



# **SIGNS AND SYMPTOMS OF BLEEDING DISORDERS**

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# OBJECTIVES

## Recognize

Recognize signs and symptoms of inherited bleeding disorders

## Discuss

Discuss different management strategies for these symptoms

## Review

Review the current treatment options for girls and women with heavy menstrual bleeding

## Review

Review a case with heavy menstrual bleeding

# FIRST LET'S REVIEW ....

## HOW DO WE STOP BLEEDING?

- **Normal clotting** requires the following:
  - Vasoconstriction
  - Platelet plug formation
  - Activation of the coagulation cascade
  - Fibrin clot formation

# CAUSES OF ABNORMAL BLEEDING

- **Hereditary**, spontaneous or acquired abnormalities
  - Platelets (abnormal quantity and/or function)
  - Clotting Factor Proteins (deficiency or absence)
    - Von Willebrand Factor (quantitative and qualitative)
    - Factors I-XIII

# SIGNS & SYMPTOMS



## **Frequent nose bleeds (epistaxis)**

Lasting longer than 10  
minutes



## **Prolonged bleeding from mouth or gums**

Dental procedures/extractions  
Trauma  
Tooth eruption

## SIGNS & SYMPTOMS (CONT.)



- **Easy and/or unusual bruising**
  - Little trauma
  - Large and raised
  - Uncommon sites
  - Bruising with vaccinations

## **SIGNS & SYMPTOMS (CONT.)**

- **Heavy or prolonged menses**
  - Menstrual cycle lasts more than 7 days
  - Clots (larger than a grape or 1 inch in diameter)
  - Anemia (due to blood loss)
  - Frequent pad/tampon change (flooding)






## **SIGNS & SYMPTOMS (CONT.)**

- **Profuse bleeding with surgical procedures**
- **Bleeding from small cuts take longer to stop**
- **Hemorrhage at time of childbirth**
- **Prolonged bleeding during post partum period**
- **Blood in urine and/or GI tract**
- **Joint bleeds**
- **Muscle bleeds**



# BEIGHTON HYPERMOBILITY SCORE

- 9 point scoring system
- Hypermobility is present when the score is greater than or equal to 6/9 in children 6-12 yrs. and 5/9 in adult
- Joint hypermobility is associated with Ehlers-Danlos Syndrome
- Generalized joint hypermobility is often associated with increased bleeding tendencies

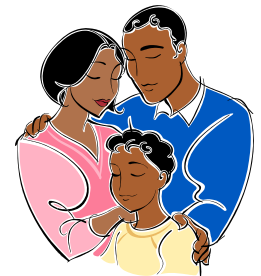
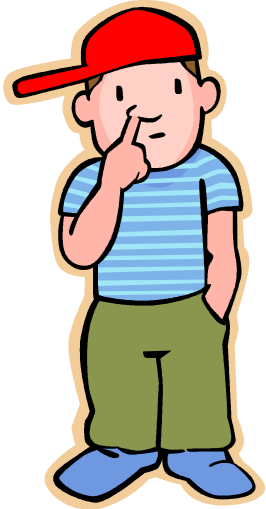
Maneuver	Example	Number of Points
Apposition of the thumb to the flexor aspect of the forearm		1 point for each side (Maximum Score of 2)
Passive dorsiflexion of the metacarpophalangeal joint to 90 degrees		1 point for each side (Maximum Score of 2)
Passive hyperextension of the elbow greater than 10 degrees		1 point for each side (Maximum Score of 2)
Passive hyperextension of the knee greater than 10 degrees		1 point for each side (Maximum Score of 2)
Forward flexion with the hands flat on floor and knees extended		1 point for the procedure (Maximum Score of 1)

## RECOMMENDATIONS

# Management of Symptoms



# COMMON CAUSES



# FIRST AID FOR NOSE BLEEDS



**Sit straight up with the head slightly tilted forward.**



**Pinch the bridge of the nose using firm continuous pressure for 10-20 minutes.**

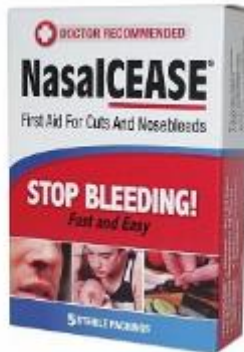


**Apply a cold pack to the bridge of the nose.**



**If the bleeding has not stopped after 20 minutes, administer medication if prescribed and notify your doctor**

# PREVENTION AND TREATMENT



# ANTIFIBRINOLYTIC THERAPY

**Amicar (aminocaproic acid) 4x per day for 7 days**

**Lysteda (tranexamic acid) 3x per day for 5 days**

- **Keeps clots in place (mucosal bleeding)**
  - Mouth bleeds
  - Nose bleeds
  - GI bleeding
  - Menstrual bleeding

