

B

ETH'S TRAUMA
& HER WISH

*"Through the
looking glass"*



THE BALTIMORE SUN

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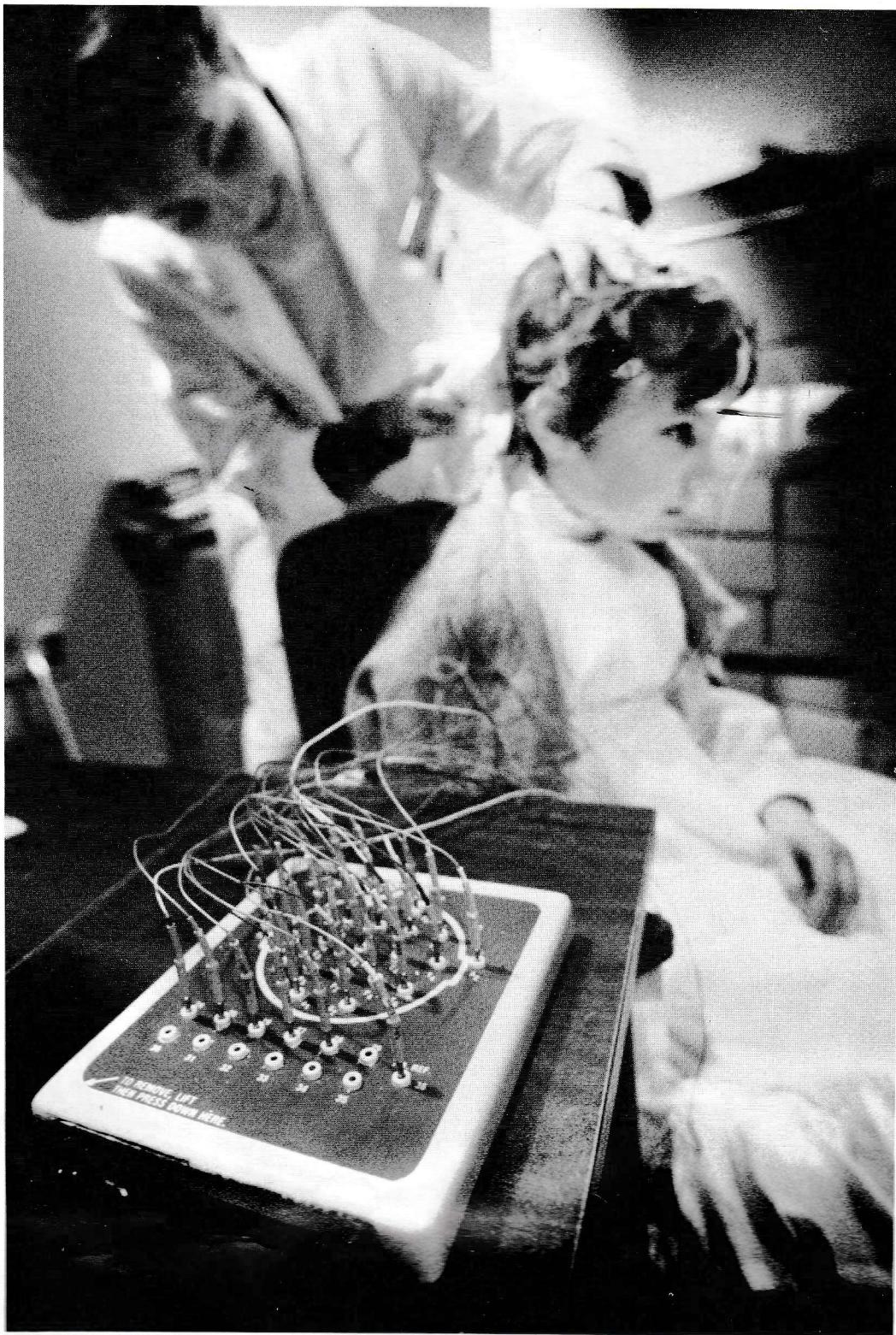
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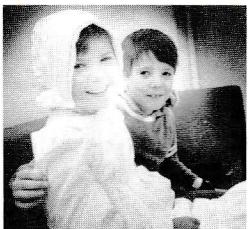
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PART I
CAUSE,
CURE
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REPRINTED FROM A SERIES WHICH APPEARED ON MARCH 29 - 31, 1987 IN THE BALTIMORE SUN



Disease had first cast its shadow across Beth Usher's life one September afternoon in 1984, when Kathy and Brian Usher were summoned by the school principal and found their 5-year-old daughter mute, terrified and trembling, the right side of her body in the grip of a seizure.

Two and a half years later, Elizabeth Catherine Usher, 7, of Storrs, Conn., was being rolled into a Johns Hopkins Hospital operating room in Baltimore for a grueling, high-stakes operation in which neurosurgeons would remove the entire left half of her brain.

"Hemispherectomy," the doctors called it. They would cut away the diseased left cerebral hemisphere, hoping on the basis of earlier cases that the right hemisphere would take over its functions.

The risks of the operation included paralysis, coma and death. But the doctors said it was Beth's only chance of a future.

Months of testing showed that she had an extremely rare brain disease called Rasmussen's encephalitis that causes progressively more frequent and more severe seizures.

Without surgery, the doctors said, she would slide irreversibly downhill as the disease gradually tore its way through her brain. Within a few years, they predicted, she would become a candidate for a nursing home.

Beth's parents had to take that grim prognosis on faith. Their child's seizures unmistakably were growing worse, but she still was loving, active, articulate.

She limped slightly — but she still could run and play. Her memory was growing weaker — but she still could learn in school. Deciding that radical surgery was the best course for their daughter was the most difficult thing Brian and Kathy Usher had ever done.

A year earlier, Beth had been reading Lewis Carroll, and had said wistfully to her mother that she'd like to go through the looking glass and come out "a different kind of person."

"What kind of person?" Kathy Usher asked.

"The kind of person," Beth replied, "who doesn't have seizures."

For 12 months, neither Beth nor her mother had mentioned the girl's fantasy again. Then, the morning of Feb. 4, 1987, when the long trip under the fluorescent lights of the hospital corridors ended at the double doors of the operating room, Beth turned to her tense parents.

"Mom. Remember?" she said, peering up from the antiseptic sheets of the gurney. "Through the looking glass."

One lucky family

A few years earlier, when Kathy Usher volunteered to drive cancer patients to the hospital

for tests and chemotherapy, she made sure her children went along for the ride. She felt her family was so free of misfortune, that Brian Jr., born in 1977, and Beth, born in 1979, needed to know just how lucky they were.

Kathy, one of seven sisters, had experienced catastrophic illness in her family. Her mother had died of cancer; a year later, her father had been seriously injured in a car accident and spent three months in the hospital recovering from brain surgery.

But years later, after her marriage to Brian and the birth of their two children, the young family's life seemed a cliche of small-town American happiness. Both parents worked at the University of Connecticut, a quick bike ride from home. Brian was an assistant football coach; Kathy helped run the athletic department's fund-raising club.

Their children were healthy, popular, successful in school. Brian Jr. was entranced by dolphins and wanted to be a marine biologist. Beth was a standout in dance classes. The nursery school at the university singled out her memory and physical coordination as unusual for her age.

"She had all these boy cousins and she'd run after 'em and tackle 'em," her mother said. "She was the one who wanted to be the football player."

