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It takes a Village

Conjoined twins from
Africa are separated by
a talented team of
health professionals.

Dr. Cindy Howard
with twins Loice and
Christine Onziga

FOR ALUMNI & FRIENDS OF THE DENTAL, GRADUATE, LAW, MEDICAL, NURSING, PHARMACY, AND SOCIAL WORK SCHOOLS

The MIRACLE of Modern Medicine

BY BRUCE GOLDFARB

THE EXTRAORDINARY JOURNEY OF CHRISTINE AND LOICE ONZIGA, CONJOINED TWINS SEPARATED AT THE UNIVERSITY OF MARYLAND MEDICAL CENTER IN APRIL 2002, BEGAN ON THE OTHER SIDE OF THE WORLD, MORE THAN 7,000 MILES AWAY IN THE REMOTE VILLAGE OF LEIKO IN THE KOBOKO AREA OF WESTERN UGANDA.

It is one of the most impoverished regions on earth, still recovering from strife and guerrilla warfare that wracked the country during the 1970s and 1980s. “The area that the twins are from is the poorest and most unstable area in Uganda,” says Sherri Shubin, MD, who was the girls’ pediatrician in the United States. “There’s no running water, no electricity, and no paved roads.”

As a senior resident in the University of Maryland Medical Center, Shubin participated in an exchange program with the Makerere Medical School in Uganda’s capital, Kampala. The medical school’s 1,500-bed Mulago Hospital, the only teaching hospital in the country, serves a region plagued with a high infant mortality rate, malaria, measles, and diseases that thrive in crowded, unsanitary conditions.

“The high incidence of acute and serious illness among children is beyond anything we see in this country,” says Cindy Howard, MD, clinical professor of pediatrics in the School of Medicine, who has supervised groups of medical students and residents since 1997.

“Practicing medicine in Uganda is a great opportunity to learn how to work with limited resources. It reminds us all why we went into medicine in the first place,” says Howard.

Senior residents in the exchange program spend two months at a time at Mulago Hospital working with Makerere Medical School faculty and conducting research. They are not usually involved directly in patient care. “Our primary responsibility is to help teach the residents there,” says Howard.

“Mulago Hospital has a great faculty, but they are completely overwhelmed by the demand for care.”

THE EXPECTANT FAMILY

Loice and Christine’s parents, Gordon and Margret Onziga, live on a farm with their four-year-old daughter, Noelle, Gordon’s parents, and his five younger siblings. The family earns about a dollar a day raising sweet potatoes, corn, and the starchy cassava root.

In October, Margret was pregnant with what the couple thought would be their second child—and planned to deliver at home. But after nearly two days of labor, exhausted and slipping into delirium, Margret was carried nearly a mile on her father’s back to the nearest taxi stand. From there, she traveled 18 miles to a hospital across the Congolese border in Aru. The hospital was unable to handle her delivery and put Margret on a bus to a hospital across the border in a town 15 miles away.

Christine and Loice were delivered Oct. 28, 2001, by cesarean section, to the amazement of everybody involved—particularly their parents, who were not expecting twins, much less conjoined twins.

Conjoined twins are extremely rare, occurring in about one of every 200,000 live births in the United States. About 200 pairs of conjoined twins are born annually around the world. For reasons that aren’t entirely clear, they are more common in Africa and India. The condition results when a single fertilized egg fails to separate into identical twins.

Like identical twins, conjoined twins are always the same gender. About 70 percent are female.

Loice and Christine were connected by tissue running from their breastbone to their navel. This condition, known as thoracopagus, is the most common form of conjoined twins. It occurs in about 35 percent of cases and almost always affects the heart.

“When I first saw that they were connected, I was surprised,” Gordon says. “It was the first time for me to see such babies. I had no idea they could be separated.”

The Onzigas intended to return home and let the babies live out their natural lives. Without surgery, about 50 percent of conjoined twins die before their first birthday. Because life expectancy is shorter in develop-



Gordon and Margret Onziga hold their daughters before surgery.



