

48. Behcet's disease and blood clots

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Q: "I have Behcet's syndrome. I almost died at the age of 37 from multiple PE's. In the 3 months before the diagnosis I had experienced shortness of breath. I was taking birth control pills and was flying frequently within the US (under 4 hours per trip) and occasionally to Europe. My question is whether I have an additional risk factor for clotting due to Behcet's syndrome. My doctors are not in agreement as to whether I should remain on coumadin® "for life". Some feel the birth control pills were the primary cause, others are concerned about the role Behcet's may have played. I am struggling with the decision whether I should come off coumadin or not. Can you provide a perspective of clotting risk for Behcet's patients?"

A: A decision on length of warfarin treatment is indeed very difficult in this patient, because no clinical studies exist that tell us what the risk of a second clot is in patients with Behcet's disease, once warfarin is stopped.

For a full risk assessment of the above patient a physician needs to know whether:

- a. the PEs occurred at a time when the Behcet's disease was active,
- b. what thrombophilic laboratory abnormalities were examined and found,
- c. whether there is a family history of clots, and
- d. whether the patient is overweight or smokes (see [Q/A 9](#)),
- e. whether the patient has residual pulmonary hypertension (measured by cardiac echo).

The 3 treatment options are:

1. discontinuation of warfarin now,
2. indefinite treatment with low-dose warfarin, target INR 1.5-2.0,
3. indefinite treatment with full-dose warfarin, target INR 2.0-3.0,

I recommend evaluation of the patient by a physician in a Thrombophilia Center (see [Q/A 37](#)). An argument for a lower risk of recurrence may be that the previous PE was triggered by birth control pill. Furthermore, if the PE occurred at a time when the Behcet was active the risk of recurrence may be lower if the Behcet is now inactive. If the patient now has pulmonary hypertension one may want to choose indefinite warfarin. Also, a D-dimer could be obtained and factored into the decision making (also see [Q/A 19](#)).

Behcet's disease is a rare inflammatory disease of unknown cause with a variety of organ involvements, most noticeably mouth and genital ulcers, and eye. Ten to 25 % of patients with Behcet's disease develop thrombosis, most commonly superficial thrombophlebitis, but also deep vein thrombosis. Men seem to develop clots more frequently than women. It is poorly understood why patients have an increased risk for clots. However, it appears that patients with Behcet's disease who also have factor V Leiden are at particularly increased risk for clots [J Rheumatol 1997;24:2196-8; Br J Rheumatol 1996;35:1178-80]. Unfortunately, it has not been studied how long patients with Behcet's disease who have had one blood clot should stay on warfarin, and what their risk of another blood clot is, once they come off warfarin. One could speculate that the risk for clots is highest when the disease is active; thus, as long as the disease is active one would continue warfarin therapy. One could then also argue that the risk of recurrence decreases if the disease is well controlled; thus, one could then consider discontinuation of warfarin. Clearly, the risk of recurrence and the risk factors leading to recurrence in patients with Behcet's disease need to be studied in a large, multicenter trial.

References:

1. J Rheumatol 1997;24:2196-8
2. Br J Rheumatol 1996;35:1178-80