Beth and Brian were unusually close. When one got in trouble with their parents, the other would leap to the defense. When they ate pizza, Beth, who was allergic to wheat, would eat the top; Brian, who didn't like cheese, would eat the crust.

When Beth started at Northwest School in Storrs, she quickly became known as an affectionate kid. "She'd walk down the hall, and everyone would get kisses and hugs," said Sharon Bartlett, one of her favorite teachers.

But she also showed a mischievous streak. She liked to make up stories with shocking endings, like the one she told her mother that began with a handsome prince marrying a beautiful princess. "And then," Beth said at the end, "they got divorced." Beth burst into laughter, watching her mother's face for a reaction.

Into the Ushers' life Beth's first seizure came as a frightening and mysterious intruder. Kathy Usher got a call at work and rushed to the school. Brian was out jogging; a fellow coach drove after him and alerted him, and he followed his wife.

They found Beth looking frightened, her mother recalled. "She gasped. She looked like she was in pain." When her father arrived, she recognized him and reached out to him with her left arm, which appeared unaffected.

At Hartford Hospital, the seizure subsided. A brain scan showed some unusual shrinkage of parts of the left hemisphere, and one of the doctors suggested cerebral palsy. Kathy called

her family doctor, who said cerebral palsy was out of the question; there would have been symptoms before age 5.

Then perhaps, they thought, the seizure had been triggered by Beth's fall from the seesaw that morning. A bigger kid had jumped off and Beth's end had crashed to the ground, the metal handle giving her a nasty knock in the face.

Whatever it was, after a couple of hours the seizure subsided and Beth, on a small dose of phenobarbital, was back to normal.

"We were shaken up," Mrs. Usher recalled. "They couldn't say what caused it, but they told us it probably wouldn't happen again."

For two months, it did not.

Then, the day before Thanksgiving 1984, as Beth was getting ready for school, the specter returned. Again her right side went rigid, her right arm jerking in front of her, her eyes rolling out of control.

Whatever it was, it wasn't going away. This time, the doctors said it was probably garden-variety epilepsy, "which seemed like the worst thing in the world to us then," Kathy Usher remembered.

Gradually, over the ensuing months, the episodes grew more frequent, eventually occurring five to 10 times a day and lasting several minutes each time. Beth was given higher and higher doses of phenobarbital to control the seizures, requiring the little girl to swallow a dozen or more pills each morning. But not long after the doctor increased the dose, the seizures would "break through" and begin again.

Beth's family physician, Dr. Nelson Walker, was baffled by the failure of the drugs to control the seizures. He referred the family to a neurologist, who in turn referred them to a second specialist.

The second neurologist mentioned Rasmussen's encephalitis. He said it was difficult to control, and that he knew one patient in a nursing home as a result of it. But he was tentative about the diagnosis, and the Ushers left his office believing he was just laying out the worst possibility.

Not satisfied without a diagnosis, but not wanting to accept the dire prognosis of Rasmussen's, Kathy Usher began gathering books on neurology, on seizures, on epilepsy and the brain and anything else that seemed even vaguely relevant. Eventually, the books and the boxes of Beth's medical records would fill the Ushers' spare room and consume their every free moment.

The Ushers had their well water tested, remembering that the previous owner of their house had died of cancer and wondering whether some unsuspected toxin might somehow be responsible for Beth's condition. The test showed nothing abnormal. Then, despite doctors' assurances that it was not a possibility, they arranged a test for Lyme disease, which is

associated with tick bites and relatively common in New England. It was negative.

Meanwhile, the seizures were occurring several times a day, and the right side of Beth's body was beginning to grow weaker. Though always right-handed, she began to switch to her left hand for many tasks.

She could pick up her lunch box with her right hand, but unless she concentrated on keeping her hand tightly closed, her grip would loosen and the box would drop to the ground.

Her smile sometimes drooped on the right side. Her speech slurred when she was tired. She began to limp on the right and to let her right arm hang limp.

Beth's memory began to slip. Despite the best efforts of supportive teachers, her schoolwork began to suffer from the combined effects of the seizures and the massive doses of medicine she had to take to combat them.

The Ushers called the neurologist again, and he arranged for another brain scan. That night he called the family at home and told them coolly that the X-ray and the symptoms made the diagnosis of Rasmussen's all but certain.

"He said, 'She'll just get worse and worse, and there's nothing that can be done about it,'" Mrs. Usher said. He said Beth probably would decline over about four years, then have to be institutionalized, and they should get used to the idea.

The Ushers were shattered, but they found it impossible simply to do nothing and wait for their daughter's decline. They heard about a prominent pediatric neurologist at Boston Children's Hospital and contacted him. Beth was admitted for eight days and given test after test to identify blocked arteries or other possible causes for the seizures.

The family returned home from Boston with no new diagnosis, no miracle cure and, despite a comprehensive insurance policy, thousands of dollars of medical bills. It still looked like Rasmussen's.

Mrs. Usher called the Epilepsy Foundation of America to ask about treatments for Rasmussen's encephalitis. No one had heard of the condition, but a helpful librarian took her name and promised to let her know whether she could find any information about the disease.

Finally, last June, a note arrived from the librarian. She'd come across a newspaper article that said doctors at Johns Hopkins Hospital in Baltimore were trying radical surgery for Rasmussen's. It mentioned a neurologist named John Freeman.

Nervously, hopefully, Mrs. Usher dialed the number an information operator gave her for Johns Hopkins, reached Dr. Freeman's office and left a message. A few minutes later, the phone rang. John Freeman listened to Kathy Usher's story and invited her to bring Beth to Baltimore for evaluation.

"They lose all function"

Like many a medical term, Rasmussen's encephalitis is precise-sounding language for something doctors know little about.

Named for the Montreal neurosurgeon who discovered it three decades ago, the disease has proved extraordinarily resistant to study, partly because it is so rare, striking one in several million children. Most neurologists never see a case.

Rasmussen's can be definitively diagnosed only from examination of brain tissue, which shows severe inflammation and scarring around blood vessels. Surgeons say the inflamed tissue feels diseased. But scientists have never identified the virus, bacteria or other organism that does the damage.

"If it's a virus, it's a very strange virus," said Dr. Freeman, chief of pediatric neurology at Johns Hopkins. "It might be a part of a virus. The way I conceptualize it, it starts somewhere in the brain and eats its way out."

Without actual brain tissue, doctors diagnose Rasmussen's chiefly by eliminating all the other possibilities: no tumors, no major blood vessel problems. In addition, an X-ray of the brain using a computer-driven machine called a CT (computerized tomography) scanner will show a distinctive pattern of atrophy, or shrinkage, of parts of the affected hemisphere.

What doctors do know is that a child with Rasmussen's has a bleak future. The spreading inflammation triggers seizures — essentially electrical storms of the cortex, random discharges that overwhelm the normal electrical activity of the brain.

With the spreading inflammation comes not only debilitating seizures, but steady loss of muscle control, speech and memory.

"The encephalitic process, after taking one hemisphere, spreads to the other, and they lose all function," said Dr. Benjamin S. Carson, chief of pediatric neurosurgery at Hopkins. After an average of about four years on a downhill course, all the reported patients have ended up having seizures so constant they can no longer walk, speak, hear or see and often need to be on life-support machinery.

