysphoric youth. Internalizing psychopathologies rather than externalizing psychopathologies are more common among this population. In one study of 120 Dutch children age 4-11 presenting to a gender identity clinic, 31% had anxiety, with the most prevalent disorder being social phobia, and 6% had mood disorder. (2) Regarding, adolescents and young adults, one recent study showed that 35% of transgender youth aged 12-24 years presenting to a large urban transgender clinic reported depressed feelings, 51% reported ever thinking about suicide, and 30% attempted at least once in their lives. (3) The elevated rates of psychopathology may be due to years of enduring prejudice, discrimination, stigma, conflicts between their stated identity and outward appearance, as well as social rejection from family, friends, and peers. A recent study in 2016 published in Pediatrics demonstrated that socially transitioned young children aged 3-12 years old who are supported in their transition showed no elevations in depression and only slightly higher anxiety symptoms compared to the control group. (4) This study argues for the early social transition of persistent and insistent young children with gender dysphoria.

MEDICAL MANAGEMENT

For some youth with gender dysphoria, psychological evaluation and support may be useful and sufficient. Pubertal suppression (halting the development of secondary sexual characteristics) should be considered in children with gender dysphoria in the early stages of puberty, specifically when a natal female has breast buds or a natal male has testicular enlargement of 4cc in volume or 2.5cm in length or greater. (5)

Pubertal suppression is used for many reasons. First, pubertal suppression allows the youth prolonged time to explore their gender identity without committing to the development of the sexual anatomy of either sex. In addition, for many adolescents with GD, the distress and psychologic harm worsen while watching their bodies sexually mature in a way that does not align with their gender identity. Finally, certain body changes that occur during puberty are irreversible, such as breast development in a female or widening of the jaw, an Adams apple, and lowering of the voice in a male. If these features develop and the youth eventually decides later to completely transition to the opposite gender, it is very difficult to physically pass as the opposite gender. (5)

Pubertal suppression is accomplished with the use of GnRH analog, such as leuprolide acetate depot or histrelin acetate, which are long acting agonists that suppress the hypothalamic-pituitary-gonadal axis temporarily. An adolescent is eligible for GnRH agonist therapy when they have reached tanner stage 2, a qualified mental health professional has confirmed that the individual has persistent gender dysphoria that has worsened with the onset of puberty, the adolescent and the parent have provided informed consent, and there are no other contraindications. (1) In a prospective follow up study, de Vries, et al has shown that with pubertal suppression, the adolescent’s overall functioning improves and behavioral/emotional problems and depressive symptoms decrease, however gender dysphoria continues. (1)

However, a follow up study by the same leader in 2014 showed that after cross-sex hormone therapy and gender reassignment surgery, the gender dysphoria and overall psychological functioning improves over time, and “well-being” in many realms was similar to that of their cisgender peers. (6) The side effects of GnRH agonist therapy include, low bone mineral density, unknown effects on brain development, impaired fertility if subsequent cross-sex therapy is initiated, and high cost if not covered by insurance.

For adolescents with persistent gender dysphoria during adolescence, cross-sex hormone therapy is considered at age 16 with the guidance of a qualified mental health professional and a multidisciplinary team including a provider competent in monitoring pubertal maturation and prescribing hormone therapy, such as a pediatric endocrinologist. By age 16 years, the teen is thought to have a reasonable level of competence to consent for this therapy. There may be compelling reasons to administer cross-sex hormone therapy prior to age 16 years, but there is minimal published data on this experience. (2)

To be eligible for cross-sex hormone treatment, a qualified mental health professional has to confirm the persistence of gender dysphoria and the adolescent must be psychologically, medically and socially ready to start treatment. In addition, prior to starting this therapy, the teen and the family need to be given informed consent regarding the risks and benefits of treatment. (2)

