RHEUMATOLOGY NURSE PRACTICE

Accredited education for registered nurses and advanced practice providers

WHAT ADULT PROVIDERS SHOULD KNOW ABOUT

PEDIATRIC RHEUMATIC DISEASES

Inside this Issue

VOLUME **5** / ISSUE **6**

- *How are pediatric rheumatic diseases similar and different from adult conditions?*
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ACTIVITY DESCRIPTION

In this issue of *Rheumatology Nurse Practice*, we'll review some of the most recent data concerning pediatric rheumatic diseases and their management, as well as offer a variety of helpful tools to help providers ensure that young patients are able to successfully transfer to adult care.

LEARNING OBJECTIVES

After participating in the activity, learners should be better able to:

- Review the most common rheumatic conditions that affect the pediatric population
- Discuss the impact of pediatric rheumatic diseases on patients' social, emotional, and intellectual development
- Identify the most common biologic therapies currently used to treat pediatric rheumatic diseases
- Assess your practice's pediatric-to-adult rheumatology practice transition plan and make updates as needed based on best current evidence

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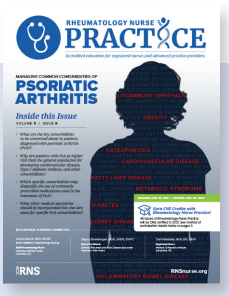

In Volume 5, Issue 5, of Rheumatology Nurse Practice, Table 2 on page 7 (Treatment Selection in the Presence of Comorbidities) contained some information that was outdated. An updated version of the table is located in the electronic version of this issue within the RNS Learning Center. Please refer to that version for more recent data:

rnsnurse.org/courses/rheumatology-nurse-practice-managing-the-common-comorbidities-of-psoriatic-arthritis/

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WHAT ADULT PROVIDERS SHOULD KNOW

ABOUT PEDIATRIC RHEUMATIC DISEASES

At one time, many pediatric rheumatic diseases were associated with considerable early mortality.[1] Today, thanks to medical advances, the majority of patients with pediatric rheumatic diseases not only live to adulthood, but many flourish unimpeded by their condition.[1] As they become adults, most pediatric patients continue to require rheumatologic care. In fact, roughly half of young people with pediatric-onset rheumatic and musculoskeletal diseases enter adulthood with active disease or develop disease flares as adults.[2,3] In addition, it has been estimated that roughly half of U.S. children with rheumatic diseases are currently being cared for by rheumatologists who specialize in adult care due to a national shortage of pediatric rheumatologists.[4]

For these reasons, adult providers of rheumatology care must be familiar with pediatric rheumatic diseases and the most important aspects of their treatment. In this issue of *Rheumatology Nurse Practice*, we will start by providing a brief overview of the pediatric rheumatic diseases that adult providers are most likely to encounter, whether in adult patients who have transitioned out of pediatric care or pediatric patients receiving care at an adult practice.

Juvenile Idiopathic Arthritis

Juvenile idiopathic arthritis (JIA) is the most common childhood chronic rheumatic disease.[5] Studies in North America and Europe have found that it affects approximately 4-16 of every 10,000 children.[6] In the United States, this translates into roughly 294,000 children with the disease.[6]

JIA is actually an umbrella term for a heterogeneous group of 7 diseases: systemic arthritis, rheumatoid factor (RF)-positive polyarthritis (which is very similar to adult RF-positive rheumatoid arthritis), RF-negative polyarthritis, oligoarthritis, psoriatic arthritis, enthesitis-related arthritis, and undifferentiated arthritis.[7] In all of these diseases, arthritis of an unknown origin persists for more than 6 weeks, and onset occurs before 16 years of age.[8]

All types of JIA are associated with decreased health-related quality of life and a risk of permanent joint damage.[8] In one recent study of patients diagnosed with JIA, the average age at onset was 6.4 years, and the symptoms that most commonly led families to seek medical attention were arthralgia, arthritis, fever, skin rash, abdominal pain, and uveitis.[9] Because JIA is the most common of the pediatric rheumatic diseases, it receives special attention in this issue.

Juvenile Ankylosing Spondylitis

Juvenile ankylosing spondylitis (JAS) is a type of inflammatory arthritis that begins before age 16 years and primarily affects the spine and the spots in the body where tendons, muscles, and ligaments attach to bone.[10,11] Over time, a patient's vertebrae may fuse, impeding movement, as well as the rib cage, making it difficult to breathe. JAS is more common among boys than girls. It is associated with the human leukocyte antigen (HLA)-B27 allele, a characteristic it shares with adult patients with ankylosing spondylitis.[10] In one study of JAS patients, the average age at onset was 13.5 years, with an onset range between 5 and 16 years of age.[12] Many patients with JAS are initially diagnosed with enthesitis-related arthritis, a type of JIA, until their spine or lower back becomes involved. This often happens years after their first symptoms appear.[13] One study has found that, relative to adult patients with ankylosing spondylitis, patients with JAS are more likely to present with peripheral enthesopathies and arthritis.[14]

Rheumatic Fever

Rheumatic fever is an inflammatory disease that typically develops 1 to 5 weeks after an untreated streptococcal infection such as strep throat or scarlet fever.[15] It most often affects children between the ages of 5 and 15 years, and it can cause permanent damage to the heart.[16,17] The most common major manifestations of acute rheumatic fever include carditis, polyarthritis, chorea, erythema marginatum, and subcutaneous nodules. The most common minor manifestations include arthralgia, fever, raised erythrocyte sedimentation rate or C-reactive protein concentrations, and prolonged PR interval on electrocardiogram.[16] In the United States, acute rheumatic fever is rare, with just 0.61 cases occurring per 100,000 children.[18] Globally, however, rheumatic heart disease, which is a sequela of acute rheumatic fever, remains a notable source of mortality and disability.[19] Rheumatic fever can be prevented by treating streptococcal infections promptly; the condition is treated by managing inflammation and using antibiotics to treat the underlying bacterial infection.[15]

