

REAL PATIENTS WITH REAL JOURNEYS

Marsha



Marsha's life before CTEPH

Marsha is a nurse who works in the emergency department of a hospital. Before she was diagnosed with CTEPH, she was a single mother of three children—the youngest still at home, while the others had made it out on their own. *"I think most people would say I was just extremely energetic,"* she says. *"I could pretty much do whatever I needed to do, when I needed to do it."*

Raising three kids by herself was challenging, and her job was equally so. She relied on good stamina and a lack of physical constraints when it came to her work as a nurse. *"I could transport people around on carts and beds and wheelchairs and I could do anything. And then things changed."*



"I could do anything. And then things changed."

IMPORTANT SAFETY INFORMATION

Adempas can cause serious birth defects if taken during pregnancy.

- Females must not be pregnant when they start taking Adempas (riociguat) tablets or become pregnant during treatment with Adempas.
- Females who are able to get pregnant must have a negative pregnancy test before beginning treatment with Adempas, each month during treatment, and 1 month after you stop treatment with Adempas. Talk to your doctor about your menstrual cycle. Your doctor will decide when to do the test.
- Females who are able to get pregnant are females who:
 - Have entered puberty, even if they have not started their period, have a uterus, and have not gone through menopause (have not had a period for at least 12 months for natural reasons, or have had their ovaries removed).
- Females who are not able to get pregnant are females who:
 - Have not yet entered puberty, do not have a uterus, or have gone through menopause.

Please see additional Important Safety Information throughout and full Prescribing Information, including Boxed Warning and Medication Guide, [here](#).

The winding road to diagnosis

Before going to the ER, Marsha thought she had a collapsed lung that turned out to be a pulmonary embolism (PE). She was prescribed blood thinners for nine months and felt fine when her time on them ended.

But two years after the PE, she felt a change. Her stamina was failing, and she was having trouble walking. She called her doctor repeatedly, but wasn't getting a response. Finally, she got an appointment. *"He told me I had anxiety and prescribed me some anxiety medicine,"* she recalls. *"But I knew something else was wrong."*

The anxiety medication didn't improve her condition, and her stamina continued to deteriorate. When she went to work one day and could barely walk, her coworker insisted on her seeing a Specialist. Upon examination, it was discovered that she had another PE—one that she, in her words, *"did not bounce back from."*

She continued to work, with difficulty. *"I was just very limited, and they [her coworkers] were very kind and were working around my physical limitations,"* she said. One of her colleagues, an ER physician, suggested she see a pulmonologist, and she decided to give it a try. A battery of tests were conducted, including a right heart catheterization, and Marsha was diagnosed with pulmonary arterial hypertension (PAH), (WHO Group 1). *"I feel like that pulmonologist really kind of saved my life,"* she said.

"I was just very limited, and they [her coworkers] were very kind and were working around my physical limitations."

INDICATION

Adempas is a prescription medicine used to treat adults with:

- chronic thromboembolic pulmonary hypertension (CTEPH) (WHO Group 4) treated with surgery but who continue to have high pulmonary blood pressure (persistent) or it comes back after surgery (recurrent), or that cannot be treated with surgery.
 - CTEPH is a type of high blood pressure in the arteries of your lungs caused by blood clots that narrow or block blood flow. Adempas can improve your ability to exercise (measured by 6-minute walk distance) and can help to improve some of your symptoms.
- pulmonary arterial hypertension (PAH) (WHO Group 1)
 - PAH is a type of high blood pressure in the arteries of your lungs. Adempas can improve your ability to exercise (measured by 6-minute walk distance), improve some of your symptoms, and help slow down the worsening of your physical condition [this includes death, heart/lung transplant, a surgery to reduce pressure in your lung, hospitalization for worsening of pulmonary hypertension, start of a new PAH-specific treatment, continued decline in your ability to exercise (measured by 6-minute walk distance) and some of your symptoms].

