APHON and ASPHO Position Paper on the Transition of Patients with Sickle Cell Disease from Pediatric to Adult Health Care

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ASSOCIATIONS' POSITION

Transition in health care involves the process of changing from a pediatric-focused to an adult-focused model of care, whether with or without transfer of the patient from a pediatric care provider to an adult care provider, and this is especially important for patients who have sickle cell disease (SCD). As the professional voices of pediatric hematology/oncology healthcare practice, the Association of Pediatric Hematology/Oncology Nurses (APHON) and the American Society of Pediatric Hematology/Oncology (ASPHO) recommend the following: that the process of transitioning patients with SCD from pediatric to adult care begins early and is presented as part of the natural process of becoming an adult; that patients, providers, and families are all involved in creating a transition plan and assessing transition preparedness annually; that the transfer of care involves direct communication between the pediatric team and the adult providers accepting the patient; and that adult services focus on helping young adults integrate into the adult model of care.

BACKGROUND AND SIGNIFICANCE

Sickle cell disease (SCD) refers to a group of inherited blood disorders that affect approximately 100,000 individuals in the United States and millions worldwide at any given time (Hassell, 2010; Sedrak & Kondamudi, 2020; Wastnedge et al., 2018). Complications of SCD result from hemolysis and vaso-occlusion, which cause acute and chronic multisystem complications that lead to significant morbidity and mortality (National Heart, Lung, and Blood Institute [NHLBI], 2014a). With improved medical interventions such as universal newborn screening, prophylactic antibiotics, targeted immunizations, and optimization of hydroxyurea (Lanzkron et al., 2013), childhood mortality has declined, and more than 90% of children with SCD in the United States now survive to adulthood (Quinn et al., 2010). Despite these advances, little improvement has been seen in outcomes for adults with SCD (Lanzkron et al., 2013).

The time of transition between pediatric and adult care is a high-risk period, with increased rates for utilization of acute care, reduced access to preventative maintenance, and risk of early mortality (Blinder et al., 2013; Hemker et al., 2011). Factors contributing to these poor outcomes include the lack of continuity of care, disease-specific comorbidities such as cognitive impairment due to cerebrovascular complications, and psychosocial vulnerabilities inherent in adolescent development (Crosby et al., 2015). Systems in which care is provided to adolescents and young adults (AYAs) with SCD must provide structured transition support for this vulnerable population.

This position paper addresses the transition of care from a pediatric-oriented healthcare system to an adult-oriented healthcare system, with or without a change in provider. Transition support goes beyond the coordination of transfer between different locations or levels of care; it encompasses support for the patient's self-management and assistance with reaching educational and vocational milestones. It is essential to realize the significant differences between pediatric-driven care and adult-driven care and their effect on overall medical management. According to Castillo and Kitsos (2017), "pediatric care is family-oriented and relies on significant parental involvement in decision making; however, adult care is patient-specific and requires autonomous, independent skills of patients without many interdisciplinary resources." The pediatric-driven care dynamic nonetheless plays a major role in how patients are able to face transition. A separate obstacle during transition is the heightened risk of the loss of insurance coverage, which can contribute to worse health outcomes (Sawicki et al., 2017).

Adolescents and young adults with SCD may face additional barriers during transition because of the disease's cerebrovascular complications, which can include stroke, silent cerebral infarctions, and cognitive impairment (Strouse, 2016). In some cases, patients with developmental delay may require decision-making support from a third party through guardianship or a power of attorney (White et al., 2018). It is incumbent upon the pediatric team to introduce a dialogue about transition early and identify and address any barriers before the transfer of care occurs. This dialogue should include encouraging the patient to self-advocate and highlighting the need to practice autonomy in care practices. Providers should aim to equip patients with realistic expectations about the level of support that they are likely to find in the adult healthcare setting.

SUMMARY OF THE PROBLEM

Health care transition is defined as a "purposeful, planned" process with a goal of maximizing "lifelong functioning and potential through the provision of high quality, developmentally appropriate healthcare services that continue uninterrupted as the individual moves from adolescence to adulthood" (White et al., 2018). Although a transfer from pediatric to adult providers is often a component of transition, the process must start well before the transfer and continue until the young adult is well established in an adult-focused system of care (DeBaun & Telfair, 2012; Saulsberry et al., 2019). Early preparation for the transition helps reduce gaps in care by ensuring that evidence-based routine healthcare maintenance continues uninterrupted.

Despite increasing recognition of the importance of transition as part of the continuum of health care for AYAs with SCD, established, effective strategies are scarce. Barriers to providing support during a patient's transition include limited staff training, a lack of identified personnel responsible for transition, and financial limitations (Lebensburger et al., 2012; Sobota et al., 2011). Professional organizations such as APHON and ASPHO, which represent providers caring for these AYA patients, should advocate for better reimbursement for transition planning and services than what is currently in place (McManus et al., 2021).

The limited availability of adult providers with expertise in SCD is a significant barrier to a smooth transition (Lebensburger et al., 2015; Sobota et al., 2011). Pediatric providers should identify specialists or interested generalists in their area who are willing to collaborate on care (Kanter et al., 2020). Providers of adult primary care can use resources such as the NHLBI's evidence-based guidelines or take advantage of regional learning consortiums to remain up-to-date on current recommendations for treating SCD (NHLBI, 2014b).

