

My story began in July 2007 when I was advised by a dietician to use a diet pill to lose some weight. I was trying to get pregnant and already stopped using the contraceptive pill 18 months before that. I did not want to gain any more weight with another pregnancy, so I used the diet pill for only one month. Suddenly I had shortness of breath, was easily getting tired. I went to my GP to do an ECG as my cousin died 3 year prior of heart failure after giving birth to her son. I knew that I had a wonderful pregnancy with Janke, but was afraid that something could go wrong as on my mother's side of the family, they had heart problems; my great-grandmother, grandfather and two of his brothers died of heart attacks.

My GP did a ECG, but could not found anything wrong; he also send me for an X-ray of my lungs and heart to see if I possibly had asthma. He said I was overweight and need to do more exercise. On the X-ray he did not pick up anything. After a month my symptoms got worst and my GP referred me to a physician and I told her my story. She also looked at the X-ray and could not see anything of concern. She then told me that she just want to do a check-up and listen to my heart. She told me that she heard a second sound on my heart and that she wants me to go for an ultrasound. They did an ECG again and also an ultrasound. The physician told me that the ultrasound showed that the right side of my heart was enlarged and that I must not get pregnant before they determine what caused the enlargement. They then did a CT scan and I was diagnosed with Primary Pulmonary Hypertension on 2007-09-11.

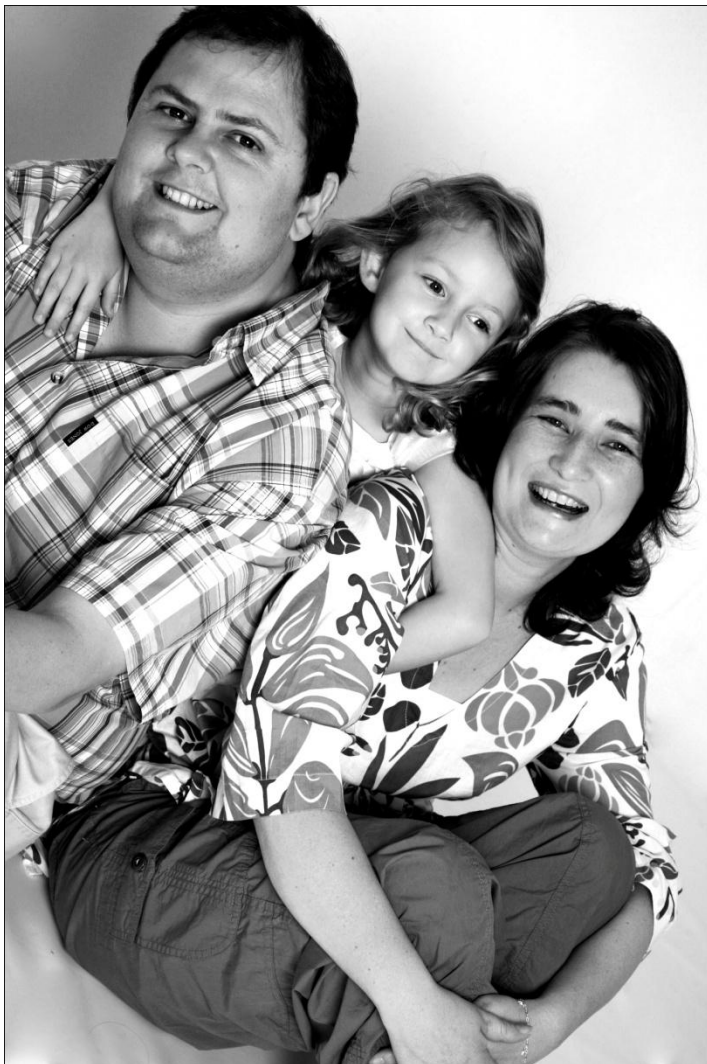
The physician first put me on high blood pressure pills, but it could not tolerate it. She then called a cardiologist and he suggested that I must be given Cialis. As I have low blood pressure I only used a half of a tablet of the Cialis. We went for a second opinion at a Cape Town Pulmonologist, which confirmed the diagnosis. I also started using Warfarin.

