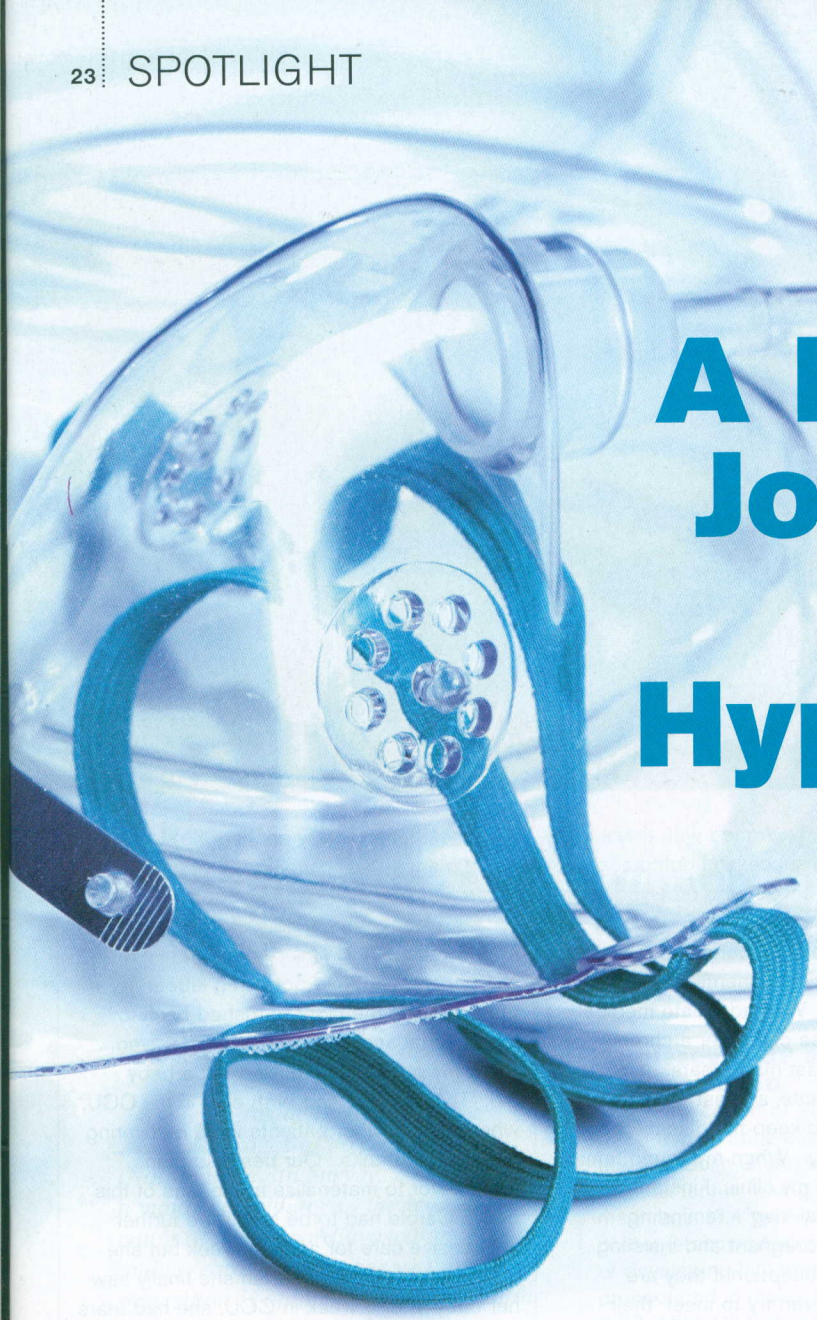


A Physician's Journey with 'The Other Hypertension'

