"AM I DIFFERENT??"

Reproductive Block, PBL; Case 1

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Endocrine Block, PBL; Case 1
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Color Guide:

- **Red:** Relatively important & mentioned in case tutorials.
- **Black:** Questions.
- **Blue:** Answers (mentioned in case tutorials).
- **Green:** Additional answers/notes.
- **Orange:** Explanation.

Learning Objectives

- Link the structure and function of the different parts of the female reproductive system.
- Discuss the physiological basis and the role of the hypothalamic-pituitary-ovarian axis in the regulation of the ovarian and the uterine cycle.
- Discuss the biological changes and physiological mechanisms underlying the occurrence of puberty.
- Discuss the biological effects of estrogen and progesterone during female puberty.

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CASE SCENARIO

Key information:
Lila Ali, a 15 year-old student.

Presenting problems:
1. No period.
2. No change in her body for the last 2 years.
3. No change in her breasts.

History:
Past Medical History: Nil
Family History: Her sisters and mother have their period when they are at 14 years old.
Medication and Allergy: Nil
Alcohol and Smoking: Nil
Social History: No close friends, Unhappy and stressed.

Clinical Examination:
Vital signs: Normal.
External genitalia and pelvic examination: few dark hair & Minimal development of labia minora and labia majora.
Abdominal examination: Normal.
Cardiovascular and respiratory examination: Normal.

Investigation:
1. Full blood count: Normal.
2. Hormonal assays:
- Low FSH
- Low LH
- Low estradiol

3. Thyroid function tests: Normal.
4. Chromosomal studies: Normal.

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QUESTIONS

Before answering the questions below, please read tutorials 1 and 2.

1. What is your diagnosis of this case?
Delayed puberty.

2. What are some possible causes of her delayed puberty?
Severe stress, family history and low bodyweight.

3. Mention the hormones that would probably be decreased in this case?
FSH, LH and estradiol.

4. Why does she have low FSH, LH and estradiol?
Due to lack of stimulation from the hypothalamus.

5. Mechanism involved in regulation of menstrual period?
Stimulation of hypothalamus causes an increase in blood levels of FSH & LH and hence the stimulation of the ovaries. As a result, the ovaries secrete estrogen & progesterone. With these hormonal changes, the lining of uterus undergoes periodic changes resulting in the occurrence of menstrual period.

6. How is puberty triggered?
Possible causes include:
1. Stimuli to the hypothalamus.
2. Hormonal changes.

7. What is the meaning of:
Menarche: First period.
Hymen: A thin, elastic membrane stretched partly or completely across the entrance of the vagina (vestibule).
Webbing of neck: extra skinfold at the neck base.

8. Investigations used in her case?
1. **Full blood count.**
2. **Hormonal assays.**
3. **Thyroid function test.**
4. **Chromosomal analysis.**

GENERAL INFORMATION

Delayed puberty	**When an organism has passed the usual age of onset of puberty with no physical or hormonal signs that it is beginning.**
Normal timing	usually between 10 and 15 years of age.
Possible causes of delayed puberty	• Variation of normal (constitutional delay) • In females, prolonged high level of physical exertion, *e.g.* from being an athlete • Systemic disease, *e.g.* Inflammatory bowel disease, chronic renal failure • Undernutrition *e.g*. anorexia nervosa, zinc deficiency • Hypothalamic defects and diseases e.g. Prader-Willi syndrome, Kallmann syndrome • Pituitary defects and diseases e.g. hypopituitarism • Gonadal defects and diseases e.g. Turner syndrome, Klinefelter syndrome, Testicular failure due to mumps orchitis, Coxsackievirus B, irradiation, chemotherapy, or trauma. Testicular failure is treated with testosterone replacement, Ovarian failure.
Constitutional delay	Children who are healthy but have a slower rate of physical development thar average have constitutional delay in growth and adolescence. These children have a history of stature shorter than their age-matched peers throughout childhood, but their height is appropriate for bone age, and skeletal development is delayed more than 2.5 SD. They usually are thin and often hav a family history of delayed puberty. Children with a combination of a family tendency toward short stature and constitutional delay are the most likely to seek evaluation. They quite often seek evaluation when classmates or friends undergo pubertal development and growth, thereby accentuating their delay.
Management	If a child is healthy but simply late, reassurance and prediction based on the bone age can be provided. No other intervention is usually necessary. In more extreme cases of delay, or cases where the delay is more extremely distressin to the child, a low dose of testosterone or estrogen for a few months may brir the first reassuring changes of normal puberty.

More information:
http://en.wikipedia.org/wiki/Delayed_puberty