In August 2008 we got an appointment with Dr Paul Williams, a Critical Care Specialist at Milpark Hospital, to be worked up for a possible lung transplant. When we arrived at the hospital Dr Williams told us that the SA Medical Board approved a trail drug for our disease and that he wanted to put me on the trail drug. We did not know if I was on the placebo, 3mg or 10mg of the trail drug. Back home my life changed for the worse as my health deteriorated rapidly. I had to stop working; I was 24 hours in bed and on oxygen.

In November 2008 my dad and I visited Dr Williams for my first 3-monthly appointment. As we got to Johannesburg, I passed out. The same happened that evening and the next morning. Dr Williams was not available and I saw a colleague of him, namely Dr Pahad. He told us that it looks like that I maybe on the placebo and that he was going to speak to Dr Williams when he returns, so that they can ask Actelion (the trail drug company) to put me onto the open label of the trail. Then they would know that I get the active drug. At that stage I could only walk 120m during my six minute walk.

We returned to Milpark two weeks later and was placed on the open label of the trial drug. After one day of use, Drs. Williams and Pahad could already see a difference as my lips were not blue as it used to be. As I started the active drug, I go better and better. At our visit in March 2009 I walked 300m in six minutes; in May 420m, in August 440m and in November 460m. I only use my oxygen when I go to sleep. I only use my wheelchair when we have to walk long distances and when we use the airplane (to fly to Johannesburg for my visits to Dr Williams).

Firstly I would like to thank God for this miracle as I received a second chance on life. Secondly I would want to thank Dr Williams - there is no better doctor in the world! Then I would like to thank Cindy Hill (the Clinical Trail co-ordinator) for all her assistance and help. Currently I am connected to more than 70 people via Facebook from all around the world who has the same disease as me. It is wonderful to link up with others and to realise that you are not alone in the fight against Primary Pulmonary Hypertension.



In this picture is my Husband Nico, my daughter Janke, and me Nicolene

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Here is some info I received today via one of my facebook friends. I don't know if you maybe can use it on the website.

"Get to know your PH" have the following details at www.knowyourph.org.

A

Alveoli

The smallest lung units that include thin-walled air sacs directly linked with a network of small blood vessels (capillaries) where gas exchange occurs

Angina

Chest pain that is caused by parts of the heart not getting enough blood; usually the result of atherosclerosis of the coronary arteries.

Anticoagulants

Medications that thin the blood make the formation of blood clots less likely.

Anticoagulation

The act of receiving therapy with anticoagulants

Aorta

The largest artery in the body; carries all the oxygenated blood pumped out by the left ventricle to the rest of the body via a branching network of smaller arteries

Aortic valve

A valve in the heart that separates the left ventricle from the aorta preventing blood from flowing backwards into the ventricle.

Arginine

An essential amino acid whose natural breakdown and metabolism generates the potent vasodilator nitric oxide

Asthma

A disease in which the muscles surrounding the airways of the lungs spasm and result in reversible narrowing of the airways, making it difficult to exhale.

Atrium

The two smaller chambers (one on each side) of the heart; the right atrium accepts blood from the veins throughout the body and the left atrium accepts freshly oxygenated blood from the lungs

B

BNP (b-type or brain natriuretic peptide)

A protein that is released by the muscle tissue of the heart, particularly the ventricles; the amount that is released increases as the heart is stretched or strained.

Bronchi

Branching airways in the lungs that carry air from the trachea to the alveoli

Bubble study

A specialized echocardiogram that looks for holes between the right and left sides of the heart by injecting bubbles (agitated saline) into the bloodstream and watching to see if they cross from the one side of the heart to the other

C

Calcium channel blocker

A type of medication that is a vasodilator and is occasionally used to treat PAH (e.g. amlodipine); also, often used for patients who have systemic hypertension

Capillaries

The smallest type of blood vessel, located throughout the body including lungs; the place where oxygen, nutrients, and waste products are exchanged from bloodstream to cells and vice versa.

Carbon Dioxide

A gaseous molecule that is one of the waste products generated by the cells of the body.

Cardiac output

The amount of blood (measured in liters per minute) being pumped out by the heart.

Cardiologist

A doctor who is specially trained in diseases of the heart and blood vessels.

Central venous catheter

An intravenous line that is placed in one of the larger veins of your body (usually in the chest) to deliver medications

Cirrhosis

End-stage liver disease due to scarring; often caused by alcohol or viral hepatitis over many years

Congenital heart defects

A defect in the structure of the heart that is present at birth

Congestive heart failure

A disease in which the heart is stiff or does not pump adequately causing a back up of blood and fluid.