That bleak outlook provoked doctors to consider radical surgery for severe seizure disorders such as Rasmussen's as long ago as the 1930s. Hemispherectomy was first tried for brain tumors during the 1920s by the pioneering Hopkins neurosurgeon Walter Dandy, among others.

Since the first attempts, doctors' belief in hemispherectomy for seizures has waxed and waned in response to reported successes and failures. Long-term complications of the opera-

tion, including bleeding and buildup of fluid or broken-down red blood cells in the skull cavity, discouraged surgeons during the 1960s.

But Drs. Freeman and Carson decided about two years ago that such advanced diagnostic tools as CT scanning and magnetic resonance imaging, as well as improvements in surgical technique, made hemispherectomy a risk worth taking.

Their results have been striking. Before Beth Usher's case, they had performed hemispherectomy on eight patients, seven of them with Dr. Carson as lead surgeon. Six turned out to be Rasmussen's; one was a tumor and one was never clearly diagnosed.

In every case, the seizures had stopped, IQ had remained the same or even improved slightly, and the children had regained the ability to walk and talk, though sometimes only after extensive therapy. The flexibility of a young child's brain is such that the remaining nerve cells can learn to take over jobs previously performed by cells destroyed by disease or removed by the surgeon's knife.

If hemispherectomy and its results appear incredible to laymen, they are not much less surprising to physicians. Dr. Carson said many doctors attending a national neurosurgery conference in Pittsburgh last December were surprised to learn from him that the operation is being performed and that paralysis of one side of the body is not an inevitable complication.

"The Murphy's Law kid"

At 10 in the morning on Feb. 2, Beth Usher seated herself on the lap of Dr. John Freeman.

She was dressed in a frilly, colonial-style dress she had begged her parents to buy. The dress had a matching bonnet, preparation for coping with the shaved head that would precede the operation two days later.

"What's a seizure feel like?" Dr. Freeman asked.

"It feels funny in my head," said Beth. On her fourth visit to Hopkins, they were old friends, and she seemed perfectly at ease.

"It feels funny like a buzz, or what?" the doctor asked.

Beth sighed, like a teacher with a particularly dense student. "It feels funny like a SEIZURE," she told him, her eyebrows in a teasing frown beneath her brown bangs.

The difference between Beth and the other Rasmussen's patients who had undergone surgery at Hopkins, Dr. Freeman said later, was that she was still in good shape.

The limp, the drooping right arm, the slight slurring of speech were the only outward signs of the disease. She mastered each seizure, gripping the nearest person and waiting for it to pass; it was an old, familiar enemy, frequent but

not constant, annoying but no longer scary. People meeting her remarked on her wit, her animation, her energy, her generosity, not on her disabilities, if indeed they noticed any.

One of the earlier Rasmussen's patients had been scheduled to be institutionalized the day she had surgery. Another had come in on a respirator, unable to breathe unassisted. Others had near-constant seizures that kept them from talking, playing, learning, even eating.

Yet, because the operation generally could not restore function already lost on the "weak" side of the body, Drs. Freeman, Carson and Eileen P. G. Vining, another Hopkins seizure specialist, had decided that the earlier the surgery took place, the better.

"It's absolutely crazy to treat a viral infection by taking out half the brain," Dr. Freeman said, "but that's the best thing we have."

"It's very hard to do a big operation on a kid at any stage. If the kid's about to die, it's an easier decision. If the kid's in pretty good shape, it's much, much harder," he said. "This one really tugs your heartstrings."

In July, when Beth had first come to Hopkins, Kathy and Brian Usher believed there was no way they could send their daughter off for such radical surgery. So they went home and waited, and watched her slide slowly but unmistakably downhill.

In October, they drove to Baltimore again, still skeptical. In an attempt to determine whether Beth's speech control had transferred to her good hemisphere, doctors tried to put the diseased hemisphere to sleep with an injection. But the drug crossed over and put the entire brain to sleep, so they couldn't know for certain whether the operation would take away her ability to talk.

The doctors still believed hemispherectomy was the only option, the sooner the better. They suggested to the Ushears that they go home and enjoy Thanksgiving, Christmas, New Year's with Beth — but not to delay surgery past January.

"We have this child," Kathy Usher, 33, said at the end of January, "who has seizures but goes to school and has a wonderful life. We asked Dr. Carson about the risks of the surgery, and he said coma, paralysis, death. . . . We knew in our hearts that she had to have the surgery. But we wanted to squeeze out every precious minute with her that we could."

"You look at Bethie," said Brian Usher, 34, "and you see that she does so many things so well. But we've seen her deteriorate — dramatically. We trust Dr. Freeman and Dr. Carson about her prognosis. We have to trust them."

So Beth had arrived at Hopkins on Groundhog Day to prepare for surgery.

As usual, the doctors wanted to perform more tests — videotaping her answering questions and performing psychological tests with

the 16 probes of an electroencephalograph attached to her scalp, monitoring the brain's electrical activity. There were walking tests and breathing tests and blood tests and all kinds of tests that it seemed she must have suffered through a hundred times before.

How did she like all these tests? she was asked, as a nurse removed the EEG probes.

"I hate them," Beth replied. She submitted to the nurses and doctors politely, almost cheerfully, but she had her opinions.

As had been usual with Beth's hospital experience — her mother called her "the Murphy's Law kid" — nothing went smoothly. The bleeding test on Monday, Feb. 2, indicated a possible clotting problem, the same problem that had postponed surgery two weeks before.

So Beth's private tour of the White House — arranged by a Washington foundation called Make-A-Wish that does nice things for seriously ill children — had to be canceled. Beth, who had endured the interminable torture of medical tests without a tear, cried for an hour when her parents told her.

Tuesday, there were more blood tests, tests that indicated the first test, which showed a problem with clotting, was wrong — probably. To further complicate matters, Beth had developed a cold, which would make the lengthy anesthesia for hemispherectomy somewhat riskier. But she'd be fine, the doctors said — probably.

As the sun set Tuesday night, a half-dozen doctors huddled to discuss whether the first abnormal bleeding test or Beth's sniffles were reason to postpone the surgery. They knew a second postponement would be enormously difficult and inconvenient for the family, and operating room schedules on several other cases had been shuffled to free an operating room all day Wednesday. Yet they did not want to compound the risks of a horrendously risky procedure.

After 45 minutes, the conference broke up, the doctors in agreement that no delay was warranted. "We're on," Dr. Freeman told the Ushears, who waited nervously in the playroom of the pediatric neurology unit on the fourth floor of the Johns Hopkins Children's Center. Brian and Kathy looked at one another with a mixture of relief and dread.

Beth was getting tired of questions. When Dr. Freeman invited her over to the little table in the playroom, she went to him reluctantly.

He told her one last time about the surgery — not the gory details, but the purpose, the "real short haircut," the fact that she'd be asleep the whole time, that she'd have headaches after she woke up.

"Do you have any questions about the operation?" Dr. Freeman asked, seriously, gazing into the little girl's eyes.

Beth shook her head, and smiled a one-sided smile.



PART II

FAMILY
HAS
LONG
DAY OF
HOPING



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First, there was the light -- intense, white, bathing the entire surgical team in the perfect clarity of a movie set.

Then, there was the noise — the loud whine of a drill, indistinguishable from the sound of a workshop power drill, singing out above the beat of a radio's pop music. Dr. Ben Carson prefers classical, but the surgical team almost always overrules him.

