PAH Doctor Discussion Guide for Newly Diagnosed Patients

If you have been recently diagnosed with Pulmonary Arterial Hypertension (PAH), you may be feeling nervous and uncertain about how to approach the first steps in learning about this condition and your treatment options. It's important to know what questions to ask your doctor and what goals to keep in mind.

This guide provides questions that you can ask your doctor to help guide your discussion and get the most out of your next visit.

Questions to ask your doctor

Li	ving with PAH		
	What are typical symptoms of PAH and how can I better manage them?		What types of daily activities should I try to participate in?
	How do I approach making my work or school environment more PAH friendly? What are some things I could do to help make my home environment more PAH friendly?		Are there any activities I should avoid?
X			How is my PAH compared to other patients you have seen?
			Will it be safe for me to get pregnant?
Т	reating PAH		
	Is PAH curable?		How do I set and maintain a schedule for my medication?
	What treatment options could help improve my symptoms?		What do I do if I am not reacting well to my medication?
	What should I expect when beginning my treatment?	\cap	What happens if I miss a dose of my medication?
	Are there side effects that I should look out for while on my medicine?		Will my dose ever change?
	Will I be on treatment long term?		Are there any vitamins or over-the-counter medications that would be helpful?
	Poctor Visits		
	How often should I come in for checkups?		What kind of goals should I set when it comes to improving my PAH symptoms?
	Is there anything I can do to prepare for my appointments with you?		Are there any additional doctors I should be seeing?
	How do I track my progress with PAH?		
R	esources and Support		
	Is there a support program that can help patients like me?		Are there organized events that I can attend to learn more about my PAH?
	How do I connect with others who can share their PAH journey with me?		Do you have any brochures or information you can provide me about PAH?

Please see Important Safety Information on the following pages. For important risk and use information, click here for the full Prescribing Information, including Boxed Warning and Medication Guide.

Notes from your visit with your doctor

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IMPORTANT SAFETY INFORMATION

Adempas® (riociguat) 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 <u>are able</u> 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 <u>are not able</u> to get pregnant are females who:
 - Have not yet entered puberty, do not have a uterus, or have gone through menopause.

INDICATION

Adempas is a prescription medicine used to treat adults with:

- 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.

Aim Patient Support Program



Aim Nursing Support

- 24/7 hotline provided by the specialty pharmacy
- Virtual visits in consultation with your healthcare professional



Aim Access and Financial Support

Co-pay Assistance Program*

- Reduce out-of-pocket costs: up to 100% of co-pays or coinsurance paid directly to the insurer Patient Assistance Program (PAP)⁺
- Eligible individuals can apply to receive medication free of cost for up to one year. Eligibility and criteria may vary

*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.

†Medicare Part D patients will be enrolled based on calendar year.



myAim Education & Encouragement

Helpful tools and personalized communications

- Educational support and personalized treatment resources through some of the key stages of treatment
- Tips on what to discuss with your doctor and information about other online resources and support



Real-life stories of patients like you living with PAH and CTEPH

Learn more at www.adempas-us.com/myjourney

IMPORTANT SAFETY INFORMATION (continued)

Females who are able to get pregnant must use two acceptable forms of birth control, during Adempas® (riociguat) 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.



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.

Before you take Adempas® (riociguat), 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.

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.

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.

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.

For important risk and use information, please see the 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.

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