Juvenile Systemic Lupus Erythematosus

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease that can involve any organ system in the body. It has a wide range of manifestations.[20] Roughly 15-20% of patients with SLE are diagnosed during childhood. Juvenile SLE typically has a more aggressive course than adult-onset SLE, with higher rates of organ involvement.[1] However, juvenile SLE is rare, affecting 3.3-8.8 per 100,000 children, with a median age of onset between 11-12 years. Because it shares characteristics with many other autoimmune diseases, juvenile SLE has been called "the great mimicker." Some of the most common clinical features of juvenile SLE include constitutional and generalized symptoms (eg, fever, lymphadenopathy, weight loss), malar or butterfly rash, painless oral ulcers, arthralgias and arthritis, renal disease, and cytopenias. Similar to patients with adult-onset SLE, roughly 80% of patients with juvenile SLE are female.[20]

Kawasaki Disease

Kawasaki disease is a rare, acute pediatric vasculitis whose main complication is coronary artery aneurysms.[21] In the United States, it occurs in only 8-20 per 100,000 children under 5 years of age, but it is a leading cause of acquired heart disease.[22] Its clinical features include fever, rash, mucosal changes, conjunctival erythema, and cervical lymphadenopathy. Available evidence strongly supports an infectious etiology.[23] Virtually 100% of cases occur in children, with 80% occurring in children <5 years of age and 50% occurring in children <2 years of age.[23] Recently, Kawasaki-like disease has emerged as a rare complication of COVID-19, drawing increased attention to this rheumatic condition.[21] Standard treatment consists of intravenous immunoglobulin and aspirin.[22]

Treatment of Pediatric Patients with Rheumatic Disease

With the advent of biologics, the prognosis for patients with pediatric rheumatic diseases such as JIA has improved substantially (see Table 1). One recent study of JIA comparing outcomes in the methotrexate (pre-2000) era vs. the biologic era found that patients diagnosed in the latter period had consistently higher frequencies of inactive disease and low-disease activity, as well as lower levels of disease damage and impairment.[24] Not surprisingly, the use of biologics to treat pediatric rheumatic disease has become increasingly common. One recent study conducted at Children's Hospital of Philadelphia found that 74% of patients with early polyarticular JIA received biologic therapy.[25] In fact, many patients with JIA will receive multiple biologics during the course of their childhood. A recent study that followed pediatric patients with JIA for a median of 2.2 years found that 23% of those who initiated biologic therapy received at least 2 biologics, 5% received at least 3, and 1% received at least 4.[26]

A treat-to-target (T2T) approach is recommended for patients with JIA, with the primary treatment target being clinical remission.[27] Intervening early and effectively seems to be especially important for pediatric rheumatic diseases. Several studies have demonstrated the effectiveness of early intervention with biologics. One found that when patients with new-onset systemic JIA were treated-to-target with the IL-1 inhibitor anakinra, the median time to achieve inactive disease was only 33 days.[28] After 1 year, 76% of patients had inactive disease and 52% had inactive disease that did not require medication. After 5 years, 96% of patients had inactive disease and 75% had inactive disease that did not require medication. Another study compared outcomes for patients with newly diagnosed polyarticular JIA who received either (1) early, aggressive use of biologics in combination with conventional synthetic disease-modifying antirheumatic drugs (DMARDs) such as methotrexate, or (2) delayed use

Table 1 *Biologics Approved by the FDA to Treat Pediatric Rheumatic Diseases*[a]

Agent	Class	Indication
Belimumab	B-lymphocyte stimulator-specific inhibitor	Active, autoantibody-positive SLE in patients aged 5 years and older who are receiving standard therapy
Canakinumab	Interleukin-1b inhibitor	Active systemic JIA in patients aged 2 years and older
Tocilizumab	Interleukin-6 inhibitor	Active polyarticular JIA in patients aged 2 years or older Active systemic JIA in patients aged 2 years or older
Abatacept	Selective T-cell costimulation modulator	Moderately to severely active polyarticular JIA in patients 6 years of age and older
Adalimumab	TNF inhibitor	Moderately to severely active polyarticular JIA in patients 2 years of age and older
Etanercept	TNF inhibitor	Polyarticular JIA in patients aged 2 years or older

[a] *Many patients with pediatric rheumatic diseases receive other biologics off-label*
Abbreviations: JIA, juvenile idiopathic arthritis; SLE, systemic lupus erythematosus

of biologics following an initial conventional synthetic DMARD.[29] This study found that early, aggressive use of biologics was more effective at reducing disease activity by 6 months. Moreover, adding a biologic to the initial conventional synthetic DMARD after 6 months was found to provide little added benefit.

However, not all evidence supports the early use of biologics over conventional synthetic DMARDs. A recent randomized study compared the T2T approach in DMARD-naïve JIA patients using 3 different therapies: sequential conventional synthetic DMARD monotherapy (sulfasalazine or methotrexate), methotrexate plus 6 weeks of prednisolone, or methotrexate plus the TNF inhibitor etanercept.[30] In this study, 71% of recent-onset JIA patients were able to achieve inactive disease by the end of the 24-month study period, and 39% were drug-free. Outcomes and adverse events were similar regardless of the therapy received, suggesting that perhaps the T2T approach—treating pediatric rheumatic disease as quickly and effectively as possible to preserve long-term joint health—is more important than whether a conventional synthetic DMARD or biologic is used.

