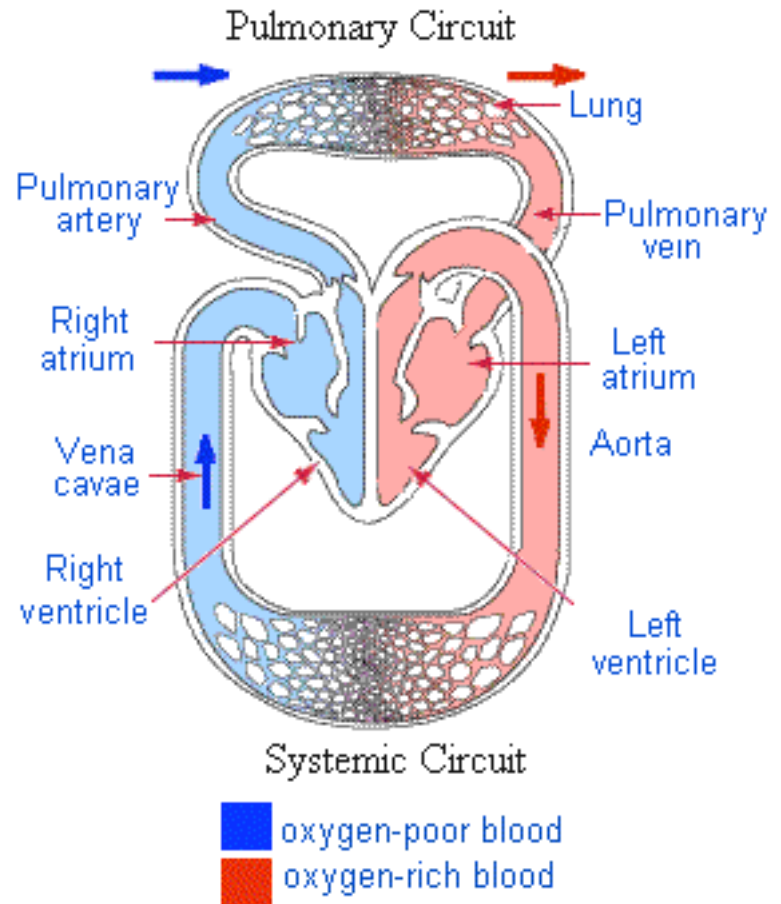


Treprostinil (Remodulin) Therapy

Your pulmonary hypertension physician/nurse practitioner believes that you may benefit from treprostinil (Remodulin) therapy for your pulmonary hypertension. This information is intended to be a guide for what to think about when you consider starting this medication. This decision will be made with your pulmonary hypertension team. This guide also explains what you can expect when starting this medication.

What is Pulmonary Hypertension?

Under normal conditions, the pulmonary arteries carry 'oxygen-poor' blood from the right side of your heart to your lungs. The blood picks up oxygen from your lungs and is then pumped into the left side of your heart to be circulated throughout your body. Pulmonary hypertension (PH) is a rare condition in which the blood pressure in the pulmonary arteries rises above normal levels. The pulmonary arteries become thickened and narrowed. As a result, the right ventricle works harder to pump enough blood through the lungs, and becomes enlarged.



What is treprostinil (Remodulin)?

People with pulmonary hypertension may not produce enough prostacyclin, a substance in your body that causes the blood vessels in your body to relax (vasodilate). Remodlin is a man made prostacyclin that works to relax the blood vessels, and reduce the build up of blood cells (platelets) within the blood vessels. This reduces the work of the right ventricle, and allows the blood to flow more easily from your heart into your lungs. The purpose of this therapy is to better control your symptoms and improve your quality of life. It can take several months for you experience this.

How is treprostnil (Remodulin) taken?

Treprostnil is active in the body for approximately 4 hours. It is given continuously through a blood vessel (intravenous/IV) or under the skin (subcutaneous/SC). It is administered with a portable infusion pump. You will have to take it 24 hours a day, 7 days a week.

For the intravenous form, you will have to have a tunnelled intravenous catheter inserted into your chest. This catheter, known as a Hickman line, will remain in place for the duration of your therapy.

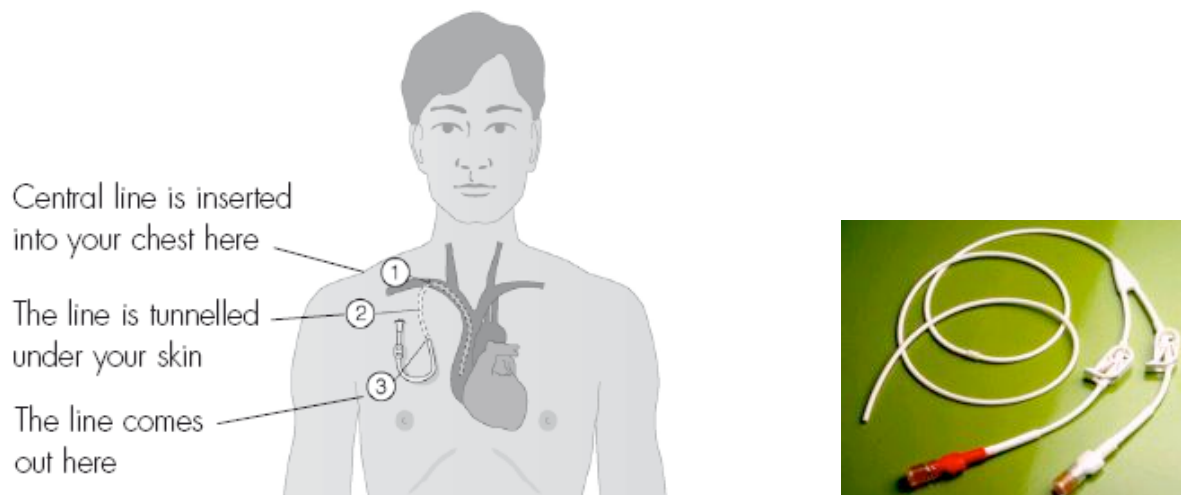
For the subcutaneous form, you will have to learn how to insert the catheter under your skin. The catheter is changed every 3 days. Some patients are able to maintain an infusion site for up to 30 days.

