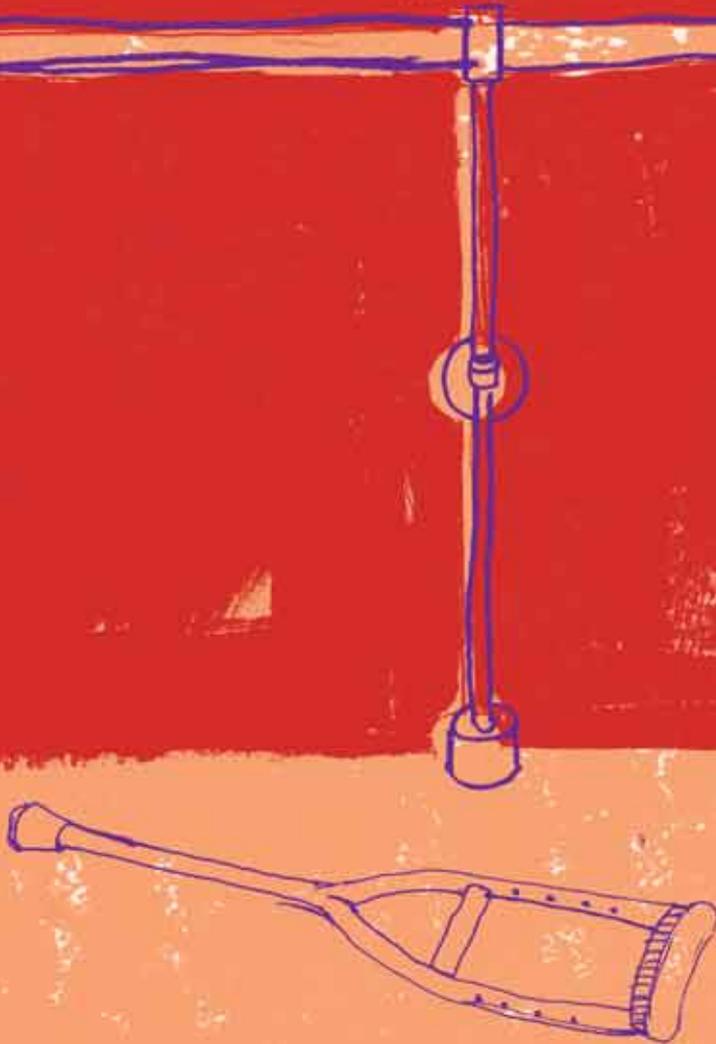
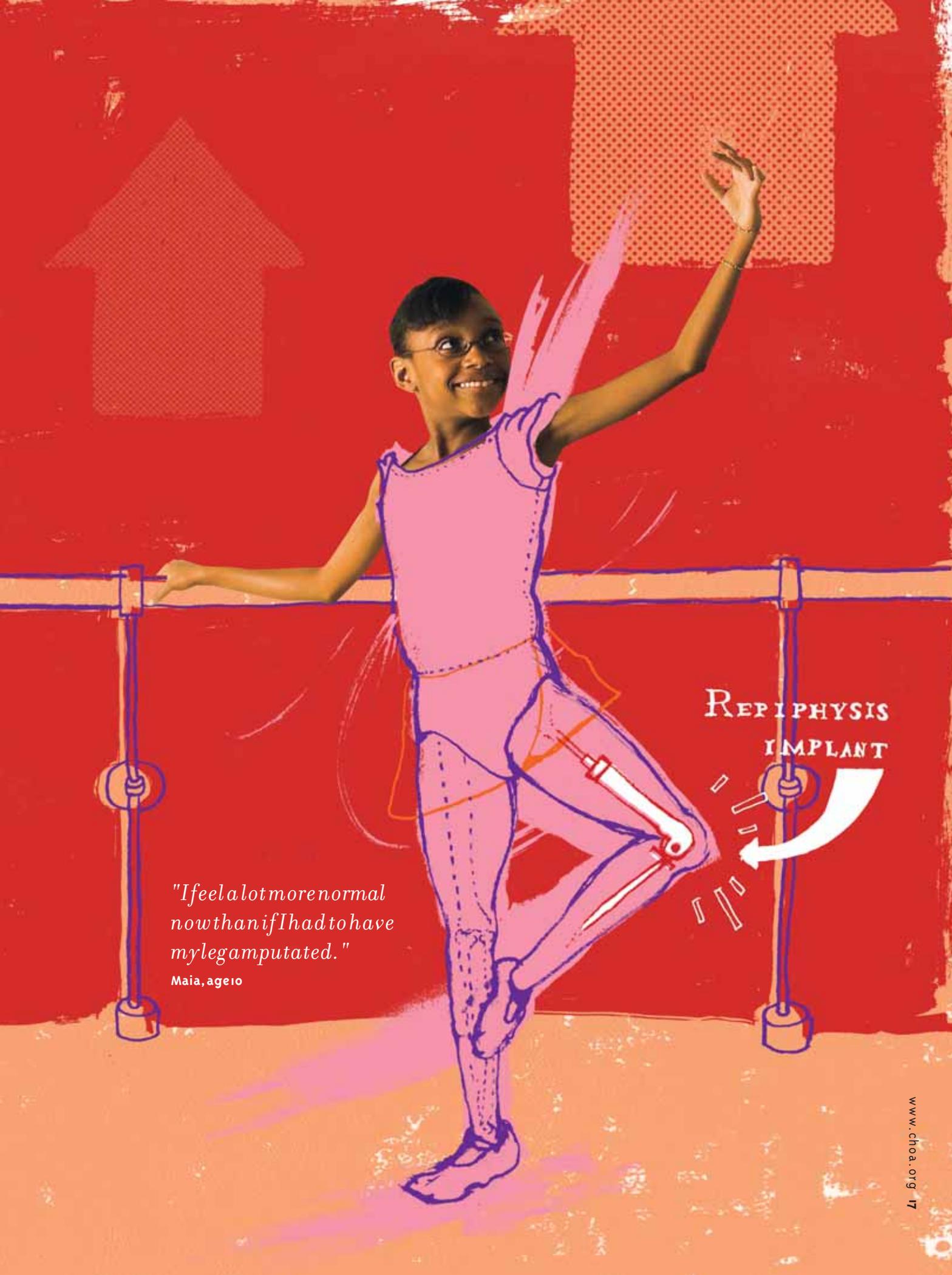