Currently, the American College of Rheumatology (ACR) conditionally recommends initial therapy with a conventional synthetic DMARD over a biologic for patients with JIA.[8] The 2019 ACR guidelines for JIA do acknowledge situations when initial therapy with a biologic may be preferred, including when risk factors are present, high-risk joints (cervical spine, wrist, or hip) are involved, high disease activity is observed, or patients are judged by their healthcare providers to be at high risk for disabling joint damage. Here, risk factors include the presence of positive anti-cyclic citrullinated peptide antibodies, positive RF status, or the presence of joint damage. In addition, the ACR guidelines note that the majority of their "Parent and Patient Panel" voted against use of conventional synthetic DMARDs as initial therapy due to methotrexate's adverse effect profile and the participants' perspective that biologics resulted in better outcomes. Finally, the guidelines conditionally recommend that when patients with JIA and polyarthritis do initiate treatment with a biologic, combination therapy with a conventional synthetic DMARD should be used over biologic monotherapy. Of note, due to a lack of data, the ACR guidelines do not recommend any one class of biologic over another for initial therapy for JIA.

Because of the conditional ACR recommendation in favor of initial therapy with a conventional synthetic DMARD, many pediatric patients with JIA or other pediatric rheumatic diseases may not receive biologics until later in the course of their disease. The cost of biologic therapy, as well as insurance requirements, may also discourage initial therapy with biologics. Finally, safety concerns may deter some pediatric providers from prescribing initial therapy with biologics. It was once thought that the use of TNF inhibitors, the most frequently prescribed biologic class for JIA,[26] might be accompanied by a small increase in the risk of lymphoma and other types of cancer in children.[5] However, the most recent data suggest that patients with JIA experience an elevated risk of malignancies absent TNF inhibitor therapy and that the risk experienced by those taking TNF inhibitors is not appreciably higher.[31] JIA itself is also associated with a higher rate of infection than is observed in age-matched controls; even so, the absolute rate of serious infections among children with JIA taking TNF inhibitors, which are also associated with greater susceptibility to infection, is low.[31]

Impact of Pediatric Rheumatic Diseases on Social, Emotional, and Intellectual Development

Pediatric rheumatic diseases such as JIA can affect virtually every aspect of a child or adolescent's development. Thus, in addition to considering disease-related outcomes, providers must consider young patients' psychological, social, and vocational outcomes,[32] as well as their health-related quality of life. One recent study found that 8-14% of patients with JIA experience persistent major impairment in their health-related quality of life.[33] High initial levels of disease activity increase the odds that a patient will experience this negative outcome, emphasizing the connection between disease control and quality of life. Of note, this study found that health-related quality of life is slower to improve than outcomes such as disease activity, pain, or disability.

Research has established a bidirectional relationship between chronic physical disease and mental illness.[34] Young adults with chronic rheumatic conditions such as JIA are at higher risk than their peers of developing mental health disorders such as major depressive disorder (MDD).[35] Indeed, children with rheumatic diseases appear to be more vulnerable to MDD than adults with rheumatic diseases. One study found that patients with juvenile SLE had a significantly higher risk of a major depressive episode or recurrent major depressive episodes than individuals with adult-onset SLE.[36] Indeed, in one study of patients with juvenile SLE, 59% screened positive for depression, and increasing severity of depression symptoms was associated with medication nonadherence.[37] Risk factors for depression among individuals with juvenile SLE include higher disease activity and poorer physical functioning.[36]

Research has also shown that adolescents who experience pain, as many patients with pediatric rheumatic diseases do, are also more likely to experience suicidality (eg, suicidal ideation, behaviors, and death). A recent systematic review found that experiencing either acute or chronic pain approximately doubled adolescents' risk of suicidality.[38] Although MDD played an important role, it did not entirely explain the association between pain and suicidality.

"Ideally, the process of transitioning a patient from pediatric to adult care should begin in early adolescence. At that time, pediatric providers can start preparing young adults and their families for the greater independence and responsibility required after the transition to an adult practice.[46]"

Because of the high rate of MDD among patients with pediatric rheumatic diseases, rheumatologists can play an important role in screening for the disorder.[36] The American Academy of Pediatrics (AAP) recommends screening all adolescents for MDD annually,[39] and the U.S. Preventive Services Task Force recommends screening adults for MDD.[40] Adult providers treating pediatric or young adult patients may want to consider incorporating MDD screening into their visits. The Patient Health Questionnaire (PHQ)-2 is a simple and fast tool to administer. If possible symptoms of depression are identified using this tool, the provider can follow-up by asking the patient to complete the more in-depth PHQ-9 or conducting a clinical interview.[41]

Regularly asking young patients about their social and functional wellbeing is also an essential part of pediatric rheumatology care. In a recent survey of patients with JIA aged 8 to 17 years, 32% reported being unable to participate in activities inside or outside class with their peers at school, and 41% reported the same for gym class.[42] Some respondents reported feeling social anxiety or embarrassment as a result. Another study found that, although school sports participation among children and adolescents with JIA has increased significantly over the past 15 years, roughly 20% still participate in school sports only sometimes and more than 15% remain fully exempt.[43] Of note, one study found that even children and adolescents with low levels of JIA disease activity have significantly lower levels of physical activity than their healthy peers, primarily because they spend less time participating in club sports.[44]

An especially important disruptor of patients' social and functional participation in life is fatigue. Research shows that roughly 25% of 10- to 18-year-olds with rheumatic disease experience severe fatigue, which is associated with significantly lower levels of functioning and significantly more school absences.[45] By asking about young people's ability to function, particularly at school, providers can ensure that these patients receive the support they need to live full and satisfying lives.

