Hoof Beats Sometimes Do Mean Zebras MMXXII

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Disclosure Slide

- We have nothing to disclose

Objectives

- Develop a practical approach to adolescents presenting with unusual clinical signs, symptoms, or confusing diagnostic tests
- Learn when to ask for help
- Review the benefits of a comprehensive risk assessment with all patients

Morning report

Our Morning report

THIS IS AN INTERACTIVE PROGRAM

1. Present history
2. Provide PMHx
3. SHEADDDSSS
4. Exam findings
5. Test results/imaging
6. Diagnosis and further information

Refine ideas and diagnosis at every step of the process

“Heavy Menstrual Flow”

- 6/6/22 - 17year 9mo female referred from Hematology Clinic for “Iron Deficiency, AUB and Heavy menstrual Flow and Enuresis for months.”
- Seeing patient in multispecialty Adolescent Medicine-Hematology “GABS” Clinic.
- Seen in Hematology 3/21/22
  - Noted to have 7days of bleeding with menses
  - Using 8 or more tampons + pads per day
  - Overflow bleeding overnight with pads
  - Had RBC anemia, started on Iron referred to GABS
    - Sent for U/S and MRI?
- U/S and MRI read largely as normal?

“Heavy Menstrual Flow”

- Differential?

TABLE 10-1

Differential Diagnosis of Abnormal Vaginal Bleeding in the Adolescent Girl

- Anovulatory uterine bleeding
- Pregnancy-related complications
  - Threatened abortion
  - Spontaneous, incomplete, or missed abortion
  - Ectopic pregnancy
  - Gestational trophoblastic disease
  - Complications of termination procedures
- Infection
  - Pelvic inflammatory disease
  - Endometritis
  - Cervicitis
  - Vaginitis
- Bleeding disorders
  - Thrombocytopenia (e.g., idiopathic thrombocytopenic purpura, leukemia, aplastic anemia, hypersplenism, chemotherapy)
  - Coagulation disorders (e.g., von Willebrand disease, other disorders of platelet function, liver dysfunction, vitamin K deficiency)
- Endocrine disorders
  - Hypo- or hyperthyroidism
  - Adrenal disease
  - Hyperprolactinemia
  - Polycystic ovary syndrome
  - Primary ovarian insufficiency
- Vaginal abnormalities
  - Carcinoma or sarcoma
  - Laceration
- Cervical problems
  - Cervicitis (including cystic fibrosis)
  - Polyp
  - Hemangioma
  - Carcinoma or sarcoma
- Uterine problems
  - Submucous myoma
  - Congenital anomalies
  - Polyp
  - Carcinoma
  - Use of intrauterine device
  - Breakthrough bleeding associated with hormonal contraceptives
  - Ovulation bleeding
- Ovarian problems
  - Cyst
  - Tumor (benign, malignant)
- Endometriosis
- Trauma
- Foreign body (e.g., retained tampon)
- Systemic diseases
  - Diabetes mellitus
  - Renal disease
  - Systemic lupus erythematosus
- Medications
  - Hormonal contraceptives
  - Anticoagulants
  - Platelet inhibitors
  - Androgens
  - Spironolactone
  - Antipsychotics

“Heavy Menstrual Flow”

