Advances in Hydrocephalus Research

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On behalf of the children and families we are privileged to serve, we express our deep appreciation for One Small Voice Foundation's longstanding support of Ann & Robert H. Lurie Children's Hospital of Chicago. For more than a decade, your philanthropic contributions have helped Lurie Children's conduct cutting edge research to improve the scientific understanding of hydrocephalus with the goal of improving children's outcomes. We are pleased to update you on recent research underway in this field.

THE IMPACT OF HYDROCEPHALUS

Hydrocephalus is a congenital condition that affects a baby's brain. In particular, it leads to the build up for cerebrospinal fluid (CSF) in the ventricular system of the brain. The resulting lack of absorption, blockage of flow or overproduction of CSF may increase pressure in the head and cause the skull bones to expand.

Hydrocephalus can be detected by the routine ultrasound examination done by many obstetricians as part of women's prenatal care. If it is detected on ultrasound, the mother may undergo a fetal brain MRI (magnetic resonance imaging) to determine the severity of the condition and make a plan for treatment.

Ann & Robert H. Lurie
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Treatment of hydrocephalus depends on the type and severity of the condition. It can range from medical management to procedures that draw out the extra CSF. One type of surgery involves placing a shunt, or tube, into the child's head to drain the CSF and redirect the additional fluid to another part of the body to be absorbed. The other type of surgery that may be performed is called endoscopic third ventriculostomy (ETV). In this procedure, the neurosurgeon creates a small hole in the bottom of one of the ventricles (or spaces in the brain) causing the CSF to bypass the obstruction and flow into the natural pathways.

Hydrocephalus can affect the brain and a child's development to varying degrees. The long-term outlook for a child born with hydrocephalus depends greatly on the severity of the problem and the presence of other associated abnormalities.

TREATING HYDROCEPHALUS IN UTERO

For over 40 years, Lurie Children's has provided fetal health consultations and expert medical and surgical care to fragile babies. During that time, the field has grown tremendously. Treatments that were once unimaginable have become a reality, providing new options for families whose babies have severe congenital defects.

In July 2017, Lurie Children's welcomed Aimen Shaaban, MD, one of the most highly respected fetal surgeons in the world, to lead The Chicago Institute for Fetal Health. His leadership and the growth of The Chicago Institute will help position Lurie Children's as a national and international leader in providing complex maternal, fetal and newborn care. The Chicago Institute has a multidisciplinary, multi-institutional mandate to provide a complete spectrum of care for the fetus and mother ranging from prevention of disease to in utero fetal surgery.

Dr. Shaaban's clinical research contributions surround the diagnosis and treatment of congenital diseases that affect the fetus' spine, lungs, and gastrointestinal tract, among others. Among his research interests is exploring new approaches to treating hydrocephalus. In fact, hydrocephalus was one of the first procedures approached via fetal surgery. In utero treatment of hydrocephalus was attempted decades ago, but did not achieve improved outcomes and work in this area stopped. Given the dramatic advances in medicine and technology since then, there is now a renewed interest in the field of fetal medicine to address this condition while the baby is still in the womb.

Dr. Shaaban is part of a study group looking at various options on how to improve fetal surgery for hydrocephalus and reduce the impact on the baby's brain development. Areas of investigation include:

- Identifying noninvasive ways to decompress the brain in utero
- Exploring ways to insert shunts in utero at 25 weeks gestation, as they are done on preemies born at 25 weeks
- Developing new types of fetal shunts and new methods to insert shunts
- Conducting studies to determine which patients are the best candidates for surgery and following patients after surgery to assess their long-term medical outcomes
- Discovering the effects of hydrocephalus on the brain and if the impact of the disease can be reversed

A particular interest of Dr. Shaaban's is the development of high intensity focused ultrasound (HIFU) to treat hydrocephalus in utero. HIFU is an early stage medical technology that is being explored to treat a range of disorders. The mechanism is similar to using a magnifying glass to focus sunlight. Focused ultrasound concentrates multiple intersecting beams of ultrasound on a target. Each individual beam passes through tissue with little effect but at the focal point where the beams converge, the energy can have useful thermal or mechanical effects. HIFU is typically performed with real-time imaging via ultrasound or MRI so the clinicians can carefully target and monitor the beam.

Using HIFU to treat fetal hydrocephalus would provide a noninvasive method for fetal surgeons to create a small hole to relieve pressure on the brain without damaging any other tissue in the fetus or the mother. This intervention would be similar to the ETV procedure that is used in some hydrocephalus cases after the baby is born. Dr. Shaaban is currently involved in research using HIFU in an animal model. This exciting work represents the greatest promise for therapy and could be in clinical applications in the next several years.

UNDERSTANDING THE UNDERLYING CAUSES OF HYDROCEPHALUS

Lurie Children's Division of Neurosurgery recently concluded a study that aimed to better understand molecular mechanisms to develop an improved and alternative treatment for hydrocephalus. The water transport process regulated Aquaporin channels have been implicated in the pathogenesis of hydrocephalus. By targeting Aquaporin water channel (AQP-4) in ventricular ependymal cells, the team investigated the flux of CSF out of cerebral ventricle in order to reduce ventricle enlargement, a key mechanism to resolve hydrocephalic disorder. The team recently submitted a paper detailing the findings of this study that reveal a promising way to manage hydrocephalus without shunt hardware. It requires a revision before publication, which is currently underway.

THANK YOU!

We are honored that the One Small Voice Foundation supports Lurie Children's efforts to improve treatment options and quality of life for children with hydrocephalus. We are grateful for your dedication to the patients and families we serve and for your belief in the incredible work done by our clinicians and scientists.