AYA SCD Intervention Article Summaries

Abel, R. A., Cho, E., Chadwick-Mansker, K. R., D'Souza, N., Housten, A. J., & King, A. A. (2015). Transition needs of adolescents with sickle cell disease. *The American Journal of Occupational Therapy, 69*(2), 1–5. https://doi.org/10.5014/ajot.2015.013730

Adolescents (N = 122) ages 13 to 21 years with Sβ thalassemia disease, sickle cell disease, sickle cell anemia, or another subtype completed a single arm, retrospective study about independent living skills for transition to adulthood. Participants self-reported their perceived ability to perform everyday tasks by rating activities as "can do already" or "needs practice" across several categories. Adolescents needed the most practice in living arrangements, money management, vocational skills, and healthcare skills, with needs decreasing with age. Adolescents who had overt strokes needed more practice in housekeeping, personal, and healthcare skills than adolescents without cerebrovascular injury. Due to the self-report method of this study, adolescents may have underestimated their needs; including parent reports in future studies could indicate adolescent needs more accurately. Programs to help adolescents with sickle cell disease develop independent living skills, especially self-advocacy and vocational skills, are critical to successful transition to adulthood.

Barakat, L. P., Schwartz, L. A., Salamon, K. S., & Radcliffe, J. (2010). A family-based randomized controlled trial of pain intervention for adolescents with sickle cell disease. *Journal of Pediatric Hematology/Oncology, 32*(7), 540–547. https://doi.org/10.1097/MPH.0b013e3181e793f9

In an RCT, adolescents (N = 53) ages 12 to 18 years with SCD-SS or Sβ-thalassemia and their caregivers completed a family-based pain CBT or a disease education intervention. The pain CBT intervention was not more effective than the disease education intervention in improving pain and health-related outcomes (e.g. percentage of days with pain, health service use, pain-related hindrance of goals) or psychosocial functioning (e.g. disease self-efficacy). However, small effect sizes indicate that pain CBT has some effect on percentage of days with pain, health services use, and disease knowledge. Recruitment and retention are challenging in African American adolescents and families, in part due to mistrust of research and lack of perceived benefits. Future work should investigate decision making surrounding enrollment in clinical trials as well as perceived barriers and benefits to participating in interventions.

Chen, E., Cole, S. W., & Kato, P. M. (2004). A review of empirically supported psychosocial interventions for pain and adherence outcomes in sickle cell disease. *Journal of Pediatric Psychology, 29*(3), 197-209. https://doi.org/10.1093/jpepsy/jsh021

A review of existing interventions indicated that CBT interventions are likely the most efficacious on pain and adherence behavior for individuals with SCD. While CBT interventions appear to be more empirically supported than behavioral change or social support interventions, this review is limited because many of the studies reviewed were qualitative. Many studies also lacked control groups or did not measure consistent outcomes variables. Future work should comprehensively evaluate existing interventions as well as investigate the effects of interventions on outcome variables, such as daily pain management practices (e.g. drinking fluids, resting), pain outcomes over time (e.g. severity of pain, ER visits), and psychosocial functioning (e.g. coping skills).

Clay, O. J., & Telfair, J. (2007). Evaluation of a disease-specific self-efficacy instrument in adolescents with sickle cell disease and its relationship to adjustment. *Child Neuropsychology, 13*(2), 188–203. https://doi.org/10.1080/09297040600770746

The Sickle Cell Self-Efficacy Scale (SCSES) is a nine-item scale that measures how much an adolescent or adult with SCD believes they have control over their daily lives despite having SCD. Adolescents (N = 131) ages 11 to 19 years with HbSS, Hb SC, Hb Sβ thalassemia, or another subtype completed the SCSES measures of physical and psychological symptoms and a measure of personal health care (e.g. drinking fluids, understanding medical instructions). Greater self-efficacy was associated with fewer physical and psychological symptoms. Greater self-efficacy was also associated with drinking fluids, but not other personal health care activities. The SCSES is clinically useful because it is brief and could be used to predict adherence and effectiveness of interventions. Longitudinal studies need to determine if increased self-efficacy improves adjustment to SCD.

