Essential Thrombocythemia (ET)

WHAT YOU NEED TO KNOW

You or your loved one has been diagnosed with essential thrombocythemia (ET). What does it mean and how will it affect you?

This fact sheet will help you:

- Learn about ET and how it is diagnosed
- Get an overview of treatment options
- Understand what happens next
What is essential thrombocythemia (ET)?

ET is a type of myeloproliferative neoplasm (MPN). This group of blood cancers features too many blood cells of a particular type made in the bone marrow. MPNs begin with one or more changes to the DNA of a single stem cell in the bone marrow. These changes cause the stem cell to create more and more abnormal stem cells.

People with ET have too many platelets, a type of blood cell. Too many platelets can result in a blood clot (thrombosis) forming in a blood vessel. This can cause serious health problems such as a stroke, heart attack, or pulmonary embolism (blockage of a major blood vessel in the lung).

About ET
- Most cases of ET feature genetic mutations to a bone marrow stem cell.
- May mean that your body makes too many platelets (blood clotting cells).
- Occurs most often in adults and is more common in women.
- The median age at diagnosis is 65 years.

Signs and symptoms
ET is often detected during a routine blood test before you notice any symptoms. One of the first signs can be a blood clot. You may experience:
- Weakness, fatigue, and fainting
  - Your blood cell counts are off
- Burning or throbbing pain in the hands or feet
  - When your blood flow decreases
- Weight loss
  - When you are eating less or using more energy
- Low-grade fevers and night sweats
  - Possibly a response from your immune system
- Feeling of fullness or bloating in the left upper abdomen
  - When you have an enlarged spleen
- Headache, itching

People with ET may be at an increased risk for:
- Deep vein thrombosis
- Heart attack
- Pulmonary embolism
- Stroke
- Transient ischemic attack (TIA)
It's important to understand the signs and symptoms of these serious issues. Speak to your doctor about your risk and what you need to know.

ET may cause bleeding for a small number of people with a very high platelet count. Signs and symptoms include:

- Easy bruising and nose bleeds
- Gastrointestinal (digestive tract) bleeding
- Bloody stools and blood in your urine

The disease may transform into other types of cancers in some cases. These include myelofibrosis, acute myeloid leukemia, or myelodysplastic syndrome.

### After your diagnosis

With your diagnosis, your doctor can determine the right treatment for you. Your test results help your doctor predict how your ET will likely progress and how you may respond to treatment.

<table>
<thead>
<tr>
<th>Name of test</th>
<th>Description</th>
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<tbody>
<tr>
<td><strong>Medical history and physical exam</strong></td>
<td>Your doctor reviews past illnesses, injuries, and symptoms. They examine your lungs, heart, and other organs.</td>
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<tr>
<td><strong>Complete blood count (CBC)</strong></td>
<td>This test measures the number of red blood cells, white blood cells, and platelets in your blood.</td>
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<td><strong>Peripheral blood smear</strong></td>
<td>This test looks at blood cells under a microscope to see the number, size, shape, type, and pattern of cells. If you have ET, your platelets may appear enlarged and/or clumped together.</td>
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<td><strong>Bone marrow aspiration and biopsy</strong></td>
<td>These two tests look for the presence and number of abnormal cells. They also look for fibrosis (scar tissue) in your bone marrow. The tests are usually done at the same time.</td>
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<tr>
<td><strong>Molecular testing</strong></td>
<td>This genetic test looks for a specific mutation of the genes known as JAK2, MPL, and CALR.</td>
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ET treatment

People with ET usually have a normal life expectancy. It's important that you're monitored and treated. Your treatment is focused on preventing blood clots and bleeding.

Factors that affect treatment

Discuss your treatment options with your doctor to make sure you understand the benefits and risks of each approach. Your treatment plan is based on:

• Age and overall health status
• History of thrombosis and platelet count
• Whether you are considered low risk or high risk

Types of treatment

Drug therapy is the main form of treatment. It may include:

• Low-dose aspirin reduces the risk of a blood clot.
• Hydroxyurea decreases the number of blood cells made in your bone marrow.
• Anagrelide decreases your body's production of platelets.
• Interferon alpha lowers your platelet count.

If you are low risk, you may be prescribed low-dose aspirin or no therapy at all. Your doctor will monitor you closely through regular exams to look for any signs that your cancer is progressing.

If you are high risk, you may be prescribed low-dose aspirin to prevent thrombosis or other medications to reduce high platelet counts.
Treatment side effects

When you begin your treatment for ET, you may experience mild to severe side effects. New drugs and therapies can help control side effects. Speak to your doctor if you are having side effects.

<table>
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<tr>
<th>Common side effects</th>
<th>You may experience side effects such as:</th>
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<tr>
<td></td>
<td>• Upset stomach, heartburn, and an increased risk of bleeding from low-dose aspirin</td>
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<tr>
<td></td>
<td>• Mouth ulcers (open sores), changes in your sense of taste, skin ulcers, or rash from hydroxyurea treatments</td>
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<tr>
<td></td>
<td>• Headaches, dizziness, nausea, diarrhea, fluid retention, and heart and blood pressure problems from anagrelide treatments</td>
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<tr>
<td></td>
<td>• Depression and flu-like symptoms from interferon alpha treatments</td>
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Living with ET

Medical follow-up is important with ET. Your medical team should provide you with a care plan listing the frequency of follow-up visits and the tests you will have at those visits.

Living with ET can be overwhelming. Seek medical help if you are feeling “down” or “blue” or don’t want to do anything – and your mood does not improve over time. These could be signs of depression, an illness that should be treated even when you’re undergoing treatment for ET. Treatment for depression has important benefits for people living with cancer. Remember, you are not alone.

This fact sheet was reviewed by:

Dr Lynda Foltz, MD, FRCPC
St. Paul’s Hospital
Clinical Professor
University of British Columbia

Never hesitate to contact us, we’re here to help!
1 833 222-4884 • info@bloodcancers.ca • bloodcancers.ca