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skulpture

DESPITE RESISTANCE FROM PEERS, A HUSBAND-WIFE SURGERY TEAM TRANSFORMS THE TREATMENT FOR AN INFANT SKULL DEFORMITY. PARENTS FROM ALL OVER THE WORLD BRING THEIR BABIES TO MU FOR A NEW PROCEDURE THAT ALLOWS A CHILD'S BRAIN TO GROW NORMALLY.

THE 19-INCH VIDEO MONITOR reveals the landscape inside Katherine Knapp's head. A nurse dims the operating room lights, and the neurosurgeon's tiny camera glides across the infant's slimy dura, the rubbery shell that houses her brain. A small blood vessel bursts, and blood oozes up as from a bubbling swamp at night. The surgeon zaps the spot with an electrified silver wand and sings the rupture closed. He moves on, freeing the dura from the thin skull above. He flirts with disaster as he nears Katherine's sagittal sinus, the giant vein that channels blood from her brain back to her heart. Now the plastic surgeon takes over. Her scissors relieve Katherine of the section of her skull that has formed prematurely and hindered her brain's natural growth.

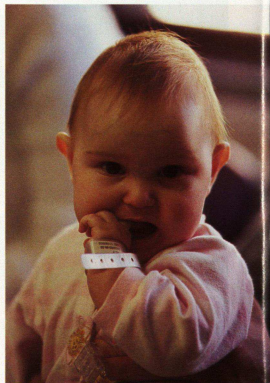
Katherine Knapp is one in a thousand. The 7-month-old girl suffers from a rare infant skull deformity called craniosynostosis (KRA ne o SIN os TOE sis). Today her parents, Heather Sollars and Dustin Knapp, have brought her from St. Joseph, Mo., to University Hospitals and Clinics to receive a treatment more rare than her condition.

Using tiny cameras called endoscopes

and custom-made molding helmets, MU surgeons David Jimenez and Constance Barone have revolutionized the procedure for correcting the skull deformity. Most doctors who perform craniectomies still cling to some variety of the "old way." A traditional craniectomy can involve cutting across the baby's scalp from ear to ear, folding down the skin to expose the entire skull, removing part of the skull for re-sculpting and then putting it all back together. At its worst, the process takes eight hours and requires several blood transfusions. A baby sometimes will lose its body's entire volume of blood — about 16 ounces, like a bottle of Coke — up to five times.

"It's putting them through hell," Jimenez says of the traditional procedure. Katherine's mom, Heather, got the same sense when she learned about the grueling surgery: "It sounded extremely scary. And we saw some pictures of what the babies looked like afterward, with their swollen eyes. It's a scary procedure."

No one knows what causes craniosynostosis, although some suspect a genetic malfunction. The problem occurs when two or more of the individual bone plates that make up a baby's skull fuse before



Above, Katherine Knapp has a mild case of craniosynostosis, a deformity in which two or more of an infant's skull bones fuse prematurely. The condition prevents Katherine's brain from growing normally and makes her skull long and narrow instead of round. About a quarter of such patients have pressure inside their heads that sometimes leads to serious brain damage.

Right, surgeons and spouses Constance Barone, left, and David Jimenez operate on Katherine using their new technique for correcting craniosynostosis. Jimenez views the inside of Katherine's head using tiny cameras called endoscopes, which allow for a minimally invasive procedure. Most doctors still perform open craniectomies, which use large incisions and can cause significant blood loss.





In the hands of her father, Dustin Knapp, Katherine gets checked out by a nurse on the morning of the operation. Some doctors disagree about the effectiveness of the new procedure, but of Jimenez and Barone's 230-plus patients, only one family has sought follow-up surgery.

they're supposed to. Like pieces of a jigsaw puzzle, six major plates cover and protect almost all of the brain. The small uncovered regions are fontanels, or soft spots. The skull bones form before the sixth month of pregnancy but remain separate so the skull can fit through the birth canal and the brain can grow freely.