**\*\*Contraindicated in patients with hematuria**

# INJURIES



# HEAD INJURIES

- **Wear protective equipment when riding bikes or scooters and when playing sports.**
- **If a traumatic head injury occurs, treatment may be necessary**
- **Transport to hospital for imaging such as a CT SCAN to check for intracranial bleeding**
- **Identifying expecting mother's carrier status for hemophilia or other bleeding disorders is crucial to avoid unintentional head trauma to the newborn**



# PREVENTION IS KEY!!



# PROTECTION FROM INJURIES



# HEAVY MENSES

- **According to The National Institute for Health and Care Excellence in the U.K.,**

**“Heavy menstrual bleeding is defined as excessive blood loss that interferes with a women’s physical, social, emotional and/or quality of life. It can occur alone or in combination with other symptoms.”**

# CHARACTERISTICS OF HEAVY MENSES




- **Bleeding for more than 7 days**
- **Blood loss (>80 ml ) Bleeding that soaks through one or more tampons or pads in a row for several hours**
- **Passage of clots > 1 inch in diameter (Quarter size)**
- **Flooding, staining of clothes and bedding**

# HEAVY MENSES




- Tracking/keeping a log or calendar

*Pictorial Blood Loss Assessment Chart (PBAC)*  
Menstrual chart and scoring system

Date of start    Score   
day month year

Towel	1	2	3	4	5	6	7	8
								
								
								
Clots/looding Clots: size								

Tampon	1	2	3	4	5	6	7	8
								
								
								
Clots/looding Clots: size								

## Scoring System

### Towels

1 point for each lightly stained towel

5 points for each moderately soiled towel

20 points if the towel is completely saturated with blood

### Tampons

1 point for each lightly stained tampon

5 points for each moderately soiled tampon

10 points if the tampon is completely saturated with blood

### Clots

1 point for small clots

5 points for large clots

Source: U.K. Haemophilia Society, A Guide For Women Living with von Willebrand's

# SISTERHOOD

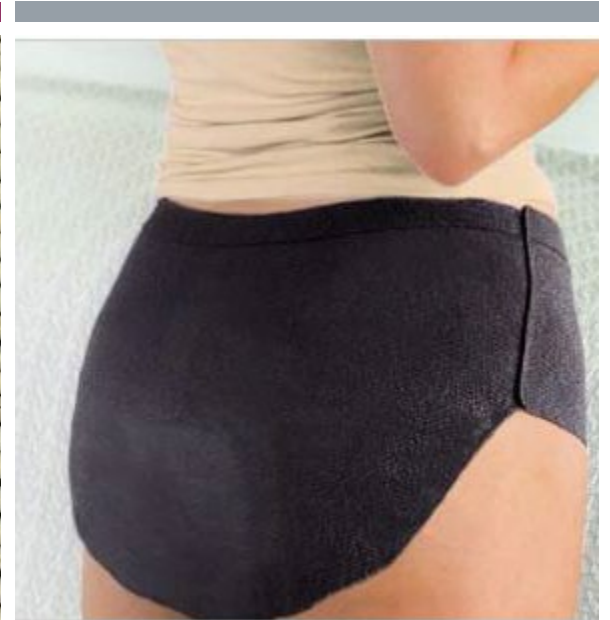


# CHALLENGES

- Difficulty in obtaining a PBAC score
  - Variety of disposable products with a range of absorbency capability
  - New Eco-friendly products
    - Reusable pads
    - Period underwear
- Patient resistance or forgetfulness to share information about their periods



# CHALLENGES





# THE MENSTRUAL CUP



# IRON DEFICIENCY ANEMIA

## Signs and symptoms

- Weakness and fatigue
- Poor concentration
- Pallor
- Headaches
- Dizziness
- Fast heart rate
- Shortness of breath
- Pica
- Restless leg syndrome

# IRON DEFICIENCY MANAGEMENT

- **Iron therapy**
  - Orally (Once daily dosing to QOD dosing)
  - Intravenously (Iron sucrose IV weekly x 4 doses)
- **Correct or manage the root cause**
- **Education**

# IRON-RICH FOODS

- Eat iron rich diet
- Take prescribed iron supplement with orange juice or other juice that contains Vitamin C
- Avoid taking iron with milk or dairy (one hour prior or two hours after)

