First Advanced Pulmonary Hypertension Treatment in African History

A House Call to Cape Town
by Todd Neff - UCH Insider

Imagine that your child, healthy and athletic until a year ago, was now dying of a degenerative condition before your eyes. Imagine a proven therapy had been available for 20 years, but that no one in your country – or even on your continent – had ever been availed of it. Imagine further that even if you could get treatment, the drug alone cost $100,000 a year and that obtaining and administering it would take a government waiver and a tricky combination of drugs, equipment and supplies from three countries, the closest being 6,000 miles away.

Here’s what one individual faced with these real-world dilemmas did.

In Cape Town, South Africa, Gabi Lowe’s daughter Jenna danced and swam and did well in school. But when she was 17, a misdiagnosis of asthma turned into something far more serious: pulmonary hypertension (PH). The disease narrows the arteries from the right side of the heart into the lungs, starves the body of oxygen, and dangerously enlarges the cardiac muscle.

Many problems can cause PH: heart and blood vessel diseases; lung diseases; liver diseases; connective tissue diseases, such as lupus and scleroderma; thyroid diseases; and HIV infection, to name a few. But sometimes, as in Jenna’s case, the cause is unclear. Without treatment, the average life expectancy is three years, according to the U.S. Centers for Disease Control and Prevention.

With the right therapies, however, patients live longer. The U.S. Food and Drug Administration has approved a dozen PH drugs since the early 1990s. For those in advanced stages of the disease like Jenna, GlaxoSmithKline’s Flolan (epoprostenol) is the gold standard. That’s the $100,000-a-year drug.

David Badesch, MD, knows Flolan well. The director of University of Colorado Hospital’s Pulmonary Hypertension Program was involved in the trials that led to its approval in 1995. The center has about 100 patients on Flolan, he said, from all over the Mountain West. His center is one of the largest in the world, having been involved in the clinical trials leading to the approval of all 12 of the approved PH therapies. Jenna’s case would be among the most challenging Badesch had ever faced.

Connecting. Gabi Lowe found Badesch in mid-2013 as she sought out the world’s top PH physicians. They exchanged hundreds of emails, some with medical reports and images, as Badesch “tried to assess the patient’s clinical situation from halfway around the world,” as he put it.

Badesch is a busy man. If you happen to be doing a story that involves the Pulmonary Hypertension Center, he will conscientiously call you back, but probably well after typical office hours. He will still be in the office, and he will probably also urge you to mention the excellent work of his colleagues.
It was quickly clear to him that Jenna needed to be on Flolan. Badesch and his team considered flying her to Colorado for treatment. The problem, he said, was that “she might get started on something and go back there and no one would know how to take care of her.”

That’s because Jenna is the only patient in all of Africa on the drug. But Badesch reasoned that if he and a nurse savvy in the complexities of mixing and administering the drug went to South Africa, they could teach others how to handle Flolan and help pave the way for the drug’s use in South Africa.

Debra Zupancic, FNP, a Pulmonary Hypertension Center nurse practitioner, was enthusiastic about the idea. She and Badesch decided they would go to South Africa in December 2013. But they would have to have a drug to administer first.

Gabi had already contacted Frank Gray, MD, GlaxoSmithKline’s lead PH physician in the United Kingdom. After Badesch followed up and explained the circumstances, GlaxoSmithKline decided to supply Jenna with Flolan for the rest of her life.

**Herding time.** But that was only a crucial first step. Gabi had to convince the South African Medicines Control Council to approve importing the drug, and she had to find a pharmaceutical importer willing to transport it for a single patient. She also had to find a suitable pump, which eventually came from Australia, and tubing and other supplies, ultimately sourced in England and the United States, courtesy of Zupancic.

There was also the challenge of administering Flolan, which must be mixed once a day in a 35-step process done in sterile conditions and administered continuously through a central line. It must be kept cold even as it flows. Lowe’s family installed a sink and otherwise modified a spare room across the hall from Jenna’s bedroom and turned it into what they called the “drug den.” Gabi didn’t stop there.

“To help Jenna get what she needs, I’m going to have to change the face of pulmonary hypertension in South Africa,” she told Badesch.