When the plates fuse prematurely, the brain grows away from the closed spot and pushes out on the skull plates that haven't fused yet. The effects sometimes include a dramatic physical deformity, such as a football-shaped head, and pressure on the infant's developing brain. Pressure causes mild fussiness at best or serious brain damage at worst. Studies suggest that about 25 percent of babies with craniosynostosis have pressure inside their heads.

A BETTER WAY

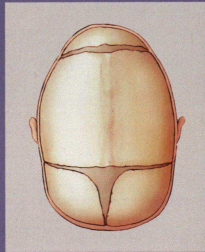
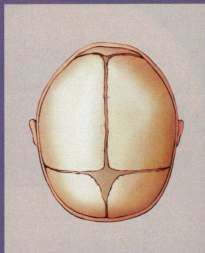
JIMENEZ AND BARONE HAVE PERFORMED hundreds of traditional craniectomies. But when they came to MU in 1992, they started their own craniofacial program and began rethinking the conventional

procedure, which was first performed in the 1800s. "We got into it because we don't think the traditional surgery is the answer," Barone says. "We don't like the way the kids look. We wanted them to have a more natural appearance, a more normal forehead contour, a less destructive operation, less blood loss and less recovery time. We were displeased with how many people across the country were showing their great results, and I thought they weren't great. I thought they looked operated on."

Another factor left Jimenez and Barone determined to find a better way: The husband-and-wife surgery team had a child of their own. "I had just had our son," Barone says, "and we really got into it at that point." They asked themselves: If we wouldn't put our own child through such a dangerous, invasive surgery, why should anyone else?

"David, there's got to be a better way," Barone said to her husband.

"I think we could do it endoscopically," Jimenez replied. But he wondered how they would keep the brain from pushing out in the wrong direction.

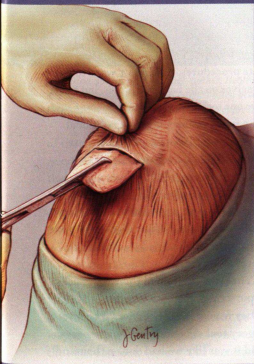
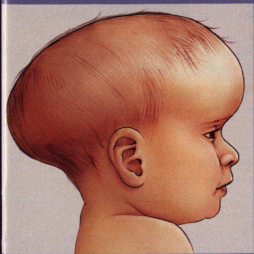
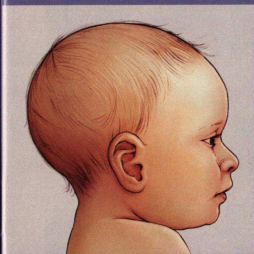


Top left, six major plates make up the skull of a normal baby. Above, a fused sagittal suture. Genetic malfunctions may cause skull plates to fuse prematurely.

Top right, a normal baby's head. Middle right, the head of a baby with sagittal suture craniosynostosis. The fused bones on top of the head force the skull to take a long, narrow shape as the brain grows.

Right, the surgeon removes the strip of bone where skull plates have fused. Rather than simply cutting the plates apart, Barone takes out a wide strip (4 centimeters in Katherine's case) to prevent the plates from immediately re-fusing.





If doctors perform the surgery before 6 months of age, the rapidly growing brain can play a helpful role in reshaping the skull. After the premature fusion has been removed, the growing brain pushes out on the skull and forces it to take a new shape. "The brain wants to be round," Jimenez says. But how would the surgeons make sure the skull would take a natural, round shape rather than a different but equally deformed one?

That's when Barone came up with the idea for the molding helmet. For about a year after the procedure, patients must wear a soft plastic helmet to force the skull into a natural shape. After 6 months of age, there usually isn't enough brain growth to reshape the skull. So there's the catch: If craniosynostosis is detected after 6 months, the patient may have to undergo the traditional procedure.

"We thought we were going to be stuck with the traditional," Heather says. Katherine was already 5 months old when her pediatrician said she couldn't feel a soft spot anymore. She sent the family to Children's Mercy Hospital in Kansas City, Mo., where Katherine was diagnosed with craniosynostosis. "They just said she was going to have to have surgery," Heather says.