Collins, M. H., Loundy, M. R., Brown, F. L., Hollins, L. D., Aldridge, Y., Eckman, J., & Kaslow, N. J. (1997). Applicability of criteria for empirically validated treatments to family interventions for pediatric sickle cell disease. *Journal of Developmental and Physical Disabilities, 9*(4), 293–309. https://doi.org/10.1023/A:1024973811042

Developing and implementing an SCD family intervention for use in low SES African American populations requires balancing methodological rigor with cultural sensitivity and flexibility with individual families. Several factors were considered in evaluating the efficacy of a manualized, family-oriented psychoeducational intervention conducted with patients ages 6 to 18 years (N = 34). Using a treatment manual for a fixed duration intervention allowed more providers to be trained in administering the intervention and was more attractive to families than an unstandardized, open-ended treatment. Random assignment to intervention and control conditions was methodologically optimal, but not always clinically optimal as families assigned to the control condition were concerned that they did not receive the intervention. Collecting follow-up data would have been useful in determining long term effects of interventions; however, collecting this data was difficult in low SES families.

Crosby, L. E., Joffe, N. E., Peugh, J., Ware, R. E., & Britto, M. T. (2017). Pilot of the chronic disease self-management program for adolescents and young adults with sickle cell disease. *Journal of Adolescent Health, 60*(1), 120–123. https://doi.org/10.1016/j.jadohealth.2016.08.022

Adolescents (N = 22) ages 16 to 24 years completed a pilot group intervention focused on understanding sickle cell disease, managing symptoms, and learning action planning and problem-solving skills. Chronic disease self-efficacy, SCD-specific self-efficacy, and self-management behavior (e.g. managing medications, talking with providers) were measured before, immediately after, three months after, and six months after the program. Chronic disease self-efficacy improved following the intervention, but SCD-specific self-efficacy and self-management behavior did not. Although participants had high satisfaction with the program, only 64% of participants completed the study, highlighting the need for shorter or web-based interventions. Future work should also investigate long term effects of group intervention programs.

Crosby, L. E., Quinn, C. T., & Kalinyak, K. A. (2015). A biopsychosocial model for the management of patients with sickle-cell disease transitioning to adult medical care. *Advances in Therapy, 32*(4), 293–305. https://doi.org/10.1007/s12325-015-0197-1

Multidisciplinary, biopsychosocial transition models are critical for adolescents with SCD who are transitioning from pediatric care to adult care. In reviewing factors contributing to transition, three factors emerge: biological factors (chronic pain, organ damage, neurocognitive effects of brain injury), psychosocial factors (lack of readiness to transition, managing chronic pain), and sociological factors of (changing insurance coverage, schooling/employment challenges). Adult SCD care has fewer multidisciplinary services compared to pediatric SCD care, particularly because adult care often falls on general internists who do not have SCD expertise. To facilitate transition to adult care that improve health outcomes, transition teams should include pediatric and adult specialists as well as use flexible and patient focused timings for transition. Biopsychosocial transition models for SCD should be modeled after cystic fibrosis and hemophilia transition programs and should be evaluated in comparison to traditional transition models.

Crosby, L. E., Ware, R. E., Goldstein, A., Walton, A., Joffe, N. E., Vogel, C., & Britto, M. T. (2016). Development and evaluation of iManage: A self-management app co-designed by adolescents with sickle cell disease. ***Pediatric Blood & Cancer, 64*****(1), 139–145. https://doi.org/10.1002/pbc.26177**

iManage is an app designed to improve self-management behaviors in AYAs with SCD. Adolescents (N = 70) ages 13 to 24 years with mostly subtype HbSS completed three phases to develop the app. First, they completed surveys regarding their internet usage, then interviews about perceived barriers to self-management, and finally participated in protype testing of iManage. Self-management barriers, including low disease self-efficacy and lack of peer support, informed the design of iManage such that the app contained features to track daily symptoms, visualize customizable self-management goals, and interact with peers. Adolescents rated iManage as easy to use, motivating, and beneficial. Future research should field test iManage to determine its efficacy in tracking health and self-management behavior.

