Indications for thrombophilia and antiphospholipid syndrome testing in VENOUS Thrombosis

NOT INDICATED Testing will not alter management	**DISCUSS WITH HAEMATOLOGY** Testing in selected cases may alter management	**INDICATED** Thrombophilia testing is likely to alter management
Unselected patients with acute VTE	**Unprovoked VTE, or associated with weak environmental risk factors and all the following:** • **age < 40 years** • **At least one first-degree relative similarly affected** • **Testing will alter management** *Consider hereditary thrombophilia testing (PC, PS, AT) +/- R97 gene panel*	**Unprovoked VTE, or associated with weak environmental risk factors** *Request Antiphospholipid syndrome screening** *Request PNH testing if anaemic or evidence of haemolysis* *Request MPN panel if FBC suggestive of MPN*
Patients with provoked VTE	**Asymptomatic first-degree relatives of people with protein C, protein S or antithrombin deficiency** *Consider selective testing if this will influence management and life choices, depending on personal circumstances*	**Venous thrombosis at unusual site (e.g. CVST, splanchnic vein thrombosis), either unprovoked or associated with weak environmental risk factors** *Request antiphospholipid syndrome screening** *Request PNH testing if anaemic or evidence of haemolysis* *Request MPN panel if FBC suggestive of MPN, or in patients with unprovoked splanchnic vein thrombosis or CVST*
Patients with central venous catheter-related VTE	**Retinal vein occlusion without any other risk factors (e.g. diabetes, hypertension, hypercholesterolaemia)** *Consider antiphospholipid syndrome screening**	**Suspected antiphospholipid syndrome**** *Request Antiphospholipid syndrome screening**
Case finding asymptomatic relatives with low-risk thrombophilia *e.g. factor V Leiden and prothrombin gene mutation*		**Children with purpura fulminans**
Prior to prescribing combined oral contraceptive or hormone replacement therapy in patients with a first-degree relative with a history of VTE *Women should be offered an alternative method of contraception regardless of identification of a thrombophilia tendency*		
Asymptomatic first-degree relatives of people with a history of VTE		

		Urgent testing for protein C and S deficiency **Skin necrosis in associated with warfarin** *Test for protein C and S deficiency after warfarin is discontinued for 4 weeks*

* Antiphospholipid screening: Request anti-cardiolipin and anti-B2 glucoprotein-1 antibody testing. If positive, repeat in 12 weeks and consider testing for lupus anticoagulant after holding anticoagulation. If anticoagulation can't be held, discuss options with haematology (which sending samples to Oxford for DOAC-stop).

** Indications for antiphospholipid syndrome testing:

- Unprovoked VTE or minor provoking factors and VTE at unusual sites such as splanchnic vein thrombosis (which includes portal vein, mesenteric vein and splenic vein thrombosis, and the Budd–Chiari syndrome) and cerebral venous sinus thrombosis without clear risk factors
- Arterial thrombosis in patients <50 years of age without clear risk factors
- History of systemic lupus erythematosus (SLE) or other autoimmune disease developing thrombosis or pregnancy complications
- Unexplained microvascular thrombosis
- Presence of livedo reticularis/livedoid vasculopathy
- Unexplained prolonged PT or APTT prior to starting anticoagulation
- Recurrent thrombosis despite therapeutic anticoagulation not explained by non-adherence or other clear risk factors
- Thrombocytopenia
- Recurrent miscarriages/stillbirths/severe pre-eclampsia or evidence of placental insufficiency <34 weeks of onset
- Cardiac valve abnormalities in the absence of other explanation

Indications for thrombophilia and antiphospholipid syndrome testing in ARTERIAL Thrombosis

Testing for heritable thrombophilia is not recommended for patients with arterial thrombosis.

NOT INDICATED Testing will not alter management	**DISCUSS WITH HAEMATOLOGY** Testing in selected cases may alter management	**INDICATED** Thrombophilia testing is likely to alter management
Unselected patients with isolated arterial thrombosis	**Concurrent anaemia or evidence of haemolysis** *Consider PNH testing* **FBC suggestive of MPN** *Request MPN panel*	**Arterial thrombosis in patients <50 years of age in the absence of other vascular risk factors** *Request antiphospholipid syndrome screening** **Cryptogenic ischaemia stroke in patients <60 years of age** *Request antiphospholipid syndrome screening** **Suspected antiphospholipid syndrome**** *Request Antiphospholipid syndrome screening**

* Antiphospholipid screening: Request anti-cardiolipin and anti-B2 glucoprotein-1 antibody testing. If positive, repeat in 12 weeks and consider testing for lupus anticoagulant after holding anticoagulation. If anticoagulation can't be held, discuss options with haematology (which sending samples to Oxford for DOAC-stop).

** Indications for antiphospholipid syndrome testing:

- Unprovoked VTE or minor provoking factors and VTE at unusual sites such as splanchnic vein thrombosis (which includes portal vein, mesenteric vein and splenic vein thrombosis, and the Budd–Chiari syndrome) and cerebral venous sinus thrombosis without clear risk factors
- Arterial thrombosis in patients <50 years of age without clear risk factors
- History of systemic lupus erythematosus (SLE) or other autoimmune disease developing thrombosis or pregnancy complications
- Unexplained microvascular thrombosis
- Presence of livedo reticularis/livedoid vasculopathy
- Unexplained prolonged PT or APTT prior to starting anticoagulation
- Recurrent thrombosis despite therapeutic anticoagulation not explained by non-adherence or other clear risk factors
- Thrombocytopenia
- Recurrent miscarriages/stillbirths/severe pre-eclampsia or evidence of placental insufficiency <34 weeks of onset
- Cardiac valve abnormalities in the absence of other explanation

Indications for thrombophilia and antiphospholipid syndrome testing in relation to pregnancy and pregnancy morbidity

Women should not be screened for thrombophilia unless the result will influence recommendations regarding antepartum and postpartum thromboprophylaxis.

NOT INDICATED	DISCUSS WITH HAEMATOLOGY	INDICATED
Women with a prior provoked VTE *Should be considered for thromboprophylaxis and hence testing not routinely required*	**Confirmed family history of antithrombin deficiency or evidence of heparin resistance** *Consider antithrombin testing*	**Women with a previous unprovoked VTE, or associated with weak environmental risk factors** *Request antiphospholipid syndrome screening* outside of pregnancy* *See guidance on thrombophilia testing in VENOUS thrombosis*
Women with a history of recurrent miscarriage or adverse pregnancy outcomes *Thrombophilia testing not recommended (consider antiphospholipid testing for recurrent or late pregnancy loss)*	**Asymptomatic women with a first degree relative with protein C, protein S or antithrombin deficiency** *Consider selected testing if it alters a decision regarding thromboprophylaxis*	**Women with recurrent miscarriages, stillbirths, severe pre-eclampsia or evidence of placental insufficiency <34 weeks of onset** *Request antiphospholipid syndrome screening***

* Antiphospholipid screening: Request anti-cardiolipin and anti-B2 glucoprotein-1 antibody testing. If positive, repeat in 12 weeks and consider testing for lupus anticoagulant after holding anticoagulation. If anticoagulation can't be held, discuss options with haematology (which sending samples to Oxford for DOAC-stop).

** Antiphospholipid screening in patients with pregnancy morbidity:

- 6 weeks postpartum: Request anti-cardiolipin, anti-B2 glucoprotein-1 antibody and lupus anticoagulant testing. Do not check lupus anticoagulant if taking anticoagulation.
- If positive, repeat in 12 weeks
- If anticoagulated, consider testing for lupus anticoagulant after holding anticoagulation or switching to LMWH