ARTICHOKES



EGG YOLK



SCALLOPS



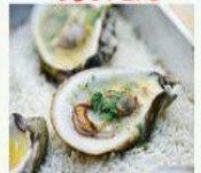
RED MEAT



DARK GREENS



OYSTERS



DRIED FRUIT



LEGUMES



# PHARMACOLOGICAL MANAGEMENT

- **Antifibrinolytic therapy (oral)**
  - Tranexamic Acid or Aminocaproic Acid
- **Desmopressin Acetate (DDAVP<sup>®</sup>)**
  - IV
- **Factor concentrates (IV)**
- **Hormonal Therapy**
  - IUD (Mirena, Liletta)
  - Implanted (Nexplanon)
  - Oral (many combinations)

# DDAVP

## Desmopressin Acetate

- Synthetic vasopressin analogue that causes the release of von Willebrand Factor from endothelium.
- Makes platelets stick better to the vessel wall to aid in clot formation
- Usually effective for treatment of bleeding in mild Hemophilia A, vWD Type 1 and some Type 2

## Trial is Recommended

## Weight based dosing

- 0.3 mcg/kg, max dose of 20 mcg

# SIDE EFFECTS



**Flushing**



**Nasal congestion**



**Headaches**



**Mild abdominal cramping**



**Low sodium due to water retention**

Fluid restriction necessary for 24 hours\*  
Low sodium can cause **seizures**

# FLUID RESTRICTION INSTRUCTIONS

- **Only drink electrolyte containing fluids for 24 hours post DDAVP administration**
  - Gatorade, juices, milk
  - Only drink to quench thirst
  - Signs of low sodium are nausea and confusion