The family set up the Jenna Lowe Trust to help defray costs and advance pulmonary hypertension care in their country. Jenna’s younger sister Kristi co-wrote with Jenna and then recorded a song, “I Need More Time,” which hit number three on South African iTunes when it came out. Since, there have been media appearances and profiles of the family’s courage in the face of PH.

Gabi arranged sponsorships to cover Badesch’s and Zupancic’s flights to Cape Town and organized the first pulmonary hypertension symposia ever held in the country; one in Cape Town, the other in Johannesburg.

Badesch headlined them and Zupancic was a featured speaker during their week-long visit in December.

**On the ground.** Badesch and Zupancic had little time to see the sights of a land 9,400 miles from the Anschutz Medical Campus. Zupancic taught the Lowe family and UCT Private Academic Hospital caregivers the intricacies of mixing and dispensing Flolan, sorting out quirks with pump programming and catheter lines along the way. Badesch worked with Lowe, but also did patient rounds. One medical resident, an African, observed the attention being showered on this one patient with a rare disease and asked Badesch pointblank how he reconciled that with the plight of the 6 million HIV/AIDS patients and all the tuberculosis patients and so many others receiving insufficient medical care in South Africa.

Badesch didn’t have an immediate answer, besides noting the medications were free. But he thought for a moment and said, “How can you deny a young woman with a disease of no fault of her own just because it’s expensive?”

In an email, Gabi described Badesch as “selfless, calm and dedicated” to the best treatment for her daughter. “His commitment from across the miles has been extraordinary to say the least,” she said.

Jenna, who has been on Flolan for about six months now, now rides a motorized three-wheeler she named “Chase” (her primary oxygen machine goes by “Thunder”) around the University of Cape Town, where she’s a scholarship student who occasionally asks men to please carry her up the stairs. Now 19, her breathing, though still a struggle, has stabilized, and there are “very small, incremental signs of improvement,” as Gabi put it.

Jenna bears the indignities of side effects ranging from flushed cheeks to joint pain. She aims to raise awareness of the disease and help others with PH. She has been approved for a double lung transplant, though Gabi said the family “hope to buy as much time as possible with the Flolan first.”

Badesch and Zupancic both described their efforts in South Africa as some of the best, most productive work they’ve ever done. Neither intends to stop now. Badesch is headed back in late May to attend a two-day meeting in Johannesburg, the goal being to develop guidelines for pulmonary hypertension patients.

“With Dr. Badesch’s help, we really are changing the landscape of PH in South Africa,” Gabi said.

*This article was written by Todd Neff and initially appeared in the May 13, 2014 issue of the “University of Colorado Health Insider.”*
The Pulmonary Hypertension Association Visited Indianapolis for International PH Conference
June 20-22, 2014

An earlier version of this article, by Rebecca Gifford, a PHA Meeting Planning Associate, appeared in the Pulmonary Hypertension Association Newsletter.

PHA's 2014 International PH Conference and Scientific Sessions took place June 20-22, 2014, in Indianapolis, Indiana. Known as the “Racing Capital of the World,” Indianapolis was the perfect setting to host the 2014 Conference, themed Racing Toward a Cure. We celebrated and acknowledged the determination and progress our PH community has made toward the ultimate finish line: a cure.

PHA’s Conference, the largest gathering of the pulmonary hypertension community in the world, brings together patients, caregivers and family members, nurses and other allied health professionals, physicians, researchers and leaders of PH associations from around the globe. The Conference provides three days of education, networking and support for the global PH community.

For many attendees, Conference is the first time they’ve met another PH patient, another caregiver, another kid with PH. For those who are newly diagnosed, Conference is the starting line at which they begin their race to combat this disease.

“PHA has incredible momentum right now. There is a hustle. So much is being done, but we have to do more,” says Diane Ramirez, a PH patient and co-chair of the 2014 Conference Communications Committee.

Conference is the perfect time to celebrate this forward momentum and build on our advances for the future. And we have a lot to celebrate and build on! The PH community has been active in all aspects of the race toward a cure this year, from the debut of three new drugs to the introduction of a PH-specific bill in Congress and everything in between; we continue to persevere, not once taking our foot off the gas.

Kristine Green, a 2012 Conference attendee, describes the vitality of the event and the PH community. “Everyone truly embraced the opportunity to interact, learn and share,” Kristine says. “PHA has created a community of patients, caregivers, researchers and physicians dedicated to helping each other and making advances to finding a cure.”

