

## Abstract

- Women with Severe Von Willebrand disease (VWD), including women with Type 3 VWD, Severe Type 1/1C VWD (VWF ristocetin cofactor and antigen <10%), and Severe Type 2A (VWF ristocetin cofactor and antigen <10%) are at high risk for bleeding-related complications, including gynecological and obstetrical bleeding.
- The prevalence of gynecological and obstetrical complications in this population is unknown, as are optimum management strategies.

## Introduction

- VWD Connect Annual meeting is the largest gathering in North America of individuals with Severe VWD and their caregivers.
- Based on feedback from previous meetings, at the 2019 VWD Connect Meeting, an educational half-day Period Symposium was held to discuss management of gynecologic issues in women with bleeding disorders.
- Each participant was assigned an Audience Response System (ARS) for the duration of the meeting.

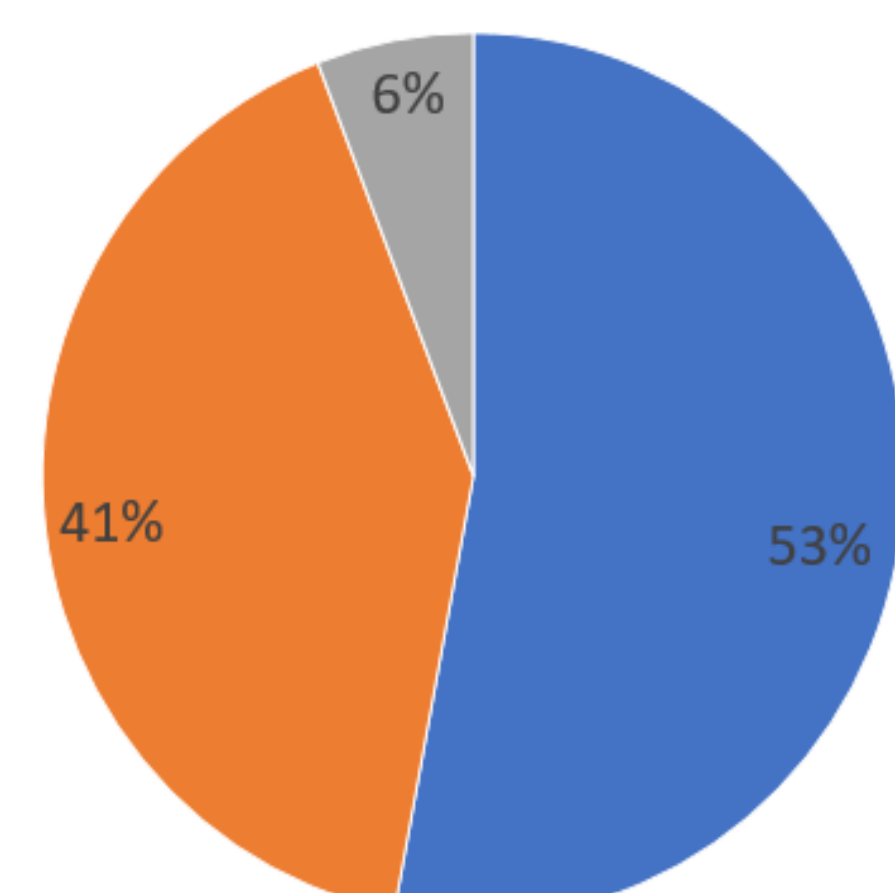
## Methods and Materials

- During the Period Symposium, participants were surveyed and their responses were recorded.
- An ARS was given to participants with bleeding disorder or their caregiver in the case of minors