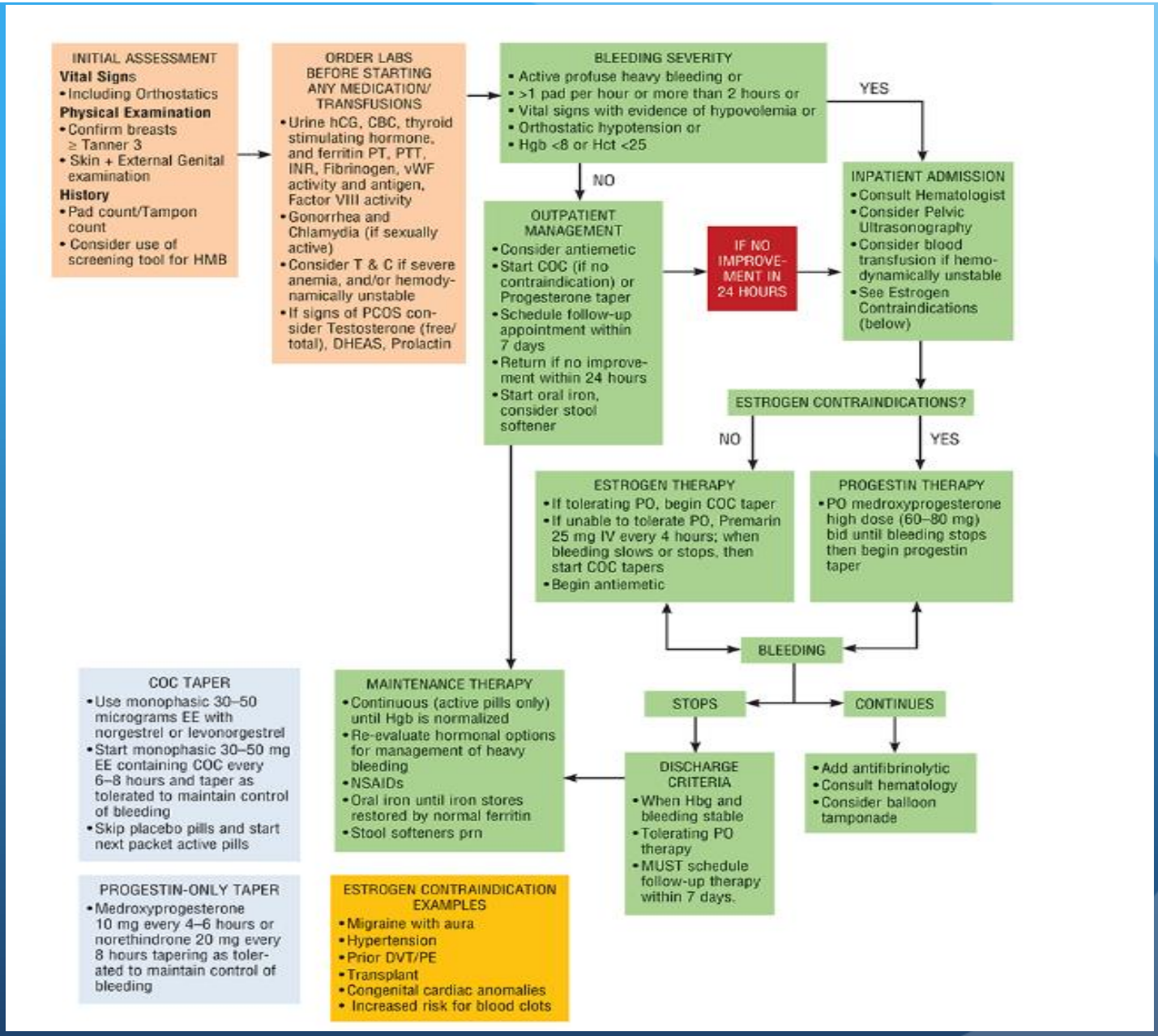
- Diagnosis: ***Heavy menstrual bleeding*** **is defined as excessive menstrual blood loss that interferes with a woman's physical, social, emotional, or material quality of life. It can occur alone or in combination with other symptoms**
- Differential?
- **Heavy menstrual bleeding should be classified according to:**
  - **PALM–COEIN system: Polyp, Adenomyosis, Leiomyoma, Malignancy and hyperplasia, Coagulopathy, Ovulatory dysfunction, Endometrial, Iatrogenic, and Not otherwise classified**

Table 1. Common Bleeding Disorders in Adolescents Who Present With Heavy Menstrual Bleeding

Bleeding Disorder	Physiologic Description	Recommended Laboratory Tests
Platelet Function Disorders (Defects of platelet adhesion, aggregation, or secretion)		
von Willebrand Disease		aPTT, von Willebrand antigen, von Willebrand activity, vWF:RCo, factor VIII level
Type 1	Quantitative deficiency of vWF, autosomal dominant inheritance	
Type 2 (multiple subtypes)	Qualitative defect in vWF activity in platelet adhesion or binding factor VIII, most commonly autosomal dominant inheritance	
Type 3	Absent vWF, autosomal recessive	
Glanzmann thrombasthenia	Abnormalities of the platelet membrane glycoproteins IIb or IIIa (GPIIb/IIIa) that mediate binding to fibrinogen, resulting in reduced platelet aggregation or clumping	Platelet aggregation studies
Bernard–Soulier syndrome	Inherited deficiency in platelet membrane glycoprotein complex Ib-IX causing defective adhesion of the platelets to subendothelial matrix	Platelet aggregation studies
Delta storage pool disorders	Disorder of platelet secretion due to defects in platelet activation factors	Platelet aggregation and secretion studies
Other Disorders		
Clotting factor deficiencies	Deficiencies of any major clotting factor, factor VIII deficiency (hemophilia A) and factor IX deficiency (hemophilia B) are symptomatic carriers due to X-linked inheritance, but can occur due to inactivation of the X chromosome carrying the normal gene	Isolated prolonged PT detects factor VII deficiency; Isolated prolonged aPTT detects VIII, IX, XII deficiencies; Combined prolonged PT or aPTT detects deficiencies in factors II, V, X, fibrinogen
Thrombocytopenia	Low platelet count. Can be associated with idiopathic thrombocytopenia, or immune-mediated thrombocytopenic purpura	Platelet count
Fibrinolytic pathway defects	Dysfibrinogenemia or plasminogen deficiency	Fibrinogen or thrombin time