A Leg up

A new surgical technique lets children with cancer keep limbs for life





*"I feel a lot more normal
now than if I had to have
my leg amputated."*

Maia, age 10

REPHYSIS
IMPLANT



Maia Brookes, a sprightly pixie of a girl with a disarming smile, loves to dance. So she was upset when she started feeling sharp pains during her practices. One Tuesday in late 2004, her mother took her to a satellite campus of Children's Healthcare of Atlanta in Duluth, Ga., for an examination.

» After examining Maia, the pediatrician ordered X-rays to be taken that evening. That night, the same pediatrician called Maia's mother, Jan, and asked to see her the next day. The films showed a mass that could be cancerous. Suddenly, time was of the essence. Within two days, Maia had received several more tests. A few days later, a biopsy confirmed what doctors had

suspected; Maia had bone cancer.

» As recently as two decades ago, Maia's leg would likely have been amputated and she would not have survived. Today, children whose bone cancer has not metastasized have a 70 percent to 90 percent chance of survival. And thanks to Children's, patients with bone cancer have a new treatment option. Maia was the first patient in Georgia to receive a magnetic expandable implant which prevented amputation. The implant, known as the REPIPHYSIS™, contains a mechanism that allows it to grow without requiring Maia to submit to multiple surgeries. "I feel a lot more normal now than if I had to have my leg amputated," Maia said

A Big Decision

Before they dealt with Maia's leg, the first concern of Children's doctors was to save her life. She underwent further tests to ascertain the size of the tumor and to examine whether it had metastasized elsewhere in her body. (Fortunately, it had not.) Then oncologists designed an aggressive chemotherapy plan: three months of chemotherapy, removal of the tumor and then three more months of chemo.

Maia and her mother, Jan, spent much of the next six months at the Aflac Cancer Center and Blood Disorders Service of Children's Healthcare of Atlanta. Jan said, she was impressed at the center's ability to keep Maia distracted from the hair loss and nausea that accompanied chemotherapy. Maia speaks affectionately of her time spent listening to music, playing board games with her mother and the other children, and attending parties for holidays and birthdays. "It was really fun," Maia said—a somewhat shocking statement

about a chemotherapy experience. "I liked it, but I didn't like the chemostuff."

After a few weeks of chemotherapy, the Brookes family met with orthopaedic surgeon Shervin Oskouei, M.D. and physical therapist Colleen Coulter-O'Berry. Dr. Oskouei and Coulter-O'Berry used dolls, skeletons, diagrams and photographs to describe the three options that the Children's team had decided were most viable for Maia's leg. (The Children's Limb Deficiency Team includes surgeons, nurses, prosthetists, physical therapists and social workers. It's distinguished by close collaboration between surgeons and the rest of the team to maximize options and optimize outcomes for patients, Coulter-O'Berry said.)

The first option was to amputate Maia's leg above the knee and replace it with a prosthesis. The procedure requires only one surgery, and recovery is often quick. Prosthetic technology

appearance outside of the prosthesis. But a child with a below-knee prosthesis can engage in almost any sport.