Transitioning from Pediatric to Adult Rheumatology Care: Keys to Success

Ideally, the process of transitioning a patient from pediatric to adult care should begin in early adolescence. At that time, pediatric providers can start preparing young adults and their families for the greater independence and responsibility required after the transition to an adult practice.[46] Unfortunately, too often the transition process is neglected, and the care of many young patients suffers as a result. In one recent study of 152 young adults with a rheumatologic diagnosis transitioning from pediatric to adult providers, 14% did not make it to their first adult appointment and 57% did not make it to their second adult appointment.[47] In another study, only 23% of patients with rheumatologic diagnoses who transitioned to adult care using the typical referral process visited their adult provider more than once in the 6-8 months following transition.[48] The risks associated with a rocky transition include gaps in continuity of care, decreased quality of care, increased healthcare costs, poor treatment adherence, self-management challenges, and poor health.[46] Here, we explore the elements of a successful transition, as well as the actions an adult provider can take to maximize a young patient's chances of treatment success after the care transition.

Special Needs of Patients with Pediatric Disease

Patients with pediatric rheumatic disorders such as JIA should be recognized as a distinct patient group as they transition to an adult practice due to their special characteristics and needs.[32] These patients are likely to pose unique challenges for the adult provider. Such challenges may include having to manage an unfamiliar pediatric rheumatic disease; interact with young adults who are still learning to take responsibility for their health; interact with their parents, whose level of involvement may seem excessive; and provide care despite insufficient medical records from pediatric providers.[49] Providers should be cognizant that, during the period when young adults typically transition to adult care, they are at a uniquely vulnerable place in their lives. They are likely to be preoccupied with their body image; relationships with family, peers, and lovers; and school and recreational activities.[49] At this stage in life, it is all too easy for patients to relegate health concerns to the back burner, particularly if their parents have stepped back from managing their care.

Practical considerations can also complicate care. Young patients are more likely than older patients to experience "churn" in insurance coverage, forcing them to change providers more frequently.[46] Some may age out of being covered by their parents' health insurance plan and have to find another source of coverage. These circumstances can make providing continuous care and access to much-needed medications challenging. In addition, the modes of communication that are most effective for young patients may differ from those that work well for older patients. For example, young patients may be more comfortable with electronic portals or texting and less responsive to calls or mail than older patients.[46]

Establishing a Transition Protocol

Despite the importance of the transition process, many adult rheumatology practices do not have established protocols for integrating young adults into their practice.[46] Establishing such protocols is essential for making sure that all young patients receive quality care. An AAP/American College of Physicians/American Academy of

Table 2 *The Six Core Elements of Transitioning Young Patients with Chronic Illnesses Into Adult Practices*[55]

Core element	Pediatric provider	Adult provider
1. Transition and/or care policy	Create and discuss with youth and/or family	Create and discuss with young adult and guardian, if needed
2. Tracking and monitoring	Track progress of youth and/or family transition preparation and transfer	Track progress of young adult's integration into adult care
3. Transition readiness and/or orientation to adult practice	Conduct transition readiness assessments	Share and discuss welcome and FAQs with young adult and guardian, if needed
4. Transition planning and/or integration into adult approach to care or practice	Develop transition plan, including needed readiness assessment skills and medical summary, prepare youth for adult approach to care, and communicate with new clinician	Communicate with previous clinician, ensure receipt of transfer package
5. Transfer of care and/or initial visit	Transfer of care with information and communication, including residual pediatric clinician's responsibility	Review transfer package, address young adult's needs and concerns at initial visit, update self-care assessment and medical summary
6. Transition completion or ongoing care	Obtain feedback on the transition process and confirm young adult has been seen by the new clinician	Confirm transfer completion with previous clinician, provide ongoing care with self-care skill building and link to needed specialists

Family Physicians joint clinical report outlines the six core elements for transitioning young patients with chronic diseases from pediatric to adult practices, distinguishing between the responsibilities of each type of provider (see Table 2). In addition, the European League Against Rheumatism (EULAR) and the Paediatric Rheumatology European Society (PReS) have released helpful guidance specifically focused on the transitional care of young people with pediatric rheumatic diseases (see Figure 1).[2] Fortunately, ACR offers resources to support adult providers seeking to implement the six core elements and EULAR/PReS recommendations, including drafts of patient welcome letters, patient self-assessments, medical summaries, and condition fact sheets.[50]

If an adult rheumatology practice frequently receives transfers from one or more pediatric rheumatology practices, it is likely worthwhile for the adult practice to reach out to the pediatric practice to establish a relationship if one doesn't already exist. The adult provider can initiate a discussion of how the two practices can work together to facilitate smooth transitions for patients. By building a strong relationship with pediatric providers, adult providers can set the stage for a free and open communication of information that can benefit transitioning patients, as described in Table 2. Pediatric practices may also find the transition resources that ACR offers helpful, including draft transition policies, patient transfer letters, and medical summaries.[50]

One important aspect of transition planning is determining whether a young patient is ready to transition to adult care. Got Transition, the federally funded national resource center on healthcare transitions, recommends that transitions to an adult provider happen no later than age 22 years, although some patients will be ready to transfer earlier than others.[51] Before young patients can successfully transition to adult care, they must be comfortable interacting independently with providers, obtaining and taking their medication consistently, and negotiating the insurance and health care system.[46] Self-assessments are available to help determine whether a young patient is ready to transition—and if not, to identify the areas in which they need help building skills. One such self-assessment tool is the Transition Readiness Assessment Questionnaire (TRAQ), one of the transition resources available on the ACR website.[50] This 1-page, 20-question form evaluates a patient's ability to manage medications, keep appointments, track health issues, talk with providers, and manage daily activities.

In addition to considering a patient's maturity level when determining whether the time is right for a transition, providers should consider several other important factors. First, transfer of care should optimally occur when a patient's disease is well controlled.[46] Attempting a transfer during the middle of a flare could compromise the care team's ability to manage a patient's disease. Second, if a young person needs to make transitions for multiple provider types, it is best to stagger the transitions.[46] This will help ensure that the patient does not become overwhelmed by simultaneous changes.

