Focus on PAH and Body Chemistry
At Actelion, we believe that knowledge is power. When you understand your condition, you have the power to ask the right questions, make informed decisions, and get the most from your healthcare team and your therapeutic options. That’s our goal in PAH education and that’s KNOWLEDGE IN ACTION.

Your body’s chemistry: a delicate balance

Changes in your body’s chemistry can affect every aspect of your health. Whether it’s your ability to fight disease, digest food, exercise, or tap into your brain’s processing power—complex chemical reactions are critical to proper bodily function.

Generally, a healthy lifestyle that includes proper diet and exercise can keep body chemistry working properly. For people with pulmonary arterial hypertension (PAH), a combination of things—ranging from genetic variations to other illnesses or conditions—can cause an ongoing imbalance in certain aspects of body chemistry, which in turn affects the health of the blood vessels and heart.¹
There are 3 chemicals that are currently known to be out of balance in patients with PAH: endothelin, nitric oxide, and prostacyclin. In patients with either too much or not enough of these chemicals, the health and function of pulmonary blood vessels are affected.⁴,⁶
Nitric oxide is a gas that is also present in the bloodstream. Nitric oxide promotes the widening of blood vessels and prevents blood cells from sticking together and forming clots. In people without PAH, nitric oxide is constantly released from the lining of the blood vessels.\textsuperscript{4,5,9}

In many patients with PAH, the body produces less nitric oxide, which causes the blood vessels to tighten, which can lead to stiffening and possible blockage of the arteries.\textsuperscript{4,5}

Endothelin (en-doe-THEE-linn) is a chemical known as a signaling peptide, which is present in everyone’s bloodstream. In most people, endothelin triggers the blood vessels to constrict (get narrower) or dilate (get wider), depending on current body chemistry and activity.\textsuperscript{4,7}

In many patients with PAH, there is too much endothelin in the bloodstream, and the body cannot process it quickly enough. In addition, the endothelin “signal” that tells blood vessels to tighten or narrow is overactive in these patients, leaving the pulmonary arteries in a constant narrow state that does not allow for normal blood flow or circulation of oxygen.\textsuperscript{2,7,8}

Over time, overstimulation with endothelin will lead to inflammation (swelling), fibrosis (scarring), and hypertrophy (thickening and stiffening) of the blood vessel walls.\textsuperscript{2,8}

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Prostacyclin (pros-tuh-SY-klin) is another chemical produced in the lining of the blood vessels. It causes the widening of vessels and increased blood flow by relaxing the muscle cells of the vessels.\textsuperscript{4,5}

Some patients with PAH don’t produce enough prostacyclin, which keeps the blood vessels from relaxing and widening.\textsuperscript{1}
NOTES
Use this section to record important notes about PAH or your treatment.
REFERENCES

Medications are available, by prescription only, to address each of these chemical imbalances. Talk to your doctor about which medications may be right for you.

To learn more about PAH and your options for treatment, visit the following organizations online:

Pulmonary Hypertension Association
www.phassociation.org

Scleroderma Foundation
www.scleroderma.org