**\*Call provider if unable to urinate for more than 8 hours**

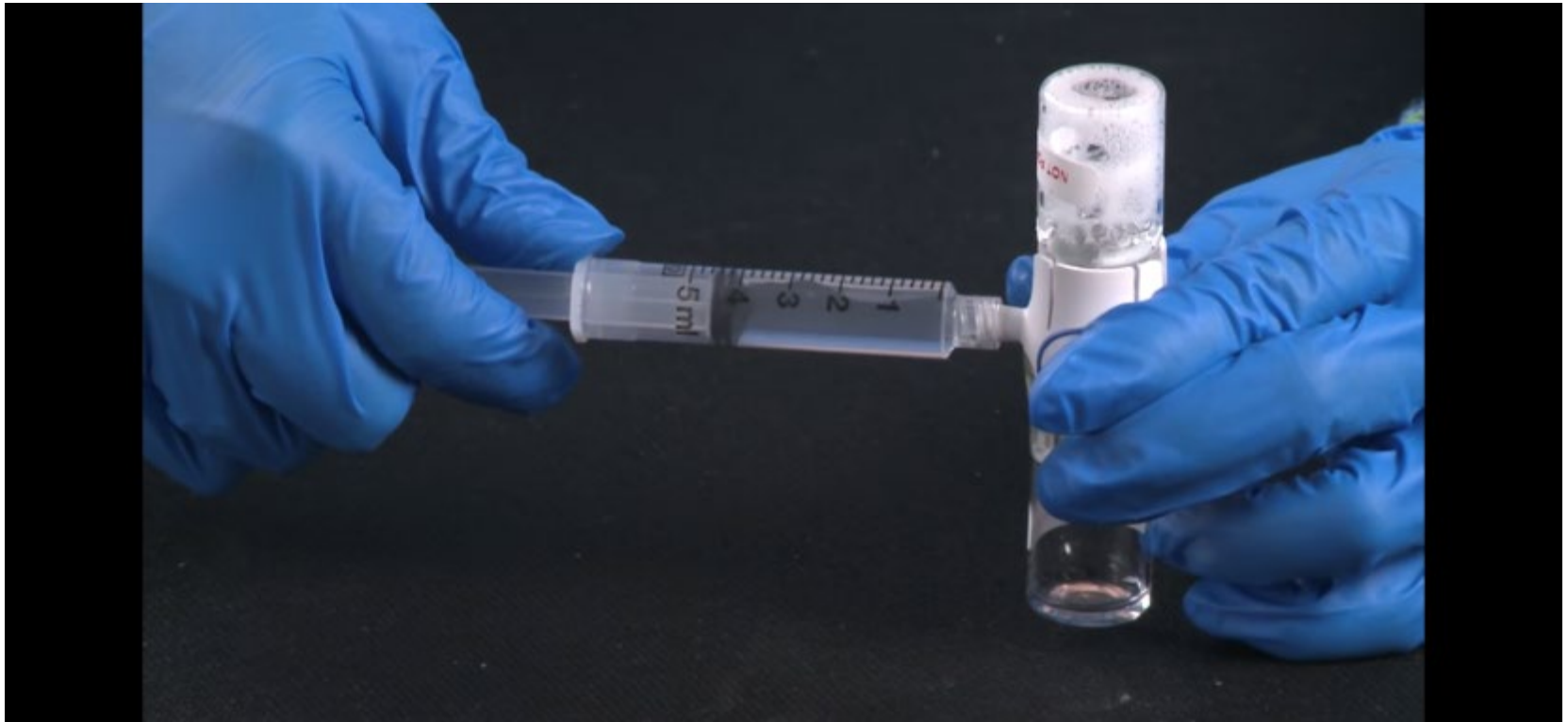


# FACTOR REPLACEMENT

- Replaces missing factor protein
- Treatment for bleeding, after serious trauma or prior to surgical procedures
- Given by intravenous infusion
- Weight based dosing
- Trial is necessary



# FACTOR CONCENTRATES



# FACTOR REPLACEMENT THERAPY

- **Diagnoses treated with Factor concentrates**
  - Factor deficiencies I,VII,VIII, IX, X, XIII
  - Von Willebrand Disease
  - Bernard-Soulier Syndrome
  - Glanzmann's thrombasthenia

## CASE STUDY

- “Luisa” is a 12 y/o girl, with von Willebrand Disease type 2A who presents with heavy vaginal bleeding on the fourth day of menarche.

## CASE STUDY

- She has a history of frequent epistaxis with chronic iron deficiency anemia. She was diagnosed with vWD at age 4 after presenting to the ED with a hemoglobin of 5gm/dl secondary to reoccurring epistaxis for an entire month. The mother and paternal grandfather also have vWD. Luisa was treated with vWF concentrate, PRBC transfusion, oral iron therapy and Amicar.
- She established care with the HTC shortly after ED visit.

## CASE STUDY

- Despite preventative measures, DDAVP and courses of Amicar, she continued to have episodic epistaxis and chronic anemia due to blood loss. By 9 years of age, she was started factor replacement therapy 2-3 times/ week. She also saw an allergist and completed iron therapy.
- Her mother eventually became independent with IV access and continued prophylaxis therapy for approximately 2 years. Luisa improved and eventually stopped prophylaxis.

## CASE STUDY

- Luisa started menarche on 8/28. She woke up during the night with soaked undergarments. She continued to change fully saturated pads every 1 ½ -2 hours. Her mother administered vWF concentrate daily x2 days and started tranexamic acid. By the third day she was experiencing dizziness and headaches. She was taken to the ED. Her hemoglobin was 7.5 gm/dl. Oral Fe was started. She was given an appt. the next day.

## CASE STUDY

- She came to the HTC the following day. Careful history, physical exam and PBAC scores were reviewed. She also reported having epistaxis. Advised to restart prophylaxis factor infusions, complete course of tranexamic acid and take iron as prescribed.