Abbreviations: aPTT, activated partial thromboplastin time; PT, prothrombin time; vWF, von Willebrand factor; vWF:RCo, von Willebrand factor ristocetin cofactor.

https://www.acog.org/clinical/clinical-guidance/committee-opinion/articles/2019/09/screening-and-management-of-bleeding-disorders-in-adolescents-with-heavy-menstrual-bleeding



https://www.acog.org/clinical/clinical-guidance/committee-opinion/articles/2019/09/screening-and-management-of-bleeding-disorders-in-adolescents-with-heavy-menstrual-bleeding

“Heavy Menstrual Flow”

Screening Tool

How to Use the Screening Tool

The screening tool is considered to be positive if 1 of the following 4 criteria were met:

1. **The duration of menses was greater than or equal to 7 days and the woman reported either “flooding” or bleeding through a tampon or napkin in 2 hours or less with most periods;**

2. **A history of treatment of anemia;**

3. **A family history of a diagnosed bleeding disorder; or**

4. **A history of excessive bleeding with tooth extraction, delivery or miscarriage, or surgery**

Adapted from Philipp CS, Faiz A, Dowling NF, Beckman M, Owens S, Ayers C, et al. Development of a screening tool for identifying women with menorrhagia for hemostatic evaluation. Am J Obstet Gynecol 2008;198:163.e1–8; Philipp CS, Faiz A, Heit JA, Kouides PA, Lukes A, Stein SF, et al.

“Heavy Menstrual Flow”

Screening Tool to Identify Adolescents With Heavy Menstrual Bleeding for Testing and Evaluation for Underlying Bleeding*

1. **How many days did your period usually last, from the time bleeding began until it completely stopped?**
    1. **Less than 7 days**
    2. **Greater than or equal to 7 days**
    3. **Don’t know**

2. **How often did you experience a sensation of “flooding” or “gushing” during your period?**
    1. **Never, rarely, or some periods**
    2. **Every or most periods**
    3. **Don’t know**

3. **During your period did you ever have bleeding where you would bleed through a tampon or napkin in 2 hours or less?**
    1. **Never, rarely, or some periods**
    2. **Every or most periods**
    3. **Don’t know**

“Heavy Menstrual Flow”

4. Have you ever been treated for anemia?

1. No
2. Yes
3. Don’t know

5. Has anyone in your family ever been diagnosed with a bleeding disorder?

1. i. No
2. Yes
3. Don’t know

6. Have you ever had a tooth extracted or had dental surgery?

1. No (If no, go to question 7)
2. Yes
3. Don’t know

“Heavy Menstrual Flow”

6a. Did you have a problem with bleeding after tooth extraction or dental surgery?

1. No
2. Yes
3. Don’t know

7. Have you ever had surgery other than dental surgery?

1. No (If no, go to question 8)
2. Yes
3. Don’t know

7a. Did you have bleeding problems after surgery?

1. No
2. ii. Yes
3. Don’t know

“Heavy Menstrual Flow”

8. Have you ever been pregnant?

1.i. No

2.Yes

3.Don’t know

8a. Have you ever had a bleeding problem following delivery or after a miscarriage?

1.i. No

2.Yes

3.Don’t know

“Heavy Menstrual Flow”

- What Else?

“Heavy Menstrual Flow”

- What Else?
  - Infection, STIs etc
  - Foreign Body
  - Endocrine/Thyroid
  - …

“Heavy Menstrual Flow”

- SHEADDDSS

SHEADDDSS Assessment

- Strengths: Good friend
- Home: Lives with Mom, Dad and younger sister
- Education: -> 12th Grade has friends at school, plans to be a paramedic
- Activities: Works part-time 10hrs/wk, plays tennis with friends
- Diet: “Eat healthy” No weight loss
- Drugs: Daily Vaping and wants to quit. No THC, No EtOH, No drugs, +Smokers in the home
- Depression: Somewhat sad about bleeding and anemia PHQ-9 <7, no SI
- Safety: No physical or sexual abuse, +Guns at home in a safe
- Sexual Health: Given/received oral sex, NO vaginal sex, no contraception. Interested in Contraception and Emergency Contraception. No history of STI or screening

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“Heavy Menstrual Flow”

- ROS?