Figure 1

Select Recommendations from EULAR/ PReS Standards for the Transitional Care of Young People with Juvenile-Onset Rheumatic Diseases[2]

1. Young people should have access to high-quality, coordinated transitional care, delivered through partnership with healthcare professionals, young people, and their families, to address needs on an individual basis
2. The transition process should start as early as possible, in early adolescence or directly after the diagnosis in adolescent-onset disease
3. There must be "direct" communication between the key participants (to include, at minimum, the young person, parent/caregiver, and a member each of the pediatric and adult rheumatologist teams) during the process of transition. Before and after the actual transfer, there should be "direct" contacts between pediatric and adult rheumatology teams.
4. Individual transition processes and progress should be carefully documented in the medical record and planned with young people and their families
5. Every rheumatology service and clinical network—pediatric and adult—must have a written, agreed, and regularly updated transition policy
6. There should be a clear description of the multidisciplinary team involved in transitional care, locally and in the clinical network. The multidisciplinary team should include a designated transition coordinator.
7. Transition services must be young-person-focused, developmentally appropriate, and address the complexity of young person development
8. There must be a transfer document

Figure 2

Helpful Resources for Supporting Successful Transitions to Adult Rheumatology Care

Got Transition website:

www.gottransition.org

Got Transition is the federally funded national resource center on healthcare transition. Though not specific to rheumatology care, it provides helpful resources for supporting successful transitions, including the following:

- Implementation guides for providers seeking to integrate the six core elements of successful transitions into clinical practice
- A transition coding and reimbursement tip sheet
- FAQs and resources for youth & young adults about the transition process
- FAQs and resources for parents & caregivers about the transition process

The American College of Rheumatology's Pediatric to Adult Rheumatology Care Transition website:

www.rheumatology.org/Practice-Quality/Pediatric-to-Adult-Rheumatology-Care-Transition

This website includes a wealth of resources specific to rheumatology, including sample transition policies, transfer and welcome letters, condition medical summaries, transition readiness assessments, and more.

Teens Taking Charge: Managing Juvenile Idiopathic Arthritis (JIA) Online website:

teenstakingcharge.carragroup.org/en/jiateen

This website, created by the Childhood Arthritis and Rheumatology Research Alliance, contains helpful information written especially for teens about JIA; JIA medications and non-medical therapies; strategies for managing symptoms, coping, and self-monitoring; and more. Young patients who are beginning to take charge of their own care may find it especially useful.

Of course, much of the time, adult providers have little input into when a patient transitions into their practice. In these cases, self-assessments such as the TRAQ can still offer important insights into a new patient's ability to adapt to adult care, as well as the areas in which they may need special support. In some situations, adult providers may want to discuss whether a transition is occurring prematurely with a patient, their caregivers, and the pediatric practice from which they are being transferred. It is important to keep in mind that seemingly small age differences can make big differences in young patients' transition readiness. In one study of young patients, only 27% of 17-year-olds scored above the behavior cutoff for transition readiness, but 62% of 18-year-olds did so.[52]

Adult providers may wonder how they can realistically incorporate the six core elements of successful transitions into their practice. After all, the activities needed to support successful transitions take time, and time is in short supply for most busy rheumatology practices. Fortunately, providers can bill transition assessments as health care assessments.[46] In addition, the Got Transition website (www.gottransition.org) includes a coding and reimbursement tip sheet for the transition from pediatric to adult health care (see Figure 2).

The Initial Visit

A young patient's initial visit to an adult provider is a key opportunity to establish a solid foundation for future treatment. It is vital for the provider to take the time to get to know young adult patients and gain their trust and confidence.[46] It is also important for adult providers to realize that young patients and their families often have negative expectations regarding the transition to adult care.[46] Over time, the pediatric rheumatology office may have come to feel like a second home to these patients, and the providers there may have come to feel like family members.[53] It can be challenging for patients to leave this comforting and familiar environment behind. Whereas adult providers may perceive young adult patients as ill-prepared and "needy," young adults may view adult providers as disinterested, distant, disease-focused, and rushed.[46] By establishing a warm relationship from the onset, providers can help ease the transition to adult care and ensure that young adults do not feel abandoned by their care team.

During the first visit, the adult provider can also set realistic expectations for the nature of the care they provide. Pediatric rheumatologists often assume primary care duties, such as overseeing immunizations.[46] Young patients who are transitioning into an adult practice may be unaware that these services are not a typical part of adult rheumatology care, and they may need referrals to appropriate providers who can offer these services.

Special Challenges for Young Adults: Adherence and Reproductive Health

Young adults, whose risk-taking behaviors are often more pronounced than those of older patients, often struggle

with adherence.[46] Here, struggling with adherence may include failure to take medications; show up for visits; or comply with laboratory monitoring, exercise regimens, or other behavioral recommendations. To address a patient's nonadherence, providers must talk with them about the cause. Is the patient forgetting to take a medication, in which case some type of reminder system or app may help? Are they having difficulty filling prescriptions—and if so, why? Are the out-of-pocket costs for their medication too high, and if so, are there programs that could help, or would another medication be more affordable? Is their regimen too complex, and might it be simplified in some way? Do they lack transportation to visits, and if so, could some type of assistance be offered? By helping young patients troubleshoot adherence problems, providers can ensure they will receive maximum benefit from their treatment plan.

Providers should also be aware that reproductive health issues are an important consideration for young women of reproductive age. Some medications used to treat pediatric rheumatic diseases are teratogenic. The risk-taking behaviors of young adults may make it difficult for them to behave responsibly in the context of these medications,[46] so candid conversations about the risks associated with treatment, as well as effective means of birth control, are essential. In addition, many adolescent and young adult women with pediatric rheumatic diseases have concerns about the potential impact of their condition and potential medications on motherhood, as well as obtaining safe and effective contraceptives.[54] An adult provider cannot assume that a pediatric provider has already discussed these issues with their patients: conversations about reproductive health are an essential part of caring for young women with rheumatic diseases.