Gil, K. M., Carson, J. W., Sedway, J. A., Porter, L. S., Schaeffer, J. J. W., & Orringer, E. (2000). Follow-up of coping skills training in adults with sickle cell disease: Analysis of daily pain and coping practice diaries. ***Health Psychology, 19*****(1), 85-90. http://doi.org/10.1037/0278-6133.19.1.85**

Adults (N = 67) with a mean age of 33.18 years were randomly assigned to complete an individual, cognitive-based pain coping skills intervention or a disease education control intervention. The cognitive pain coping intervention included relaxation and distraction exercises. At a three-month follow-up, participants in the coping skills condition had lower pain as measured in the lab and reported higher coping attempts compared to disease education condition participants. Analyses of daily pain diaries also indicated that patients in the coping skills condition used coping skills more often on high pain days. However, at three months, neither the coping skills intervention nor the disease education intervention affected pain severity, medication use, or healthcare contacts. Future work should investigate interventions that promote daily practice of coping skills.

Gil, K. M., Wilson, J. J., Edens, J. L., Workman, E., Ready, J., Sedway, J., … Daeschner, C. W. (1997). Cognitive coping skills training in children with sickle cell disease pain. ***International Journal of Behavioral Medicine, 4*****(4), 364-377. https://doi.org/10.1207/s15327558ijbm0404_7**

Children and adolescents (N = 49, mean age = 11.9 years) with HbSS, HbSC, and HbSβ thalassemia were randomly assigned to a brief cognitive coping skills intervention or to a standard care control. Participants who learned cognitive coping skills (e.g. relaxation, pleasant imagery,

calming self-statements) reported less pain when measured with a lab pain stimulation. On a coping strategies questionnaire that measured coping attempts, negative thinking, and illness focused strategies, participants in the coping skills condition indicated decreased negative thinking compared to the control. Future work should investigate the utility of teaching coping skills to parents so that they can serve as models for their children.

Hankins, J. S., Osarogiagbon, R., Adams-Graves, P., McHugh, L., Steele, V., Smeltzer, M. P., & Anderson, S. M. (2012). A transition pilot program for adolescents with sickle cell disease. ***Journal of Pediatric Health Care, 26*****(6), e45-e49. https://doi.org/10.1016/j.pedhc.2012.06.004**

Adolescents (N = 83) aged 17 to 19 years with SCD subtypes: HbSS, HbSβ^0 thalassemia, HbSC, HbSβ^+ thalassemia HbS/HPFH) were recruited to participate in a pilot transition program. In this transition program, adolescents and their parents toured an adult SCD program, discussed transitioning with pediatric staff, and scheduled a first appointment with the adult program. Adolescents who volunteered to complete the program (N = 43) rated the program as helpful and attended first appointments with adult hematologists at a higher rate than adolescents who did not participate in the program. Seamless, continued care from pediatric to adult settings is essential for AYAs with SCD, indicating that future work to determine the efficacy of transition programs on behavior and medical outcomes (e.g. ER visits, self-management) is imperative.

Kaslow, N. J., & Brown, F. (1995). Culturally sensitive family interventions for chronically ill youth: Sickle cell disease as an example. ***Family Systems Medicine, 13*****(2), 201–213. https://doi.org/10.1037/h0089393**

Strong extended family network, including nonblood relatives, as well as low socioeconomic (SES) and desire for a sense of empowerment characterize many African American families and are important factors to consider in the development of interventions for sickle cell disease. In a six-session intervention, SCD patients, their primary caregiver, and sometimes additional family members participated in sessions on disease education, problem solving and disease management skills, pain and stress management skills, communication and expressing feelings, and family relationships. Although this program was not empirically tested for efficacy, it emphasizes the importance of awareness of and sensitivity to the cultural and socioeconomic background of the family in providing therapeutic interventions

