



A-T Clinical Center at Johns Hopkins

IMPORTANT FACTS ABOUT: Ataxia-Telangiectasia (A-T) and Estrogen Replacement in Females



Many young women with A-T have irregular menstrual periods or stop having menses at an early age. A few never complete puberty development to the point of starting menstrual periods. There are two general risks for all females with irregular or no menstrual periods:

1. There may be accumulation of the tissue that lines the uterus (endometrium). If this tissue is not periodically shed, there is an increased risk for the development of endometrial cancer.
2. If the lack of menstrual periods is caused by a deficiency of estrogen, there can be a long-term risk for development of osteoporosis (thinning of the bones).

With the help of our pediatric endocrinology and gynecology colleagues at the Johns Hopkins Hospital we have developed the following general guidelines:



In post-pubertal women without menstrual periods, a gynecologist or endocrinologist should be consulted about prescribing a 10-14 day trial of progesterone. This hormone causes the endometrial lining to be shed.

If a menstrual period is induced:

- there is sufficient estrogen production for bone protection.
- treatment with progesterone (as above) every 1-3 months is recommended to prevent accumulation of endometrial tissue.

If a menstrual period is NOT induced:

- there is insufficient estrogen production.
- on-going therapy is recommended to provide normal cycles of estrogen and progesterone.
- this can be accomplished by using standard birth control pills.
- as an alternative, one may use estrogen (Premarin 0.625mg given every day or 5mg/day for 12 consecutive days each month).

In young women who (a) produce their own estrogen or receive it from birth control pills and (b) have a normal diet, there is usually no need for supplemental calcium or vitamin D if dietary intake is adequate. The recommended daily allowance of Calcium (1200 mg) and vitamin D (400 units) can be supplied by 4 eight ounce glasses of milk  or other dairy products. 

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