Conclusion

Navigating the transition from pediatric to adult care is difficult for many patients with pediatric rheumatic diseases. Adult providers can help young people build a foundation for success by establishing effective transition protocols and recognizing this group of patients' unique needs. With the array of effective medications now available to treat patients with pediatric rheumatic diseases, adult providers have the potential to meaningfully improve their patients' health and quality of life. In addition, helping young adults become independent and take control of their own healthcare decisions can prove immensely rewarding.

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A Mile(s) in Their Shoes

by Carrie Beach, BSN, RN-BC

I understand that transitioning from a pediatric to an adult rheumatologist can be a very scary moment for many of our young adult patients. Often, these patients have been with their providers for many years and have built strong relationships built on trust. What these young adults don't know is that I have a secret, too… as providers, sometimes we're just as scared as they are!

As an adult rheumatology provider, I am always focused on learning and keeping myself up to date about every rheumatologic condition that affect adults as well as the medications that are used to treat these conditions. It's a lot of information to keep up with, and it can be overwhelming at times, so it's not a big surprise that I sometimes forget that these diseases can affect kids too. And sometimes, these diseases present in a very different manner than we are used to.

Heather is a longtime patient of ours that was diagnosed with erosive rheumatoid arthritis (RA) at age 34. It was a pretty classic case of adult-onset RA as she presented with synovitis and joint pain in several joints, a positive rheumatoid factor, and a family history that included both RA and systemic lupus erythematosus. During the course of her treatment, Heather responded reasonably well to several disease modifying anti-rheumatic drugs, and then eventually to biologic therapy once those were needed to help keep her disease under control. All in all, she was a pretty typical RA patient for us—no big surprises.

Over the years, Heather's appointments were part medical and part social check-ins. We had built a great rapport and, since her condition rarely changed, we always had time to discuss our families and what our kids were up to. Years ago, Heather told me about some of the health issues her son, Miles, was experiencing when he was about 4 years old. He had recently been quite ill, requiring a feeding tube at one point, and she had been extremely worried about him. Following his recovery, he went on to develop joint pain in several different joints, which raised concerns for Heather that her son was experiencing autoimmune arthritic symptoms.

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During the next 3 years as Miles started school, Heather would fill me in periodically on some of the battles she had been having with her pediatrician over her son's symptoms. The pediatrician was, I was told, attributing Miles' joint issues to "growing pains" that he would eventually outgrow. It was only after swelling in his left knee appeared years after these "growing pains" had first emerged that Miles was finally referred to a pediatric rheumatologist for evaluation. He was 7 years old when he was initially diagnosed with polyarticular JIA, which was later changed to enthesitis-related JIA.

I would hear from Heather on and off about Miles' health. He had done well on infliximab for many years and was now preparing to head off to college (man, I feel old!). Heather told me that Miles was planning to continue with infliximab infusions while he was away at school, but she was concerned the current plan might not be realistic. The closest pediatric rheumatologist to Miles' college was several hours away in a rural community. That would be hard for any patient, much less a college student. Heather asked if it would be possible to have Miles transition to our practice to take over his infusion schedule and overall care. I didn't think twice before saying yes and quickly set up a tentative date for him to come for an initial visit during his holiday break from school.

It was more than a year later that I realized that Miles never came in for that visit. We're all busy, and it's not always a priority for us to follow up with patients who haven't established their presence in our practice. Because Miles wasn't in our electronic medical record, there was nothing reminding me that he was supposed to have come to see us. It wasn't until an "urgent" request came into our scheduling department for a new patient visit that Miles reappeared on my radar. As with all urgent requests, I asked about the patient's circumstances to be sure this was truly an urgent case. I was told that the patient's mother had called in a panic because her son was about to lose his health insurance and his pediatric rheumatologist was retiring. He needed to establish himself as a new patient within our practice ASAP in order to continue with his longstanding treatment regimen. I knew right away this "urgent patient" was Miles.

Miles' first visit was a tough one for me. Due to the urgency of his situation, we saw him before any records had arrived from his pediatric rheumatologist. And while I wasn't expecting a 20-year-old male college student to open up and tell me his life story on the day we met, Miles truly was a man of few words! I got a few mumbles and nods from him, but not much else. Fortunately, his mom accompanied him to this visit and filled in a lot of details that were missing. I knew I couldn't possibly immediately fill the shoes of the pediatric nurse practitioner who had managed Miles' disease since childhood, but I was determined to slowly build up a level of trust with him and get him to open up, at least a little.

Building rapport and establishing trusting relationships with patients is always my personal goal as a rheumatology nurse. When I see new patients, I always feel confident in building that trust from the very first visit. However, when it comes to pediatric patients transitioning to an adult rheumatology practice, I am always a little timid. These were kids who had put a lot of trust in their pediatric provider, as did their parents. In addition, I am admittedly no expert in pediatric rheumatic diseases (though reading this issue of *Rheumatology Nurse Practice* will surely help!). After Miles' first visit, I took a crash course on his specific pediatric diagnoses to try to figure out how those translated to adult disease and possible treatment regimens.

As more pediatric patients are diagnosed with a rheumatic disease, they will inevitably start arriving more and more frequently in adult practices as they get older. It's not an easy transition—for us or them—so it's important to be prepared. Learn what you can about the most common pediatric conditions. Develop a formal transition plan, with flexibility built in to account for the individualism of the patient. While these are "new patients" to us, they are not new to rheumatology, and they likely know more that we give them credit for.

