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.***

Congenital Adrenal Hyperplasia (CAH)

Condition Description: Congenital adrenal hyperplasia is a group of autosomal recessive genetic defects of steroidogenesis leading to ACTH-induced hyperplasia of the adrenal cortex. Greater than 90% are due to 21-hydroxylase deficiency. There are three main phenotypes: classical salt-wasting, classical simple virilizing and non-classical (late-onset).

Clinical Considerations: During periods of stress, both males and females are subject to adrenal crisis and glucocorticoid therapy may have to be increased. The condition may result in precocious puberty, short stature, masculinization in females, sexual dysfunction, infertility, and psychosocial difficulties. Ovarian cysts, anovulation, and irregular menses may be seen in females with CAH. Sexual functioning in females may be affected (due to anatomical and/or psychosocial issues) and may require appropriate consultation. Pregnancies in women with CAH should be considered high risk. Males should be monitored periodically for overgrowth of ectopic adrenal tissue in the testes (testicular adrenal rest tumors).

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 an endocrinologist experienced with CAH (ideally the pediatric endocrinologist who has been caring for the patient) to establish a co-management plan, including input from the patient/family. The plan should include:
  - Continued maintenance of treatment with glucocorticoids and mineralocorticoids
  - Appropriate response to acute illness, physical trauma/stress
  - For females, consultation to address gynecologic, psychologic, urologic, dermatologic, and reproductive issues
  - For males, consultation to address testicular adrenal rest tumors and fertility
  - Nutritional counseling in overweight individuals
- Review the medical record and previous laboratory studies.
- Follow laboratory studies as ordered by an endocrinologist, such as 17-hydroxyprogesterone (17-OHP), androstenedione (Δ4A), free and total testosterone (and plasma renin activity in salt-wasters).
- Identify the patient's health care coverage (including insurance) and access to care.
- Assess and address the patient's psychological, behavioral, and social service needs.
- Offer health education and genetic counseling concerning future reproductive decisions.

Additional Information:

AAP/AAFP/ACP Transition Clinical Report
American Association of Clinical Endocrinologists
Endocrine Society
Androgen Excess Society
Society of Reproductive Endocrinology and Infertility
American Urological Association
Got Transition

Referral (local, state, regional and national):

Clinical Services

Pediatric Endocrine Society "Find A Doc"
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
https://pediatrics.aappublications.org/content/pediatrics/early/2011/06/23/peds.2011-0969.full.pdf

New England Consortium of Metabolic Programs Transition Toolkit
https://www.newenglandconsortium.org/printable-transition-toolkits

American Association of Clinical Endocrinologists
https://www.aace.com/

Endocrine Society
http://www.endo-society.org/

Androgen Excess Society
http://www.ae-society.org/

Society of Reproductive Endocrinology and Infertility
http://www.socrei.org/

American Urological Association
http://www.auanet.org/

Got Transition
http://www.gottransition.org

Referral *(local, state, regional and national)**:***

Clinical Services

Pediatric Endocrine Society "Find A Doc"
http://lwpes.org/

Find Genetic Services
https://clinics.acmg.net/