The only part of Beth Usher not covered by the blue drapes was an oblong, convex, 5-by-7-inch patch of skull. Her scalp was peeled back on each side like the pages of a book. When the drill finished its work, the bone was pulled away to reveal the dura, the tough, white covering of the brain.

It was a few minutes after 10 a.m., Wednesday, the 4th of February.

Coma. Paralysis. Death. Those were the risks of hemispherectomy, the surgical removal of half of the cerebral cortex. Yet it was the only treatment for Rasmussen's encephalitis, a severe, progressive seizure disorder that otherwise would make Beth Usher, 7, a candidate for a nursing home within a few years.

On one side of the drapes, Dr. Carson, chief of pediatric neurosurgery at Johns Hopkins Hospital, and Dr. Nathan Moskowitz, senior resident in neurosurgery, went to work on the left hemisphere of Beth's brain. They wore blue caps, masks and surgical loupes, special glasses with magnifying lenses and fiber-optic headlights for perfect illumination of the area being worked.

On the other side of the drapes, a team of anesthesiologists headed by Dr. Stephen Derrer, working with a beeping, flashing tangle of electronic equipment and intravenous lines, concentrated on keeping the rest of Beth alive.

It was Dr. Carson's second operation of the day. Earlier, he'd stopped by another operating room for a 15-minute procedure, installing a tiny reservoir near the cancerous lesion on the brain of another little girl, to be used for taking samples and applying chemotherapy.

With Beth's brain open, the surgeons attached a "tiara montage" — an 18-electrode crown hooked up to an electroencephalograph, a machine that records electrical activity in the brain. On a chart of the brain, Dr. Carson drew in the locations of the electrodes. Neurologists John Freeman and Eileen P. G. Vining stopped by to look at the EEG results.

To no one's surprise, it showed glaring abnormalities, especially in the areas controlling speech and movement. Paradoxically, the severity of the damage was reassuring to the surgeons, who now felt certain that removing 40 percent of Beth Usher's brain tissue was the right medical course.

Working steadily but without hurry, changing tools every minute or two, constantly suc-

tioning off the blood to keep the view clear, the surgeons moved ahead. Cutting down through an area called the Sylvian fissure, they located the middle cerebral artery and put a tiny silver clip on it, taking care not to damage the branching vessels that feed the brain stem and basal ganglia, portions of the lower brain that were to remain untouched.

Next, they cut between the two hemispheres, hugging the fibrous divider called the falx. But they quickly ran into swollen veins that bled at the touch and seemed to be under extremely high pressure. Dr. Carson realized they had to back off, tackling that area only after most of the tissue was removed.

They traced the anterior cerebral artery and clipped it, then worked under the temporal lobe to find the posterior cerebral artery. Once all three major arteries were clipped, the color of the brain tissue changed, growing darker as it lost its supply of oxygen-carrying blood.

Now, with the major arteries taken care of, they could start to free the tissue, cauterizing vein after vein, slowly separating the most complex human organ from its myriad ties to the body.



"Kids are neat"

Coma, paralysis, death.

But Dr. Carson, 35, had said he doesn't get nervous about such operations. A religious man, he said he believes God helps him in the OR. His colleagues say he remains calm, cheerful, imperturbable, even in the midst of medical catastrophe.

The night before, Dr. Carson had rated hemispherectomy "maybe a seven and a half" in difficulty, on a scale of one to 10. On the one hand, separating a tumor from vital brain tissue can be far trickier. On the other hand, hemispherectomy is an all-day, marathon affair, with ample opportunity for error, repeatedly putting the scalpel within millimeters of critical parts of the lower brain.

Fatigue? "It just never happens," he said. "Most surgeons will tell you you get a surge of adrenalin when you're going into a big operation. Your bladder goes on vacation. Your stomach goes to sleep. The time just zips by. Six hours, when you're really concentrating, can seem like one hour."

Dr. Carson had chosen to specialize in children partly because he was drawn by their straightforwardness. In adult patients, by contrast, the physical and psychological components of illness are often inextricably tangled, he said.



"The nice thing about kids is they're not malingeringers. What you see is what you get. It's pure pathology," Dr. Carson said. Plus, he added, "Kids are neat."

Dr. Carson's co-workers insist they have never seen him mad, never seen him truly worried. "Even when I feel I'm going into a situation where there's a very high risk of mortality, I don't get nervous," he said. "Getting nervous doesn't help you."

But Dr. Carson, the father of three young sons, admitted that it's harder for him to keep his equanimity when the patient is his own child. One night a few weeks before Beth's surgery, one of his sons had begun vomiting repeatedly.

"Being a neurosurgeon, I immediately began to think 'brain tumor,'" he said. "Naturally, it turned out to be the flu."

Knock-knock jokes

Coma. Paralysis. Death.

A hundred yards of twisting corridors from the operating room where Beth Usher lay, the surgical waiting room was jammed with relatives of a score of patients, sitting tensely, waiting for some word.

Brian and Kathy Usher and Beth's 10-year-old brother, Brian Jr., had been joined for the vigil by Kathy's father, William McNamara, her sisters, Maureen McNamara and Sarah Liguori, and Sarah's husband, Vinny. A sign in the waiting room set a limit of two family members per patient, so the rest of the family sat down the hall on some benches next to the seventh-floor elevators.

They tried to cheer one another up, telling and retelling how Beth had regaled the nurses with knock-knock and why-did-the-chicken-cross-the-road jokes up to the moment the anesthesia knocked her out. But there were long silences. No one really felt like talking.

Every half hour or so, an emissary from the operating room would phone or stop by with a progress report. Each time, Brian and Kathy would sit up and stiffen, fearing the worst. Each time, the message was reassuring: Everything is going well. Everything is as they expected. There are no problems.

Out in the corridor, every so often, a patient would roll by on the way out of surgery, an IV bag dripping into his veins, a heart monitor flashing and beeping in cardiac rhythm. An intern would pass, looking sleepless. A nurse would walk by, balancing lunch on a tray.

They were hospital people, for whom other people's surgery is a workday routine.

But in the waiting room the family members existed in a wholly different world, watching the clock, wanting the minutes to speed by, wanting the day to be over.

By the late afternoon, most of the family had left to take Brian Jr. home to Connecticut. The waiting room had gradually emptied, as patients undergoing less lengthy operations came out of the surgery suites and headed for a recovery room or intensive care.

Only the Ushers remained. Kathy slumped in Brian's arms, exhausted with the waiting. Maureen paced nearby.

Waves in a pond

In the operating room, a hole gaped where Beth's brain had been.

The whitish skull bones were visible. A large piece of brain tissue sat in a bowl nearby. It was hard to believe that the child beneath the blue drapes was still alive.

"We're just about finished with the resection [removal of tissue]," said Dr. Carson. "We just have some high-pressure veins that we have to close off, and then I think we'll be in Fat City." It looked as if he might be smiling underneath the blue mask.

Gone were the four lobes of the left hemisphere: frontal, controlling intellect and personality, speech and motion; temporal, responsible for processing sound, calculating and musical ability, and emotion; parietal, in charge of the senses of touch and balance; and occipital, controlling vision.

But most of the tissue of the left brain looked so damaged that the doctors believed it no longer could have been doing much work. The billions of nerve cells of the right hemisphere must have taken over; otherwise, she could never have functioned so well before surgery.

