



Feminine Health Issues and FOP

BEAU PARK MD

MONA AL MUKADDAM MD, MS

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Hypothetical Case 1

- ▶ 25 yr old female with FOP diagnosed at the age of 12 yr
- ▶ Primary amenorrhea ie never had a menstrual cycle
- ▶ Last flare up was 12 months ago, she received a short course of prednisone
- ▶ Normal breast development and does have some axillary and pubic hair

Definition of Amenorrhea

- ▶ **Primary**

- ▶ Absence of menarche by age 15 years or thereafter

- ▶ **Secondary**

- ▶ Absence of menses for more than three months in girls or women who previously had regular menstrual cycles or six months in girls or women who had irregular menses)

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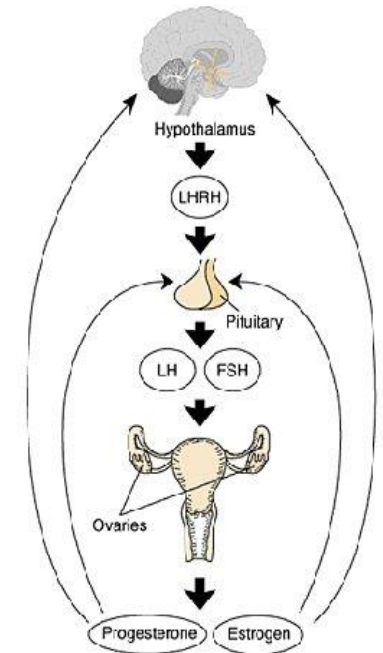
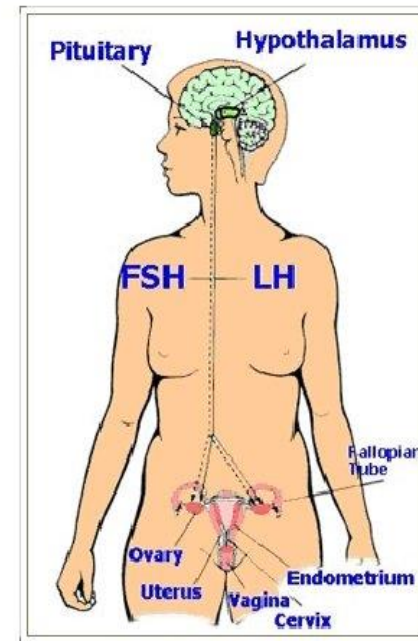
What other information would
you need?

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Cause of Amenorrhea

- ▶ Anatomical
 - ▶ Absent uterus
 - ▶ Adhesions in the uterus
 - ▶ Congenital defect of urogenital outlet
- ▶ Disorders of hypothalamic-pituitary-ovarian axis
 - ▶ Polycystic ovary syndrome (PCOS)
 - ▶ Ovarian insufficiency or agenesis
 - ▶ Hypothalamic as seen stress/chronic illness
 - ▶ Pituitary example prolactin secreting pituitary adenoma

Hypothalamic-Pituitary-Ovarian Axis



Previous work up

- ▶ FSH, LH and estradiol: low
- ▶ Pelvic imaging: Uterus present, no evidence for outlet obstruction
- ▶ MRI of pituitary: no abnormalities noted
- ▶ Normal thyroid levels, normal prolactin, normal testosterone

Treatment options

- ▶ Depends on the etiology
- ▶ Providing estrogen replacement is important for bone health (with progesterone with present uterus)
- ▶ Oral contraceptives and hormone replacement therapy have been used safely in FOP patients
- ▶ Monitor for blood clots

Hypothetical case 2

- ▶ 24 yr old female with FOP
- ▶ High CAJIS score of 24 with both shoulders, elbows, hips and knees are fully ankylosed
- ▶ Getting menstrual cycle every 5-7 weeks and with heavy menstrual bleeding
- ▶ What would you recommend to decrease the burden of monthly cycles and assist with hygiene

Medications to decrease or stop menstrual cycle?

