Follow-up of congenital dysfibrinogenemia

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Casini A, Blondon M, Lebreton A, Koegel J, Tintillier V, de Maistre E, Gautier P, Biron C, Neerman-Arbez M, de Moerloose P. Natural history of patients with congenital dysfibrinogenemia. *Blood.* 2015;125(3):553-561.

1. Your patient is a 30-year-old woman with congenital dysfibrinogenemia (CD). According to the multicenter study by Dr Casini et al, which of the following statements about complications of major bleeding and thrombosis in persons with CD and their affected relatives is correct?

- ☐ At the time of diagnosis, ~40% had experienced major bleeding
- ☐ At the time of diagnosis, ~40% had experienced thrombotic events
- ☐ Fatality rate of major bleeding episodes was 15%
- ☐ At age 50 years, the estimated cumulative incidence was 19.2% for major bleeding and 30.1% for thrombotic events

2. According to the multicenter study by Dr Casini et al, which of the following statements about complications of pregnancy and surgery in persons with CD and their affected relatives is correct?

- ☐ More than half of bleeding episodes in this series were postsurgical bleeding
- ☐ ~10% of pregnancies ended in spontaneous abortion
- ☐ ~10% of pregnancies were complicated by postpartum hemorrhage (PPH)
- ☐ Among 137 surgical procedures analyzed, 9 (6.5%) were complicated by abnormal bleeding

3. According to the multicenter study by Dr Casini et al, which of the following clinical, laboratory, and genetic factors would *most likely* affect the risk for thrombotic or bleeding outcomes in persons with CD and their affected relatives?

- ☐ Having a previously identified bleeding phenotype
- ☐ Hotspot mutations
- ☐ Fibrinogen levels
- ☐ Activity:antigen ratios

Activity Evaluation (where 1 is strongly disagree and 5 is strongly agree)

1. The activity supported the learning objectives.
   1 2 3 4 5
2. The material was organized clearly for learning to occur.
   1 2 3 4 5
3. The content learned from this activity will impact my practice.
   1 2 3 4 5
4. The activity was presented objectively and free of commercial bias.
   1 2 3 4 5