“Wait, We Gotta Go Back!”

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- 6/6/22 - 17year 9mo female referred from Hematology Clinic for “Iron Deficiency, AUB and Heavy menstrual Flow and **Enuresis,** for months since starting her period.”
- PMHx: She was seen in an outside rural ER “Out Lady of the Sea,” at age 13yo, in 2019 for RLQ, LLQ pelvic pain with “deep” radiation to the back repeatedly over 4 days

“Heavy Menstrual Flow”

- Differential?

“Heavy Menstrual Flow”

- PMHx: She was seen in an outside rural ER “Out Lady of the Sea,” at age 13yo, in 2019 for RLQ, LLQ pelvic pain with “deep” radiation to the back repeatedly over 4 days
- The pain is described as cramping, pressure-like and sharp. She went to LOSH and CT scan was done. Mom reports that the ER said, "it looks like her period is about to start, but it can't" and recommended GYN follow up. She then went to TGMC and St. Anne over the weekend secondary to worsening pain. She reports that she has never received relief with any pain medication. She reports "feeling a knot" in her vagina. She has never had a period and has never been sexually active. The pain has gotten so bad that she has been unable to tolerate anything PO for about 1 day.

“Heavy Menstrual Flow”

- Differential?

“Heavy Menstrual Flow”

- Genitourinary:
  Genitourinary Comments: Imperforate hymen, complete
  Bulging hymen membrane

- OPERATIVE NOTE:
  The patient was taken to the operating room where MAC anesthesia was found to be adequate. She was prepped and draped in the normal sterile fashion in the dorsal lithotomy position. An in and out catheter was used to drain the bladder. The catheter was kept in place for visualization of urethra location. The hymen-vulvar border was outlined with the needle Bovie. The hymen was opened with the needle Bovie and 700 cc of dark blood was spontaneously expressed. The hymen was then grasped and excised with the Bovie. The area was sutured with 2-0 Vicryl in a running fashion for hemostasis. The vagina was inspected by placing a speculum in the posterior vagina. The cervix was visualized but noted to be dilated. The vagina was irrigated with sterile water. The vagina was examined and I was now open 2 finger breadths. All instruments were then removed. The patient tolerated the procedure well. Instrument, needle, and lap counts were correct x2. The patient was taken to recovery room in stable condition.

“Heavy Menstrual Flow”

- Imperforate hymen (IH) is an uncommon congenital anomaly of the female genital tract, with the hymen completely obstructing the vaginal opening. Despite the simple diagnosis and treatment of IH, missed or delayed diagnosis is often a clinical problem owing to its low incidence, nonspecific symptoms, or insufficient physical examination.
- Occurs in 0.05% to 0.1% of infant girls

Lee KH, Hong JS, Jung HJ, et al. Imperforate Hymen: A Comprehensive Systematic Review. *J Clin Med*. 2019;8(1):56. Published 2019 Jan 7. doi:10.3390/jcm8010056

Mou J.W., Tang P.M., Chan K.W., Tam Y.H., Lee K.H. Imperforate hymen: Cause of lower abdominal pain in teenage girls. *Singap. Med. J.* 2009;50:378-379.

“Heavy Menstrual Flow”

https://www.researchgate.net/figure/Normal-anatomy-of-vulva-and-variations-in-the-hymenal-appearance_fig1_260578888

“Heavy Menstrual Flow”

- **Etiology:** During urogenital development, the cloaca is divided by the urorectal septum to separate the ventral urogenital sinus from the dorsal anal canal. The Wolffian ducts (mesonephric ducts) and Mullerian ducts (paramesonephric ducts) are the two main ductal systems that give rise to specific structures in the male and female genital and urinary tracts. The Wolffian duct develops from the paired urogenital ridges and grows caudally to induce the formation of the mesonephric tubules. Additionally, the Wolffian duct induces the development of the Mullerian ducts from the coelomic epithelium of the paired urogenital ridges. The Mullerian ducts grow caudally using the Wolffian ducts as guides. The close association of the Mullerian ducts and Wolffian ducts during development explains the high incidence of urinary tract anomalies in women who also have Mullerian anomalies. The fusion of the Mullerian ducts forms the uterovaginal canal that gives rise to the uterus, cervix, and upper two-thirds of the vagina. The distal point of contact of the Mullerian ducts with the urogenital sinus is the site of the proliferation of the urogenital sinus epithelium, thereby forming the sinovaginal bulb. The sinovaginal bulb develops into the vaginal plate.