New Dedicated Pulmonary Hypertension Nurse Phone Line

Beginning September 15th, (720) 848-6622 will be the new direct phone number to reach a pulmonary hypertension triage nurse. Office hours are 9am to 5:30pm. Messages left after 5:30pm will be returned the next business day. If you have a life-threatening emergency, please call 911. If your call is urgent (and you need to speak to the pulmonary hypertension MD/RN on call), they can be reached through the UCH operator at (720) 848-0000.

For medication refills or oxygen requests, please direct your pharmacy or oxygen company to contact fax (720) 848-0480. To seek release of your medical records, HIPPA regulations require that you please contact the UCH Medical Records Department at (720) 848-1031 or fax (720) 848-5551. Patients needing to schedule/cancel an appointment are asked to call the Cardiac and Vascular Center at (720) 848-5300, option 1.

Pulmonary Hypertension Patients to Receive Their Medication List at Check-In

One improvement that pulmonary hypertension patients will notice when they check in at the clinic is that they will receive a printout of their current medications. The reason for this is that patients will be asked to review the list and bring it up to date, adding any new medications, crossing out any discontinued medications and noting any change in dosages. This updated list will accompany the patient to the examination room to ensure the patient’s electronic health file is updated. It is always best to bring all the medications in their original containers so drug, strength and dosing can be accurately captured.
Deb McCollister to Help Launch University’s New “Clinical Trial Management System”

Pulmonary Hypertension Advocate to Assume New Position at University

The good news for Deb McCollister is that she has accepted an exciting new position with the team that will be implementing the university’s recently-purchased “clinical trial management system.” The bad news is that she will be leaving the pulmonary hypertension department, where she has been such a reliable, driving force these past ten years.

“I’ve really loved every minute of my time here in pulmonary hypertension, working with all the staff and patients, but this was too good an opportunity to pass up.”

A Clinical Trial Management System (CTMS) is a customizable software system used by clinical research institutions such as the University of Colorado to manage the large amounts of information in a clinical trial. Deb is intimately familiar with clinical trials because she has been running them for Dr. Badesch and his colleagues for many years. “When I started with the program in 2003, I had maybe two or three PH studies going on at the same time, and now over the years it’s blossomed to 15+ trials going on at any one time.”

As background, Deb explained that the cost of transitioning a new drug from discovery through clinical trials to approval is complex—and expensive. A full series of trials can cost many millions of dollars, and consequently pharmaceutical companies don’t entrust their drug trials to just anyone. Physicians are evaluated to ensure that they have an established number of pulmonary hypertension patients and adequate staff to support the projects, and that they’ll provide reliable, accurate data. “Dr. Badesch is well known as a very conservative, thoughtful physician, and he is invited to participate in virtually every single pulmonary hypertension trial. As interest in the field has grown, so has the number of PH clinical trials. This ultimately helps our patients in the long run as more drugs are approved through this process. I’ve been fortunate to play a part in drug development.”

When asked about her new job, she says “I will be part of a team implementing the new clinical trial software across the entire university enterprise. My specific responsibility will be to work with the cancer center, using this new software system to fit into the unique way in which they do their work. Additional members of the team will do the same for other University programs, such as neurology, cardiology, orthopedics and ophthalmology.”

During her time in the PH Program, Deb was a very active member of PHA and the PH community. She was the recipient of the 2010 Outstanding Medical Professional Award, presented at PHA’s 9th International PH Conference and Scientific Sessions in Garden Grove, Calif. She has also served on the PH Professional Network (formerly PH Resource Network) Executive Committee and helped to plan both the 2007 and 2009 PH Resource Network Symposia.

When asked what she was proudest of in her work in pulmonary hypertension, she replied: “Co-chairing two symposia for pulmonary hypertension was very satisfying, but what I am proudest of is helping start our first Colorado PH fundraiser in 2008.” The fundraiser took the form of a dinner gala held at Belmar in Lakewood and had an Italian theme. “And then in 2010 we launched our first “PHun Run” (Pulmonary Hypertension Run). It was held on the Anschutz Medical Campus. I liked the fact that more patients and family members could join in. I’ve been impressed that the event has grown in volume year after year by at least 20%.”