# PBAC SCORES

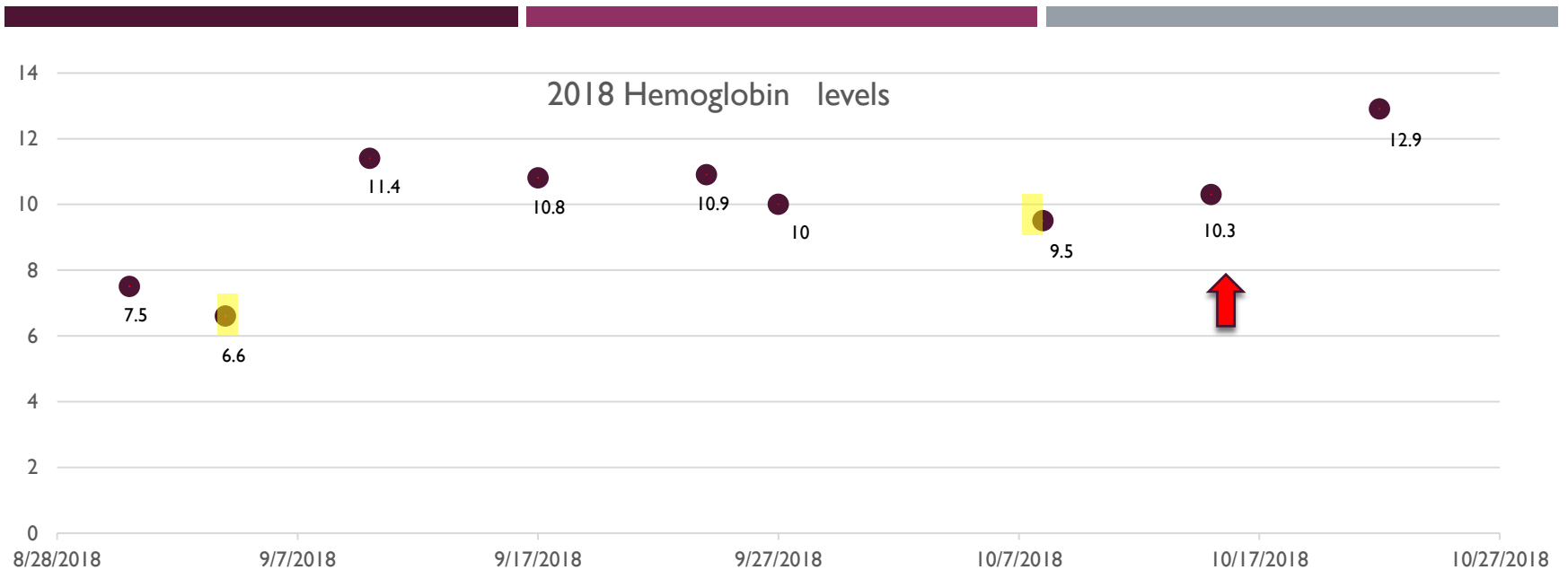
Day 1: >240 vWF concentrate daily x2, Tranexamic Acid 3x/d for 5 days and oral iron initiated

Day 2: 100

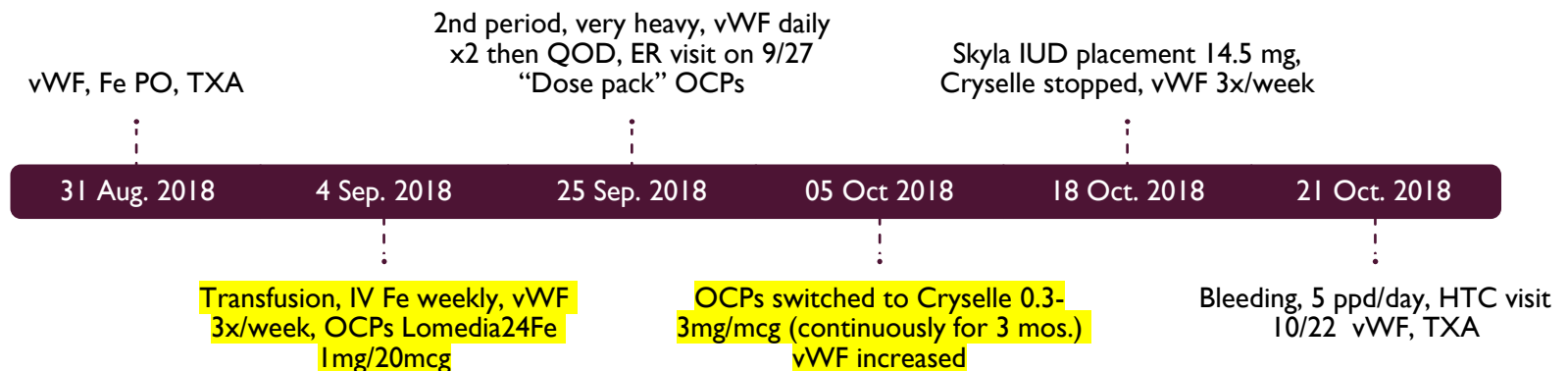
Day 3: 100 Hemoglobin 7.5 gm/dl

Day 4: Day of visit PBAC score was 35 before 12 pm \*\*By the ninth day, hemoglobin dropped further to 6.6gm/dl