The Transition In, and Then Out

by Nancy Eisenberger, MSN, APRN, FNP-C

Transitioning from pediatric to adult rheumatology can be quite a traumatic experience for many patients with rheumatic disease. Some have seen their pediatric rheumatologist since their earliest days of childhood and cannot recall a year going by without a handful of visits to their office. Most have built special bonds with their team of providers who have watched them grow from toddlers into teenagers, offering guidance and a helping hand to manage the ups and downs of their disease.

While I do not often see patients until they reach their early adulthood in my current rheumatology practice, there have been a handful of times in my career when I have become involved with patients at an earlier age. Abby is the patient I always think about most.

Abby was 15 years old when she first came to our adult rheumatology practice. Since her diagnosis of juvenile idiopathic arthritis at age 4, Abby had been treated by a wonderful pediatric rheumatologist near a leading academic center for several years, but the 5-hour roundtrip drive for her busy family was becoming a growing burden. Her pediatric rheumatologist had called our practice a year before we first met Abby asking if we would be willing to take over her care. He explained that Abby was an unusually mature patient whose family was simply worn out by the long commute, and he felt an early transition to an adult practice would be the best thing for them. Despite our reluctance to take on such a young patient, we eventually agreed.

Prior to her arrival, our team reviewed Abby's medication history and recent lab results. We were lucky to have been provided with such thorough documentation from her pediatric practice. That does not always happen.

Yet even though we thought we knew what to expect, we were completely blown away when Abby first came into our office. She was an exceptional young woman. Vibrant yet soft spoken, Abby oozed a positive attitude despite the toll her disease had taken over the years. She had contracture deformities across her entire left hand and ulnar drift of her right hand, despite technically being in clinical remission (her disease had remained stable for >2 years). Abby also had some minor bone changes in her left knee.

At the time we first met her, Abby was being treated with a regimen of weekly subcutaneous etanercept 50 mg, weekly subcutaneous methotrexate 25 mg, daily folic acid 1 mg, and daily hydroxychloroquine 200 mg daily. She was also taking oral birth control.

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I knew that I was going to have to take things slowly with Abby. No matter the age of the patient, going from a longtime provider to a new team can be scary. Our first discussion therefore focused on Abby's feelings surrounding the change in providers. She admitted being nervous about suddenly having to share her life with a new team. We were grateful to her pediatric rheumatologist in that he told Abby he would still remain available as a resource if she had any urgent questions that she was uncomfortable discussing with her new team of providers.

We then started talking about the things she liked to do. Abby told me that she was currently running for class president (she won). She served on a variety of school committees. Her relationship with her parents seemed rock solid. She emphasized to me that she was not going to let her arthritis slow her down. This was all good information to hear and made me feel more comfortable knowing that this was a motivated patient who seemed to have her life in order.

Once we covered her personal history, our discussion moved into the expectations of our practice. I explained to Abby the frequency we expected to have her labs monitored, which fortunately mirrored the expectations of her pediatric practice. We set a mutual goal for her disease, which was the maintenance of disease remission.

Over the next few months and years, I continued to see Abby regularly, and our relationship matured. I saw her through her high school years and witnessed a variety of milestones, including prom, high school graduation, her college years, and then a few years ago, her wedding.

Two years ago, Abby came to me looking a little more nervous than usual. I asked her what was wrong. Was her disease flaring? Was she having problems with her personal life or job?

"No, it's none of that," she told me. "I'm moving."

It always hurts a little when we "lose" a patient we've spent so much time with over the years, but once I got over the initial shock, I thought about all of the ways I could help Abby with her next healthcare transition. Just like her pediatric rheumatologist had done so many years ago, I gave Abby my phone number and told her to call me if she had any questions.

I talked to Abby a few months later once she was settled into her new home and had seen her new rheumatologist. Abby told me she and her husband had decided it was time to start a family, and Abby wanted to make sure the changes her rheumatologist had suggested were appropriate. Yes, I told her, he was right to stop her methotrexate. Six months later, Abby discontinued all of her medications, which is not what we typically recommend but was a personal decision she felt most comfortable with. She fortunately soon got pregnant.

I next heard from Abby 2 months into her pregnancy. Unfortunately, her disease was flaring despite intra-articular injections and a pulse of low-dose steroids. Her new rheumatologist suggested that she restart etanercept. Before agreeing to do so, Abby called me to get my thoughts. This put me in a tough situation. I did not want Abby to continue to second-guess her current provider and make her feel like she was always looking over his shoulder for my approval, but I also felt loyalty to Abby. Fortunately, it was a moot point in this circumstance, as I explained to Abby that controlling her arthritis was vital during the early stages of pregnancy and that we knew enough about the use of tumor necrosis factor inhibitors such as etanercept during pregnancy to feel comfortable with its safety. If I were still her provider, I told Abby, I would make the same recommendation as her current rheumatologist.

My hope is that Abby's comfort level with her new provider is blossoming, just like it did with me so many years ago. I do not know how many times she called her pediatric rheumatologist after transitioning into my care—I have never asked—but her current reticence is certainly not surprising.

The transitioning of patients, whether it is from a pediatric to adult practice, or from one adult practice to another, is so important for our patients. Due to life circumstances, patients with chronic diseases often to go through a variety of healthcare stops, with every change bringing in unknown variables. While we cannot control the recommendations of the new practice, it is our job to provide the new provider team with whatever assistance we can to best serve our patients. In the case of Abby, her transition into and then out of our practice is a model I often think about when I am either about to say hello to a new patient or goodbye to an old one. We should always think about the ways in which every transition affects our patients and how we can do the best possible job to impact their future in a positive way.

