

## A Rare Case of Bobble-Head Doll Syndrome



A three-year-old boy diagnosed as having Bobble-Head Doll Syndrome underwent minimally invasive endoscopic neurosurgery at MUSC, resulting in an incision less than an inch long (left).

At the age of two to three years, when most kids are entrenched in “the terrible twos,” Mickey was strangely agreeable, frequently bobbing his head back and forth in a gesture of seeming affirmation. It was only when the bobbing became constant, from the time he woke up in the morning until the time he went to sleep at night, that his parents suspected a problem and took him to see a physician.

When magnetic resonance imaging (MRI) revealed hydrocephalus (the accumulation of fluid in the brain), the local physician feared a brain tumor and recommended immediate surgery to remove it, surgery that would involve a craniotomy, the opening of the skull to allow surgical access to the brain.

Wanting a second opinion before subjecting their child to major, invasive surgery, Mickey’s parents brought him to MUSC for further evaluation. He was seen by **Sunil J. Patel, M.D.**, Chair of Neurosurgery, Department of Neurosciences at MUSC, who ordered a contrast-enhanced MRI, which ruled out a tumor but revealed an arachnoid cyst in the third ventricle, a very deep part of the brain. The cyst was benign but was preventing cerebrospinal fluid from draining appropriately, resulting in hydrocephalus.

Dr. Patel shared the unusual case with his colleagues **Rebecca K. Lehman, M.D.**, Assistant Professor of Neurosciences, and **David J. Walsh, M.D.**, Professor of Neurology, who diagnosed Mickey as having Bobble-Head Doll Syndrome.

Only a handful of cases of Bobble-Head Doll Syndrome have been reported. In his 30 years as a neurosurgeon, Dr. Patel had never seen a case until Mickey. The condition is named for its characteristic symptom, a “yes-yes” head bob, which is often associated with cystic abnormalities in the third ventricle that also produce hydrocephalus.

If left untreated, the structural changes that cause Bobble-Head Doll Syndrome can result in permanent neurologic dysfunction, which had not yet manifested in Mickey.

Fortunately for Mickey, MUSC specializes in minimally invasive endoscopic neurosurgery, which decreases the risk of surgical

complications and requires a much shorter recovery time. This was very good news for Mickey’s parents, who strongly preferred a less invasive approach because they, like Dr. Patel, “did not want to risk Mickey losing one drop of who he was.”

Instead of performing the traditional craniotomy to gain access to the third ventricle, Dr. Patel drained the cyst endoscopically, removing as much of the capsule as possible—a procedure that lasted 15 minutes and required an incision that was less than an inch long. Mickey was able to leave the hospital within 24 hours of the operation and to resume his normal life quickly.

Mickey will be followed up closely, with periodic brain imaging as he grows, to ensure that the cyst does not come back and that his mild residual head bobbing resolves, according to MUSC pediatric neurosurgeon **Edward J. Kosnik, M.D.**

Although Mickey’s parents are grateful for the depth of diagnostic expertise and the minimally invasive treatment options that they found at MUSC, they are perhaps most grateful for the warmth and compassion of the physicians and staff: “The people at MUSC were so great. They really did look after him. He wasn’t another number; they treated him like he was a human being.”

Dr. Patel is well aware that “when you have a sick child, you also have very, very anxious parents” and is pleased to be able to offer his patients the services of a dedicated pediatric hospital. In addition to specialized pediatric nursing care and a child-friendly infrastructure, the Children’s Hospital of South Carolina at MUSC provides parental support and easy access to social workers. In short, it is a place where “everything is dedicated to the care of the child and the well-being of the family.”

Mickey today is a happy, healthy, outgoing 3-year-old boy who enjoys being able to play with his siblings and jump on his trampoline every day, realizing his parents’ dream that not even one drop of his personality be lost.