Framework

Johns Hopkins Orthopaedic Surgery

JOHNS HOPKINS MEDICINE

NEWS FROM JOHNS HOPKINS MEDICINE

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Milestones Preserve Walking for Pediatric Patients with Cerebral Palsy

"The right procedure at the right time is critical when treating children with cerebral palsy," says **Ranjit Varghese**, Johns Hopkins orthopaedic surgeon and medical director of the Ortho-Cerebral Palsy program at Kennedy Krieger Institute. Varghese understands the importance of this concept from his specialized single-event multilevel surgery (SEMLS) approach to treating children with spastic diplegia and his work with multidisciplinary programs that focuses on tone management and continuity of care of such patients from infancy through adulthood.

Caleb Schneider (left) is one of Ranjit Varghese's patients.

"Most children with CP will need some kind of orthopaedic intervention, so we have a path of treatment that reduces the total number of procedures they need."
–Ranjit Varghese

During early childhood, Varghese sets his patients on a trajectory for surgical success and improved quality of life by creating treatment milestones. "Most children with CP will need some kind of orthopaedic intervention, so we have a path of treatment that reduces the total number of procedures they need," says Varghese.

Varghese believes that the management of patients with cerebral palsy must be individualized according to the child's clinical presentation and that it requires a multidisciplinary approach. At Kennedy Krieger Institute, he is involved in a multidisciplinary spasticity management program that includes pediatric specialists from the departments of neurosurgery and physical medicine and rehabilitation.

"We review ambulant children at 3 to 4 years of age who will benefit from surgery that reduces muscle tone," explains Varghese.

Some of these children may benefit from selective dorsal rhizotomy—a neurosurgical procedure that reduces muscle tone permanently. Once the muscle tone is reduced, the child begins rehabilitation to learn to walk again. Approximately two years later, the child's gait is analyzed to determine whether orthopaedic surgery is needed.

"As the child grows, the bones may grow in an abnormal manner. That doesn't go away when you do selective dorsal rhizotomy, so we realign the lower extremities via SEMLS," says Varghese. "I can do all of the lower extremity surgeries at once. Then hopefully we do not have to intervene again in the future."

The goals of these early surgeries are to improve children's quality of life as they transition through puberty and into adulthood and to preserve their ability to walk. For children who do not receive early intervention, CP can lead to many orthopaedic surgeries throughout adulthood, as well as physical therapy and rehabilitation.

Varghese and his team provide caregivers the security of knowing that their child not only has a long-term treatment plan but also a team of doctors who will see him or her through to adulthood.

"Sometimes as a parent or a referring physician, you are not sure what the individual child needs," says Varghese, "but this system enables us to connect the child to the right specialists with the right treatments to minimize the burden on patients and families."■

Remarkable Recovery
Inspires One Family's Giving

In 2005, Kerry Burton gave birth to her third child, Benjamin. She'd had a healthy, full-term pregnancy but then, Burton recounts, "about two hours after Benjamin was born, we noticed he wasn't coming back to my room." To her surprise, she learned that Benjamin had temporarily stopped breathing and was being admitted to the NICU.

Soon, doctors discovered that Benjamin was missing several ribs and that the bones in his neck were malformed. "We sat in the doctor's office literally looking through books, trying to figure out, what does this mean?" says Burton.

The family was referred to **Paul Sponseller**, chief of pediatric orthopaedics at Johns Hopkins. "Ben had malformations of all of his cervical vertebrae," Sponseller explains. The decision was made not to intervene surgically at that point. "We decided to monitor him and wait until he had enough growth in height to correct his head, which over time became very tilted."

Sponseller evaluated Benjamin's growth and scoliosis regularly. The plan was to delay surgery until he had neared skeletal maturity, in the teen years.

"Ben had malformations of all of his cervical vertebrae… We decided to monitor him and wait until he had enough growth in height to correct his head, which over time became very tilted."
–Paul Sponseller

However, at age 6, Benjamin's scoliosis had worsened, forcing his head forward and down, and his spinal cord was vulnerable.