Kaslow, Nadine J, Collins, M. H., Loundy, M. R., Brown, F., Hollins, L. D., & Eckman, J. (1997). Empirically Validated Family Interventions for Pediatric Psychology: Sickle Cell Disease as an Exemplar. ***Journal of Pediatric Psychology, 22*****(2), 213–227. Retrieved from https://academic.oup.com/jpepsy/article-abstract/22/2/213/944638**

Designing manualized family-based interventions for chronically ill children presents several challenges. In the context of a family psychoeducation intervention for N = 28 families with children ages 6-18 with SCD compared to a treatment as usual control, families randomly assigned to the intervention condition improved on disease knowledge (measured quantitatively) and family functioning (measured qualitatively). Cultural sensitivity, specifically attention to the African American family structure and economic hardships of poor families, as well as establishing trust between the therapist and the family, is critical for SCD family-based interventions. Balancing methodological rigor (e.g. random assignment to intervention and control groups, selection and operationalization of outcome measures) and good clinical practice (e.g. benefits to family

functioning hat may not be captured through quantitative measures only) is key challenge in developing and implementing these interventions.

Kaslow, N. J., Collins, M. H., Rashid, F. L., Baskin, M. L., Griffith, J. R., Hollins, L., & Eckman, J. E. (2000). The efficacy of a pilot family psychoeducational intervention for pediatric sickle cell disease (SCD). *Families, Systems, & Health, 18*(4), 381-404. http://dx.doi.org/10.1037/h0091863

Children (N = 39) aged 7 to 16 years with HbSS, HbSC, Sβ thalassemia and their caregivers were randomly assigned to a family psychoeducational intervention or to a treatment as usual control. In the intervention condition, African American therapists taught families about SCD, coping with complications and pain, and fostering positive interpersonal relationships with family. Outcome measures, including a test of disease knowledge, psychological functioning (e.g. depression, internalizing and externalizing behaviors), and family cohesion, were assessed pre-, post-, and six months after the intervention. Relative to the controls, children and caregivers in the intervention indicated increased disease knowledge post-intervention but did not differ from controls in any other outcomes. Children also maintained increased disease knowledge six months after the intervention. Many families dropped out of the study as it progresses, indicating that future work should develop and evaluate strategies to provide family psychoeducation to treatment resistant families.

Mulchan, S. S., Valenzuela, J. M., Crosby, L. E., & Diaz Pow Sang, C. (2016). Applicability of the SMART model of transition readiness for sickle-cell disease. *Journal of Pediatric Psychology, 41*(5), 543–554. https://doi.org/10.1093/jpepsy/jsv120

In a qualitative study, AYAs (N = 14, ages 14-24 years, HbSS, HbSC, Sβ thalassemia) and clinical experts (N = 10) completed semi-structured interviews and questionnaires to identify the factors that influence transition readiness. AYAs and clinical experts discussed objective (not easily modified) factors, such as neurocognitive impairments that may be undetected and societal stigma/lack of awareness about SCD, especially stigma in relation to racial bias. They also discussed modifiable factors including pain management skills, communication about pain to providers, and health care navigation skills (e.g. acquiring adequate insurance). Future work can use the factors identified in this study to inform transition-readiness interventions and measures related to both individual AYAs' skills and socio-ecological factors (e.g. provider racial bias).

Porter, J. S., Wesley, K. M., Zhao, M. S., Rupff, R. J., & Hankins, J. S. (2017). Pediatric to adult care transition: perspectives of young adults with sickle cell disease. *Journal of Pediatric Psychology, 42*(9), 1016–1027. https://doi.org/10.1093/jpepsy/jsx088

In a qualitative study, young adults (N = 19) ages 18 to 30 years participated in focus groups to discuss factors that influence transition. Factors discussed were mostly consistent with factors in the SMART (Social-ecological Model of Adolescent and Young Adult Readiness to Transition) model. Young adults indicated that negative medical experiences, such as pain not being taken seriously by ER providers, relationships with adult providers, and taking responsibility for self-management and self-advocacy skills affected their transitions. They also rated finding an adult provider, knowing when to seek emergency care, and understanding the impacts of healthy (e.g. medication adherence) and unhealthy (e.g. smoking) behaviors as important to transitioning. Transition interventions should focus on teaching and assessing transition skills and include peer support from adults who have successfully transitioned to adult healthcare.