While Dr. Badesch is sad to see Deb go, he is also thankful for having her help in taking the University of Colorado PH Program to the next level. “Deb has provided outstanding leadership for our clinical research program over the past 10 years. She has overseen a period of tremendous growth in the program. What has been most impressive, though, are her contributions above and beyond managing our clinical trials. She's been a leader within the Pulmonary Hypertension Association at a national level, organizing two large conferences with attendees from across the country, and locally she's helped to lead our annual awareness and fundraiser event. That terrific event will undoubtedly be her legacy. We'll miss her, and we wish her all the best in her new role.”

Deb McCollister, RN, BSN

Robin Hohsfield, Deb McCollister, Kelly Moulden and Lisa Nicotera (left to right) at the special event PHA On The Road: PH Patients and Families Educational Forum held in Denver on June 1, 2013.

Jason Alexander and Deb McCollister at the Pulmonary Hypertension meeting in New York City in 2009. For many years Jason has unselfishly lent his star power to help in the fight against the disease.

Beth Coleman (left) of Children’s Hospital with Robin Hohsfield (center) and Deb McCollister (right) of University Hospital, organizers of the first “Run for PHun,” held in 2010 on the Anschutz Medical Campus.
Debra Zupancic receives “Doctor of Nursing Practice” degree from the University

On Friday, May 23rd, Debra Zupancic received her degree of “Doctor of Nursing Practice” from the University in an outdoor ceremony presided over by Bruce Benson, President of the University of Colorado and Don Elliman, the Chancellor. The Doctor of Nursing Practice (DNP) is a terminal professional degree that focuses on the clinical aspects of a disease process. The curriculum for the DNP degree generally includes advanced practice, diagnoses, and treatment of diseases. The DNP is intended to prepare a registered nurse to become an independent primary care provider and is intended to be a parity degree with other health care doctorates such as psychology, medicine, and dentistry.

As a part of her degree program, Debra produced a research paper entitled: “Reducing 30-Day Hospital Readmissions for Patients with Pulmonary Hypertension and Right Heart Failure: A Quality Improvement Project.”

“All hospitals are trying to reduce readmission rates,” says Debra Zupancic. As she wrote in her paper: There are certain hospital readmissions—such as for myocardial infarction, pneumonia, and heart failure—that occur more frequently than others and are inordinately costly to the entire healthcare system. We in the pulmonary hypertension program have recently conducted an in-depth quality-improvement initiative with the aim of reducing 30-day readmission rates for our patients admitted for pulmonary hypertension and right heart failure, a subset of heart failure. Our aim is to reduce readmission rates and to improve our patients’ lives. The first step in reducing hospital readmissions was to conduct a comprehensive survey; the survey results highlighted several important steps for improving our comprehensive discharge practice: 1) Increase our patient-centered education regarding appropriate lifestyle modifications—such as sodium and fluid restrictions; 2) Emphasize increased physical activity, as appropriate; 3) Initiate follow-up phone calls within 24-48 hours of discharge to enable rapid detection of problems before they become so severe as to require re-hospitalization; this also allows for early intervention if necessary; 4) Schedule the patient for rapid post-hospitalization follow-up visits. The adoption of the above measures was successful in reducing our 30-day hospital readmission rates for patients with pulmonary hypertension/right heart failure from 12.4% to 7% in the first quarter of 2014. Though the information above refers specifically to pulmonary hypertension patients who were admitted to the hospital on an inpatient basis, the self-care management and education information they received is beneficial to all pulmonary hypertension patients, and can be summarized as follows.

Pulmonary hypertension causes the body to hold onto excess fluid, which causes a big problem for the strained right ventricle of the heart. Accordingly, it is common for pulmonary hypertension patients to be instructed to eat low-salt diets or to consume less than two grams of salt per day. Pulmonary hypertension patients are also instructed to restrict their total fluid intake to less than two liters (2000cc) per day to help avoid swelling and fluid retention. Many patients are surprised to find out they actually drink too much water. In the past they had always heard that drinking lots of water was the key to staying healthy. Prior to discharge, the patients received a weekly and a monthly chart designed to help them track their daily intake of sodium and fluid, as well as their weight. Many patients indicated that a better understanding of their dietary restrictions—both sodium and fluid—affecting their condition made them feel “more secure” during their follow-up appointments.