Sponseller recommended treating Benjamin with halo traction, using weight to straighten the spine, followed by fusion of his cervical vertebrae. The family agreed, and in 2011, after two weeks of traction in the hospital, Benjamin underwent a four-hour surgery by Sponseller and colleague George Jallo, clinical practice director of pediatric neurosurgery. Sponseller used bone from Benjamin's hip and titanium rods to stabilize his neck. "When Dr. Jallo saw Benjamin's spinal cord," Burton says, "he was amazed he was able to walk and develop as well as he had. We feel strongly that God was protecting Benjamin throughout all this."

During Benjamin's recovery, Burton says, "Dr. Sponseller was that solid ground for us. He explained things so well and was so good with Benjamin." She recalls that, while Benjamin was sleeping in the hospital, he lost a tooth. "Dr. Sponseller took out his wallet," she says. "It was made of duct tape, obviously by one of his kids, and he put money under Benjamin's pillow. He said the tooth fairy even comes at Hopkins."

The surgery was a success. Today, Benjamin is in the seventh grade. "He plays soccer," his mother says. "He doesn't like to sit still. He's doing great." Sponseller believes that Benjamin's family is a big part of his excellent result. "His whole extended and caring family was amazingly supportive in helping him through the recovery," he says.

Inspired by her gratitude to Sponseller and the team at Hopkins, Burton and her family's philanthropy, the Lundy Family Foundation, support a Johns Hopkins orthopaedic outreach program, which sends orthopaedic surgery residents to volunteer in underserved communities in the U.S. and abroad. "I have an interest in mission and have been to Uganda through my church several times," says Burton. "Dr. Sponseller talked about how they send surgeons all over the world to perform surgeries that kids would never be able to have otherwise. That lines up exactly with what our family thinks is so important. That was a no-brainer for us." ■


Benjamin Burton

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A Clearer Path to Axon Regeneration
for Brain and Spinal Cord Injuries

Research at The Johns Hopkins University School of Medicine may ultimately change the course of treatment for brain and spinal cord injuries. **Feng-Quan Zhou**, orthopaedic surgery and neuroscience researcher, leads the Zhou Lab in the exploration of molecular neuroscience. His current research focuses on epigenetic regulation of neuronal morphogenesis during development and regeneration.

"We are looking for the mechanism that promotes axon regeneration in the peripheral nervous system so we can apply that knowledge to the central nervous system and enable repair of optic nerve and spinal cord injuries," Zhou explains.