- ▶ Goals of treatment
 - ▶ Predictability of menses
 - ▶ Reduction in duration or frequency
 - ▶ Decrease menstrually-associated symptoms
 - ▶ Complete amenorrhea (no periods)

Medications to decrease or stop menstrual cycle?

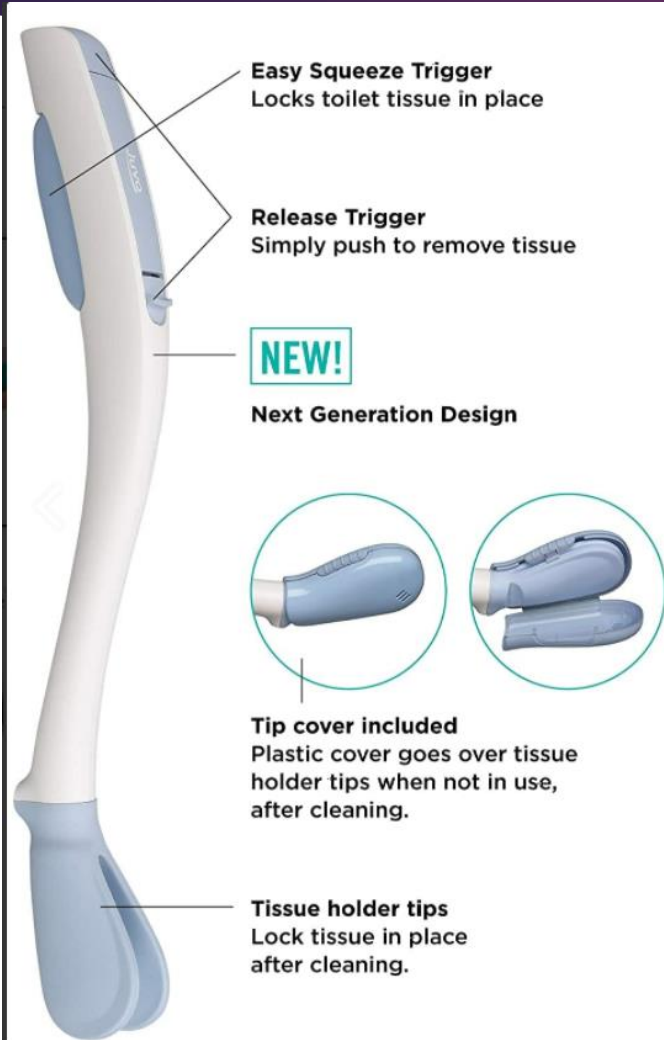
- ▶ Combined estrogen/progestin contraceptive pills
- ▶ Combined estrogen/progestin patches, vaginal rings
- ▶ Progestin only pills (may not offer contraception)
- ▶ Progestin IUD



Medications to decrease or stop menstrual cycle?

- ▶ Non-hormonal methods
 - ▶ NSAIDS (ibuprofen) can decrease menstrual flow and menstrual cramping
 - ▶ Tranexamic acid can reduce menstrual bleeding

Ability Toolbox Guidebook (ifopa.org)



Tip cover included
Plastic cover goes over tissue holder tips when not in use, after cleaning.



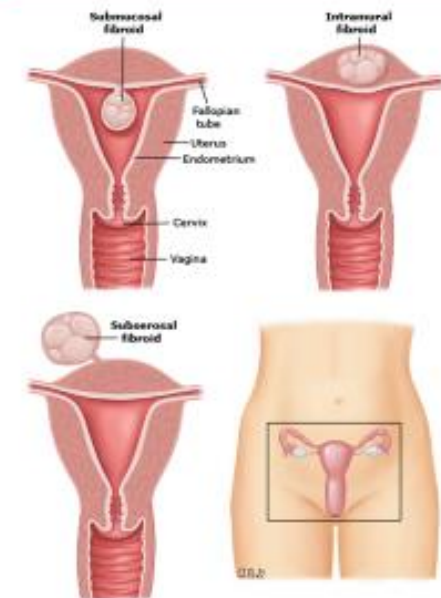
Hypothetical Case 3

- ▶ 34 yr old female with FOP diagnosed with a large uterine fibroid on evaluation of irregular and heavy menstrual cycles
- ▶ The uterine fibroid is causing abdominal discomfort but tolerable and the bleeding is not bothersome

What is a uterine fibroid?