The pattern of inflammation and scarring was in concentric circles, "like you'd thrown a pebble in a pond," Dr. Carson said later. "The entire hemisphere was extremely abnormal in feel — very knobby, almost as if there were pebbles or peas under the surface."

With the same calm, steady motions they had been using for six hours, Dr. Carson and Dr. Moskowitz sealed off veins with a heated, two-pronged, fine-tipped instrument.

They discovered a tiny hole through to the sinus, made accidentally as they were cauterizing veins. They cut a piece from the temporalis muscle, which controls chewing, and used it to patch the gap.

Then they cleaned up the empty space, flooding the entire skull with saline solution to wash out blood, which might cause infection. They

covered the skull lining, the sinuses and the exposed parts of the lower brain with a gel containing a natural clotting factor, as a precaution against bleeding.

They stitched the dura together, filling the cavity again with saline before putting the final suture in place. The hemispheres' fibrous divider would hold the right brain in place, and the empty hemisphere would eventually fill with spinal fluid.

The skull bone was retrieved from the table, where it had been wrapped in a damp sponge, and wired back in place. Then the scalp was stitched together.

The surgery was over, 10 hours after it had begun.

In the corridor, the gurney paused for a few minutes on the way to the pediatric intensive-care unit. Beth's parents wordlessly embraced the child's unconscious form, then the doctors, then one another, the tears coming after the day's tension.

Dr. Carson was talking, but they could take in only phrases: "stable as a rock . . . went just fine . . . wait and see . . ."

A couple of the earlier hemispherectomy patients had made meteoric recoveries, saying their first words on the way out of the operating room. That was clearly not going to happen with Beth, and the doctors had decided to leave her on the respirator overnight because of her cold.

But she'd survived. When Dr. Carson and Dr. Freeman returned from intensive care an hour later and said she was responding to commands, Brian, Kathy and Maureen were ecstatic.

"If you ask her to squeeze her left hand, she squeezes it," Dr. Freeman said. "If you ask her to wiggle her toes, she wiggles her toes."

"The main thing I always worry about is the brain stem," the critical part of the lower brain that controls breathing and heartbeat, Dr. Carson said, "and that's working."

"She's not out of the woods yet, but she's nearing the clearing. We still have the potential for bleeds. We'll just take it day by day."

A terrible trembling

But by late that night, the immediate euphoria following the surgery was gone.

After the good initial signs — responding to commands, moving all four limbs — Beth had gradually become unresponsive, slipping deeper and deeper into unconsciousness.

Coma, paralysis, death.

At midnight, the doctors rushed her to the basement for an emergency X-ray scan of the brain. It showed swelling of the brain stem, ap-

parently in response to the trauma of surgery. She was given drugs in an attempt to control the swelling, since increased pressure inside the skull could be dangerous, even fatal.

All night, Brian and Kathy sat at Beth's bedside in the pediatric intensive-care unit, where the machinery loomed over her pale and swollen body. Starting at about 2 a.m., she began shaking, a frightening, terrible trembling of her entire body that did not stop for hours.

Her temperature and blood pressure rose. She seemed to her parents to be in pain. They played a tape of Mr. Rogers next to her pillow to cover the beeping and buzzing of the life-support machinery, but they had no confidence that she could hear his soothing voice.

"It's been very, very scary," Kathy's sister, Maureen, said Thursday morning, Feb. 5, near tears outside the wide electric door to the pediatric intensive-care unit. "They say the worst is yet to come, because of the swelling. They keep saying, 'It's all expected, it's all expected.' But I don't think so. I think it's not going well."

"If we could see just a little sign that she's getting better. If just one eye would open . . ."

That afternoon, in a tiny conference room, Diana J. Pillas, a counselor in the pediatric neurology program, spoke for an hour with Kathy, Brian and Maureen, urging them to get food and sleep, reminding them that they could be in for a long wait.

Kathy found herself remembering her mother's death from cancer. She recalled how every time the ailing woman tried to talk about her impending death, well-intentioned family members would cut her off and try to cheer her up.

"She knew," Kathy said. "She knew she was dying. That's my only regret now, that we didn't let her talk about it."

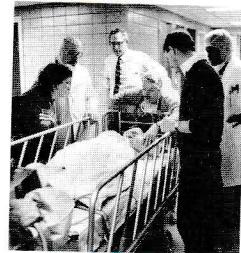
The parents, seeing the puffy, tremulous, comatose figure of their daughter and remembering the bright, energetic child who had entered the operating room the day before, began to wonder aloud about whether they had done the right thing.

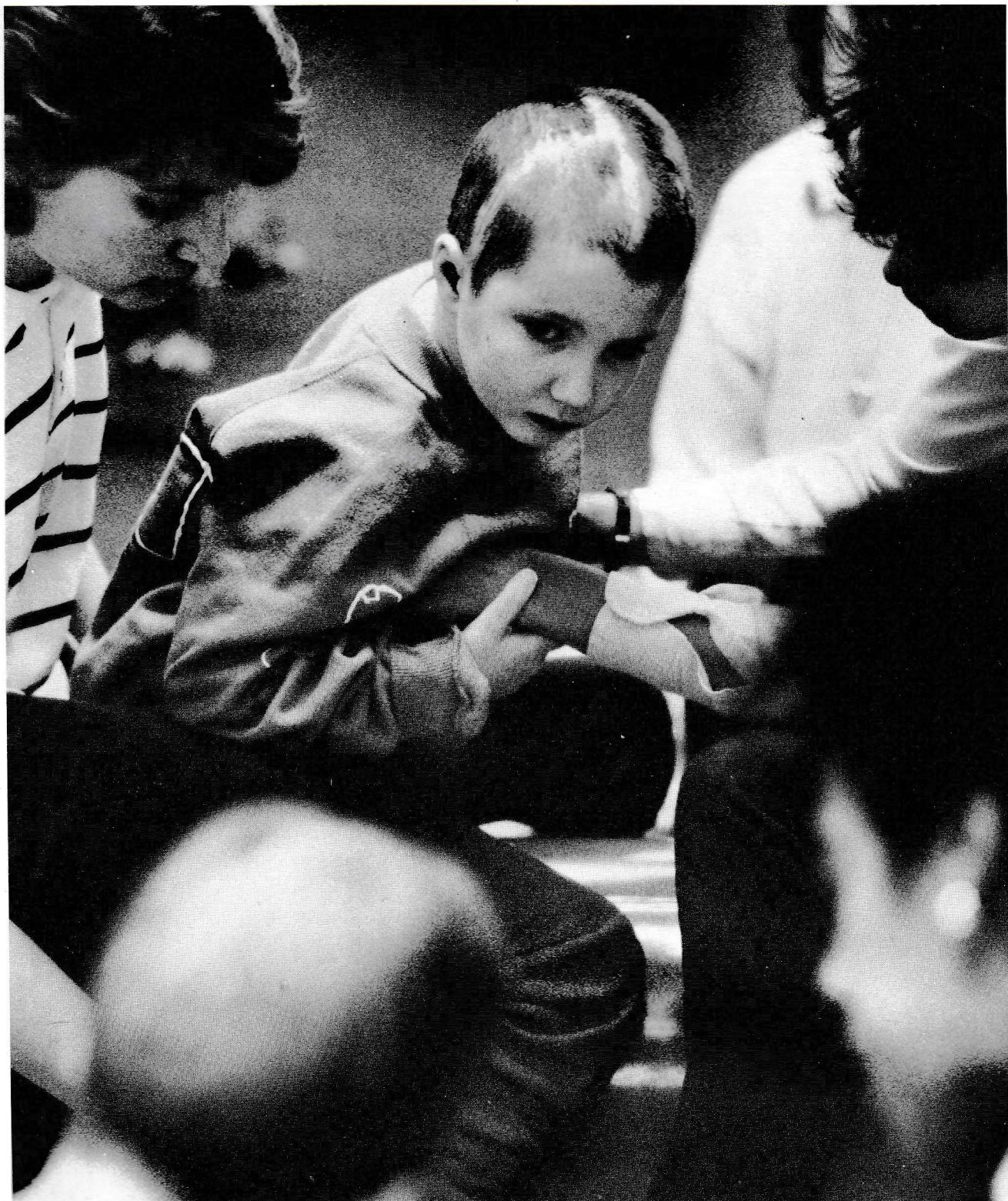
Had they rushed the surgery? Had they sentenced their daughter to a tortuous semi-consciousness, perhaps even to death?