For transgender females, 17 β-estradiol in the form of pill or patch is given in escalating doses if puberty is being initiated, or in higher doses initially if the teen is already post-pubertal. The GnRH agonist should be continued until gonadectomy, because the endogenous testosterone interferes with the desired estrogen effects. The side effects of estrogen therapy include, thromboembolic disease, liver dysfunction, hypertension, and infertility. For transgender males, testosterone intramuscular or subcutaneous is given in increasing doses to induce puberty or initially in higher doses if already post-pubertal. The side effects of testosterone therapy include, erythrocytosis, hypertension, lipid changes, cistic acne, and infertility. (7) Prior to starting cross-sex hormone therapy, the provider should counsel the youth and family on the negative effects on fertility and discuss the option for fertility preservation. In one transgender clinical program, despite fertility preservation counseling provided by a dedicated advanced practitioner, out of 105 transgender adolescents, only 4 complete sperm cryopreservation and 1 completed oocyte preservation prior to cross-sex therapy. (8)

GENDER AFFIRMING SURGERY

Gender affirming surgeries fall within two main categories, procedures that effect fertility, such as removal of penis, uterus, and gonads, and other gender conforming procedures, such as breast augmentation, facial surgery, and hair removal in natal males and mastectomy in natal females. Gender affirming surgery is considered after age 18 years, and is associated with improved psychological functioning and low levels of regret. (9,10,12,22)

CONCLUSIONS

While there is an increasing body of literature to support providers in the treatment of transgender youth, there are many unanswered long-term questions that need to be addressed with prospective studies. Transgender children and young adults are a victimized and marginalized group, but providers can take measures to make the health care experience more welcoming and comfortable, such as using preferred names and pronouns, conducting staff training, and researching resources for transgender youth in their area.

REFERENCES

CASE REPORT

A previously healthy 13-year-old male presented to the emergency department for evaluation of four days of fever (maximum temperature 101.4 F), abdominal pain, non-bloody and non-bilious emesis, diarrhea, and one day of rash. The abdominal pain was mild in severity, diffuse, and not worsened by movement. Stools were loose, watery, and non-bloody. One day prior to admission (the fourth day of illness), he developed a non-pruritic, erythematous, raised rash on his abdomen that spread to his chest and proximal extremities, but spared his palms and soles. He denied neck pain, shortness of breath, dysuria, or blood in his urine.

Review of systems revealed a recent history of preseptal cellulitis two weeks prior to his current illness that resolved with oral trimethoprim-sulfamethoxazole and erythromycin eye ointment. The patient lived with his mother and step-father in a wooded area in rural Central Florida. Animal exposures included several family pets including a dog, a ferret, and 4 outdoor adult cats, all of which were healthy and vaccinated. He denied any animal scratches or bites from these animals. The family reported seeing a tick on his body 6 months prior to admission and did not recall more recent exposures. The remaining past medical, social, family and exposure history was non-contributory.

On physical examination, the patient had bilateral, non-exudative conjunctival injection and his abdomen was diffusely tender to palpation without guarding, rebound tenderness or hepatosplenomegaly. He had a patchy, blanching, erythematous, pinpoint papular rash over his trunk, abdomen, and bilateral proximal upper and lower extremities without excoriation. The remainder of the examination was normal.

Laboratory studies were significant for leukopenia with WBC 2500 cu/mm with absolute lymphocyte count of 1225 cu/mm (normal 1400-3800 cu/mm) and the presence of frequent atypical lymphocytes. Platelets were decreased to 83 thou/cu mm and transaminases were mildly elevated (AST 62 U/L, ALT 82 U/L). Hemoglobin and hematocrit were normal. Coagulation
Ehrlichiosis is transmitted by the lone star tick, *Amblyomma americanum*, which is mainly found in white-tailed deer, dogs, rabbits, foxes, and raccoons. The rash appeared more erythematous. His absolute leukocyte count and platelet count continued to decrease. Initial viral studies showed a slightly elevated prothrombin time of 15.3 seconds, INR of 1.2, and PTT of 36 seconds. A chest radiograph and abdominal ultrasound were normal. Viral hepatitis serologies were negative.

**HOSPITAL COURSE**

The morning after admission, the patient continued to have fever, abdominal pain, vomiting, conjunctivitis, and anorexia. The rash appeared more erythematous. His absolute leukocyte count and platelet count continued to decrease. Initial viral testing for enterovirus, parvovirus, adenovirus, cytomegalovirus, and Epstein-Barr virus was negative. The vague symptoms upon presentation, laboratory findings of transaminases with worsening leukopenia and thrombocytopenia, and his history of previous tick exposure increased the concern that his illness was due to ehrlichiosis. Due to the risk of morbidity and mortality from ehrlichiosis and the low risk of therapy with antibiotics, doxycycline was empirically started. Within 24 hours of starting antibiotics, the fever and rash resolved, and laboratory data improved (see Table 1). Ehrlichia PCR testing was positive for *Ehrlichia chaffeensis*.