Heather and Dustin went back to their pediatrician, who referred them to Jimenez and Barone. But because Katherine was 7 months old by then, her parents weren't sure whether the new technique would be an option. "Here could be a chance to not have to put her through such a big, invasive surgery," Heather thought. "It was kind of like, 'Please let her be able to do that.'"

A CHALLENGING CHANGE

KATHERINE AND HER PARENTS MADE A TRIP to Columbia to see the doctors in their surgery clinic. "Dr. Jimenez spent a lot of time with us," Heather says. After the

Back in her mother's arms after the surgery, Katherine will spend just one night in the hospital and go home the next day.

consultation, he gave his recommendation: Because Katherine's case was mild, the procedure, called an endoscopic strip craniectomy, would likely be successful despite her age. "We felt extremely elated after he told us she was going to be able to have this surgery," Heather says.

Katherine was lucky, not just because she was going to be able to have the endoscopic surgery, but also because her pediatrician, Michelle Cebulko, MD '97, happened to be an MU graduate who knew of Jimenez and Barone's work. Other craniosynostosis patients are not so lucky. Many pediatricians and family practice doctors simply don't have the experience to diagnose craniosynostosis early enough for infants to benefit from the new technique, Jimenez says: "A lot of it has to do with medical education. Say you want to be a pediatrician. How much time do you think the medical students at MU spend checking out neurosurgical patients? If there are 96 students in a year, I get up to 12 in my clinic. For a half-day. The rest never come by. Now, how much are they going to know about craniosynostosis?"

Knowing the symptoms and getting an



early diagnosis are crucial to making an infant eligible for the new procedure. That's the first problem craniostylosis patients face, but it's not the biggest one. The endoscopic strip craniectomy may be a blessing for the patients who receive it, but because fewer than a dozen surgeons perform it, most patients are still stuck with the traditional procedure. And as Jimenez and Barone spread the word about their success, they are met with both enthusiasm and skepticism.

"Medicine is very slow to change," Jimenez says. "If something works for a physician, it's going to take hell and high water to get him to change." At a 2001 conference attended by all of the 120 pediatric neurosurgeons in the United States, Jimenez used eight Carousels of slides to show every one of his patients to date. "At the end of the meeting, I thought they were going to start throwing tomatoes at me," he says. "As far as they're concerned, this is sacrilegious."

Much of the resistance, Jimenez says, comes from surgeons who don't use endoscopes. "Maybe 10 percent of neurosurgeons have experience with endoscopes," he says. "So we're asking people not only to change their operation but also to deal with a technique they're not familiar with." And it's not easy to learn to use endoscopes, says John Oro, chief of the Division of Neurological Surgery at MU: "Yes, you can go out and train, but it's more of a commitment than you would think."

That's why hope lies largely with the surgeons of tomorrow, the ones being trained in endoscopy and other cutting-edge techniques. But even many of today's surgeons see the promise of the endoscopic procedure. As pediatricians learn about the new procedure, demand for it will increase. "The surgery is a lot easier on the babies, and their recovery is a lot better," says Cebulko, Katherine's pediatrician. "Jimenez and Barone are pioneers in their field, and I'm sure this is the way that the surgery is going to be going."



Katherine gets fitted for her molding helmet by Phillip Bates, orthotist with Hanger Prosthetics and Orthotics. Bates has fitted more than 700 helmets for endoscopic strip craniectomy patients. Children typically require three helmets as their heads grow over the course of their treatment.

SPREADING THE WORD

WHILE SOME DOCTORS REMAIN SKEPTICAL or unaware of the new technique, parents love it. More than 230 babies have received the endoscopic strip craniectomy at MU since 1996. They've come to Jimenez and Barone from Norway, El Salvador, Canada, Spain, Ecuador and 39 of the United States, including Alaska. The doctors say they run the second-busiest cranial surgery center in the world after the Dallas Craniofacial Center. The eight surgeons in Dallas worked on 104 cases in 2002, while Jimenez and Barone alone did 90.

But the doctors in Dallas are skeptical of the new procedure, says David Genecov, a surgeon at the Dallas Craniofacial Center: "As I tell my patients today, 10 years from now that may be the way to do it, but at this point I'm not willing to allow my patients to undergo an operation that I don't think is necessarily proven."