Coronary arteries

The blood vessels that supply oxygenated blood to the heart itself; there are three main coronary arteries.

Coronary artery disease

Narrowing of the coronary arteries due to atherosclerosis (build up of cholesterol plaques in the walls of the arteries).

CT Scan

A specialized X-ray machine that can provide detailed images of internal organs including the lungs and their blood vessels.

D

Diaphragm

A muscle that sits below the lungs; when it contracts it helps the lungs expand and take a breath in

Diastole

The period of time when the heart is relaxing, filling with blood and getting ready to contract again

Diastolic blood pressure

(the bottom number of the reading) is measured during diastole.

Diffusing capacity (also known as DLCO)

One of the PFTs which measures how well oxygen and carbon dioxide transfer from the air in your lungs into the bloodstream and vice versa.

Diffusion impairment

A characteristic of gas exchange in PAH in which the movement of oxygen from alveoli to capillaries is reduced

Diuretics

Medications used to rid the body of excess fluid

Dyspnea

Shortness of breath, or any uncomfortable sensation of breathings; typically worsens with exertion

E

Echocardiogram

An image obtained by ultrasound (a machine that creates an image from sound waves bouncing off an object) of the heart that provides information about structure, blood flow, contractility ('squeeze'), and allows estimation of the blood pressure in the pulmonary arteries.

Edema

Swelling caused by fluid build-up and fluid leaking out of blood vessels into surrounding soft tissues; usually occurs in areas that are dependent, such as the legs.

Emphysema

A lung disease usually caused by smoking in which alveoli are lost or damaged, making gas exchange and breathing more difficult

Endothelial cells

Type of cells that line all blood vessels including pulmonary arteries. Transaminitis: an elevation in liver enzymes (measured by a blood test) that indicates irritation, inflammation, or damage to the liver

Endothelin

A very potent vasoconstrictor found in the blood and lung tissue both naturally and in disease states

Endothelin Receptor Antagonists

A class of oral medications that act primarily as vasodilators, and which are used to treat PAH. There are two types of endothelin receptors and medications may block both of these or just one type preferentially.

Exercise oximetry

Measurement of the oxygen saturation in blood during exercise

F

Femoral vein

A large vein in the groin and leg that carries blood from the legs back towards the heart and can be used in right heart catheterization

Functional Class

A four-tiered system that doctors use for describing your overall ability to do activities of daily living.

H

Half-life

The period of time it takes for half of any medication to be metabolized by the body; used to measure how long a medication's effects will last and thus how frequently it must be dosed

Heart

The main organ of the circulatory system that has the function of receiving blood from and pumping blood to the rest of the body

Heart attack

Also known as a myocardial infarction (MI); a sudden event when part of the heart is deprived of blood, usually due to a blockage in one of the coronary arteries, resulting in damage to that part of the heart.

Hepatitis

Inflammation of the liver, most often caused by infection from a hepatitis virus (e.g. hepatitis A,B,C)

Hypertension

High blood pressure

Hypoxic vasoconstriction

A characteristic of the pulmonary arteries by which they constrict during periods of low oxygen concentration

I

Inferior vena cava

One of the largest veins in the body, located in the chest; all blood in the lower body (below the heart) travels through this vein which then empties into the right atrium.

INR (or pro-thrombin time)

A blood test that measures how well your blood clots; used to adjust the dose of an anti-coagulant (blood thinner) called warfarin® (coumadin)

Internal jugular vein

A large vein in the neck that carries blood from the head back to the heart and is often used in right heart catheterization

L

Lungs

The main organs of the respiratory system that have the function of transporting oxygen from the air into the blood and removing carbon dioxide from the blood; located in the chest, one on the left and one on the right.

M

Mean arterial pressure

An average of the systolic and diastolic blood pressures (mean arterial pressure = $\frac{2}{3}$ diastolic pressure + $\frac{1}{3}$ systolic pressure).

Metered-dose inhalers (MDIs)

Devices that deliver medications into the lungs in the form of a powder or liquid spray

Mitral valve

A valve in the heart that separates the left atrium from the left ventricle preventing blood from flowing from the ventricle backwards into the atrium

N

Nebulizer

A device that uses pressurized air to volatilize (turn into mist) a liquid medication that can then be inhaled.