The Adempas clinical studies included mostly patients with WHO functional class II-III symptoms. It is unknown if Adempas is safe and effective in children.

The winding road to diagnosis (continued)

She began routine medications for PAH and slowly began to gain back some stamina. *"I still was having problems, like pushing carts, and anything on an incline."* Still, she felt better than she did before her diagnosis with PAH, and she continued to take her medications.

After being a single mother for fourteen years, Marsha remarried. She moved near a large city and transferred her care to a hospital that had a pulmonary hypertension program. She attended a pulmonary hypertension event with expert thought leaders, and that's when she heard about CTEPH (WHO Group 4) for the first time.

"I did research and learned more about CTEPH and the different types of PH," she said. *"I realized that I hadn't had a V/Q scan yet, and once I did, my doctor discovered that I had CTEPH rather than PAH."* A V/Q scan (along with other assessments), as Marsha learned, is the preferred and recommended screening test for CTEPH.¹ It's the most accurate scan available for CTEPH, with a screening sensitivity greater than 96%.²

Once diagnosed with CTEPH, Marsha was assessed for surgery and determined inoperable because of how deep her clots were in her lungs. Her medications were updated, and she was prescribed the only approved medical therapy for CTEPH: Adempas.*



"I realized that I hadn't had a V/Q scan yet, and once I did, my doctor discovered that I had CTEPH rather than PAH."

IMPORTANT SAFETY INFORMATION (continued)

Females who are able to get pregnant must use two acceptable forms of birth control, during Adempas treatment and for one month after stopping Adempas.

- If you have had a tubal sterilization, have a progesterone implant, or have an IUD (intrauterine device), these methods can be used alone and no other form of birth control is needed.
- Talk with your doctor or gynecologist (a doctor who specializes in female reproduction) to find out how to prevent pregnancy during treatment with Adempas.
 - **Do not have unprotected sex. Talk to your doctor or pharmacist right away if you have unprotected sex or if you think your birth control has failed. Your doctor may tell you to use emergency birth control.**
 - **Tell your doctor right away if you miss a menstrual period or think you may be pregnant for any reason.**

If you are the parent or caregiver of a female child who started taking Adempas before reaching puberty, you should check your child regularly to see if she is developing signs of puberty. Tell your doctor right away if you notice that she is developing breast buds or any pubic hair. Your doctor should decide if your child has reached puberty. **Your child may reach puberty before having her first menstrual period.**

Females can only receive Adempas through a restricted program called the Adempas Risk Evaluation and Mitigation Strategies (REMS) program. If you are a female who can become pregnant, you must talk to your doctor, understand the benefits and risks of Adempas, and agree to all of the instructions in the Adempas REMS program. Males can receive Adempas without taking part in the Adempas REMS program.

*Adempas treats adults with CTEPH (WHO Group 4) treated with surgery but who continue to have high pulmonary blood pressure or it comes back after surgery, or that cannot be treated with surgery. The Adempas clinical studies included mostly patients with WHO Functional Class II-III symptoms. It is not known if Adempas is safe and effective in children.

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Living with CTEPH

After beginning therapy with Adempas, Marsha started to see improvements in some of her symptoms. *“When my doctor evaluated me and told me my functional class had improved, I felt better because I was improving.”*

She has continued to improve on other measures, such as her six-minute walk test, but still avoids stairs at work. *“I kind of know what my new normal is, and I avoid certain things,”* she says.

Marsha’s family is understanding of her illness, and her life at home with her husband is supportive. *“It’s kind of odd because my husband — since we’ve been married for three and a half years — he’s never known me any different. He might not have liked the old me. I would have left him in the dust maybe! But he’s very, very understanding of it.”* She is also close with her daughter and talks with her on the phone multiple times a day. *“We’re like best friends. I think we kind of were before, but maybe more so now—very close.”*



“I kind of know what my new normal is, and I avoid certain things.”

