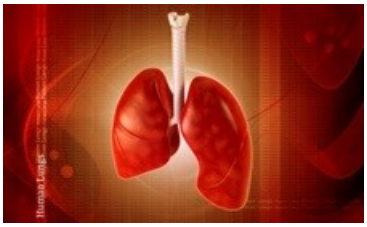
# New treatment for pulmonary arterial hypertension in SA

Nineteen-year-old Jenna Lowe of Claremont, Cape Town, has a very rare condition called primary pulmonary arterial hypertension (PAH), a degenerative, life-threatening condition. Her doctor, Professor Paul Willcox, a pulmonologist at the University of Cape Town Private Academic Hospital (UCTPAH), says PAH is a disease in which the pulmonary (lung) arteries thicken, become obstructed and stop functioning properly. This places pressure on blood circulation in the lungs and can cause the heart to become enlarged and eventually fail.



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Just a few years ago, there was no effective treatment for this condition and sufferers could expect to live just two or three years after the onset of the disease. Today, there are a number of different treatments, one of which is the medication Lowe is currently being treated with, Flolan (IV epoprostenol), which helps to open the blood vessels and keep them healthy.

Prof Willcox observes that Lowe is the first person in Africa to be treated with IV epoprostenol. "It is not a new treatment and has been approved for use in a number of countries since the mid-1990s. However, it is an extremely expensive therapy. It is also a very difficult to administer as it is a lifetime therapy that requires uninterrupted infusion. It is continuously administered intravenously through a surgically implanted catheter by a portable, battery-operated pump. These are some of the reasons why IV epoprostenol, which has long been approved by the Food and Drug Administration (FDA) in the US, has never been registered with the Medicines Control Council (MCC) in South Africa. However, the treatment can be very effective and in many cases has obviated the need for sufferers to have a lung transplant. Special permission was therefore sought from the MCC for Jenna to use it."

## International support

After she was diagnosed with primary PAH in 2012, her parents swung into action to provide whatever support they could find for their daughter. Her mother, Gabi Lowe was instrumental in pulling together a medical team that could provide the advice and care needed for her daughter.

Along with Prof Willcox this included Dr Paul Williams, a pulmonologist and transplant specialist from Netcare Milpark

Hospital in Johannesburg. She also looked overseas for help and was able to enlist the assistance of Dr Anne Keogh from St Vincent's Hospital in Sydney Australia and world-renowned PAH expert, Dr David Badesch of the University of Colorado in the US. Dr Badesch flew out to South Africa in December 2013 and, free of charge, provided the necessary expertise to get Jenna started on her IV epoprostenol treatment. He also met local physicians, who have an interest in PAH and its treatment, gave a lecture in Cape Town and participated in a symposium in Johannesburg.

"Jenna's treatment would never have been possible without this team and we are extremely grateful to them all. We are also forever in the debt of Dr Frank Gray, PAH lead physician at GlaxoSmithKline in the UK, who has so kindly provided Jenna with an unlimited supply of IV epoprostenol on a compassionate basis," says Gabi Lowe.

Prof Willcox says Lowe is doing well on her treatment, but suggests it is too early to determine conclusively whether the epoprostenol is benefitting her. "She has only been on the medication for eight weeks and it is likely to be three months or so into treatment before we are able to tell if it is improving her condition, as the dose has to be built up gradually. However, we are hopeful that it is going to make a substantial difference in the life of this young woman."

## Trust to support sufferers

Despite her debilitating condition, Lowe remains a positive and highly dynamic individual who is unwavering in her determination to live life to the full and change the society in which she lives for the better. One of her goals is to make more South Africans aware about PAH and to this end she has, with the help of her mother, established the Jenna Lowe Trust. The Trust aims to help sufferers get earlier diagnosis and have access to the treatment they need and raise funds for Lowe's medication. She says PAH is often mistakenly diagnosed as asthma, which leads to a delay in the sufferer receiving appropriate treatment.

Prof Willcox confirms that PAH is difficult to diagnose, even for experts, because screening and tests for the condition are often unreliable and inconclusive. After many years of misdiagnoses at different medical centres, Prof Willcox was able to confirm that Jenna indeed had PAH early in 2012. By this time, she could not walk more than a few metres without feeling completely exhausted.

Her efforts to create greater awareness of PAH in South Africa are paying dividends. Dr Greg Symons, a pulmonologist and clinical researcher at the UCT Lung Institute, with the assistance of Prof Willcox and Gabi Lowe, is establishing a specialist PAH unit at UCTPAH for the diagnosis and treatment of the disease. He says while the condition is rare, it is also very likely under-diagnosed in South Africa. He agrees that it is important for both physicians and the public to be made more cognisant of the disease so that it can be identified and treated early.

Lieselle Shield, manager of UCTPAH, says Lowe is a brave young woman and a role model to all South Africans. "PAH can be an extremely debilitating disease and sufferers in South Africa need to know that they are able to find help. UCTPAH is honoured to be working alongside skilled physicians such as Prof Willcox who are determined to take forward the fight against PAH in South Africa," she concludes.