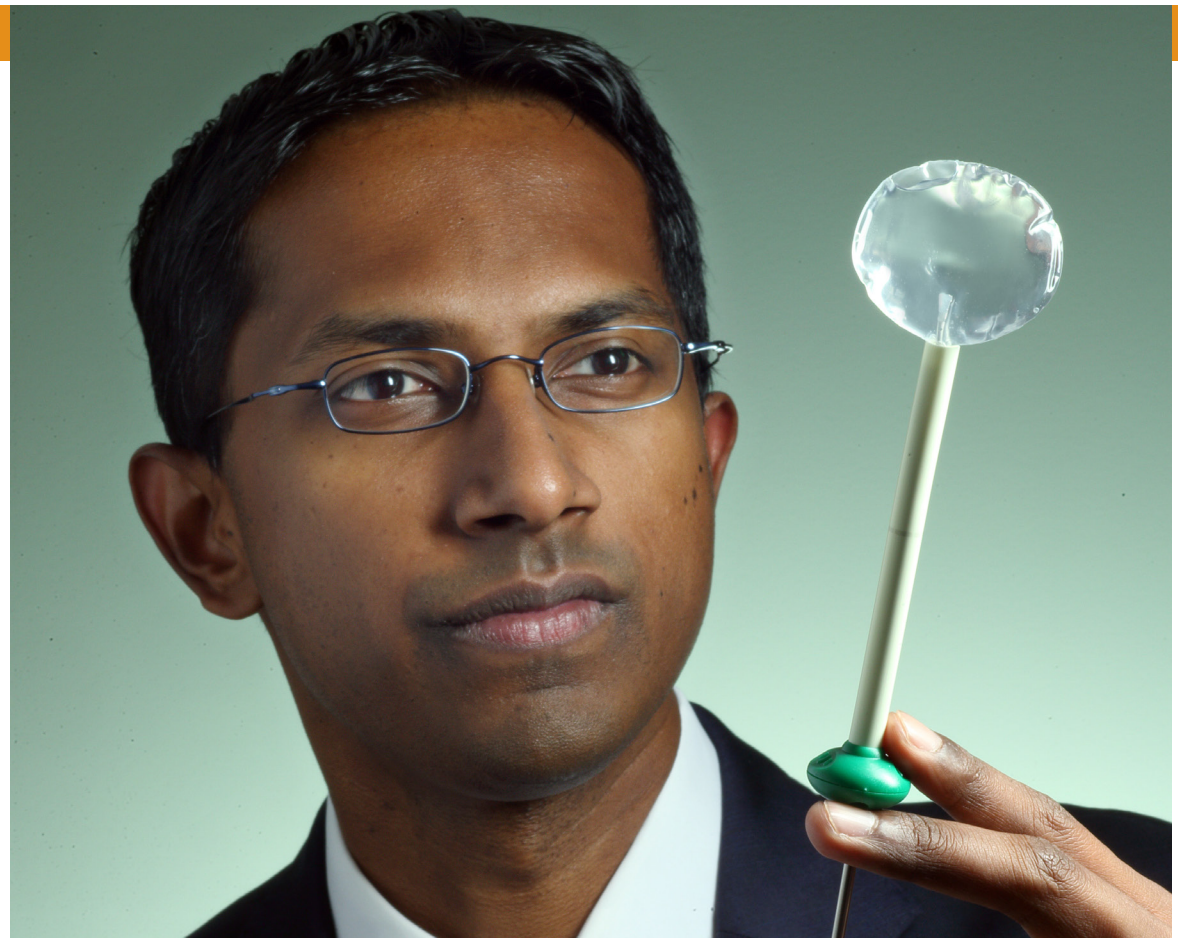
Distance is the major challenge for axon regeneration. In humans, the axon requires three to four years to regenerate because of the axon's length; axons cannot regenerate fast enough to achieve functional recovery.

By using the short optic nerves of mice, Zhou is working to bridge the distance from the optic nerve injury back to the brain and restore vision. The mouse models used in Zhou's research have also provided insight into the intrinsic and extrinsic factors of axonal regeneration.

As a neuron matures, there is an intrinsic change in the gene expression to suppress growth, which stabilizes the nervous system. The gene in a segment of DNA can be folded or loose. The loose segment of DNA allows the gene to be exposed to proteins, which bind to the gene and promote gene expression.

"We are focusing on how to get to the more

New Technologies **for Shoulder Damage**



"A lot of the research we are working on focuses on value-centered treatments. These options are faster, cheaper, easier and achieve the same or better outcomes for patients."
–Uma Srikumaran

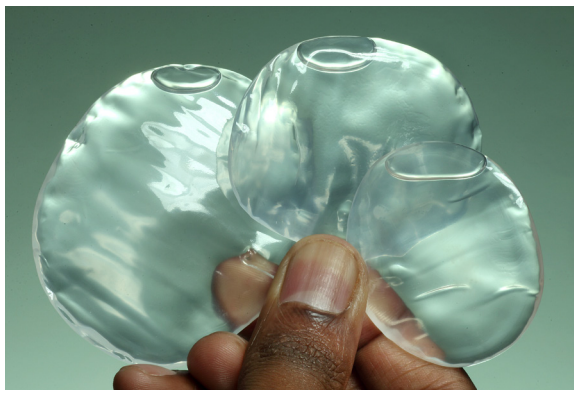
Uma Srikumaran, Johns Hopkins orthopaedic surgeon, is excited to be able to offer patients new shoulder treatments through his participation in several ongoing FDA clinical trials. He is particularly enthusiastic about helping younger patients avoid joint replacements.