Dr. David Badesch, who heads up the pulmonary hypertension program, was very complimentary of Debra’s recent efforts: “We're very fortunate to have Debbie with us. She's loved by all of our patients, and provides outstanding care for them. She always goes the extra mile to address their needs. And in addition to the time and effort she devotes to her clinical work, she's been able to pursue further education and obtain the highest degree offered in her field. Truly amazing. Congrats, Debbie!”

“Run for PHun” to be Held in Denver’s City Park on Sunday, Sept. 21, 2014

The fifth annual “PHun Run” (Pulmonary Hypertension Run) for Colorado will be held in Denver’s City Park on Sunday, September 21, 2014. The event benefits the University of Colorado Pulmonary Hypertension Program, the Pulmonary Hypertension Program at Children’s Hospital Colorado, and the Pulmonary Hypertension Association (PHA). Sign up today via the QR Code to the right or link below: https://www.firstgiving.com/phassociation/ColoradoPH_InThePark

Debra Zupancic shows her happiness on graduation day
Meet Bruce Hoskinson, the Pulmonary Hypertension Center’s ‘guardian angel’

Super Volunteer Brings Wealth of Experience to the Job
by Todd Neff - UCH Insider

If there were Oscars for hospital volunteers, Bruce Hoskinson would have a pretty good shot in two very different categories. One would be Most Interesting Background; the other, Most Organized and Thorough.

Hoskinson was, in fact, recently lauded, but he received a nice plaque rather than a little golden man. It was for his 1,000 hours of volunteering with University of Colorado Hospital’s Pulmonary Hypertension Center since mid-2012.

Hoskinson may wear the maroon vest of a volunteer, but he’s become a core member of the team, said Deb McCollister, RN, BSN, a research instructor with the group. “We offered Bruce a paid job, and he said no,” she said. “I’m a good value for the money,” Hoskinson laughed.

Man of action. Hoskinson, 70, worked for pay in Hollywood, Europe and the Middle East for decades, learning Italian and a bit of Arabic to go with his fluent French along the way. Now he would rather just help out and come and go as he pleases, he said. He occupies his cubicle in the Pulmonary Hypertension team’s fifth-floor Leprino Building haunts most of the day Tuesdays through Thursdays.

When on duty, his work is diverse. Foremost, he tracks blood tests of 114 patients from multiple commercial labs. That’s a critical task, McCollister said, because pulmonary hypertension drugs can harm the liver. Hoskinson’s careful tabulations also free up the team’s eight nurses to focus on clinical trials and patient care. He’s working on calendars with which patients can track sodium and liquid intake, too.

In addition, Hoskinson has become the team’s informal director of morale-boosting and team-building. He learned page design to create the center’s first newsletters, then applied his new skills to design posters with collages of photos — aligned in neat grids — commemorating team outings. The posters bear such titles as “Gala Artistic Evening” and “Cherry Creek Marina and Yacht Club.” Hoskinson had the posters printed on foam-boards at Staples; they’re hard to miss among the team members’ doors and cubicle walls.

Winding road. It’s remarkable that a man fluent in three languages and who has traveled so widely can derive such obvious joy from these small acts of generosity. Hoskinson was born to a mother whose travel bug had, in her late teens, taken her from her native Denver to Washington D.C., where she met a civil engineer and fell in love. After the Second World War, the young family followed dam projects – Boysen Dam in Wyoming, then Chief Joseph Dam on the Columbia River in Washington State.
They moved back to Denver when Bruce was in the sixth grade; he graduated from Denver North High School and majored in French at Stanford, graduating after a year in Nantes in 1966.

He was drafted in 1967, and spent most of 1968 and 1969 in Vietnam, applying his French skills to gathering intelligence and working on military assistance teams. He came home and took premed classes back at Stanford, but realized that the same wanderlust that had taken his mom to Washington D.C. was in him, too.

“I’ve always been interested in medicine, but I’ve been interested in almost everything,” Hoskinson said. “I just wanted to go out and explore, and you can’t do that as a physician. You’ve just got to stick there.”