Sansom-Daly, U. M., Peate, M., Wakefield, C. E., Bryant, R. A., & Cohn, R. J. (2012). A systematic review of psychological interventions for adolescents and young adults living with chronic illness. *Health Psychology, 31*(3), 380–393. https://doi.org/10.1037/a0025977

A systematic review evaluated 25 psychological intervention studies that targeted AYAs with diabetes, cancer, juvenile idiopathic arthritis, sickle cell disease, or asthma. Skill-based interventions, particularly communication skills and interventions with practical components (e.g. homework assignments, role playing), had more positive outcomes relative to peer support and educational interventions. However, many interventions displayed inconsistencies between the reported goals of the intervention and outcome measures (e.g. educational intervention lacking a disease knowledge measure). Future interventions should be developed and tested with methodological rigor to address disease-specific adjustment.

Schwartz, L. A., Tuchman, L. K., Hobbie, W. L., & Ginsberg, J. P. (2011). A social-ecological model of readiness for transition to adult-oriented care for adolescents and young adults with chronic health conditions. *Child: Care, Health and Development, 37*(6), 883–895. https://doi.org/10.1111/j.1365-2214.2011.01282.x

Evidenced based interventions that are informed by theory are necessary to support AYAs' successful transition from pediatric care to adult care. Existing transition literature focuses on patient age, disease knowledge, and disease skills as markers for transition readiness. A socio-ecological model of AYA transition readiness, the SMART model, was developed to extend beyond these variables to broader factors, including socio-cultural variables, factors related to the interconnections between patients, parents, and providers, and emphasis on variables that can modified through intervention. In pilot testing, the SMART model of transition readiness encompassed the factors that childhood cancer survivorship providers used in making transition decisions. Through this novel model, perspectives of multiple stakeholders (patients, parents, and providers) can inform transition interventions.

Schwartz, Lisa A., Brumley, L. D., Tuchman, L. K., Barakat, L. P., Hobbie, W. L., Ginsberg, J. P., … Deatrick, J. A. (2013). Stakeholder validation of a model of readiness for transition to adult care. *JAMA Pediatrics, 167*(10), 939–946. https://doi.org/10.1001/jamapediatrics.2013.2223

The SMART (Social-ecological Model of Adolescent and Young Adult Readiness to Transition) model was validated by three key groups of stakeholders: patients, parents, and providers. The SMART model includes preexisting factors (e.g. medical status, access to insurance) and modifiable variables (e.g. knowledge, self-efficacy, relationships) that affect transition readiness. AYA survivors of childhood cancer (N = 14) ages 16 to 28 years with various diagnoses, parents of these patients, and providers participated in focus groups and then rated the importance of each component of the SMART model. These three stakeholder groups rated all components of SMART as important for successful transition from pediatric to adult care. Relationships/communication (e.g. family cohesion, patient-provider communication) was a particularly highly rated, contradicting the emphasis on knowledge in past transition literature. Transition is a multifaceted, dynamic process involving several psychosocial variables, indicating the need for multidisciplinary care teams. Future research should investigate the SMART model with larger samples to further demonstrate its validity in informing transition interventions and assessments.

Schwartz, L.A., Daniel, L. C., Brumley, L. D., Barakat, L. P., Wesley, K. M., & Tuchman, L. K. (2014). Measures of readiness to transition to adult health care for youth with chronic

physical health conditions: A systematic review and recommendations for measurement testing and development. *Journal of Pediatric Psychology, 39*(6), 588–601. https://doi.org/10.1093/jpepsy/jsu028

A systematic review examined four general and six disease specific transition readiness measures. General measures included assessments of disease knowledge, self-management and self-advocacy skills. Disease-specific measures assessed these factors, and some measured factors related to adult providers, psychosocial adjustment, and emotions about transitioning. Although these measures are a valuable starting point for development and validation of transition readiness measures, most were designed as clinical tools, precluding methodologically rigorous evaluation and precise definitions of transition-readiness. Recommendations include using multiple outcome variables in evaluating transition-readiness measures, strong theoretical frameworks including perspectives of multiple stakeholders, and using transition readiness measures to improve outcomes of transition readiness over time.