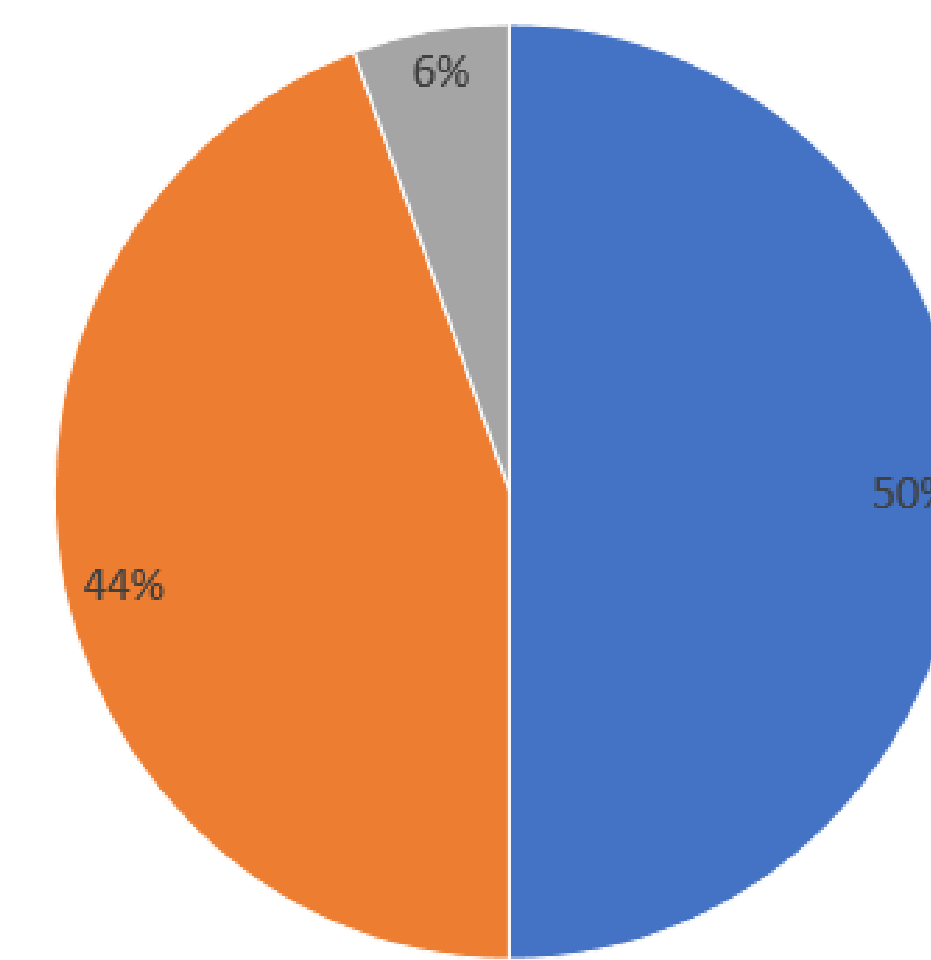
## Demographics

- 22 patients and/or caregivers participated in the period symposium; 21 had demographic information. Participants were 100% female.
- 38% of participants were patients or caregivers of patients under 18, 14% were 18-29 years, 38% 30-45 years, 9.5% over age 45.
- Diagnosis of participants was
  - Type 3 VWD 15 (31%)
  - Type 1/1C VWD 1 (5%)
  - Type 2A VWD: 4 (19%)
  - Other VWD: 1 (5%)
- Caregivers, spouses, pre-menarchal and post-menopausal women were all invited to participate in the symposium which may reflect high number of "N/A" on some questions