Movin’ on up. His first stop was Hollywood, where he worked at CBS Television’s script office services department. This was the 1970s, the days of “All in the Family,” “The Jeffersons,” “The Mary Tyler Moore Show,” and “Rhoda.” Hoskinson ultimately became night supervisor, proofreading, typing up and mimeographing 100 copies of scripts and delivering them to producers, directors, associate directors, cameramen, story editors and others. By 1976, he saw that writing for TV wasn’t his talent.

“How fortunate to learn early on that, hey, you’re going in the wrong direction,” Hoskinson said.

He had fallen in love with Florence, Italy while traveling during his college days in Nantes, and enrolled in the Italian MA program at Middlebury College’s campus there, spending summers on campus in Vermont. He taught English as a foreign language during that time and continued to teach while working on his doctorate in English at the University of Florence (he’s in the “all-but-dissertation” group). Hoskinson stayed a student so long in part so as not to have to pay for being a student, he said.

“Predicated on all this was the fact that I took out a lot of college loans,” he said. “You don’t have to pay those back if you’re still in school.”

Playing D. The turmoil in the Middle East following the 1979 Iranian Revolution opened up teaching positions at the American School of Kuwait. Like much expatriate work in the region at the time, it came with a free house and a car and no income tax. Hoskinson taught English composition, literature and physics for a few months and decided he liked the Middle East. In 1980, he moved to Riyadh, Saudi Arabia to work in administrative positions for U.S. defense contractors, ultimately spending 20 years with McDonnell Douglas (later Boeing), which had won a billion-dollar contract for 60 F-15 fighter jets and all the associated infrastructure, testing, maintenance and services. Hoskinson was staff assistant to the program managers–six in total.

“I was the corporate memory,” he said.

He traveled to dozens of countries along the way. He also learned a lot about health care. McDonnell Douglas had to establish services to handle 80,000 clinic visits a year by its own 1,200 employees as well as U.S. Air Force and Royal Saudi Air Force personnel and families. Among other efforts, he arranged to airlift blood tests to Mayo Medical Laboratories in Minnesota.

In 2001, the F-15 contract complete, he moved to the United Arab Emirates to work on a project to upgrade that country’s military health system. Then, in November 2001, he came home to Colorado to care for his mother, which a few years later brought him to UCH.

The UCH connection.

His mother had pulmonary hypertension, and by 2010 it had become life-threatening. A Mayo Clinic pulmonary hypertension expert, Michael McGoon, MD, suggested Hoskinson touch base with David Badesch, MD, who directs UCH’s Pulmonary Hypertension Center.

What Hoskinson brought to their first meeting sheds light on why he outlasted six McDonnell Douglas/Boeing program managers. Among other documentation of his mother’s care, he produced printouts of echocardiograms from the past five years and accompanying hemodynamic status reports, stress and treadmill studies; CT scans and chest X-rays; seven years of renal ultrasounds; six years of daily blood pressure readings; five years of his mother’s medication history; several academic articles on pulmonary hypertension he had printed after a visit to the University of Colorado Health Sciences Library; and the biography and publications of one David B. Badesch.

“He’s incredibly well-organized,” Badesch said. “He used his skills and interests in a way that benefitted his mother tremendously.”

Marian Hoskinson passed away at the age of 91 in May 2012. Bruce Hoskinson was so impressed with Badesch and colleagues that he asked how he could help. An Oscar-worthy volunteer was born.

Badesch likens Hoskinson to the Pulmonary Hypertension Clinic’s own version of Clarence Odbody, the guardian angel in “It’s a Wonderful Life.”

“He just drops in and does all these wonderful things to help people,” Badesch said. “He’s the model volunteer.”

This article was written by Todd Neff and initially appeared in the March 18, 2014 issue of the “University of Colorado Health Insider.”
DIRECTIONS FOR NEW PATIENTS

PLEASE ENTER ON THE GROUND FLOOR OF THE “AIP” LOBBY AND THEN FOLLOW THE RED DOTS

After entering the lobby of the Anschutz Inpatient Pavilion (AIP), turn left and continue down the main hallway until you reach Elevator E.

Take Elevator E to the 3rd floor and follow the signs to check-in counter.

VALET AND FREE PARKING ARE AVAILABLE IN ADJACENT LOTS