Transition to Adult Health Care ACT Sheet

Transition is an ongoing process that does not end with transfer of care. The goal of transition of adolescents with chronic medical conditions is to provide uninterrupted, comprehensive, culturally sensitive, coordinated, and developmentally appropriate healthcare. The transition team includes at least the patient and family, and the pediatric, adult PCP, and specialty care providers. ***For the general principles of transition, refer to the 2011 AAP/AAFP/ACP transition clinical report, which includes the recommendation that transition planning begin no later than age 12 and includes a patient readiness assessment.***

Sickle Cell Disease

Condition Description: Sickle cell disease (SCD) encompasses a group of autosomal recessive genetic disorders including sickle cell anemia (Hb SS), sickle hemoglobin C disease (Hb SC), hemoglobin S beta plus thalassemia (Hb Sβ+thal), S beta zero thalassemia (Hb Sβ0thal), and other sickling disorders involving hemoglobin S and other interacting hemoglobin variants. The sickle cell disease is characterized by hemolysis and vaso-occlusion due to abnormal deformability of red blood cells.

Clinical Considerations: As patients age, cumulative vascular damage affecting the lungs and kidneys makes pulmonary and renal insufficiency common problems. Pain episodes cause life-long morbidity, and many individuals develop chronic pain from cumulative bone, joint, and nerve damage. Ongoing communication with the specialist is essential to the management of these potential complications of sickle cell disease. Clinical concerns in adults include:

- Acute vaso-occlusive episodes (managed primarily with hydration and opioids)
- Chronic pain
- Hemolytic anemia, with episodic acute severe anemia
- Increased susceptibility to severe infections
- Chronic organ damage and failure: kidney, lungs, heart
- Increased risk of strokes and neurocognitive deficits in adolescents and adults
- Retinopathy
- Priapism
- Avascular necrosis (osteonecrosis) of hips and shoulders
- Puberty, sexual function and fertility - generally no special considerations. Pregnancies should be considered high risk.

THE TRANSITION TEAM SHOULD TAKE THE FOLLOWING ACTIONS:

- Initiate a dialogue among transition team members and establish an adult medical home.
- Facilitate consistency and coordination of care among multiple health care providers as the patient transitions to independent living (to include college, relocation, employment).
- Consult with the hemoglobin disorder specialist who cares for the patient to establish a co-management plan, including input from the patient/family.
- Confirm the diagnosis by review of medical records and laboratory studies.
- Emphasize major SCD-related events, usual pain pattern and treatment, and any end-organ damage in past history and physical examination, and order laboratory studies in consultation with hemoglobin disorder specialist.
- Develop and implement a comprehensive care plan that includes a pain management plan, preventive measures (immunizations – pneumococcal vaccine every 5 years; Hib, HBV and meningococcal vaccines, if not previously administered) and screening.
- Ensure patient has an acute illness protocol to be taken to the emergency room.
- Identify the patient's health care coverage (including insurance) and access to care.
- Assess and address the patient's psychosocial well-being and social service needs.
- Offer health education and genetic counseling concerning future reproductive decisions.

Additional Information:

AAP/AAFP/ACP Transition Clinical Report
Sickle Cell Information Center
Transition Guidelines

Referral (local, state, regional and national):

Testing
Clinical Services
Find Genetic Services

ACMG
American College of Medical Genetics and Genomics
Translating Genes Into Health®

LOCAL RESOURCES: Insert State program web site links

State Resource site *(insert program information)*

Name	
URL	
Comments	

APPENDIX: Resources with Full URL Addresses

Additional Information:

AAP/AAFP/ACP Transition Clinical Report
http://aappolicy.aappublications.org/cgi/reprint/pediatrics;128/1/182.pdf

Sickle Cell Information Center
http://www.scinfo.org/

Transition Guidelines
http://www.nhlbi.nih.gov/health/prof/blood/sickle/sc_mngt.pdf

Referral (local, state, regional and national):

Testing
http://www.ncbi.nlm.nih.gov/sites/GeneTests/lab/clinical_disease_id/2775?db=genetests&country=United%20States

http://www.rbclab.com

Clinical Services
http://www.genetests.org/

Find Genetic Services
http://www.acmg.net/gis

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