**Doxycycline**

Doxycycline is the most commonly reported tick-borne illness in the southern United States. Human ehrlichiosis refers to infections typically caused by *Ehrlichia chaffeensis*, a small intracelluar bacterium that infects monocytes. Ehrlichiosis is transmitted by the lone star tick, *Amblyomma americanum*, which is mainly found in white-tailed deer, dogs, rabbits, foxes, and raccoons. Infections are most common in the early summer months from May to July, but can occur year-round. Clinical symptoms typically develop five to ten days after a bite from an infected tick; however, many tick bites go unrecognized and are unreported by patients later found to have ehrlichiosis.

Ehrlichiosis can result in severe disease, with an estimated 40% of infected patients requiring hospitalization and a reported mortality rate of 3%. It typically presents with nonspecific symptoms of fever, malaise, myalgias, and arthralgias. Fever is often the predominant symptom affecting 97% of patients. 30% of patients with ehrlichiosis develop a rash that is maculopapular initially and becomes petechial roughly five days after the initial onset of symptoms. Complications from ehrlichiosis are rare and can occur at any point during the disease course, even after the initial infection has resolved. Complications are more common among immunocompromised hosts, and include sepsis, meningitis, renal failure, myocarditis, among others. Laboratory findings for ehrlichiosis are nonspecific and include leukopenia, thrombocytopenia, and evidence of liver disease with elevated transaminases and abnormal coagulation studies, all of which were seen in our patient. On Wright stain, one may see intracytoplasmic inclusions or morulae (stipple blue inclusions of bacteria in monocytes), but the sensitivity of peripheral smear for diagnosis is low. Serology testing showing seroconversion, or a four-fold increase in antibody titers during the convalescent phase occurring 2-4 weeks after initial onset of symptoms, is diagnostic. However, antibodies including IgM are typically negative during the first 7-10 days of illness, when most patients present with clinical symptoms. Additionally, once IgG antibodies develop, they can persist for months to years after infection, even without clinical signs or relapse.

Table 1. Laboratory Data Before and After Treatment of Ehrlichiosis

| Table 1. Laboratory Data Before and After Treatment of Ehrlichiosis
<table>
<thead>
<tr>
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<tbody>
<tr>
<td>ADMISSION</td>
<td>HOSPITAL DAY 2</td>
<td>HOSPITAL DAY 4 (~24 HOURS AFTER THERAPY STARTED)</td>
</tr>
<tr>
<td>White blood cell count (thou/mm)</td>
<td>2.5</td>
<td>2.9</td>
</tr>
<tr>
<td>Absolute leukocyte count (cu/mm)</td>
<td>1225</td>
<td>551</td>
</tr>
<tr>
<td>Platelets (thou/mm)</td>
<td>83</td>
<td>65</td>
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**REFERENCES**

ABSTRACT:
Raynaud phenomenon (RP) occurring in the pediatric population can be of particular concern due to the possibility of underlying disease as well as permanent tissue damage. This case investigates the efficacy of specific therapies used to reduce the incidence of Raynaud attacks in a 14-year-old female who had previously had a negative work up for systemic disease. Treatment was predominantly sought due to the pain and loss of fine motor movement associated with frequent attacks, which affected fingers, toes, nose, ears, and nipples. Effectively treating pediatric patients with Raynaud phenomenon can be challenging due to the side effects associated with first-line treatments. In this case, greatest success and tolerability was accomplished with sildenafil 20 mg daily and as needed prior to exposure to triggers.

CASE PRESENTATION:
A 14-year-old female with no significant past medical history presented to the rheumatology clinic, referred from pediatrics, for evaluation of RP. The patient’s RP first presented during the winter at approximately age 10 years and lasted for about 3-5 minutes once weekly in the toes and fingers. The attacks progressed in severity and frequency, and around age 13 they began occurring 6 times daily for 10-20 minutes. Even the smaller attacks were followed by a tingling pain in the affected parts for as much as one hour after the attack. Approximately twice monthly the attacks lasted 45-60 minutes. The fingers and toes were mostly affected, but some attacks also included her nose, ears, and nipples. Effectively treating pediatric patients with Raynaud phenomenon can be challenging due to the side effects associated with first-line treatments. In this case, greatest success and tolerability was accomplished with sildenafil 20 mg daily and as needed prior to exposure to triggers.