Before the separation, Christine and Loice were connected from the breastbone to the navel.

ing nations, Loice and Christine’s chances for survival were even lower.

Margret’s father said he had heard that surgery can be done to separate twins like Christine and Loice and persuaded Gordon and Margret to take the babies to Mulago Hospital, about 310 miles southeast in Kampala.

Traveling to Kampala and staying in the city would be expensive. To finance their trip, the Onzigas sold nearly everything except their modest two-room house. Gordon even sold his most prized possession: his bicycle, essential for selling their crops. Together, with contributions from neighbors and relatives, the family raised about \$350—more than their yearly income—to support their stay in Kampala.

When the twins were 10 days old, Margret, who was still recovering from the cesarean section, and Gordon left Noelle in the care of Gordon’s parents and boarded a bus for the dusty, 10-hour ride to Kampala. It was the first time either had been to the capital city.

FATEFUL ENCOUNTERS

In November 2001, Sherri Shubin, MD, was conducting a research project to study neonatal morbidity and mortality in the neonatal intensive care unit at Mulago Hospital. Working with Shubin and Howard was Sue Rhee, MD, then a pediatrics resident. The nursery was abuzz with talk of the “special” babies who had come from so far away. “We went to see them because conjoined twins are so rare and interesting,” she says.

During their first weeks, the babies showed signs of healthy growth, but not at an equal rate. Loice, the smaller infant, was not gaining weight as rapidly as Christine, because about half of Loice’s blood supply was being shunted to her sister. Also, the position of their bodies was causing scoliosis of Loice’s spinal column. As they got older, the discrepancy in their size and weight would increase until their small bodies could no longer tolerate the stress.

Gordon and Margret were devastated to learn that an



The 12-hour operation was performed on April 19, 2002.

operation to separate the twins was far beyond the rudimentary capabilities of Mulago Hospital and that no hospital on the continent was up to the task.

Howard and Shubin had met and talked to the Onzigas, and knew their options were running out. One of the doctors at Mulago asked Howard if she would take Christine and Loice to the University of Maryland Medical Center. It was their only hope.

"There was nowhere else to go," Howard says. "If we didn't take them, they would die."

Howard called Jay A. Perman, MD, chair of the Department of Pediatrics in the School of Medicine, and presented the case for the babies. "There weren't resources in Africa to do the operation," says Perman. "There weren't even resources in Kampala to determine whether surgery was feasible."

Perman took the matter to decision-makers at the School of Medicine and the University of Maryland Medical Center, and both agreed to waive charges for the surgery.

The medical center hadn't attempted such a surgery since 1986, when pediatric surgeon J. Laurance Hill, MD, professor of surgery at the School of Medicine, led a 24-member team that separated two-month-old thoracopagus conjoined girls who shared a liver, diaphragm, chest wall, and heart sac. Those twins are now healthy 16-year-olds in Baltimore, but none of the surgeons involved in the Onziga case had ever separated conjoined twins. Hill, now chief of the division of pediatric surgery, was not directly involved in the Onziga surgery, but provided advice to the team.

"Nobody has a lot of experience with these cases," says cardiac surgeon and assistant professor Marcelo Cardarelli, MD. "If you're lucky as a surgeon, you may be involved in one case like this in your entire life."

As a teaching case, Christine and Loice presented a

unique opportunity for the staff to work together as a large team, and to gain experience that can ultimately help other medical center patients.

"What is learned in a teaching institution in caring for children like these carries over to related situations," Perman says. "Another set of conjoined twins is probably going to be born in or near Maryland, and as a medical school I think we all—students, faculty, and patients—benefit from this experience."

"We're a tertiary care center, and we're supposed to teach and do complex surgery," says Eric Strauch, MD, the assistant professor of surgery at the School of Medicine who coordinated the 35-member surgical team. "The Onzigas came to our attention, and we decided that this was something we wanted to do."