Diana Pillas reassured them, reminding them of the terrible future Beth faced without surgery, reminding them that the brain tissue taken out had been riddled with disease. "She has to be better off without that! She has to! Of course you did the right thing," she said, gripping Kathy's hand.

The Ushers heard her words. But only Beth's condition could offer real reassurance that their decision had been correct, and her condition was in grave doubt.

"We could have had two good years with her —" Kathy Usher said. Her voice trailed off into tears.





REPRINTED FROM A SERIES WHICH APPEARED ON MARCH 29 - 31, 1987 IN THE BALTIMORE SUN



Beth Usher's recovery from radical brain surgery required many grueling hours of physical therapy, as her right brain hemisphere gradually learned to take over the functions previously performed by the diseased left hemisphere. Johns Hopkins Hospital therapists support Beth as she masters sitting up (upper left), practices muscle control of her hand (lower left), and takes her first, hesitant steps (upper right) about eight weeks after surgery. At lower right, a month after her return home to Connecticut, Beth gets a hug from her physical therapist, Janet Frattaroli, as her walking continues to improve.

PART III

JOURNEY
BACK
THROUGH
THE
LOOKING
GLASS



REPRINTED FROM A SERIES WHICH APPEARED ON MARCH 29 - 31, 1987 IN THE BALTIMORE SUN

"Dad."

It was only a whisper, but at 1 a.m. in the hushed neurology ward of the Johns Hopkins Children's Center, it was unmistakable.

Brian Usher, who was preparing to lie down to sleep in the chair in his daughter's hospital room, stepped over to Beth's bed and leaned over her.

"Dad.

"My ... nose ... itches ... me."

Mr. Usher hugged Beth and talked excitedly to her, trying unsuccessfully to coax her to repeat the words. Then, wearing only underwear, he ran into the hall, grabbed a passing intern and told him what his daughter had said.

It was Sunday, March 8. It had been 32 days since the neurosurgeons had removed the left hemisphere of Beth Usher's brain, in a 10-hour operation to stop a rare disease that otherwise would have left her totally disabled from constant seizures.

Since Beth was right-handed, her left hemisphere was the so-called "dominant" hemisphere, which ordinarily would be expected to control speech.

But her rare disease, called Rasmussen's encephalitis, had gradually spread across the left hemisphere, leaving inflammation and scarring in its wake. Based on experience with other cases of Rasmussen's, the doctors believed many of the functions normally located in the left side of Beth's brain would have transferred to the healthy right side — but they could not be certain.

In particular, they could not be sure about speech. A pre-surgery test designed to find out where in her brain speech was located had failed. Only Beth's progress after the operation could tell them whether she would have to learn to talk all over again at the age of 7.

So the whisper in the dark hospital room was proof that the surgeons had not excised Beth's ability to speak along with the diseased tissue. It was a giant step in the girl's long, slow journey out of coma.

It was a promise, after more than four weeks of waiting, that Beth Usher was coming back.

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Before Brian and Kathy Usher decided to go ahead with surgery for Beth, they had talked to several parents of children who had undergone hemispherectomy at Hopkins during the last two years. They had grilled the doctors, again and again, about the earlier cases.

Of the eight recent cases, only one child had been comatose for more than a few hours after the operation. She had regained consciousness after about two weeks and made rapid progress. Several of the other children had been talking virtually on the way out of the operating room.

Before surgery, moreover, most of the other children had been far more disabled by seizures than Beth, still an active, articulate, happy child. So her parents, despite the doctors' cautions, thought there was every reason to believe their daughter would recover quickly after surgery.

Instead, she fell into a coma. An emergency scan showed swelling of the brain stem and evidence of slight bleeding in the good hemisphere. Through the night after surgery and the next morning, the doctors were worried.

That afternoon, they took a second brain scan. At 3 p.m., Dr. Ben Carson, who had performed the surgery, walked into the pediatric intensive care unit, where the Ushers stood beside Beth's bed.

"I had thought the swelling was getting worse, but it's not," Dr. Carson said. "It actually looks better. A ventricle was visible that we couldn't see before. So I still have a good feeling about her."

Later, Dr. Carson said he thought it possible — though unprovable — that the brain swelling suffered by Beth and the earlier patient with the two-week coma might have been caused by slight shifting of the brain as they were moved following the operation. He and Dr. John Freeman, Beth's neurologist, agreed that future patients should be kept immobile during the first hours after surgery.

Virtually from the moment of Dr. Carson's bedside remarks about the second brain scan, there was a chasm between the doctors' view of Beth's condition and the family's view.

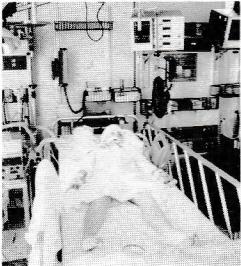
Within a few days of surgery, the doctors no longer appeared seriously concerned about Beth's long-term prognosis. They had seen innumerable patients emerge from coma produced by internal swelling from trauma to the brain. Since Beth seemed to be improving, they appeared fundamentally confident of her ultimate recovery.

"Right now, she's very much like someone who's been hit by a car," Dr. Carson said on the second day after surgery. "It's possible for a kid with brain-stem swelling to be out for weeks. I've seen it for six months. But there's every reason to believe that she'll come out of it and do well."

To Beth's parents, by contrast, her condition after surgery was cause for deep, unremitting fear.

It was their decision, after all, that had sent Beth to the operating room. She had come back prostrate, swollen and unresponsive, unable to hear or talk or see, hooked up to the tubes and wires of an intensive-care bed.

They understood, intellectually, what the doctors kept telling them. They knew, when they tried to be rational, that their decision to go ahead with surgery had been correct. But emo-



tionally, the weeks after Feb. 4 were torture.

"It requires a lot of faith on the part of the parents," Dr. Freeman said of Beth's long journey out of coma. "It's faith on their part. But it's experience on my part."

During the first days after surgery, the Ushers didn't eat. They slept little, an hour or two at a time, on cots in a conference room near intensive care. One parent or the other was always at her side. They learned to interpret the shifting numbers on blood pressure, temperature, spinal fluid protein count and a half-dozen other indicators of Beth's medical progress. They prayed.

Inevitably, there were moments when they were wounded by chance remarks of people for whom medicine, after all, is a job.