% of patients who have required ED evaluation for management of heavy menses

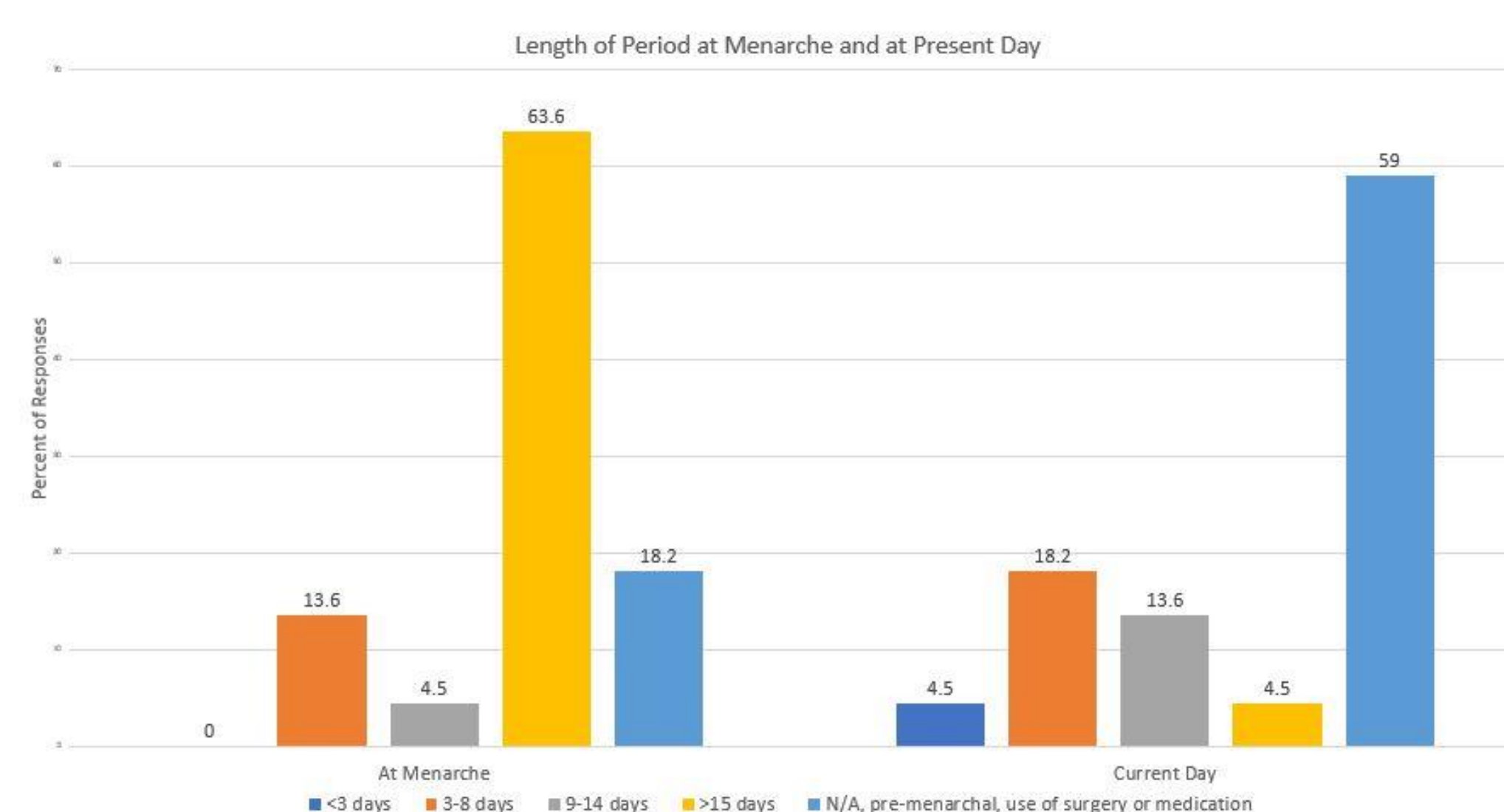


% of Patients Hospitalized due to Heavy Period

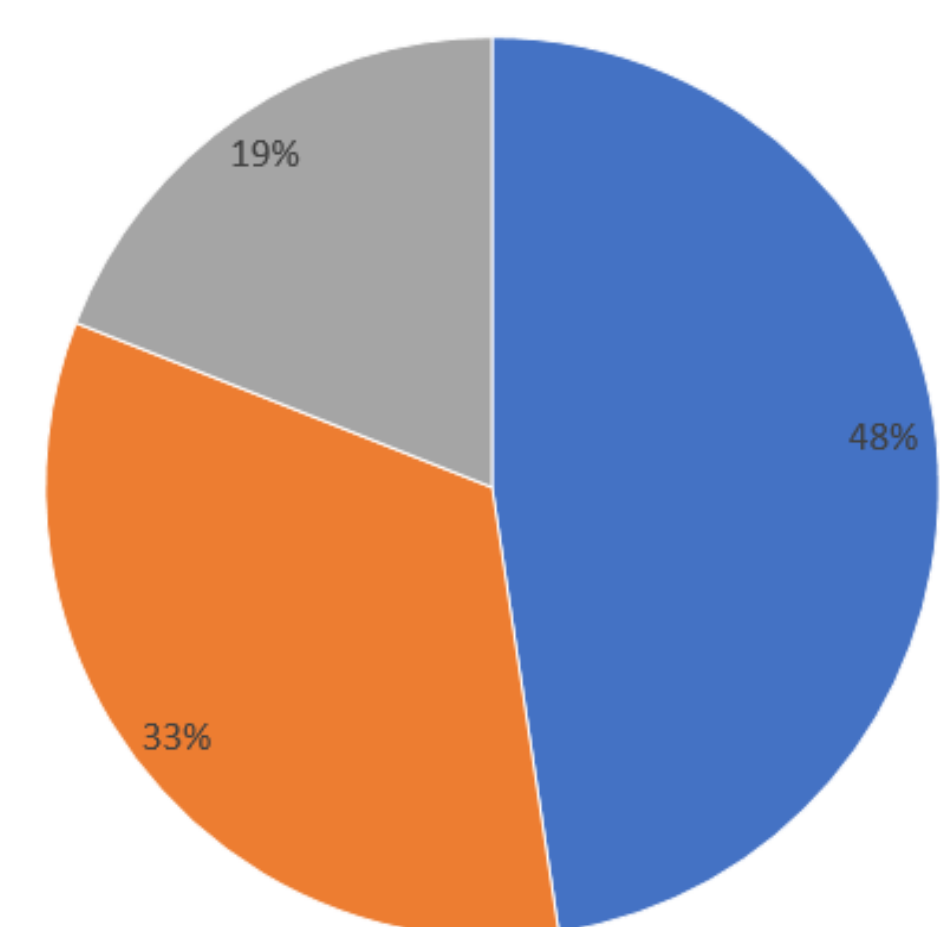