The third option was to surgically implant an expandable endoprosthesis. By doing so, the cancerous bone and surrounding tissue are removed and a custom made expandable endoprosthesis is inserted. This has the obvious benefit of salvaging the limb. But, children with implanted prostheses have suffered from problems in the past.

The Growth Dilemma

Bone cancer is a rare phenomenon, affecting about four in every million children each year, Dr. Oskouei notes. Doctors do not know the cause of bone cancer, nor are they sure what are the cells of origin. But, they do know that it often strikes during a child's growth spurts. The tumors are typically located near the growing ends of the bone, most often near

From the outside, the REPIPHYSIS™ looks much like any other internal implant that might be used in a total knee replacement, with two pieces of strong metal connected by an artificial joint.



has advanced to the point where children with amputations should lead near normal lives, participating in age appropriate sports.

The second option was a tried-and-true procedure that the Brookes family nonetheless found to be more than a little upsetting. The first step of the Van Ness Rotation plasty, as it is known, is a resection of the diseased portion of the leg. The lower part of the leg is rotated so that the foot faces back, and the calf is reattached to the thigh so that the ankle joint serves as the knee joint. The child is then fitted with a below-the-knee prosthesis offering a below the knee amputation functional level. The positive part is that you are redesigning a knee joint from the foot which would not have been an option in a traditional above the knee amputation. The potential downside of a rotation plasty is the cosmetic

of the knee. So, removing the tumor stops the bone's growth. The resulting limb length discrepancy can lead to nerve damage and other problems.

The prosthesis, therefore, must grow along with the child's leg. But, how can this be done without having to surgically enter the leg and expand the prosthesis every time the child had a growth spurt, with all the risk and recovery associated with surgery?

Dr. Oskouei told the Brookes family that he had a new kind of prosthesis in mind. He acknowledged, however, that it was the first time the procedure had been done in Georgia. It was also the first time he had performed the procedure. Would the Brookes be interested? The custom-designed endoprosthesis would have to be ordered soon—in time for the tumor removal surgery. Dr. Oskouei needed an answer right away.

The French Invention

The story of the REPIPHYSIS™ implant harks back to one day in 1986, when a pretty 13-year-old girl named Violaine Bertrand died of bone cancer in a suburb of Paris. "I was really shocked," said her cousin, Arnaud Soubeiran. "Not by the fact that she had died—because it seemed relatively normal to die of the disease at this time—but because her leg had been amputated three months before dying."

Soubeiran, then 21, was a student in aeronautical engineering and had no medical training. Yet, the subject of prosthetics and cancer stayed on the forefront of his mind. That same year, he read of a French surgeon who had treated bone cancer without amputation. The surgeon said he was in search of a prosthetic that could be expanded without surgery. Soubeiran wrote the physician that same day. Three months later, the surgeon implanted Soubeiran's first attempt at an expandable prosthetic.

At this time, Dr. Oskoue in notes, surgeons were experimenting with prosthetics that were expandable with only minimally invasive surgery. They included one that could be expanded through inserting and twisting a sort of screwdriver. But, this prosthesis tended to break as the child's weight wore down the threads of the device. Most surgeons had settled on a modular device that needed to be pulled out, taken apart and reassembled for each expansion, notes Ross Wilkins, M.D., a Denver orthopedist who helped bring the REPIPHYSIS™ to the United States.

In 1991, a French cardiac surgeon asked Soubeiran if he could design an expandable ring that would reshape the mitral valve in a child's heart as it grew. The prosthetic valve Soubeiran designed never reached the market. But, it was while pondering on this idea that he hit upon how electromagnetism could be used to expand a prosthesis—the fundamental principle behind what would become the REPIPHYSIS™.

From the outside, the REPIPHYSIS™ looks much like any other internal implant that might be used in a total knee replacement, with two pieces of strong metal connected by an artificial joint. The magic is inside, where a tightly compressed spring is locked inside a polymer tube. (Dr. Wilkins, amazed at the device's simplicity, compares the spring to the kind you find inside a toilet paper holder.)

During an expansion procedure, the child places his leg inside a metal collar. Controlled pulses of energy sent through the collar generate an electromagnetic field around the implant. The heat created by this field softens the polymer tube. This allows the spring to expand, expanding the limb in turn. Each 20-second activation is followed by a fluoroscopy (or X-ray) to monitor progress of the expansion. The procedure takes only a few activations, and while the

child may experience some discomfort, an anesthetic generally is not required.

After years of use in Europe, the U.S. Food and Drug Administration approved the REPIPHYSIS™ in 2002. Sue Henderson, of REPIPHYSIS™ manufacturer Wright Medical Technology, said about 250 of the devices have been implanted in the United States.