IMPORTANT SAFETY INFORMATION (continued)

Do not take Adempas if:

- **you are pregnant, plan to become pregnant, or become pregnant during treatment with Adempas. Adempas can cause serious birth defects.**
- **you take:**
 - another medicine called a soluble guanylate cyclase stimulator (sGC). Ask your healthcare provider if you are not sure if you are taking an sGC medicine
 - a nitrate medicine to treat high blood pressure or heart disease, such as nitroglycerin, or a medicine called a nitric oxide donor, such as amyl nitrite
 - certain other medicines that contain sildenafil (Revatio or Viagra), tadalafil (Adcirca or Cialis), vardenafil (Levitra or Staxyn), dipyridamole, or theophylline. Revatio and Adcirca are also used to treat PAH
- **you have pulmonary hypertension associated with idiopathic interstitial pneumonias (PH-IIP).**

Ask your doctor or pharmacist if you are not sure if you take any of the medicines listed above.

Throughout her journey with CTEPH, Marsha has set goals that center around spending time with her family. *“I have four grandchildren, and I try to see them every three months,”* she says. *“I have goals, though they’re probably not super long-term.”* Over time, she has come to accept her condition and gained perspective on how to think about it. *“You can’t reverse it,”* she says. *“You can’t just stop it where it is. It’s going to keep progressing.”*

Even so, she has been encouraged by the progress she has shown. *“I was kind of pleasantly surprised when I really started to notice changes in the way I feel and what I was able to do. My laundry is in the basement, and now I am able to go down there to bring a basket up.”*

For Marsha, living with CTEPH is a journey of highs and lows, but she is grateful for the time she gets to spend working towards her greatest goal: spending time with her family. *“My son is getting ready to graduate college, and my dad is still living. I’m planning to take an Alaskan cruise next summer,”* she says. *“My goals right now are all about relationships with people.”*

This is an example of one patient’s experience. Always talk with your doctor to see if Adempas is right for you.

This story is part of myAim Education & Encouragement.



For more information, visit adempas-us.com.

IMPORTANT SAFETY INFORMATION (continued)

Before you take Adempas, tell your doctor if you:

- smoke; have recently had serious bleeding from your lungs, or if you have had a medical procedure called bronchial arterial embolization to stop you from coughing up blood; have problems with your heart or blood circulation; have low blood pressure; have liver problems; have kidney problems or are on dialysis; have narrowing of the pulmonary veins, a condition called pulmonary veno-occlusive disease or PVOD; have any other medical conditions.

Tell your doctor about all the medicines you take, including prescription and non-prescription medicines, vitamins, and herbal supplements. Adempas and other medicines may affect each other causing side effects. Do not start any new medicine until you check with your doctor.

Please see additional Important Safety Information throughout and full Prescribing Information, including Boxed Warning and Medication Guide, [here](#).



With CTEPH, how far might Adempas take you?

Adempas is the first and only medicine approved by the FDA for treating adults with inoperable and recurrent/persistent CTEPH (WHO Group 4) after surgery. The Adempas clinical studies included mostly patients with World Health Organization Functional Class (WHO FC) II-III symptoms.

Adempas can help increase your ability to walk farther (as measured by 6-minute walk distance [6MWD]) and improve some of your symptoms.

A 16-week study of 261 adults with CTEPH (WHO Group 4) showed that patients who took Adempas walked farther during the 6MWD test, compared to patients who did not take Adempas. The 6MWD test determines how far you can walk in 6 minutes. Your doctor may use it periodically to measure any differences in progress.

It is important to track your progress throughout the course of your treatment.

- Talk to your doctor to set specific goals that may help you in your treatment journey



2X as many patients who took Adempas improved their WHO FC compared to patients who took placebo (no Adempas)

—33% of Adempas patients showed WHO FC improvement vs 15% of placebo patients

IMPORTANT SAFETY INFORMATION (continued)

What should I avoid while taking Adempas?