■ Yes ■ No ■ Unsure

■ Yes ■ No ■ Unsure

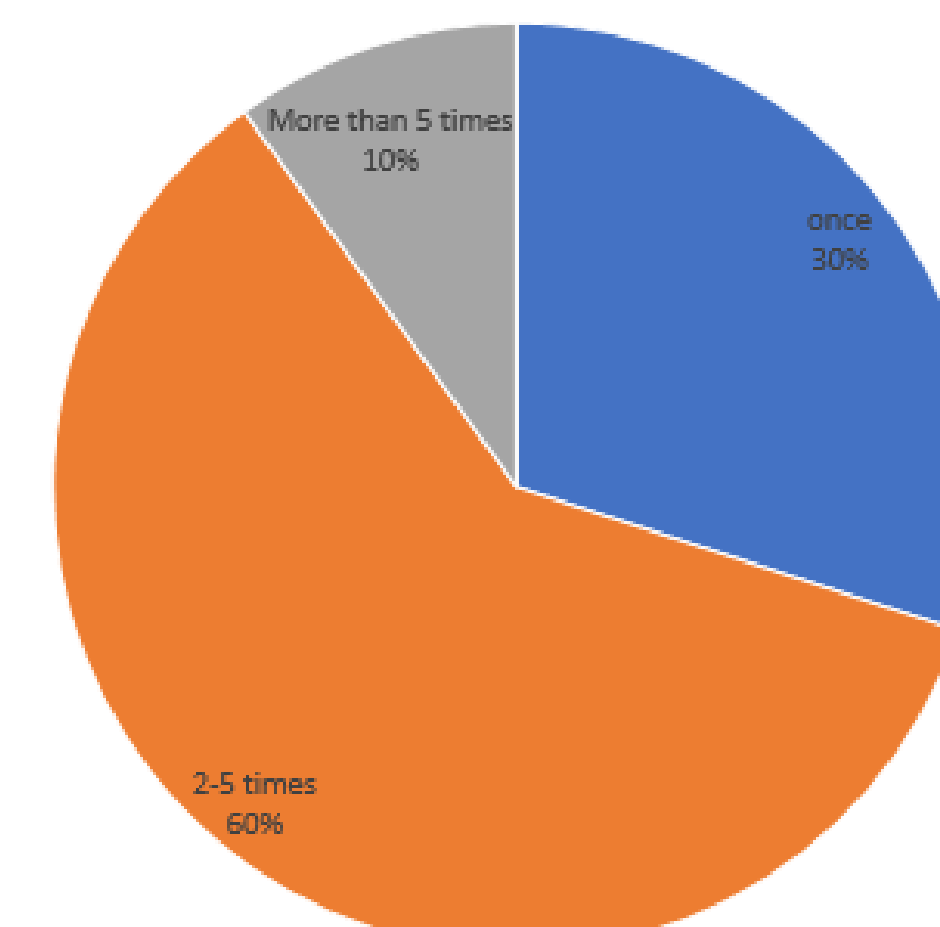