In Dallas, the surgeons feel that the traditional open surgery, called cranial vault remodeling (CVR), provides better physical results.

Genecov adds that some complex cases with multiple sutures can't be done with endoscopes. The surgeons also opt for a more conservative approach because of

the number of secondary surgeries they perform. "We see a lot of the kids who've had operations on the upswing of new procedures and new technologies. What happens is the downside isn't seen until two or three years later, then they come to us to fix it. We see a skewed patient population, so that makes us a little more wary."

So far, Barone and Jimenez have experienced success. Only one of their endoscopic strip craniectomy patients has sought follow-up surgery. In contrast, some patients who receive the traditional surgery have to undergo two or three separate procedures. "We don't want to put ourselves on the back too much, but we have our techniques down," Jimenez says. Even when performing the traditional CVR surgery, Jimenez and Barone are standouts. Their average time for that procedure is around three hours, less than half of what it takes some surgeons. And a third of their patients don't need a single blood transfusion. "Even that surgery we had already streamlined tremendously," Jimenez says.

The team performed their first endoscopic strip craniectomy in 1996 on a little boy from Brookfield, Mo. "The first surgery took 55 minutes," Jimenez says, "and it was like we had done a thousand



Katherine, shown here with her mother, Heather Sollars, will wear her new helmets about 23 hours a day for six months to a year. Parents often accessorize the helmets with stickers and ribbons. After her treatment is complete, she'll be able to enjoy her normal, round head for the rest of her life.

of them." Not long after that first surgery they began receiving national attention. Soon, they would appear on *Good Morning, America*, the Discovery Channel and in *Parenting* magazine. More important, parents spread word-of-mouth satisfaction through testimonials on countless Web sites.

BEFORE AND AFTER

KATHERINE'S PARTICULAR BRAND OF CRANIOSYNOSTOSIS is called a sagittal synostosis, the most common variety. The name tells

which suture in her skull has fused early. The sagittal is on top of the head, running from the top of the forehead straight back to the crown. It connects the two parietal skull plates, found on either side above the ears. The fused suture prevents Katherine's brain from growing up or out to the sides, so it takes the path of least resistance and pushes out in the front and back. A more dramatic case would cause a giant bulge in the forehead and a point in the back of the head, like a football.

Even though Katherine's case is mild,

Heather can tell her daughter has a problem: "You can feel it. Her head's more narrow and, around where her temples are, kind of pinched in." In the weeks before the surgery, there have been signs that Katherine can feel her brain pushing out. "She's gotten a lot more fussy in the last couple of weeks," Heather says, "and she wakes up every hour at night."

At 12:06 on a Tuesday afternoon, Jimenez makes the first of two small incisions in Katherine's head. Fifty-two minutes later, the procedure is complete. Katherine has lost only 20 cc of blood, slightly more than one tablespoon. "Isn't that better than cutting from ear to ear?" Jimenez asks proudly. He visits the parents in the waiting room. "Everything went perfect," he says. "Now she's screaming and crying for Mom. Everything went perfect. Textbook."

Katherine spends just one night in the hospital, instead of the usual four or five after a traditional craniectomy. On Wednesday, the family is back home. "It was kind of like coming home from the hospital for the first time," says Katherine's father, Dustin. "Everyone came to see her." The benefits of the surgery are immediate: "When we got home and she only got up once in the night, it was like, 'Thank you!'" Heather says. "She's a lot better." The following week, Katherine returns to Columbia to be fitted for her molding helmet, which will help her head take a round shape over the next year.

Katherine is already an example of the safety and efficacy of the endoscopic procedure. Some skeptics won't be fully convinced until Katherine is older. But as the number of successful surgeries mounts, it gets harder to ignore that the future may have already arrived. "People like to see long-term results," Jimenez says. "Now the long-term results are starting to come in. And they look great." ❁

For more information, visit <http://www.muhealth.org/~neuromedicine/craniosynostosis.shtml>.