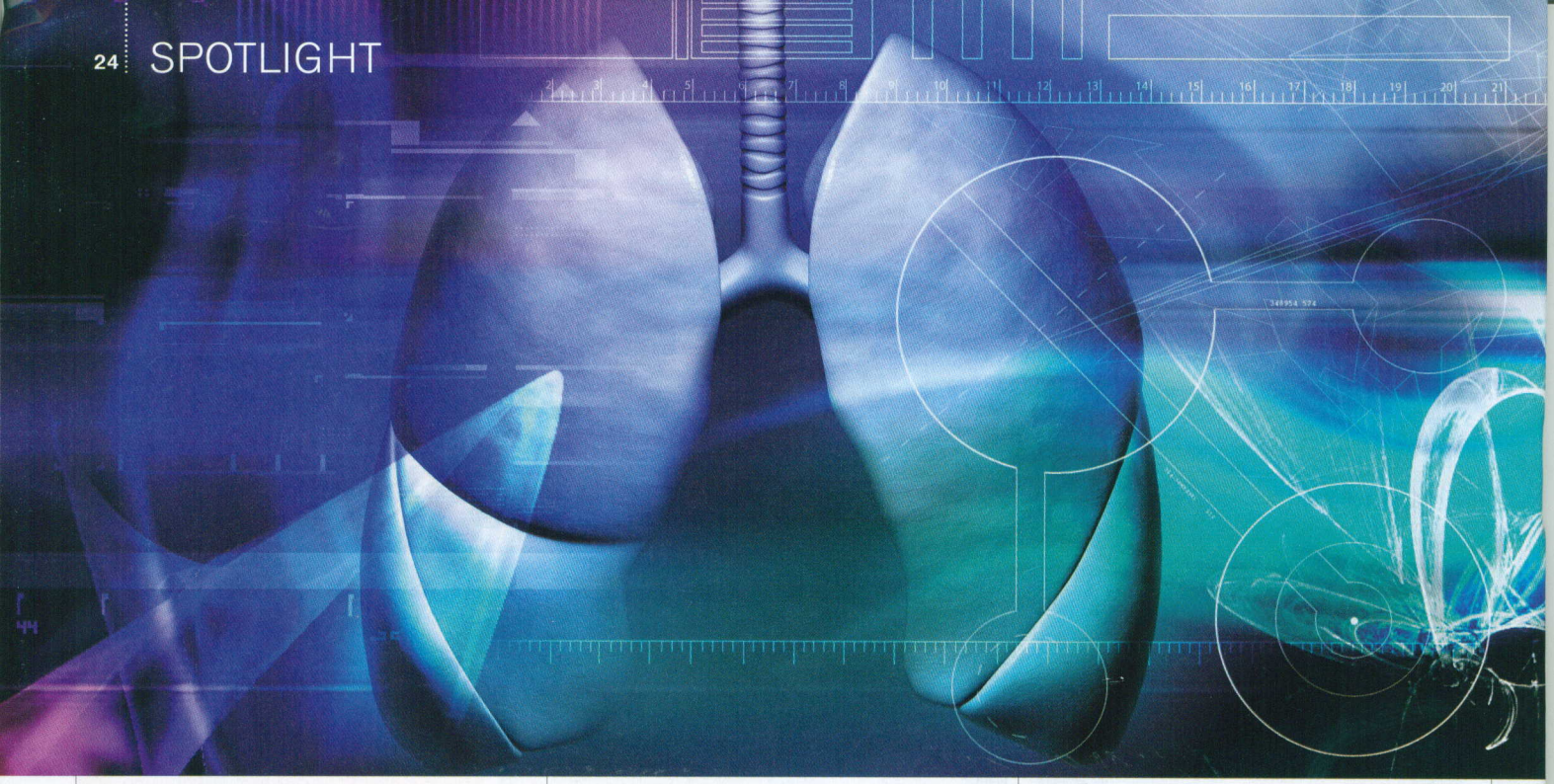
Pulmonary Arterial Hypertension

'The Other Hypertension'

Many are familiar with the humble sphygmomanometer which is present in every physician's office to measure systemic hypertension. But not many are familiar with 'the other hypertension' or pulmonsary hypertension, which is difficult to measure; hidden away - save with specialised ultrasound equipment or invasive catheter techniques. There are a myriad of affordable treatments options for systemic hypertension, but for pulmonary hypertension; until the last decade, lung transplantation remained the only option for those affected.

One of my greatest joys is to help women with heart disease realise their dreams of giving birth and becoming a mother. This might seem very basic, but for women who suffer from pulmonary hypertension, they may have a risk of dying from pregnancy which may exceed 50% in some cases. When I was a young specialist, I had a young 22-year-old Malay woman named Siti *(not her real name)* who had pulmonary hypertension who was pregnant a second time despite my advice. She appeared for review very late in pregnancy and there was very little we could do. Her heart was failing and we had to deliver the baby by caesarian section. She was in a critical condition after the baby was successfully delivered, but she never got to see her child as she passed away just 2 days later. This episode left a lasting impact on me as a doctor and I spent one and a half years training in Canada in 2001 to further my training in congenital heart disease and pulmonary hypertension.