Pediatric Raynaud Phenomenon Workup and Treatment: A Case Report
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2Chief of Rheumatology at the University of Miami

Of note her mother’s medical history was significant for RP and hypothyroidism which presented, simultaneously, around age 35. The mother’s evaluation was negative for rheumatic disease.

At the initial rheumatology clinic visit, her blood pressure was 112/78, heart rate was 72, and BMI was 17.6. Bluish discoloration of the fingers and toes was noted on initial physical exam, as well as several pitting scars at the tips of the fingers. No other physical exam findings were remarkable, including no evidence of cutaneous calcinosis. Initial workup included anti-centromere, anti-Th/To, anti-RNA polymerase II, anti-topoisomerase 1, anti-Ro, anti-La, anti-RNP, anti-Sm, C3/C4, aldolase, serum protein electrophoresis, total IgG, Rheumatoid factor, anti-dsDNA, anti-CCP, hepatitis C antibody, HIV antibody & antigen, CBC, CMP, EGFR, ESR, CRP and urinalysis. All values were within normal limits, except for ANA, which was 1:320 with a homogenous pattern. The ESR was 2 mm/hr, CRP was 0.28 mg/dL, and C3/C4 levels were 102 and 14.4 mg/dL respectively. Chest x-ray and pulmonary function tests were normal and failed to demonstrate any pattern of interstitial lung disease or pulmonary hypertension. Nailfold capillaroscopy showed tortuous, dilated capillaries and dropout [Fig. 3].

A diagnosis of RP was reached, with close follow-up for early detection of potential underlying connective tissue conditions. Since the patient already attempted lifestyle modifications, pharmacologic therapy was initiated. Amlodipine 5 mg daily was trialed for 3 months, which resulted in self-reported mild improvement in duration of attacks, but not the frequency of attacks,
during the last month of treatment. The patient, however, was not satisfied with the results, and the dose was increased to 10 mg daily. After two weeks at the higher dose, the patient discontinued the regimen due to fatigue and dizziness. Losartan 25 mg was trialed for 3 weeks, which was also discontinued due to orthostatic hypotension which failed to improve with time. Unlike the amlodipine, no improvement was seen in either frequency or duration of attacks. After two failed medications, sildenafil 20 mg twice daily was initiated.

At two months follow-up, the patient reported drastic improvement in both frequency and duration of symptoms. She reported no side effects from the medication. At one year follow-up, she self-reported improvement in quality of life and satisfaction with the medication’s ability to prevent attacks, particularly when taken about 45 minutes prior to cold exposures. While taking sildenafil therapy, attacks occurred only during exposure to rapid temperature changes, lasting less than 10 minutes, and were not observed when medication was forgotten. When medication was forgotten, the fingers and toes were cold and numb for over 10 minutes like they had previously. She currently follows up in clinic every 6 months, and reports fewer than 10 attacks per month, which each last about 5 minutes, while taking sildenafil. Overall, she has had significant symptomatic relief using sildenafil. No additional manifestations of connective tissue disease have emerged to date.

CONCLUSION:

Pediatric patients presenting with RP first require workup and, at times, ongoing scrutiny to rule out underlying connective tissue disease. When lifestyle modifications fail to provide satisfactory reduction in ischemic injury, attack frequency, and/or attack duration pharmacologic agents should be considered. If first line agents are unsatisfactory, due to inefficacy or intolerability, daily doses of sildenafil as well as dosing prior to trigger exposures should be considered. It is possible that pediatric patients may have lower blood pressure and more potential for hypotension with the usual first line agents used in adults. It may be appropriate to consider whether agents that have less effects on blood pressure should be used sooner in the therapeutic algorithm in pediatric patients. Rhodeheffer RJ, Rommer JA, Wigley F, Smith CR. - Controlled double-blind trial of sildenafil in the treatment of Raynaud’s. N Engl J Med. 1983;308(15):880-883.

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