NIGHTMARES in Rheumatology

by Teri Puhalsky, BSN, RN, CRNI

In the adult rheumatology world, the transition of a pediatric patient into our world is often tricky. We know that the patient's juvenile idiopathic arthritis (JIA) does not magically transform into adult rheumatoid arthritis (RA) with the wave of some magic wand. While in some cases, these patients will now be able to access some biologic or small molecule therapies that were out of reach for them as children and young adults, we have no idea if those will be any more effective in controlling their disease and preventing further joint damage than previous options each patient has tried.

There are a host of other questions as well. What is the family dynamic like? Are the parents ready to hand over authority for some treatment decisions to their child? How well does the family understand the disease process? How can we make sure we don't lose the patient to follow up as their lives become busier and they gain more and more independence?

There are often a lot of questions with a variety of vague answers. The stuff of nightmares!

We recently had a patient, Melanie, who unfortunately keeps us all up at night. She came to us from her pediatric rheumatology practice at the age of 16. Melanie is the oldest of three children. She is active in her school and does cheer for the football and basketball teams.

Melanie was diagnosed with JIA at the age of 12. Her prior medication history included naproxen, methotrexate, etanercept, adalimumab, prednisone, and folic acid. She was able to self-inject both the etanercept and adalimumab. Both worked for a short time before losing their efficacy.

Melanie initially transitioned to our office to begin her first infusible biologic, tocilizumab. She was accompanied by her father at her initial appointment. A physical exam showed bilateral swelling in her feet and ankles. Melanie's knees were also swollen and tender. When asked if this level of disease activity was normal, Melanie admitted that things had gotten worse recently since she had missed several injections. Her father chimed in that it was getting to be increasingly difficult to get Melanie to administer her injections without lots of nagging. The increasing general pain and stiffness had caused Melanie to step away from her school's cheer team.

With the change in her medication regimen to tocilizumab, we reiterated to Melanie the importance of adhering to her infusion schedule. The fact that Melanie did not have her

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drivers license made things a little more complicated but was perhaps a blessing in disguise as it made it more difficult for her to skip a dose without her parents knowing since they would be the ones driving her to each appointment.

Melanie's first infusion was scheduled for November 2018. She tolerated the initial infusion well and showed up on time to her first several appointments. At our initial 3-month follow-up appointment, Melanie told me she was feeling better and had returned to the cheer team. A physical exam confirmed the improvement, with resolved swelling and tenderness in most of her joints.

Unfortunately, the good times didn't last long. Melanie started missing appointments, so we scheduled a teleconference with her, as well as her mom and dad. Melanie's parents explained they were having a hard time getting all the kids where they needed to be (remember that there were two other children), but they said they understood that it was important for Melanie to not miss her infusions. We again explained to them what would happen to Melanie in the long term if she continued to miss medication doses. Over the summer, Melanie got back onto her regular appointment schedule and even began telling me about her plans for her senior year of high school and her college aspirations.

A few weeks later, it was back again on the roller coaster. Melanie was a no-show for her next appointment, and the next time we met, she was accompanied by both her parents, a dead giveaway that something was up. They told our team that the burden of the infusion schedule was simply too great and asked if Melanie could transition to tocilizumab injections. With her spotty record of injection adherence, we were a bit apprehensive, but there wasn't much we could do, so we agreed with the change.

One month later, Melanie and her parents were back again. She had stopped the injections and had discontinued use of methotrexate and naproxen. She had even begun to wean herself off of prednisone. When asked to rate her pain level, Melanie said it was a 4/10 (it had been a 2/10 at her previous visit). "Why did you decide to stop with your medications?" we asked. Melanie and her parents got into a rather heated discussion that morning after she admitted to them that she had independently decided to stop her treatment. Clearly, they were as frustrated as we were. This was a patient we had agreed to bring into our practice as a teenager with the hopes of getting her onto the right track. Instead, things were just getting bumpier and bumpier. Melanie and her family again left our office agreeing to restart the tocilizumab injections, but frankly, we had our doubts.

The next time we saw Melanie in the office was 4 months later, a month before high school graduation. It was obvious from the way she walked into the exam room that things were not going well. A physical exam showed severe loss of range of motion in several extremities. Melanie rated her pain as a 10/10. Not surprisingly, she told us she never started back on the tocilizumab injections after her last visit. She had only come back to our office because her mom insisted that our practice do something about the swelling in Melanie's left knee. We reluctantly agreed to an intra-articular injection of triamcinolone acetonide.

At this visit, we again tried to reiterate the importance of adherence to the biologic regimen we had agreed upon. And again, Melanie and her mother both said that they understood the potential repercussions and expressed concern about Melanie's potential future disabilities.

Unfortunately, but not surprisingly, we haven't seen Melanie in the six months since that last visit. During the COVID-19 pandemic, we had scheduled two telehealth visits to gauge how she was doing. She didn't show up to either one. Repeated phone calls, voicemails, and text messages both to Melanie and her parents have gone unreturned. Has she found a new provider? Is she simply "done" with trying to treat her disease? Is something else going on? We have no way to know.

Not every patient who walks into the rheumatology clinic is an "easy win." We all have those patients who make us want to bang out heads against the wall as they promise us one thing and do the complete opposite. Yet it is our responsibility to remain professional and keep trying regardless of the boulders our patients drag in front of us.

I don't know what the future holds regarding Melanie's relationship with our practice and how she will manage her disease. She should be in college by now, but with the pandemic, everything has been thrown out of whack, so we don't know for sure. We've hopefully learned a few lessons from our interactions with her—establishing short- and long-term goals for treatment at the initial visit and having independent discussions with the parents and patient to understand the family dynamic and what potential barriers to treatment might be—that will help us to be better prepared in the future.

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The RNS is a nonprofit professional nursing and advanced practice provider (APP) organization representing registered nurses (RNs), nurse practitioners (NPs), physician assistants (PAs), and other healthcare professionals who are engaged in clinical practice, education, research, and advocacy for the care of adult and pediatric patients with rheumatic diseases.

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