RECOMMENDATIONS

It is the position of APHON and ASPHO that programs caring for AYAs with SCD must provide transition preparation focused on helping patients become independent in managing their disease and integrating into an adult-focused model of care. Got Transition, a program of the National Alliance to Advance Adolescent Health, proposes using six core elements of transition as a framework to guide best practices: developing a healthcare transition policy, identifying transition-aged youth and maintaining a transition registry, assessing and tracking transition readiness, addressing healthcare transition needs with the youth and family, ensuring direct communication between pediatric and adult providers during any transfer of care, and continuing contact with the young adult to ensure successful completion of the transition (Got Transition, 2020). With financial limitations sometimes posing barriers to transition support, Got Transition and the American Academy of Pediatrics (AAP) developed a transition payment tip sheet that is updated annually and assists providers with achieving greater reimbursement for improved transition planning and utility (McManus et al., 2021).

1. Transition Policy

- The practice or institution should reference the annually updated transition payment tip sheet to support the delivery of recommended transition services (McManus et al., 2021).
- Formal discussion about transition should begin at age 12 (or when the patient is developmentally ready) and should include both the patient and the patient's family (Porter et al., 2014).
- The practice or institution should have a clear transition policy that is accessible to providers, patients, and families (e.g., posted on the organization's website, posted in exam rooms, or mailed to families).
- Whether or not a transfer of care will occur, the discussions about transition should include details about changes in privacy when the patient turns 18 and any shift to an adult model of care, which requires much greater autonomy than pediatric care (White et al., 2018).
- The transition planning team should be multidisciplinary and should include physicians, advanced practice providers, nurses, psychologists, and social workers from pediatric and adult care settings (Jordan et al., 2013).
- Pediatric providers should allow patients and caregivers to express their feelings and concerns about transition (e.g., fears about transferring to a new provider) and should include family members in the transition plan.

2. Tracking and Monitoring

- By age 14, every patient should have an individualized and documented transition plan that is updated annually (AAP et al., 2011, 2015).
- Each institution should have a way to identify and track transition-aged patients and ensure that planning has been done and documented. Integration into the electronic medical record can help improve transition planning (Sharma et al., 2018).

3. Transition Readiness

- Patients should have an assessment of their readiness for transition performed regularly, using either a generic or an SCD-specific tool (American Society of Hematology, 2021; Treadwell et al., 2015). Use of these tools can guide the patient's specific transition planning and determine the appropriate timing of the transfer of care.
- Patients should have a neurocognitive assessment to identify deficits that may affect their transition readiness (Daly et al., 2011; Saulsberry-Abate et al., 2021).

4. Transition Planning

- Consensus statements recommend that transition preparation focus on improved patient-provider communication and increased patient responsibility for self-management of the disease (AAP et al., 2011, 2015).
- Transition preparation should be addressed annually and should include medical topics (e.g., SCD complications, disease inheritance) as well as educational and vocational topics.
- Pediatric providers should directly address insurance issues and any insurance coverage changes that will accompany an increase in the patient's age.
- Nursing staff members can facilitate transition by assessing preparedness, resources, relationships, and responsibilities as part of the individual transition plan (Fegran et al., 2014).
- Providers caring for patients with SCD can normalize the transition process by helping families understand that transition to an adult-focused system of care is a normal part of development (Mahan et al., 2017).
- Medical professionals should initiate an "adult" model of care during the adolescent period by seeing patients alone for part of the visit, directing questions to the adolescent during the visits, and (as the patient nears age 18) encouraging patients to call for appointments and arrange their own medication refills.

5. Transfer of Care

- Pediatric care providers should identify local adult care providers who have expertise or an interest in SCD.
- If the transition includes a transfer of care, it is helpful to have the family meet the adult care provider prior to the transfer. Taking this step may improve the patient's access and adherence to adult care (Bloom et al., 2012; Hankins et al., 2012).
- The pediatric care providers should consider staggering the transfer so that patients are not changing primary care and subspecialty providers at the same time.
- The transfer should occur when the patient, family, and providers agree that the patient is ready to take on an adult role in their care. If the hospital has set a specific age for transfer, a policy exception should be developed for patients with cognitive delay or other special needs.
- Transfers should be planned; making an abrupt transfer (e.g., during an unplanned hospital admission or a pregnancy) should be avoided.
- Pediatric care providers should communicate directly with the adult care provider and send the new provider a written medical summary that includes the patient's medical history, baseline laboratory test results, and an individualized pain management plan.

6. Transfer Completion

- A transfer is not complete until the patient has been seen at least once in the adult care provider's office, which should happen within 3 months of the patient's leaving pediatric care.
- Pediatric care providers should contact the AYA to confirm attendance at the first adult visit and follow up with the adult practice to confirm completion of the transfer (AAP et al., 2011, 2015). Consultation can be provided as needed.
- Pediatric and adult care teams should continue to evaluate and adjust their transition process. This evaluation should include the perspective of patients, families, and all involved providers.

CONCLUSION

All AYA patients with SCD should receive transition preparation to help them maneuver from pediatric- to adult-focused health care. This preparation allows for greater independence and the development of self-management skills regardless of whether a transfer to a new provider is involved. Transition education before, during, and after the transfer of care may reduce morbidity and the high rate of mortality from SCD complications. Transition readiness should be assessed regularly because the move from pediatric- to adult-focused care occurs over time rather than in a single event. Pediatric and adult care teams should work together and continuously evaluate their transition processes.

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