What is a tunnelled intravenous catheter?

As previously mentioned, treprostnil may be given through a continuous intravenous (IV) infusion. You will require a Hickman line, which is a long IV tube that sits under the skin (tunnelled) in your chest before entering a large vein that leads to your heart. Your Hickman line will have 2 lumens (tubes). These lumens act like separate IV lines.

Placement of the Hickman line requires minor surgery. Arrangements will be made for the Hickman line to be inserted when you come to the hospital to start the Remodulin infusion. It is very likely that your Hickman line will need to be changed every few years. This is often related to infection, a break in the catheter, or blockage of a lumen).

You will learn how to take care of the Hickman line. A nurse from the IV Therapy team will work with you to assist you to become comfortable with how to manage the Hickman line as part of your daily routine.



What is an infusion pump?

As previously mentioned, treprostiniil is a continuous infusion that requires a portable, battery operated pump for delivery. You will have to carry the pump with you at all times. The pump used for intravenous form of Remodulin is a CADD Legacy Plus 6500. The pump used for the subcutaneous form of Remodulin is a CADD MS3. You will be provided with two pumps, which will be changed daily. You will learn how to use the pump. Written instructions and a video will also be provided.



*CADD Legacy Plus 6500
(for intravenous infusion)*



*CADD MS3
(for subcutaneous infusion)*

Why has my team chosen to start treprostiniil (Remodulin) therapy for my pulmonary hypertension?

Your PH team believes you may benefit from being on this medication. This decision has been made based on a number of factors, including:

- How far you can walk
- How bad your symptoms are
- Results of various tests
- Your response to other treatments so far

What is the recommended dose of treprostiniil (Remodulin)?

The dose of treprostiniil is different for each patient. The amount of medication you take will depend on the strength of the medication you are using, your weight, and your PH symptoms. There is no set dose for Remodulin. You will be started on a small dose, and it will slowly be increased over time (up-titrated). The rate of up-titration will depend on the side effects you experience.

What are the side effects of treprostiniil (Remodulin)?

People who are on treprostiniil therapy may experience side effects. These side effects are often related to the dose you are on. The side effects commonly become less severe over time. Your PH doctor or nurse will help you learn to manage the side effects. The most common side effects are:

- | | | |
|------------|-------------|------------------------------------|
| ▪ Headache | ▪ Dizziness | ▪ Pain in feet, ankles, back |
| ▪ Jaw pain | ▪ Flushing | ▪ Hypotension (low blood pressure) |
| ▪ Nausea | ▪ Diarrhea | |

For the subcutaneous infusion, infusion site pain and infusion site reaction (redness, swelling, tenderness) occurs in the majority of patients on this medication. These symptoms can be severe at times. Your pulmonary hypertension team will work closely with you to manage these symptoms.

How do I know if the treprostinil (Remodulin) is working?

Your pulmonary hypertension symptoms will begin to improve over time. You may have more energy, and less shortness of breath. It may take a couple of months for this to happen.

How much does treprostinil (Remodulin) cost?

Flofan is very expensive. One year of treatment may cost between \$60 000-\$100 000 per year (includes medication and supplies). This depends on the dosage and form of administration. Your PH physician will need to apply to Pharmacare for Special Authority approval. You will be responsible for paying your annual Pharmacare deductible. The Fair Pharmacare Program will cover the remainder of the cost for medication, and the VGH Pulmonary Hypertension Clinic will cover the remainder of the cost for supplies.

What is involved in learning to manage the treprostinil (Remodulin) at home?

Prior to starting the Remodulin, you may have to come to Vancouver General Hospital for pre-teaching sessions to learn how to mix and administer the medication. Once this has been done, the PH clinic will make arrangements for you to be admitted to Vancouver General Hospital to have a Hickman line inserted (if using the intravenous form), and to start the Remodulin infusion. Expect to be in hospital for approximately 2-4 weeks.

You will need to arrange for a family member or friend to learn how to mix and administer the Remodulin as well. That person will be responsible for maintaining the Remodulin infusion during times when you are unable to manage the Remodulin infusion yourself (ex. When you are in hospital).

Can I mix more than one cassette/cartridge at a time?

No. The IV Remodulin is stable for 48 hours after being mixed. The SC Remodulin is stable for 72 hours after being drawn up into the cartridge.

Can the pharmacy mix my treprostinil (Remodulin) for me?

No. Your Remodulin needs to be mixed at the same time every 48 hours. Many pharmacies are not able to guarantee that they will be able to have the Remodulin ready for you when you need it. Many pharmacies do not have the resources to ensure your Remodulin is mixed under sterile conditions.

Will I have to be on treprostinil (Remodulin) for the rest of my life?

Your PH team encourages you to think of Remodulin as being a life-long therapy. It is possible to wean off the Remodulin infusion. This must be done with direction and support from your pulmonary hypertension team. Suddenly stopping the infusion is dangerous.