Idiopathic pulmonary arterial hypertension (iPAH) is a rare disorder, which afflicts 2 per 10,000 individuals. Without treatment, the median life expectancy is only 2.8 years from diagnosis which is worse than some cancers. Pulmonary arterial hypertension (PAH) encompasses a much larger group with aetiologies as diverse as congenital cardiac shunts, collagen vascular disease, portal hypertension and HIV infection. Some slimming drugs like fenfluramine have been associated with idiosyncratic reactions causing pulmonary hypertension. This condition can also be inherited in an autosomal dominant fashion from parent to child in 20% of cases of iPAH. The typical affected patients are young women in their second or third decade of life, striking them even before their prime of life. Often, they present with just breathlessness on exertion, but in the later stages, would have leg and abdominal swellings or fainting spells.



Treating iPAH

The key to diagnosis is having a high index of suspicion and sending the patient for echocardiography. The gold standard for measuring pulmonary pressures is with a catheter in the pulmonary circulation. Cardiac catheterization is usually necessary to confirm the diagnosis, exclude left heart disease as a cause and to apply a pulmonary vasodilation test which identifies a small subset of PAH patients which may respond to high dosage calcium channel blockers.

The proliferation of treatment options has expanded dramatically in the past 10 years. I had very few therapeutic options to offer Siti when she first presented to us. Lung transplantation was offered as a last hope for salvation in this group of patients. In Canada, I saw the wonders of a medication called epoprostenol. Patients would come to clinic in a wheelchair, dragging a tank of oxygen behind them on one visit, and one month later after treatment - walking in without assistance and asking us when they could go back to work again. This treatment, however, had a serious drawback in that the medication had to be delivered with an infusion pump through a central line continuously (like chemotherapy) 24/7. At that time, we started trying out new oral medications like Bosentan and Sildenafil *(yes – the blue pill called Viagra® used for erectile dysfunction)* which overcame the technical problems of Epoprostenol.

Another novel mode of treatment is with an inhaled drug called iloprost which is nebulized from a battery-operated device 6-8 times a day. These medications, however, are very expensive and may cost S$500 to $6000 per month. Armed with these new therapies, the successful outcomes for PAH exceeded those who received lung transplantation in the past. Today, many patients who have been successfully treated have been permanently taken off the transplant list. With adequate medical therapy, the disease can enter a chronic phase which can last many years. In a quarter of my patients, at least 2 drugs have to be given to keep the disease from progressing rapidly. When my young female patients see me in my clinic, I instantly become a perpetual 'nag' - reminding them never to get pregnant and insisting on adequate contraception if they are sexually active. I even try to meet 'their significant other' to stress the dangers of pregnancy for this condition. Despite these measures, the urge to procreate still exists.

Pregnancy and Pulmonary Hypertension

Two years ago, another young patient, Carol *(not her real name)* with pulmonary hypertension came to me late in pregnancy. She had been seeing a doctor regularly for her antenatal checkups but was not diagnosed as having heart problems before this. She was severely breathless and oxygen levels were very low due to a hole in her heart. I admitted her to my coronary care unit (CCU) and started her on a special inhaled medication called nitric oxide and sildenafil to relax the blood vessels in the lungs. My plan was to stabilise her for a few days in the CCU, then electively plan a Caesarian section to deliver the baby. I discussed with my team of obstetricians and anesthetists every detail of how we could manage her condition to optimise her outcome. One day before our plan was to take place in the middle of the night, I received a call from my registrar that Carol, without warning, had gone into labour in the middle of the night. I rushed back to the hospital and by the time I had arrived, I saw my obstetrician pulling out a baby girl. This was the first birth ever in my CCU, where all the other patients were recovering from heart attacks. Our best-laid plans were never to materialise in the face of this baby. Carole had to be stabilised further in intensive care for another week but she made good recovery. When she finally saw her baby after a week in CCU, she had tears of joy streaming from sides of her face.

Carol is still seeing me, but her health is still frail. Her beautiful 2-year-old girl comes with her each time, reminding me of why I am still in this business. MG

Dr James Yip graduated from the National University of Singapore in 1991. He has been a Fellow of the Academy of Medicine, Singapore since 2001. He trained in Adult Cardiology at the National University Hospital (NUH) in Singapore from 1995-2001, and was a clinical fellow at Toronto Congenital Cardiac Centre for Adults 2001 to 2002. He was also a fellow in the pulmonary hypertension unit at Toronto General Hospital from 2001 to 2002.

Dr Yip is currently the Director of the Adult Congenital Heart Programme in the National University Heart Centre, Singapore (NUHCS) and Senior Consultant in the Department of Cardiology. He runs the pulmonary hypertension service and is actively involved in clinical trials for this condition. His other interests include pregnancy and heart disease, Marfan syndrome, Hypertrophic Cardiomyopathy and medical informatics. Dr Yip is also the Chief Medical Information Officer at NUH.