- **Do not get pregnant** while taking Adempas. If you miss a menstrual period, or think you might be pregnant, call your doctor right away.
- It is not known if Adempas passes into your breast milk. You should not breastfeed if you take Adempas. Talk to your doctor about the best way to feed your baby if you take Adempas.
- Adempas may make you feel dizzy. **Do not drive, operate machinery, or do other activities that require mental alertness or coordination until you know how Adempas affects you.** Talk with your doctor if you are concerned about when it is safe for you to do these activities.
- **Smoking.** Adempas may not work as well if you smoke during treatment. Tell your doctor if you stop smoking or start smoking during treatment with Adempas, because your dose of Adempas may need to be changed.

We're here for you

Aim Patient Support Program



Financial Assistance

Co-pay Assistance Program*

- Reduces your out-of-pocket responsibility; up to 100% coverage of co-pays or coinsurance directly to the insurer

Temporary Patient Assistance Program (TPAP)

- For temporary situations when immediate coverage is not available (for example, waiting for approval of a prior authorization appeal or waiting for coverage to begin) or existing coverage is lost and alternate funding is needed

Patient Assistance Program (PAP)

- If you are uninsured or underinsured (when criteria are met and alternative funding cannot be identified), you will receive your medicine free of cost for one year from the date of acceptance (Medicare Part D patients will be enrolled based on calendar year if eligible for the PAP)

* Patients must have private commercial insurance with a percentage coinsurance or co-pay requirement. Medicare or Medicaid patients are ineligible. Assistance is for one year, after which, patients must re-apply. Patients must notify the program of any change in their insurance status. Patients in certain states may be ineligible. The co-pay assistance program has an annual cap per patient year.



Aim Nursing Support

You can call toll-free 24/7 and speak with a specially trained nurse.

You and your physician can authorize virtual nurse visits through your certified pharmacy. If you and your physician decide that virtual nurse services are right for you, you can expect the following:

- Adempas information for you and your caregiver
- Explanation of titration (dose adjustment)
- Review of your status for titration of dose
- Follow-up on your specific needs
- Explanation and review of your schedule of nurse visits



myAim Education & Encouragement

A helpful resource that provides personalized communications and educational support throughout your Adempas treatment

You can expect to receive direct mail and emails about:

- Personalized treatment resources, provided how and when you want them
- Support throughout some of the key stages of your Adempas treatment and what to do along the way
- Tips on what to talk to your doctor about
- Information about other online resources and support

Keep a lookout for more patient stories from **myAim Education & Encouragement**

IMPORTANT SAFETY INFORMATION (continued)

Adempas can cause serious side effects including:

- **Serious birth defects.**
- **Reduced blood pressure.** This may cause symptoms of low blood pressure, such as lightheadedness, chest pain, and dizziness especially in people who are dehydrated, have a severe blockage of blood flow out of the heart, or have certain other medical problems. Your doctor will check you for any of these problems.
- **Increased risk of bleeding, including bleeding from the respiratory tract.** Tell your doctor right away if you cough up blood during treatment with Adempas.
- **Worsening of symptoms in people with Pulmonary Veno-Occlusive Disease (PVOD).** If you have PVOD, treatment with Adempas may cause a build-up of fluid in your lungs (pulmonary edema) which may result in shortness of breath. Your doctor may tell you to stop taking Adempas and switch you to a different medicine.

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The most common side effects of Adempas are:

- Headache; dizziness; indigestion; swelling of your hands, legs, feet, and ankles (peripheral edema); nausea; diarrhea; and vomiting

Tell your doctor if you have any side effect that bothers you or that does not go away. These are not all the possible side effects of Adempas.

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For important risk and use information please see the enclosed full Prescribing Information, including Boxed Warning and Medication Guide, [here](#).

You are encouraged to report negative side effects or quality complaints of prescription drugs to the FDA. Visit www.fda.gov/medwatch or call 1-800-FDA-1088.

References:

1. Kim NH et al. *J Am Coll Cardiol*. 2013;62(suppl D):D92-D99.
2. Tunariu N et al. *J Nucl Med*. 2007; 48(5):680-684.