A few hours after surgery, a doctor peripherally involved in Beth's case stopped by for a word with Kathy Usher. He told her he thought Beth had weathered the surgery well. Then, as he strolled away, he said, "I hope you made the right decision." He did not see the anguish in her face at the questions raised by his comment.

On another occasion, the Ushers winced when they heard a nurse refer to Beth coolly as "what's-her-name."

But most of the time, Beth's parents felt overwhelmed with gratitude for the care Beth was getting.

"The nurses here are incredible," Kathy Usher said one day. "They take care of the parents, as well as the patient. They'll ask you if you want a cup of tea. They'll bring you food. They've answered every question 10 times, and they've taught us how to help out with Bethie's care."

□

Monday, Feb. 9:

The nurses, with some fanfare, moved Beth from the intensive care unit to 4-East, the pediatric neurology floor. Kathy Usher had taped an old snapshot of her daughter to the metal rail of the hospital bed "to let them know what she really looks like."

"If you want to hear Mr. Rogers," said Brian Usher in his loud-and-clear football coach's voice, "move your foot. Give us a big movement."

Beth's left leg trembled slightly. "That's right! Good girl!" Brian said, switching on the child's tape recorder beside the pillow. But from time to time, Beth's legs moved again, making it impossible to be certain that she had responded to his words.

"It's hard," Brian said. "You're on a roller coaster. We've been with her around the clock, and we think we're seeing changes, but we're not sure."

Diana Pillas, a Children's Center counselor, stopped by to offer support. Kathy Usher began to compare Beth's progress with that of the earlier hemispherectomy patients, and Ms. Pillas

interrupted.

"It's real hard not to compare," she said. "But you ought to pretend she's the only girl who ever had this surgery."

Thursday, Feb. 19:

Mr. Rogers, of TV's Mr. Roger's Neighborhood, came to see Beth.

Kathy Usher had written to him about Beth's illness and impending surgery. To her surprise, Fred Rogers had telephoned and spoken with the little girl at length before the operation. He called several times after surgery to check on her progress. One day, out of the blue, he announced he was going to pay a visit.

Without any public announcement, he flew to Baltimore from his Pittsburgh home. He drove to Johns Hopkins Hospital, took the elevator to the fourth floor, and spent nearly an hour with Beth, talking to her and running through his repertoire of puppet characters.

Though still in a light coma, Beth was having distinct periods of sleeping and waking. While Fred Rogers stood at her bedside, she slept.

Her parents poked her, spoke in her ear and tickled her feet.

Beth slept.

Then Mr. Rogers put the puppets back in his briefcase and took the elevator back downstairs. As he left the hospital, he said Beth had made a strong impression during their phone conversation before surgery.

"It was a real treat for me to talk with her — she's such an imaginative child," he said. "My plane was late today, so I only had an hour with her." Then he drove back to the airport and flew home.

A Baltimore friend who had driven him to and from Hopkins was curious. "You get 1,000 requests a year like this. Why are you doing this?" the friend asked.

Mr. Rogers replied: "This wasn't a request. It was just something I had to do. I just had a hunch I ought to come."

Beth's personality, illness and surgery had caught imaginations other than Mr. Rogers'. Relatives, friends and friends of friends sent her cards, paintings, photos, tapes, books, flowers, candy, balloons and stuffed animals.

The president of the University of Connecticut, her parents' employer, sent a bouquet. Beth's cousins chipped in and bought a talking dog containing a computer chip programmed with "over 1,000 phrases." A class of Baltimore County second-graders heard about her and sent get-well-soon notes. The walls of Room 427 were papered with kids' bright tempera paintings, Valentines, poems and cheerful messages.

Did Beth notice any of it? It was hard to say.

At times, she seemed to be aware of her parents' presence, their words, their actions. At other times, Brian and Kathy weren't so sure.

The Ushers had come prepared for a three- or four-week hospital stay. Yet here it was almost

three weeks after Beth had entered the hospital, and there was still no hint of when she might be well enough to go home.

The University of Connecticut, where Brian worked as a football coach and Kathy as an athletic fundraiser, was being generous with paid leave, but that could not last indefinitely. Bills were piling up at home. Brian Jr., 10, who was living with relatives in Connecticut, missed his parents and sister.

But the practical difficulties posed by the lengthening hospital stay seemed trivial in comparison to the fears that Beth's slow progress was a symptom of permanent damage.

Brian had said just the day before Mr. Rogers' visit that the imperceptibly slow pace of recovery was almost frightening. "We're getting a little discouraged, a little down," he said. "I'm just afraid something's wrong, and that she won't wake up."

To counter such fears, there were always the doctors, bearing good news, impressed by changes that seemed minuscule to the Ushers.

An hour after Mr. Rogers left, Dr. Carson came by, between operations. Beth had opened her eyes. Dr. Carson chatted about Mr. Rogers while he carefully manipulated her limbs, gauging the stiffness in the muscles, a measure of brain recovery.

"That's improving a lot. That's coming along," Dr. Carson said. "She's a million times better than she was a week ago." Kathy and Brian looked encouraged.

Then, as he strolled out of the room, he added over his shoulder, "But I think we still have a couple of weeks before we're in Fat City."

Kathy looked at Brian and slowly shook her head. "A couple of weeks," she said, "is a long time."

Thursday, March 5:

Brian had taken a turn sleeping "out" in a room across town at the Ronald McDonald House, a charitable organization that provides housing to families of seriously ill children. When he awoke, he phoned Kathy in Beth's room.

Kathy, in an oft-repeated gesture that had never gotten a reaction, held the phone out to Beth. "Bethie, do you want to talk to Daddy?" she asked.

To her astonishment, Beth reached out with her left hand, brought the phone to her ear and whispered, "Dad. Dad. Dad." Then she kissed the phone.

In that moment, Beth had demonstrated a great deal: She could talk, at least in monosyllables. Memories were preserved. Emotions were intact.

Later that morning, Dr. Nathan Moskowitz, the senior neurosurgery resident who had assisted Dr. Carson in the operation, strolled in and said, "Beth, hold up two fingers."

Her left hand shaking slightly, she held up two fingers.

"Hold up one finger," he said.
She held up one finger.
Progress.

□

The doctors had always said that once Beth began to emerge from coma into consciousness, her recovery would accelerate. It did.

Four days after she said "Dad" into the phone, Beth said that first sentence: "My nose itches me." She began to sit up, to wave goodbye, to wink and smile and follow people with her eyes.

A therapist, then her mother, began to feed her with a spoon, bananas and tapioca and chocolate pudding. Finally, they took the feeding tube out of her nose, and for the first time since surgery, she had no medical attachments.

Brian Usher left his daughter for the first time and spent four days looking at rehabilitation hospitals within a few hours' drive of their Connecticut home. When he returned, he remarked that Beth had made more progress in those four days than in the previous four weeks.

For the first time, the tension was disappearing from Kathy and Brian Usher's voices. Their spirits were high. At last, they shared the doctors' optimism.

At 4 one morning not long after Beth began to talk, Brian was lying beside his daughter in her bed and discovered they both were awake. He began quizzing her, probing the limits of her mental recovery. He gave her math problems. He asked her about people, places and events at home before surgery.

Finally, Beth stopped him. "Dad," she whispered, "I remember *everything*."

