

Because life expectancy has increased greatly in the past few decades for individuals living with sickle cell disease (SCD), transition to the adult healthcare setting has become a necessity to continue disease management. Transition for young adults with SCD is associated with declining health outcomes, including increased acute care use and mortality. Nurses can assist young adults with SCD who are at risk after transition by assessing the young adult's ability to carry out disease self-management, facilitating the supportive role of the family, and recognizing young adults who may have difficulty accessing healthcare resources and providers.

#### AT A GLANCE

- Transition to adult health care for young adults with SCD poses many risks and burdens.
- Not all young adults with SCD are adequately prepared to care for their disease and independently engage with healthcare providers.
- To address barriers, nurses can assess the young adult's readiness to independently perform disease self-management and access healthcare resources.

#### KEYWORDS

transition to adult care; young adult; continuity of care; self-management

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# Young Adults With Sickle Cell Disease

## Challenges with transition to adult health care

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Sickle cell disease (SCD) is an autosomal recessive disorder that causes individuals to produce abnormal hemoglobin that can become sticky and crescent-shaped (National Heart, Lung, and Blood Institute [NHLBI], 2018). As a result, vaso-occlusion is common as the red blood cells stick to each other and blood vessel walls, leading to blocked blood flow, ischemia, and mild to severe pain (NHLBI, 2018). Over a lifetime, organ damage associated with vaso-occlusion can occur in the brain, lungs, kidneys, heart, skin, penis, and joints (NHLBI, 2018). Severe, life-threatening complications of SCD include splenic sequestration crises, infections, acute chest syndrome, aplastic crises, and strokes (NHLBI, 2018). Treatment for SCD is focused on reducing and addressing SCD complications as they develop (NHLBI, 2018).

### Transition to Adult Health Care for Young Adults With SCD Necessity

Routine screening for SCD and advances in medical treatment have rapidly increased the life expectancy of those diagnosed with SCD from 14 years in 1973 to 40–60 years (Elmariah et al., 2014; Gardner et al., 2016). With more than 93% of children living with SCD reaching adulthood, an effective transition to adult health care provides continuity between pediatric and adult healthcare settings (Quinn et al., 2010). Transition processes ensure that young adults with SCD continue to receive treatment to manage

SCD and receive preventive care to reduce long-term complications (Bryant et al., 2015). However, in the United States, young adults with SCD transitioning to the adult healthcare system face numerous barriers, and this time period is associated with increased morbidity and mortality (Blinder et al., 2013, 2015; Brousseau et al., 2010; McClish et al., 2017).

### Challenges

Challenges faced by young adults with SCD range from simple logistic issues (e.g., finding providers) to more complex challenges (e.g., lacking preparation to independently manage their SCD, discrimination). As individuals with SCD age, there are increasing rates of acute pain crises and healthcare use (Brousseau et al., 2010; Cronin et al., 2019; Pope et al., 2016). It is critical that young adults with SCD successfully establish a trusting relationship with an adult healthcare provider with the expertise to manage the long-term complications of SCD in adulthood. Even so, trouble identifying adult hematologists with SCD expertise is commonly reported by healthcare providers and individuals with SCD (Lunyera et al., 2017; Travis et al., 2019). In the United States, there is a lack of hematologists with SCD expertise, forcing young adults with SCD to turn to the emergency department for crisis management, leading to missed opportunities to prevent or mitigate severe SCD complications (Blinder et al., 2015; Lanzkron et al., 2018).