The babies arrived in Baltimore with their parents in February 2002 for extensive diagnostic imaging tests to learn about their unique internal anatomy. The babies had a fused liver and shared a diaphragm along with the breastbone and chest wall. Their hearts were twisted from their normal position and enclosed in a common sac.

"We believed that we could separate them with a fair chance of survival," says Cardarelli, "but with a 20 percent chance of them dying."

One week before the surgery, the team held a dress rehearsal in the operating room to plot the position and movement of all the principals using two life-size cloth dolls sewn together like Loice and Christine. The team had to figure out how to position the operating tables, where to place instruments, and where everybody would stand. The operating room was crowded with two of everything—two anesthesia machines, two cautery machines, two newborn warmers, and two heart-lung bypass machines.

"We went over all the possibilities, from prepping the patients to where we would move them after they were separated," says Strauch.

Nearly overwhelmed by the rapid development of events that took them so far from home, the Onzigas reassured each other that they had chosen the right course. "It was a very difficult decision," Margret says. Adds Gordon, "We told them to do the surgery because otherwise Loice and Christine couldn't live."

The 12-hour operation took place on April 19, 2002. The first part of the surgery was led by Strauch, who began the abdominal portion, separating the liver. Anesthesia was performed by Anne Savarese, MD, assistant professor of



Cardiac surgeon Marcelo Cardarelli (left) and pediatric surgeon Eric Strauch led the 35-member team that separated and cared for the girls.

anesthesiology and pediatrics and director of pediatric anesthesiology, and Monique Bellefleur, MD, assistant professor of anesthesiology. Once the heart and blood vessels were exposed, Cardarelli, along with Bartley Griffith, MD, chief of cardiac surgery, took the lead in untangling the girls' circulatory systems. In case the girls experienced heart rhythm problems and needed pacemakers, Jon Love, MD, an assistant professor and director of the pediatric cardiac catheterization lab, was also in the operating room.

Despite using diagnostic imaging to view internal structures before an operation, in the case of conjoined twins, surgeons don't know what they'll find until they're inside. As the surgeons delved deeper, they made an alarming discovery. Loice and Christine shared a vessel that connected the upper chambers of their hearts, a finding with unknown consequences.

"We knew there was blood going through the vessel from the smaller baby, Loice, to the larger baby, Christine," says Cardarelli. "But we didn't know what hemodynamic changes would happen if we cut the vessel. The girls also had exactly the same heart rate. We thought there may be some of the conduction system of the heart going

through the connection."

With pacemakers standing by in case either girl developed an irregular cardiac rhythm, the surgeons clamped and then carefully severed the vessel connecting Loice and Christine's hearts. Fortunately, their heart rate and blood pressure remained unchanged. By the fourth hour of surgery, the babies were completely separated. The remainder of the surgery consisted of closing the chest and abdominal wall with synthetic grafting material placed by plastic surgeon Bradley Robertson, MD, associate professor of surgery at the School of Medicine.

Margret recalls the first time she saw her babies after the surgery, swaddled in bandages and deeply slumbering. Two beautiful—and separate—baby girls. "I wanted to cry," she says.

After a brief stay in the medical center's six-bed transitional unit, Loice and Christine were cared for in the Pediatric Intensive Care Unit at the University of Maryland Hospital for Children. Once the twins were discharged from the hospital, Margret and Gordon stayed at the nearby Ronald McDonald House while the babies received physical therapy and additional medical treatment.

The girls underwent physical therapy to strengthen muscles



A final farewell for the Onziga family (front right) with some members of the surgical, pediatric, nursing, therapeutic, and nutritionist teams.

they were unable to use while conjoined, and have since reached normal developmental milestones. Loice had a small hole in her heart that was repaired by Love non-surgically last fall. According to their doctors, they have recovered well since the surgery and have an excellent prognosis.

Howard attributes that positive prognosis to "the huge role played by a medical team that included medical students, residents, pediatricians, nurses, therapists, and nutritionists who have cared for the girls from start to finish."

Loice and Christine returned home to Uganda in November, shortly after their first birthday, and are expected to live normal lives. No small miracle. ☞