Thomas, V. J., Dixon, A. L., & Milligan, P. (1999). Cognitive-behaviour therapy for the management of sickle cell disease pain: An evaluation of a community-based intervention. *British Journal of Health Psychology, 4*(3), 209–229. https://doi.org/10.1348/135910799168588

British adolescents and young adults (N = 59) ages 15 to 35 years with HbSS were randomly assigned to complete a CBT group therapy for sickle cell disease pain, a group attention placebo therapy (i.e. a peer support group to discuss feelings about SCD), or no intervention. Outcome measures, including positive and negative pain-related cognitions, self-efficacy, reliance on powerful others (e.g. doctors), and affective distress showed greater improvement following the CBT intervention compared to the attention placebo and the control conditions. Although this CBT intervention appears to improve SCD pain, further research is necessary to determine health outcomes such as hospital admissions and long-term pain mangement.

Treadwell, M., Telfair, J., Gibson, R. W., Johnson, S., & Osunkwo, I. (2011). Transition from pediatric to adult care in sickle cell disease: Establishing evidence-based practice and directions for research. *American Journal of Hematology, 86*(1), 116–120. https://doi.org/10.1002/ajh.21880

Studying transition requires conceptual models that are translated into practice using evidence-based interventions that are assessed using consistent measures. Treadwell, Telfair, Gibson, Johnson, and Osunkwno (2011) discuss systemic and patient factors involved in transition as well as the components of model programs. Systemic issues include limited access to specialized adult providers and limited communication between pediatric and adult providers, while patient issues include lack of independent living skills and fearful expectations of the transition process. Model transition programs should emphasize disease knowledge, self-management and independent living skills, self-advocacy, school and employment plans, include support from adults with SCD who have successfully transitioned, and facilitate communication between patients, pediatric, and adult providers.

Waldboth, V., Patch, C., Mahrer-Imhof, R., & Metcalfe, A. (2016). Living a normal life in an extraordinary way: A systematic review investigating experiences of families of young people's transition into adulthood when affected by a genetic and chronic childhood condition. *International Journal of Nursing Studies, 62*, 44-59. https://doi.org/10.1016/j.ijnurstu.2016.07.007

A systematic literature review explored the experiences of young adults with chronic, genetic diseases, including neuromuscular diseases, cystic fibrosis, and sickle cell disease who were transitioning to adult care. Across the studies reviewed, young people had a desire to feel normal; they wanted to cultivate friendships, maintain educational and employment goals, and become more independent from their families. They experienced ambivalence in establishing a personal identity separate from their families and did not feel prepared to transition. Parents experienced psychosocial difficulty adapting to their changing roles and expressed high concern for their child's future health and disease management skills. Future research should comprehensively investigate psychosocial experiences of parents and siblings of chronically ill children to better help families adapt and cope.

Wojciechowski, E. A., Hurtig, A., & Dorn, L. (2002). A natural history study of adolescents and young adults with sickle cell disease as they transfer to adult care: A need for case management services. *Journal of Pediatric Nursing, 17*(1), 18-27. https://doi.org/10.1053/jpdn.2002.30930

In a non-experimental, descriptive research study, AYAs (N = 18, ages 18 to 24 years, HbSS, HbSC, Sβ thalassemia, SG) and their providers completed interviews on transition preparation, compliance, and self-efficacy. Results indicated that most patients received little to no preparation before transferring to an adult provider and that transfer was based on age or on pregnancy (for female patients). Although patients who kept their adult care appointments had higher self-efficacy, transition preparation was not related to health-behavior compliance (e.g. drinking enough fluids). Future work should investigate patients who do not follow-up with adult providers after leaving pediatric care. Nurse care managers play an important role in successful transition, particularly through assisting patients in finding resources to develop educational/vocational goals.