- **PBAC score >100 requires evaluation**

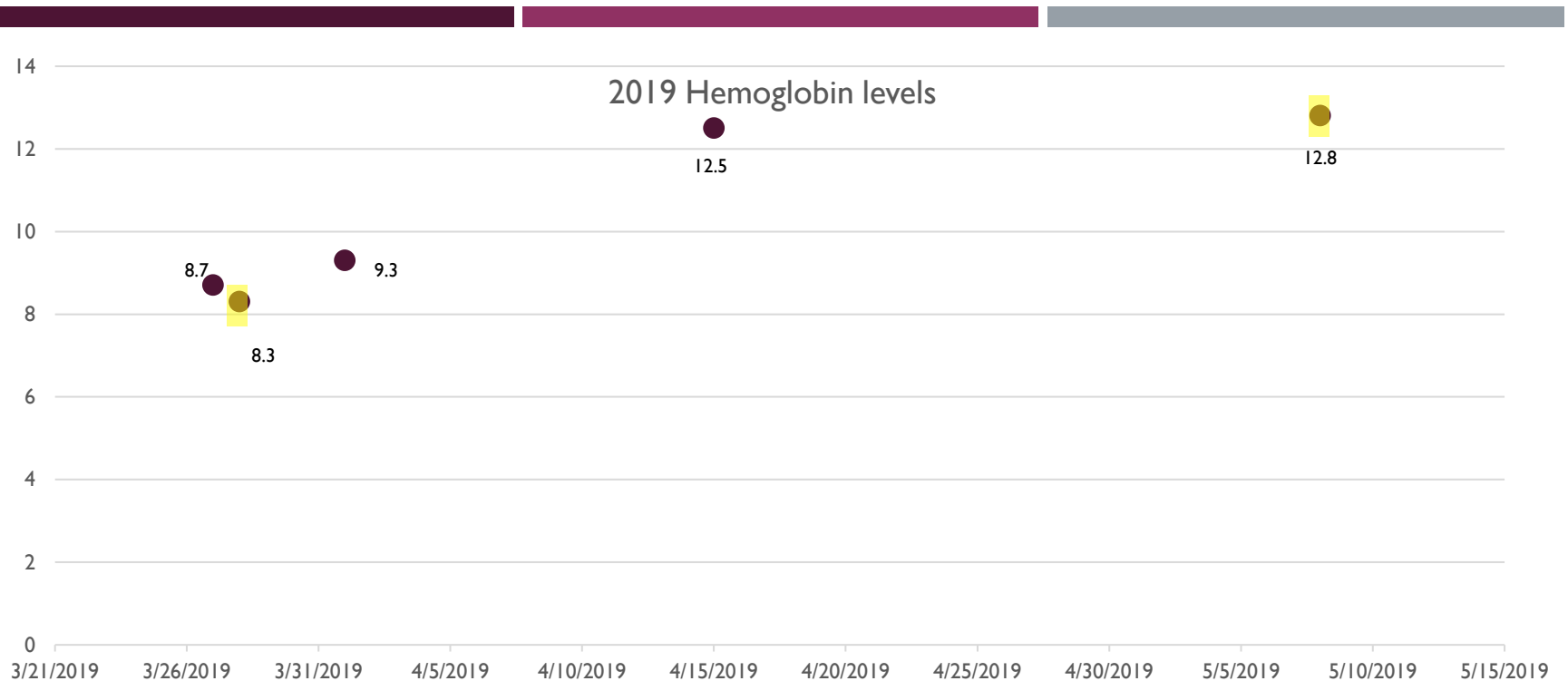


## INTERVENTIONS AND HIGHLIGHTS



## CASE STUDY: AFTER IUD PLACEMENT

- Vaginal bleeding stopped after nine days.
- Periods became “very light,” only lasting 4-5 days for the next 4 months (November through February)
- No medications administered during this time.
- On the 5<sup>th</sup> month, she had a very heavy period that began on 3/4 and ended on 3/27. She was also having episodes of epistaxis. Hemoglobin dropped to 8.3 gm/dl on 3/28.



## INTERVENTIONS AND HIGHLIGHTS

HTC visit for heavy period that began on 3/4, IV Fe, vWF, TXA initiated on 3/23

Called HTC, 1st day of heavy period, daily vWFX3, improved by 4/17



ED visit for symptomatic anemia, Loestrin 2 tabs until bleeding stops, vWF QOD

\*\*\*Weekly Fe infusions until 4/15

Skyla (noted in endocervical canal with distal end protruding through lateral aspect of the cervix) was replaced with Mirena 52mg IUD

## CASE STUDY: AFTER SECOND IUD PLACEMENT

- Skyla was displaced, noted in the endocervical canal with distal end protruding through lateral aspect of the cervix. After removal, Mirena IUD was inserted.
- Luisa is now 16 years old. She has occasional spotting since Mirena placement. She is active in sports and is no longer anemic. She still has episodic nosebleeds which she manages with tranexamic acid and vWF concentrate when needed.

## References

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