- ▶ Uterine leiomyomas (also referred to as fibroids or myomas) are the most common pelvic tumor in females
- ▶ Prevalence ranges from 7-80%, increases with age
- ▶ Abnormal growths that form in the muscle of the uterus
- ▶ NOT cancer

Fibroid locations in the uterus



These figures depict the various types and locations of fibroids. A woman may have one or more type of fibroid.

Symptoms of Fibroids

- ▶ Fibroids often cause no symptoms at all. When they do cause symptoms, they can cause:
 - ▶ Heavy periods
 - ▶ Pain, pressure, or a feeling of "fullness" in the belly
 - ▶ The need to urinate often
 - ▶ Too few bowel movements (constipation)
 - ▶ Difficulty getting pregnant

Do they need to be treated?

- ▶ Your symptoms
- ▶ Your age (most fibroids shrink or stop causing symptoms after menopause, which is when monthly periods stop)
- ▶ Whether you want to get pregnant in the future
- ▶ Whether your fibroids cause so much bleeding that you have a condition called anemia
- ▶ The size, number, and location of your fibroids
- ▶ How you feel about the risks and benefits of the different options

How can you treat uterine fibroids?

▶ **Medicine**

- ▶ Decrease bleeding
- ▶ Does not significantly decrease the size of the fibroid

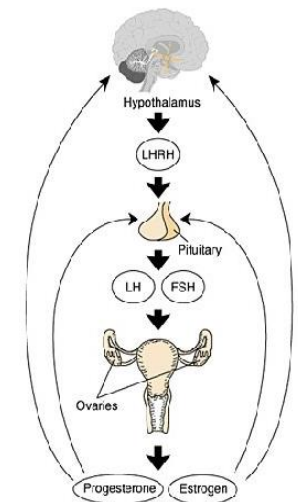
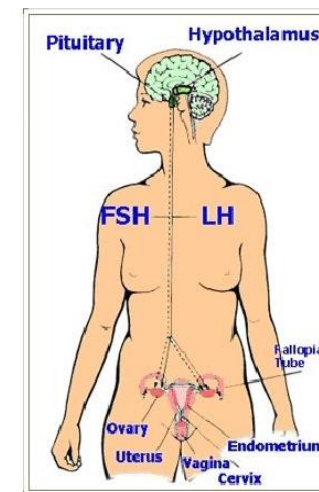
▶ **Interventional**

- ▶ Surgical removal
- ▶ Embolization
 - ▶ No experience in FOP patients (as far as we are aware)
 - ▶ Requires sedation
 - ▶ Recurrence of symptoms can occur

Which medication are safe in FOP?

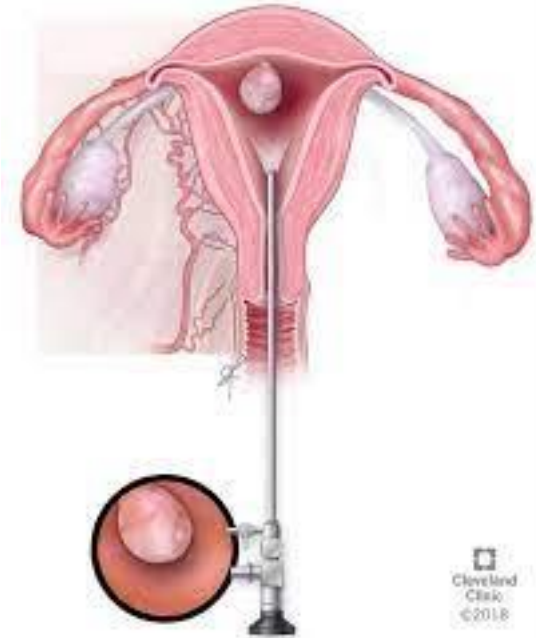
- ▶ Estrogen and Progestin hormones as mentioned earlier
- ▶ GnRH Antagonist (Orilissa, pill) –for endometriosis
- ▶ GnRH Antagonist + estrogen/progestin (Oriahnn, pill) – for fibroids
- ▶ GnRH Agonist (Zoladex, subcutaneous implant) – for endometriosis