One new treatment under trial is a subacromial balloon spacer for patients with massive rotator cuff tears. The balloon is inserted arthroscopically via a minimally invasive approach. The balloon unrolls, inflates with fluid and fills the gap where the rotator cuff should be but has torn and retracted away.

"It has been found to be very safe in European trials. It is simple, easy to do and will be lower cost than a joint replacement," Srikumaran says.

The subacromial balloon spacer is also an option for a subset of patients who are not strong candidates for reverse total shoulder replacement or who don't want to have a joint replacement. Srikumaran says the subacromial balloon spacer fills a gap in terms of available treatments, but he is currently able to offer it only through participation in the clinical trial.

As the value of care becomes increasingly important, Srikumaran says the balloon spacer fits the bill. "A lot of the research we are working on focuses on value-centered treatments," he says. "These options are faster, cheaper, easier and achieve the same or better outcomes for patients."



Subacromial balloon spacers are inserted arthroscopically via a minimally invasive approach. The balloon unrolls, inflates with fluid and fills the gap where the rotator cuff should be but has torn and retracted away.

Another new technology Srikumaran and his colleagues are studying is the Conventus CAGE PH for the treatment of complex proximal humerus fractures. The implant is a scaffold structure that is inserted into the bone and expands like a balloon cage, allowing the surgeon to place screws from different angles and support the fractured bone. The cage is intended to overcome some of the limitations of locking plates. Srikumaran expects it will be "a significant step forward in fracture care," especially for younger patients for whom arthroplasty is not a reasonable option.

Pyrocarbon hemiarthroplasty, a new alternative surface to metal for shoulder arthroplasty devices, may be an answer for young patients with arthritis or avascular necrosis of the humeral head. Srikumaran is part of another trial that is currently evaluating this new material, believed to have advantages for the joint socket in terms of better cartilage protection.

Having worked on an array of complex cases, including revision rotator cuff reconstructions and failed arthroplasties, Srikumaran is collaborating with his fellow clinician-scientists to build the clinical research program. He hopes to offer patients more treatment options before complicated endstage procedures are necessary. "We believe in the careful adoption of new technologies, and participating in multicenter clinical trials allows us to do just that." ■

detailed mechanism underlying this change of gene expression during the development of the neuron," says Zhou.

To manipulate gene expression, Zhou is working to silence the genes that suppress axon regeneration. This will change the pattern of gene expression so other suppressors are inhibited and still others are activated to promote axon regeneration.

Overcoming the inhibitory environment around the axon has led to the discovery that glial cells, which suppress axon regeneration, can be reprogrammed directly into neurons. Instead of harvesting glial cells, researchers can reprogram them via gene expression to expand their purpose. This process is driven by a combination of chemicals. By identifying the chemical combinations needed to change the genes, researchers can change glial cells into neurons that can regenerate.

"It is very exciting. We hope to apply this to patients with spinal cord injuries. We are still very far away, but it is possible."
–Feng-Quan Zhou

"It is very exciting," says Zhou. "We hope to apply this to patients with spinal cord injuries. We are still very far away, but it is possible."

Zhou's laboratory also works with the Johns Hopkins Wilmer Eye Institute to analyze the DNA sequencing data that are invaluable to his research. This enables researchers at Johns Hopkins to target multiple neuronal subsets and easily exchange information about breakthrough findings, accelerating the pace of discovery.

"That is the strength of Hopkins," says Zhou. "It is a huge and diverse institution so you can easily find the resources you need to build on your work." ■

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Framework

This newsletter is one of the many ways we seek to enhance our partnership with our thousands of friends and patients. Comments, questions and topics you would like to see covered in upcoming issues are always welcome.

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