Soubeiran believes the REPIPHYSIS™ is just the beginning of uses for his electromagnetic technique. In Europe, a smaller version of the implant is being installed on an experimental basis in children with spinal curvature. The goal is to allow spine growth without multiple surgeries. He is also working on a new generation of the implant that could be used for limb lengthening and bone transportation for adults or children.

It might seem strange that an aeronautical engineer is behind such medical innovations. But, Soubeiran said many of the materials and manufacturing approaches used in prosthetics—such as the use of titanium alloys and certain welding methods—stem from the world of aeronautics. "In both cases," he said, "you need something that will never fail. It leads to the same materials."

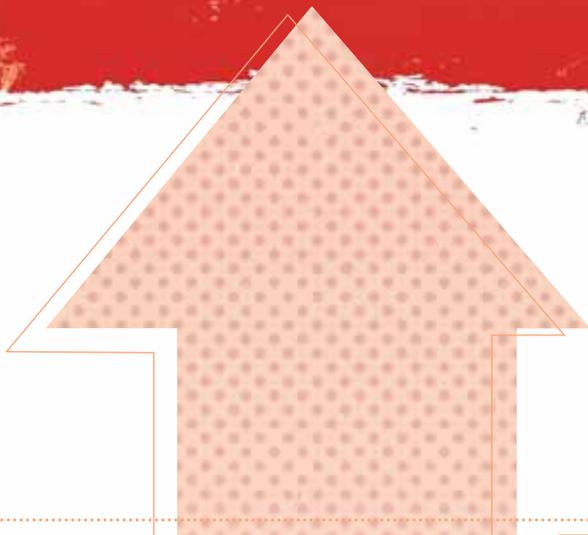
A New Future

After speaking with the Children's limb deficiency team, the Brookes family decided on the REPIPHYSIS™. "We wanted to look at the option that would save her leg and allow her to have the most normal life possible," Jan Brookessaid.

Maia got to go home for Christmas with her family while her endoprosthesis was custom-designed at Wright Medical's factory in Arlington, Tennessee. In a three-hour operation in January, Dr. Oskoue removed the cancerous bone and replaced it with the REPIPHYSIS™ implant. He also had good news for the family: the chemotherapy had almost totally destroyed the tumor.

Almost by accident, Dr. Oskoue said, medical and technological developments have led to increased survival rates for children with bone cancer. Previously, physicians would amputate the limb and then provide chemotherapy, but the patient would still die. As custom prostheses came into vogue, patients would receive chemotherapy immediately while the prosthesis was being designed. Suddenly, survival rates improved dramatically. It turned out that the delay in chemotherapy caused by the amputation surgery had been giving microscopic tumor time to metastasize throughout the body, Dr. Oskoue said.

After Maia's surgery, she had to learn to walk again. But by a June vacation, she had shed her crutches. By August she was dancing again, though her teachers have modified her routines to minimize leaping and leg bending.



*Almost by accident
developments in prosthetics
have led to increased
survival rates for bone
cancer patients.*

— SHERVIN OSKOEI, M.D.

In April 2006, 15 months after her first surgery, it was time for Maia's first leg lengthening procedure. The outpatient procedure took just a few minutes, compared to the invasive surgery required to lengthen a typical prosthetic implant. Still, Maia had to return to physical therapy afterwards, as the muscles and soft tissue in her leg had to adjust to the limb's new length.

Maia, now 10, will undergo this lengthening and rehabilitation routine every time she has a growth spurt—perhaps once or twice a year. When she reaches skeletal maturity, surgeons will insert a more durable adult prosthesis, which in turn must be replaced about once a decade.

"The good Lord is better at making [limbs] than we are," Dr. Wilkins said. "When these kids are fully grown, my vision is they'll be able to take advantage of some sort of bioimplant." Dr. Wilkins is researching ways for living tissue to take the

place of cancerous bone.

Meanwhile, Maia has returned to the busy job of being a little girl. Her hair has regrown nicely into a fetching pageboy style. She's taking jazz and modern dance classes and even participates in recitals. When her leg feels sore, she is allowed to ride in her school's elevator. The boys like to carry her books for her, she reports. That way they can ride the elevator, too. 📧

Shervin Oskoei, M.D. is an Orthopaedic Surgeon at Children's Healthcare of Atlanta and Assistant Professor of Orthopaedic Surgery and Director of the Orthopaedic Residency Program at Emory University School of Medicine.

Colleen Coulter-O'Berry, P.T., M.S., P.C.S. is Team Leader of the Limb Deficiency Center at Children's Healthcare of Atlanta.

To obtain CME credit for the articles you have read in this publication, go to www.choa.org/cme and click on the *peds* magazine module.