- ▶ GnRH Antagonists/Agonists are not long term options
- ▶ GnRH Antagonists/Agonists may have severe side effects



Which surgical interventions are safe in FOP?

- ▶ Hysteroscopy - surgery within the uterus for fibroids, polyps
 - ▶ Myomectomy – removal of fibroids
 - ▶ Hysterectomy – removal of uterus
 - ▶ Laparoscopy – surgery through small ports
-
- ▶ Need multi-team approach including surgeons, anesthesia, postoperative/ICU teams, pulmonologist that are familiar with FOP patients and their specific challenges



Has hysterectomy ever been performed in FOP patients?

- ▶ Hysterectomies have been done safely in patients
- ▶ No heterotopic bone formation in our experience and from case reports



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Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABDEF 1,2 Michelle Ho
ABEF 3 Beau Y. Park
DEF 3 Norman G. Rosenblum
DEF 4 Mona Al Mukaddam
ADEF 4 Frederick S. Kaplan
EF 5 Victor Kucherov
EF 5 Scott G. Hubosky
AD 6 Gregory Kane
DE 7 Vishal Desai
DE 7 Michael R. Kramer
AD 2,8 Bon S. Ku
EF 9 Eric S. Schwenk
EF 9 Jaime L. Baratta
EF 9 Deepti Harshavardhana
ABDEF 9 Zvi Grunwald

Surgical and Radiological Management of Complicated Uterine Leiomyoma Aided by 3D Models in a Patient with Fibrodysplasia Ossificans Progressiva

1 Sidney Kimmel Medical College, Thomas Jefferson University, Philadelphia, PA, U.S.A.
2 Health Design Lab, Thomas Jefferson University, Philadelphia, PA, U.S.A.
3 Department of Obstetrics and Gynecology, Thomas Jefferson University, Philadelphia, PA, U.S.A.
4 Departments of Medicine and Orthopedic Surgery, Center for Research in FOP and Related Disorders, The Perelman School of Medicine, University of Pennsylvania, Philadelphia, PA, U.S.A.
5 Department of Urology, Thomas Jefferson University, Philadelphia, PA, U.S.A.
6 Department of Medicine, Thomas Jefferson University, Philadelphia, PA, U.S.A.
7 Department of Radiology, Thomas Jefferson University, Philadelphia, PA, U.S.A.
8 Department of Emergency Medicine, Thomas Jefferson University, Philadelphia, PA, U.S.A.
9 Department of Anesthesiology, Sidney Kimmel Medical College at Thomas Jefferson University, Philadelphia, PA, U.S.A.

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How often to repeat imaging if patient decides to monitor only?

- ▶ In general there is no consensus on frequency of ultrasounds for fibroid surveillance
- ▶ Fibroid growth can be unpredictable
- ▶ However, we can consider yearly ultrasounds or sooner (if there are changes in symptoms such as bleeding or pain) for FOP women
 - ▶ To avoid bigger or more complicated surgeries if possible
 - ▶ To avoid any emergent or unplanned surgeries

When would you recommend intervention?

- ▶ Symptoms such as bleeding, pain, pressure, urinary frequency are bothersome
- ▶ Surgical intervention when medical therapies are not helpful and surgery is carefully planned
- ▶ Definitive therapy (hysterectomy) when done with fertility

Thank you