CT Children's CLASP Guideline

Early Onset Pubic Hair and/or Body Odor

INTRODUCTION

Premature pubarche (**PP**) refers to the early onset of pubic hair, body odor, and/or axillary hair in girls less than 8 years of age and boys less than 9 years. **Premature adrenarche** (**PA**) refers to PP that is also associated with increasing levels of adrenal androgens that fall within pubertal ranges. There is no accelerated growth velocity in either PP or PA. PP and PA need to be distinguished from other rare conditions that require treatment, including late onset congenital adrenal hyperplasia (CAH), virilizing adrenal or gonadal tumors, or exogenous exposure to androgens. In these rare conditions, there will often be rapid virilization *and* accelerated linear growth velocity.

Idiopathic PA is more commonly observed in girls by a ratio of 9:1. Although PA was once thought of as a benign condition, newer studies are linking PA with functional hyperandrogenism, polycystic ovarian syndrome, insulin resistance, Type II diabetes, and metabolic syndrome. Early identification and education of healthy lifestyle choices are encouraged.

INITIAL EVALUATION AND MANAGEMENT

INITIAL EVALUATION:

- **Careful review of the growth chart and calculation of the growth rate**
  - Linear growth rate in PA and PP is within 2.5 SDs of normal without evidence of a linear growth spurt
- **History and physical exam**
  - Should include chronologic details of the onset of the changes noted and sexual maturity staging (see **Clinician Guide: Sexual Maturity Staging**)
  - Assess for rapid virilization (e.g., rapid increase in pubertal hair growth, new acne) *and* accelerated linear growth velocity
    - May indicate rare conditions such as late onset congenital adrenal hyperplasia (CAH), virilizing adrenal or gonadal tumors, or exogenous exposure to androgens
  - Family history of precocious puberty, congenital adrenal hyperplasia, hirsutism/acne, polycystic ovarian syndrome, Type II diabetes, or early cardiovascular disease
- **Girls 8 years of age/boys 9 years of age:**
  - No laboratory evaluation needed unless there are concerns for rapid virilization, rapid height acceleration, and/or rapid pubertal advancement during the observational period
- **Laboratory evaluation:**
  - Indicated for:
    - All girls up to 7.99 years of age and all boys up to 8.99 years of age AND
    - Girls 8 years of age and boys 9 years of age with rapid virilization, rapid height acceleration, and/or rapid pubertal advancement
  - Fasting 8 AM levels of DHEA-S, total testosterone, and 17-hydroxyprogesterone
  - If there is evidence on exam of true precocious puberty (breast development in girls or testicular enlargement in boys):
    - Obtain Pediatric FSH/LH for both boys and girls
    - Add estradiol ultra-sensitive measurements for girls

- *Interpretation of laboratory values in those with PP or PA:
  - PP: normal DHEA-S level and total testosterone level <20 ng/dL
  - PA: DHEA-S level of 40-150 mcg/dL and total testosterone level <20 ng/dL
- Since radiologists interpret bone ages (BAs) differently than pediatric endocrinologists and BA measurements are often advanced in children with PA, we DO NOT recommend obtaining a BA as part of the initial evaluation. The results of the recommended basal hormone levels will suffice to distinguish children who need further evaluation by a pediatric endocrinologist.

INITIAL MANAGEMENT:

- For children with BMI >85th percentile, counseling on healthy lifestyle and obesity prevention (see **Obesity & Screening for Co-Morbidities CLASP tool**)
- **All children less than 4 years of age:**
  - Refer to Endocrine (see **When to Refer**)
- **Girls between 4-7.99 years of age/boys 4 – 8.99 years of age:**
  - If labs are normal, clinical monitoring is appropriate
  - If laboratory results consistent with diagnosis of PP or PA, patient to be followed clinically with primary care provider every 6 months
    - Monitor for evidence of height acceleration, precocious puberty (i.e., breast development in girls, testicular enlargement in boys) and/or rapid virilization
  - Offer education to patients and families (see **Family Handout: Premature Adrenarche**)
- **Girls 8 years of age/boys 9 years of age:**
  - These are the normal ages for children to develop adrenarche. Give reassurance
  - Carefully observe for signs of rapid virilization, height acceleration and/or rapid pubertal progression (i.e., significant change in breast development in girls, or rapid testicular enlargement in boys) for 4-6 months

WHEN TO REFER

All children less than 4 years of age:

- Routine referral to Endocrinology

Girls between 4-7.99 years of age/boys 4 – 8.99 years of age:

- Routine referral to Endocrinology if:
  - laboratory results above ranges for PP or PA, or
  - there are concerns for rapid virilization, rapid height acceleration, and/or rapid pubertal advancement

Girls 8 years of age/boys 9 years of age:

- Routine referral to Endocrinology is only recommended for patients for whom there are concerns for rapid virilization, rapid height acceleration, and/or rapid pubertal advancement

If patient with a BMI ≥95th percentile:

- Refer to Weight Management Program via the **Obesity & Screening for Co-Morbidities Tool**

HOW TO REFER

Referral to Endocrinology via CT Children's One Call Access Center
Phone: 833.733.7669 **Fax:** 833.226.2329
For more information on how to place referrals to Connecticut Children's, click **here**.

	Information to be included with the referral: ▪ Notes from the initial and follow-up visits with the PCP ▪ Complete growth charts ▪ Relevant laboratory and diagnostic studies
WHAT TO EXPECT	**What to expect from CT Children's Visit:** ▪ History, physical exam ▪ Evaluation of prior laboratory testing and growth chart ▪ Additional labs, if appropriate ▪ Bone Age obtained and read by pediatric endocrinologist, if appropriate ▪ Other imaging studies, if appropriate ▪ Initiation of treatment, if appropriate ▪ Comprehensive patient education