Blood Transfusion for Anemia Secondary to Menstrual Bleeding

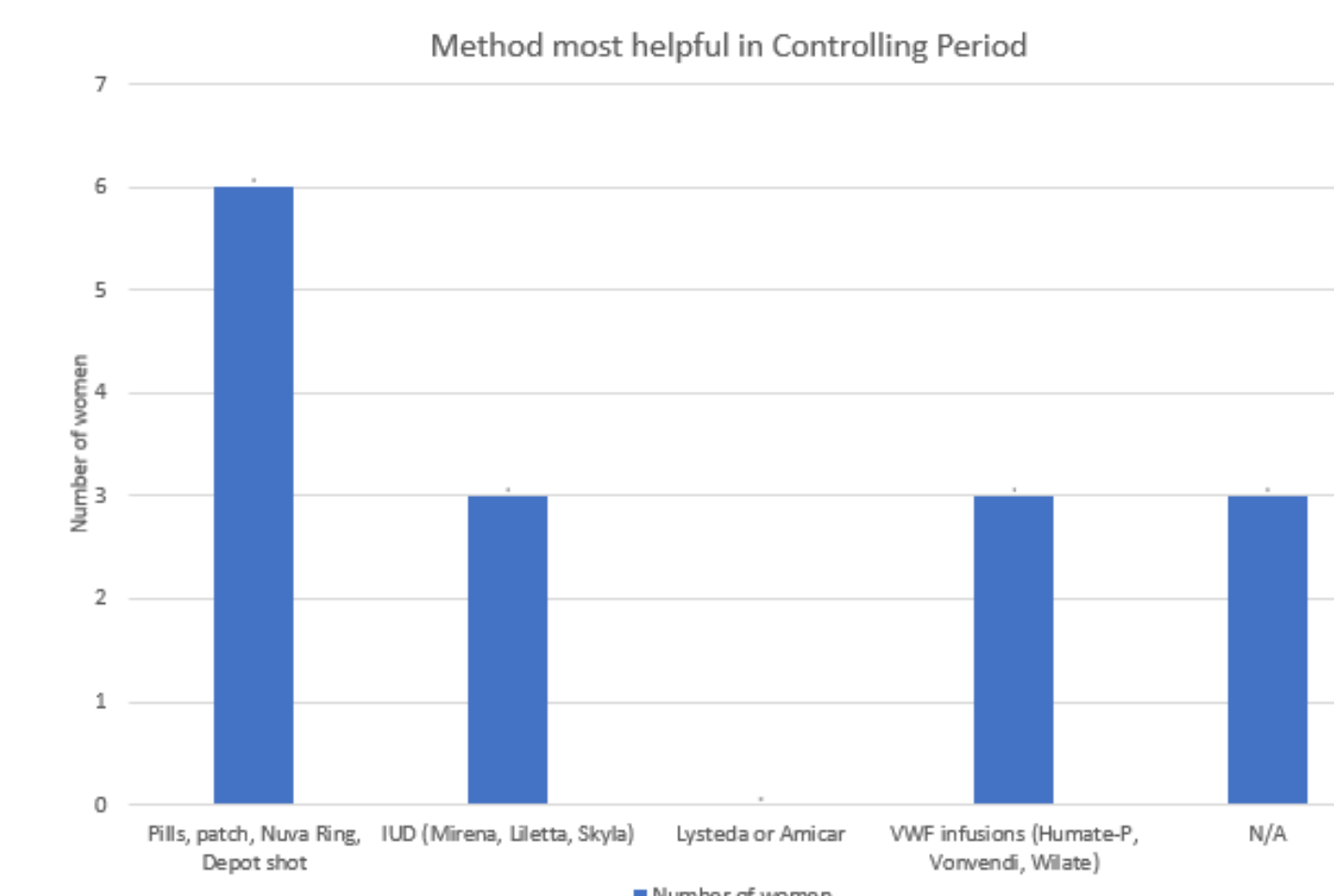