- The embryological origin of the lower third of the vagina is the urogenital sinus. The precise embryological origin of the imperforate hymen is controversial. It is thought that the hymen normally perforates during the perinatal period at approximately 22 weeks. The most popular theory is that the hymen may fail to canalize with the rest of the vagina during development when the sinovaginal bulbs canalize at the site where the uterovaginal canal meets the urogenital sinus.[4][5] The imperforate hymen results from unsuccessful canalization of the vaginal plate and failed degeneration of the hymenal epithelial cells.[4] The hymen is likely derived from urogenital sinus posterior wall invaginations.[6]

“Heavy Menstrual Flow”

- Imaging 3/21/22
- LCMC US PELVIS COMPLETE:
- There is a thickened endometrial stripe at 1.6 cm. There is a small cyst at the junction of the cervix and vagina. Right ovary appears normal with a volume of 9 mL. The left ovary is enlarged at 45 mL with a complex cystic appearing lesion.
- IMPRESSION:
- Enlarged left ovary with complex cystic appearing mass. Consider MRI with and without contrast. Otherwise follow-up study recommended in one month. Thickened endometrium.

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“Heavy Menstrual Flow”

- MRI Imaging 4/4/22
- MRI PELVIS WITH AND WITHOUT CONTRAST: Sequential imaging of the lower abdomen and pelvis was performed both before and following administration of intravenous contrast. The patient has a described palpable lower anterior right abdominal mass.
- There is moderate subcutaneous fat deposition throughout the abdomen and pelvis without evidence of associated subcutaneous mass or cyst. There are no irregularities of the subcutaneous septa and no evidence of vascular anomaly. The underlying musculoskeletal system appears normal. There is physiologic change within the ovaries and uterus. There is no additional abnormality.
- Following contrast administration there is no unusual enhancement. IMPRESSION: NORMAL STUDY

“Heavy Menstrual Flow”

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- Following contrast administration there is **no unusual enhancement**. IMPRESSION: NORMAL STUDY

“Heavy Menstrual Flow”

- Additional History:
- Past Medical History:
- Family Medical History:
  - No significant medical problems in family
  - ?
  - ?

“Heavy Menstrual Flow”

- Additional History: Bilateral lower abdominal/pelvic pain now.
- Past Medical History: As above
- Family Medical History:
  - No significant medical problems in family
  - No bleeding disorders noted
  - No history of early hysterectomy
  - Mother started menses at 12yo

“Heavy Menstrual Flow & Pain”

- Additional History: Bilateral lower abdominal/pelvic pain now.
- Past Medical History: As above
- Family Medical History:
  - No significant medical problems in family
  - No bleeding disorders noted
  - No history of early hysterectomy
  - Mother started menses at 12yo
  - Sister started menses at 12yo (younger)

“Heavy Menstrual Flow”

- What Else?

“Heavy Menstrual Flow”

- SHEADDDSS

SHEADDDSS Assessment

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- Home: Lives with Mom, Dad and younger sister
- Education: -> 12th Grade has friends at school, plans to be a paramedic
- Activities: Works part-time 10hrs/wk, plays tennis with friends
- Diet: “Eat healthy” No weight loss
- Drugs: Daily Vaping and wants to quit. No THC, No EtOH, No drugs, +Smokers in the home
- Depression: Somewhat sad about pain and anemia PHQ-9 <7, no SI
- Safety: No physical or sexual abuse, +Guns at home in a safe
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“Heavy Menstrual Flow”

- ROS?

“Heavy Menstrual Flow”

- ROS- Enuresis. NOT helped with DDAVP, fluid restriction and urination before bed. Patient uses tampons and pads to address at night, and day.
- Constitutional: Negative for chills, fever, malaise/fatigue and weight loss.
- Eyes: Negative for blurred vision. Cardiovascular: Negative for chest pain.
- Gastrointestinal: Positive for abdominal pain. Negative for blood in stool, constipation, diarrhea, heartburn, melena, nausea and vomiting.
- Genitourinary: Positive for frequency. Negative for dysuria, flank pain, hematuria and urgency.
- Neurological: Positive for headaches. Negative for dizziness, loss of consciousness and weakness. **NO Migraine with aura diagnosis and no evidence of aura for this patient.**
- Psychiatric/Behavioral: Negative for depression and substance abuse. The patient is not nervous/anxious.