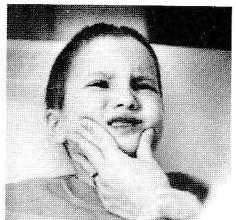
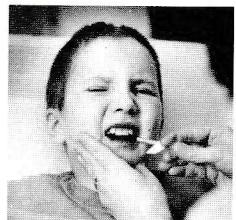
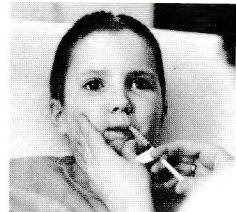
In fact, Beth was only slowly recovering her short-term memory, occasionally forgetting things that had happened hours or days earlier. But as had been the case with the earlier hemispherectomy patients, her long-term memory seemed undamaged.

"We don't know why they don't lose memories," said Dr. Carson. "It could mean the same things are routinely stored on both sides. It could mean that memories are transferable, and they get copied onto the other side as the disease progresses."

The doctors could say what was happening in the brain, Dr. Freeman noted, but they often could not say how or why it was happening.

"If you look behind our arrogance," he said, "you find there's very little we know about most things. What causes a seizure? How's aspirin work?"

Like her memories, Beth's personality seemed untouched by the surgery. She was affectionate and friendly, blowing kisses and waving to visitors. Almost from the beginning of her





return to consciousness, her sense of humor was evident.

One day, when Kathy Usher was concerned about Beth's flagging appetite, she stopped a resident physician outside the room and asked him to tell her sternly that if she didn't eat they would have to put back in the hated feeding tube.

But when the resident went in and soberly delivered his speech, Beth simply winked at him. He burst out laughing, and the plot failed.

Sharon Bartlett, a favorite teacher who had sent her a card every day of her hospitalization, arrived on the train from Connecticut for an overnight stay. She and Beth laughed over old inside jokes and talked about school.

"That humor's still there," Ms. Bartlett said. "She's got an incredible will to achieve, and that's still there, too. She never gives up. She's not a quitter."

Physically, however, Beth's progress came at the price of pain and hard work. Weeks of immobility had left her muscles stiff and shrunk. The nerve cells of her right brain hemisphere were learning to take over functions that had been handled before surgery by the diseased left hemisphere.

Beth began two hour-long physical therapy sessions each day, descending one floor in her wheelchair to a bright room with mats on the floor and mirrors on the wall.

Supported by therapists Beverly Duvall and Michelle Malanoski, she took her first, halting steps. At their urging, she would hold her upper body erect for the count of 10; grasp and replace the pieces of a simple, toddler's puzzle; swat at soap bubbles blown by her mother.

Often, her face would contort with the pain of stretching long-unused muscles. "It hurts! It hurts!" she would say in her whisper, which was slowly gaining volume. Occasionally, she'd get angry, slump to the ground and begin to cry. But at nearly every session, she made perceptible progress toward normal muscle control.

□

To a conference with the doctors during Beth's seventh week in the hospital, Brian and Kathy Usher brought a long list of questions about the future. As always, many of the doctors' answers were a little tentative; in a rare disease treated by a rare operation, medical science offered no absolute certainty about long-term consequences.

But the doctors had, as Beth put it one day, "taken the seizures out and thrown them in the garbage." For the first time in more than two years, Beth was taking no anti-seizure drugs. Her electroencephalogram showed none of the abnormal electrical activity it had recorded in her brain before the operation.

"Beth has turned the corner," Dr. Carson said. "We think she'll continue to make rapid progress. She's still going to need a lot of therapy — months, maybe even years. But we fully believe she'll be ambulatory by herself."

The doctors agreed with the Ushers' decision to choose a rehabilitation unit at Newington Children's Hospital, 40 minutes from their Storrs, Conn., home. They said Beth might need a few weeks there before switching to outpatient therapy.

Already, Beth's treatment had cost an estimated \$200,000 — half of that before she came to Hopkins and the rest for the operation and its aftermath. But the Ushers said most of the bills were covered by insurance, and friends had started a fund to help pay uncovered costs, so they were not seriously concerned about money.

The doctors said some of the pre-surgery weakness in Beth's right arm and leg probably would be permanent. The right side of her field of vision was permanently reduced, forcing her to turn her head farther than normal to see objects to her right. That visual flaw might eventually prevent her from getting a driver's license. But the doctors said they believed she would walk and talk normally, or nearly so, by summer — perhaps by the time of a planned gathering of the nine recent Hopkins hemispherectomy patients in June.

Of the possible long-term complications, the doctors said the most likely would be hydrocephalus — a buildup of fluid in the brain. The first symptoms would be lethargy and vomiting.

"That's of concern, but it's eminently treatable," Dr. Carson said. "If she gets involved in head trauma, she should have a neurological exam. I don't have any other special concerns."

As the conference ended, the atmosphere lightened. The Ushers asked whether Beth could return immediately to swimming, a favorite pastime before surgery. "No problem at all," Dr. Carson said. "I guess I would just sacrifice diving. I don't know what would happen, and I'm not sure we want to find out."

Brian Usher, who had taken some kidding from the doctors for his skepticism about their repeated reassurances, tried to pin down them one last time. "I know you can't give us a scenario, but is everyone confident she's basically going to be all right?" he asked.

"I think," Dr. Carson replied, "that Beth's going to be a superstar."

Down at the end of the conference table, nurse Maybian Pruet, one of a half-dozen nurses who had grown close to the Ushers during Beth's long recovery, began to cry.

"I'm happy," she said, smiling, in answer to the quizzical eyes that turned her way. "It's not often you get to cry for happiness around this place."

□



As Beth's discharge date drew near last week, Kathy took her daughter on her first venture outside Hopkins, hailing a taxi to the Inner Harbor. Tired of hospital food, Beth feasted on french fries, fresh lemonade and a hot fudge sundae.

Her mother wheeled her chair around Harborthouse and across the elevated walkways outside, where Beth enjoyed the spring sun. Beth picked out a harmonica, shaped like a Maryland crab, to build her wind and speed the return of normal speech.

After eight weeks at Hopkins, and despite the first-name friendliness of the nurses, the Ushers were finding the hospital increasingly oppressive.

Before, its high-tech medicine had been Beth's lifeline and the family's security. Now it was beginning to feel like a prison. Beth was getting better, but all around her were seriously ill children, some in pain, some with a bleak prognosis.

"I just want to get away from all this sadness," Kathy Usher said. "We just want to go home, and be happy and try to live a normal life for a while."

Last Friday afternoon, as her parents packed for the weekend's drive to Connecticut, Brian Jr. clowned with his sister, pushing her along the halls in the oversized stroller her parents had purchased for use at home. A steady stream of doctors, nurses, counselors, therapists, patients and parents who had heard Beth was getting out stopped by to say good bye.

In a demonstration for the neurosurgeons, Dr. Carson and Dr. Moskowitz, Beth walked a dozen steps across the neurology unit's playroom, supported on only one arm. Everyone clapped.

A few minutes before the Ushers' departure, Dr. Freeman breezed into Beth's room.

"We'll miss you, Bethie," he said, pulling her into his lap. "Thank you for being special."

"Thank you for being a special doctor," Beth replied.

"When you come back in June, you'll be walking without anyone holding you. You'll be talking loudly," the doctor said.

"I won't have any more seizures," the patient replied.

"No more seizures," Dr. Freeman said.

"No more seizures," Beth repeated.