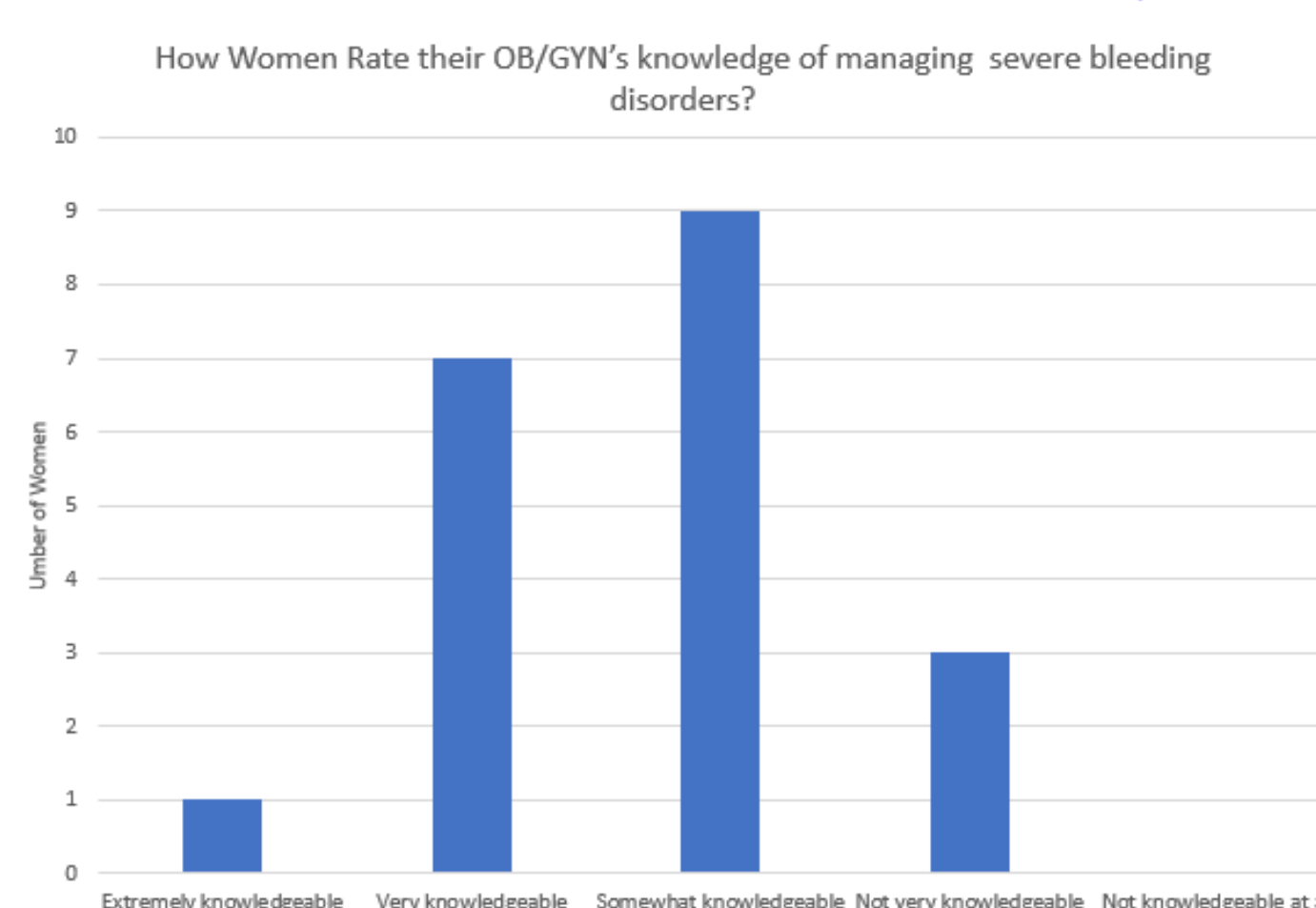
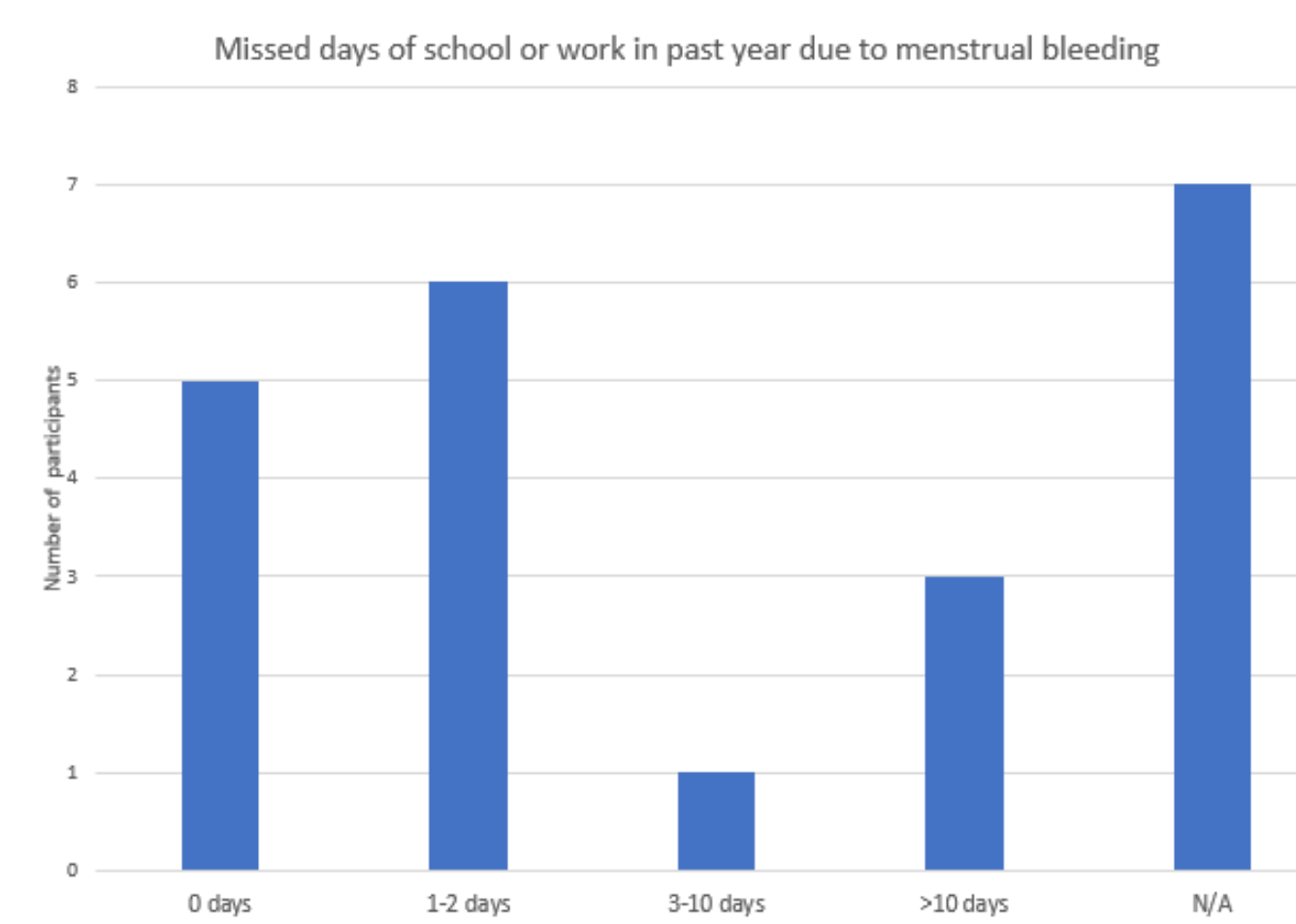