“Heavy Menstrual Flow”

- ?

“Heavy Menstrual Flow”

- Exam:

Abdominal: General: Abdomen is flat. No distension. Abdomen is soft, no mass. Tenderness: There is abdominal tenderness. There is guarding. There is no right CVA tenderness, left CVA tenderness or rebound.

Genitourinary: Vagina: Vaginal discharge present. Cervix: Discharge present. Uterus: Tender. Adnexa: Right: Tenderness present. No mass or fullness. Left: Tenderness present. No mass or fullness. Rectum: Normal.

“Heavy Menstrual Flow”

- Pelvic Exam:

Comments: Patient with urine/clear fluid at vaginal opening and labia throughout and she notes using Tampons during day to stop urine flow.

Yellow-White mucoid discharge at vagina/cervical os.

Right cervical Cyst visualized at Cervico-Vaginal Junction on exam with some discharge present and appearing similar to Cervical Os.

Cervix somewhat difficult to ascertain initially.

Patient had Bilateral Adnexal Tenderness to palpation. Mild Uterine Tenderness.

“Heavy Menstrual Flow”

- Differential?

“Heavy Menstrual Flow”

- Imaging 6/6/22
- Pelvic and Transvaginal U/S Ordered

“Heavy Menstrual Flow”

- Imaging 6/6/22
- Transvaginal U/S: Result:

“Heavy Menstrual Flow”

- Imaging 6/6/22
- Transvaginal U/S: Result:

“Heavy Menstrual Flow”

- Imaging 6/6/22, Pelvic U/S:

- PELVIC ULTRASOUND: Ultrasound of the pelvis was obtained through a distended urinary bladder. Endometrial stripe measures 0.6 cm, previously measured 1.6 cm. **Again noted a 2.1 x 1 x 1.3 cm cystic structure about the junction of the cervix and vagina.** Right and left ovaries are within normal limits with a volume of 12 mL and 7 mL respectively. Previously identified hemorrhagic cyst in the left ovary is not noted on the current study. Bilateral ovaries demonstrate normal arterial and venous flow. There is no free fluid.

- IMPRESSION: **AGAIN NOTED A CYSTIC STRUCTURE ABOUT THE CERVIX & VAGINA JUNCTION**, AS IN PRIOR. OTHERWISE UNREMARKABLE STUDY

“Heavy Menstrual Flow”

- MRI Imaging 6/6/22 (ADDENDUM)
- ======== ADDENDUM #1: The previously visualized complex process within the left ovary is represented by hemorrhage within a follicular cyst. There is no significant size discrepancy between the right and left ovaries with abundant follicular cyst noted bilaterally. **The right cervical cyst remains unchanged.**

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- Journal ofthe Royal Society of Medicine Volume 76 July 1983 Imperforate hymen and vaginal atresia and their associated anomalies 1 L M A Shaw MB BS W A Jones FRCS R J Brereton FRCS2 Department of Paediatric Surgery. Alder Hey Children's Hospital, Liverpool

- The presenting features and associated abnormalities of imperforate hymen and vaginal atresia were studied in 24 girls under the age of 16 years.

- Urinary tract abnormalities were found in all of the remaining 15 patients in whom intravenous pyelography was performed. Unilateral hypoplastic kidneys were found in 2 girls, .and 3 had unilateral renal agenesis. Duplex kidneys were found in 3 girls. Ectopic ureters were discovered in 2 infants, and one neonate had a non-functioning dysplastic kidney without a ureter. Hydronephrosis or hydroureter occurred in 8 children. One girl had a pelvic kidney.

Congenital Urethrovaginal Fistula With Imperforate Hymen: A First Case Report: https://www.jogc.com/article/S1701-2163(16)34245-1/pdf

“Heavy Menstrual Flow”

- Now what?

“Heavy Menstrual Flow”

- Treated for PID in the office:
  - Labs:
  - CBC and platelets normal, Bleeding Studies and VWD Panel normal, Mild +ANA,
  - Trichomonas Negative, GC/Chlamydia?

- Now what?
- Referred to urogynecology for additional evaluation and imaging, WITH TRANSVAGINAL U/S
- Appointment in 2 weeks