

## **Factor V Leiden (FVL) / Activated Protein C (APC) Resistance And Blood Clotting**

### **What is normal clotting?**

A blood clot (also call a thrombus or thrombosis) is one of our natural defenses against bleeding. Normally, blood is maintained in a fluid state in the body because of a delicate balance between proteins that make clots (also called coagulants or factors) and proteins that prevent clots (also called anticoagulants). Blood clotting occurs when there is damage to the blood vessel. 1<sup>st</sup>, the platelets, which are tiny blood cells like sticky jigsaw puzzle pieces, plug the injured vessel. At the same time they release chemicals that start the clotting process. The result is like a domino effect that eventually leads to the formation of a blood clot. Whenever a blood clot is formed, anticoagulants are also activated to prevent excess clot formation and start the clot breakdown process. It is a delicate balancing act between bleeding and clotting.

### **What happens when a clot is formed?**

Clots can occur in veins (venous) or arteries (arterial). Arteries are tougher, elasticized blood vessels that carry blood away from the heart and deliver oxygen to the body. Arterial blood clots may cause stroke, heart attack, or damage to limbs or other organs. Veins are thinner, collapsible blood vessels that carry blood back from the tissues to the heart. Venous blood clots are often associated with surgery, pregnancy, birth control pills, IV lines or other external factors. Other common risks of thrombosis include cigarette smoking, high blood pressure, high cholesterol, and diabetes.

### **What is Factor V Leiden and APC resistance?**

APC is a blood protein involved in the prevention of clotting. APC normally breaks down activated clotting proteins (factors V and VIII) which slow down clotting. Individuals with activated protein C resistance have an altered (mutated) factor V so that it is resistant to breakdown by APC and increasing the risk of clotting. More than 95% of people with APC resistance have the Leiden mutation. People with the factor V Leiden mutation have blood that has an increased tendency to clot. APC resistance is the most common abnormality associated with venous thrombosis.

### **How do I get Factor V Leiden?**

Factor V Leiden is not a disease; it is the presence of a specific gene that is passed on to you from your parents (inherited). As with all inherited diseases we inherit one gene from our mother and one from our father and so it is possible to inherit two normal genes or one Factor V Leiden gene and one normal or two factor V Leiden genes. Having one Factor V Leiden gene (heterozygous) will result in a slightly higher risk of developing a thrombosis (3-8 fold) than people who do not have the gene, but having two genes (homozygous) makes the risk much greater (80-140 fold). There is further risk of thrombosis if you have a second risk factor such as oral birth control pills, estrogen therapy, pregnancy, immobilization, surgery, infection, increased age, protein C deficiency or other coagulation abnormalities.

### **How do I know if I have Factor V Leiden?**

FVL is a silent abnormality. The only way to know if you have Factor V Leiden is by a blood test. The test will determine the presence or absence of the mutation and distinguishes between the heterozygous and homozygous type. The test can be performed with results in 5-7 days.

### **What are the signs of Factor V Leiden?**

There are no specific symptoms of Factor V Leiden unless you have a clot. The most common sites for a clot is in the legs (deep vein thrombosis - DVT) and in the chest (pulmonary embolism - PE). Clinical problems associated with Factor V Leiden can be extremely unpredictable. Many people with Factor V Leiden never develop thrombosis; yet some have recurrent clotting before the age of 30. The diagnosis is suspected in patients with a history of thrombosis and in those with a family history.

### **How do I know if I should be tested?**

People should be tested if they:

- Have a family history of clotting or thrombophilia
- Have a history themselves of clotting
- Have a history of transient ischemic attacks or premature stroke
- Have a history of repeated miscarriages or stillbirths

### **How do I get treated if I have Factor V Leiden?**

You should be tested for other possible inherited or acquired clotting disorders. Treatment of FVL depends on a variety of conditions such as if you have other inherited or acquired clotting abnormalities. If you have only the abnormal factor V gene you do not need treatment unless your blood starts to clot. If you have or have had a blood clot and have the abnormal factor V gene, you will be put on anticoagulant therapy for a period of time ranging from 3 months to life. The length of time you are on anticoagulants depends on several issues such as; how many blood clots you have had, how serious your clot was and how many additional risk factors you may have. Protective anticoagulation therapy may be needed in times where your risk for developing a clot is greater such as pregnancy, surgery, or long plane/car rides. Discuss this with your Hematologist.

### **How do I keep from getting a clot?**

1. Avoid long periods of bed rest. Avoid prolonged sitting or standing in one position. Don't cross ankles or legs while sitting or lying. Keep your feet higher than your hips while sitting. While resting, occasionally move your legs, ankles and toes to promote circulation
2. On long car or plane rides, get up and move around for ~15 minutes at least every 2 hours
3. Do not smoke or be around others who are smoking
4. Exercise regularly and stay well hydrated
5. Use support stockings (such as JOBST compression stockings or TED compression stockings) if you have severe varicose veins or if you have had a clot in your legs
6. Avoid knee socks or hosiery that might limit blood flow. Avoid tight diapers around the legs in infants
7. Do not take Birth Control pills without discussing it with your Hematologist
8. If you become pregnant, consult your GYN and Hematologist about the Factor V Leiden
9. Tell the doctor or surgeon prior to having surgery about having the Factor V Leiden

**If you have any questions about your diagnosis, please contact KC Clevenger, Coagulation Nurse Practitioner at 303.861.6972 or the Hematology fellow on call after hours or on the weekend at 303.861.6740.**