■ Yes ■ No ■ N/A

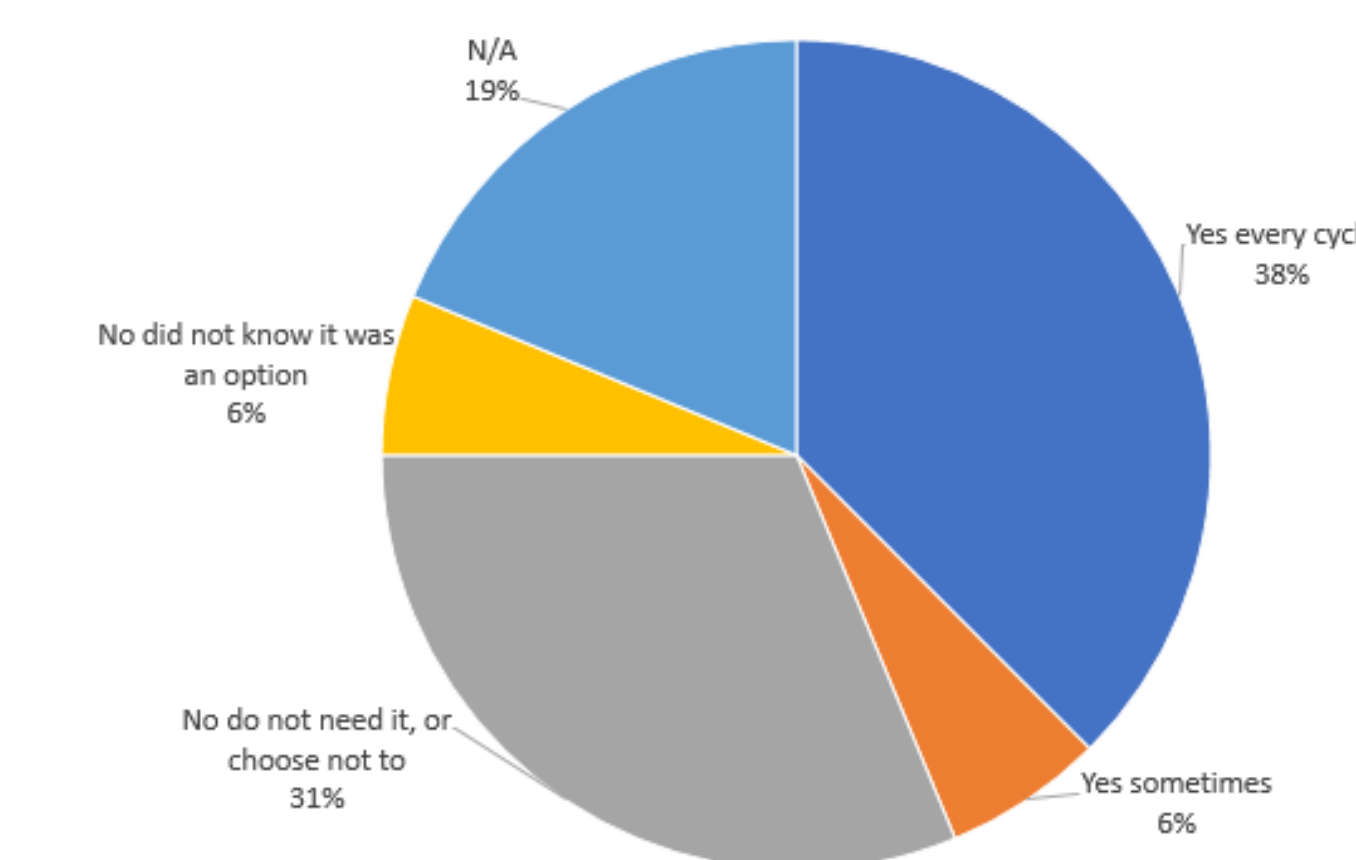
Number of Times Received a Blood Transfusion for Menstrual Bleeding



■ once ■ 2-5 times ■ More than 5 times



Current use of Factor Concentrates during Menses



## Additional Results

- 78% of respondents reported starting hormonal therapy within 2 years of starting their period.
- For women on oral hormonal contraception, 85% (n=7) reported taking as extended cycle (period every 3-6 months) compared to traditional method (3 weeks on/1 week off).
- Given the young age of this cohort, rates of surgical procedures to manage heavy menses were low including Hysterectomy (0%), Dilation and Curettage (n=1) or Ablation (n=1).
- Bleeding due to ovarian cyst rupture requiring medical treatment was common (44%, n=13).
- Iron deficiency anemia was common, with 38% of women treated with oral iron, 5% IV iron, and 33% of women had received both oral and IV iron.

## Discussion

- At menarche, 63% of women experienced significant and prolonged menses >15 days, and at the current time 18% of women still have menses lasting >9 days.
- Estrogen therapy is often employed as first line treatment, but variable methods including IV VWF concentrates have been used with success/patient satisfaction, highlighting patient variability.
- Extended cycle combined oral contraceptives were commonly prescribed.
- Menstrual bleeding continues to have a negative impact on school and work functioning.
- Other challenges identified include improving access to knowledgeable OB/GYN providers with expertise in management of bleeding disorders.

## Conclusions

- Women with Severe VWD have significant morbidity from menstrual bleeding including high rates of hospitalization, high rates of blood transfusion, and hemorrhagic ovarian cyst rupture requiring medical treatment.
- Ongoing prospective studies with a larger cohort are needed to better understand treatment effectiveness, impact on quality of life and inform practice guidelines.
- Women should consider establishing care with a gynecologist prior to menarche to help prepare for menarche and to decrease the negative impact on school/work.

The authors have no conflicts of interest to disclose