Nitric oxide

A very potent vasodilator found in the blood and in lung tissues; can be used diagnostically during cardiac catheterization as an inhaled agent to test for acute vasodilatation or 'vasoreactivity'.

O

Obstructive sleep apnea

A disorder in which the upper airway collapses or becomes too lax during sleep causing obstruction of the air passages and cessation of airflow to the lungs.

Oxygen

A gaseous molecule found in the air that serves as the fuel for all the cells in the body.

P

Phosphodiesterase inhibitors

A class of medications used to treat PAH that act as vasodilators

Pneumonia

An infection in the lung, typically causing fever, cough, and phlegm

Polysomnography

Also known as a sleep test; a test during which various parameters such as oxygenation, breathing, heart rhythm, brain waves, blood pressure are measured while you sleep.

Prostacyclin

A group of molecules naturally found in the body with various functions including vasodilation, inhibition of platelet aggregation and smooth muscle proliferation; synthetic prostacyclins are used to treat PAH.

Prostacyclin analogue

A synthetic chemical that acts in the same way that natural prostacyclins do.

Pulmonary arterial pressure

The blood pressure in your pulmonary arteries

Pulmonary arteries and veins

Blood vessels in the lungs that carry blood to (arteries) and away (veins) from the lungs

Pulmonary embolus

(pl. = emboli) a blood clot in one or more pulmonary arteries that arises somewhere else in the body and travels to the lungs; usually forms in the deep veins of the legs and travels up to the pulmonary arteries

Pulmonary function tests (PFTs)

A series of breathing tests that allow measurement of your lung function, including lung capacity, how fast you can expel air from your lungs, and how well your lungs exchange gases.

Pulmonary hypertension

Elevated blood pressure in the pulmonary arteries

Pulmonary valve

A valve in the heart that separates the right ventricle from the pulmonary arteries preventing blood from flowing backwards into the right ventricle

Pulmonologist

A doctor who is specially trained in diseases of the lungs and breathing

R

Red blood cell

A type of cell in your body found in the blood whose function it is to carry oxygen to all the tissues of the body

Resistance

The opposing force to the flow of a fluid, such as blood

Right heart (cardiac) catheterization

A procedure during which a catheter (small flexible tube) is advanced from a vein in your arm or leg to the right side of the heart and into the pulmonary arteries in order to measure pressure

S

Saturation

A measure (in percent) of how close to full your red blood cells are with respect to their oxygen-carrying capacity.

Spirometry

A measurement of how fast you can expel the air in your lungs; usually a part of PFTs.

Subclavian vein

A large vein in the upper chest (between the shoulder and neck) that carries blood back to the heart and is often used in right heart catheterization or as a site for a long-term indwelling catheter through which medication may be infused.

Superior vena cava

One of the largest veins in the body, located in the chest; all blood in the upper body (above the heart) travels through this vein which then empties into the right atrium.

Systemic blood pressure

The blood pressure in the main arteries of your body that run from the left side of your heart to the rest of your body (arms, legs, intestines, kidneys, brain, etc)

Systole

The period of time when the heart is contracting and pumping blood to the lungs and the rest of the body

Systolic blood pressure

(the top number of the reading) is measured during systole

T

Thromboemboli

Blood clots that travel from the location they originally formed to another area of the body, i.e. clots in the legs that travel to the lungs

Thrombus (thrombi)

Blood clots

Trachea

Also known as the "windpipe"; a tube-like structure that carries air from the mouth to the lungs

Tricuspid valve

A valve in the heart that separates the right atrium from the right ventricle preventing blood from flowing from the ventricle backwards into the atrium

V

V-Q (ventilation-perfusion) scan

A nuclear imaging study that looks at how well areas of the lungs that have air delivery (ventilation) are receiving blood flow (perfusion); usually used to diagnosed pulmonary emboli that cause decreased perfusion

Vasodilation

Widening or dilation of blood vessels.

Vasodilator

A chemical (natural or synthetic) that dilates blood vessels

Ventricle

The two larger chambers (one on each side) of the heart; the right ventricle pumps blood to the lungs for oxygenation and the left ventricle pumps freshly oxygenated blood to the rest of the body.

Vitamin K

A natural vitamin that is found in green leafy vegetables; it counteracts the blood thinning effects of warfarin®.