Neurosurgery Working Group:

**Introduction and Background: Neurosurgical Issues in a Prioritized Research Agenda**

On May 9-10, 2003 the US Department of Health and Human Services in collaboration with US Department of Education, Spina Bifida Association of America, the Spina Bifida Foundation and the Agency for Healthcare Research and Quality convened a meeting in Washington D.C. entitled Evidence-Based Practice in Spina Bifida: Developing a Research Agenda. The purpose of the meeting was to determine a prioritized research agenda in Spina Bifida that centered upon 15 separate domains of care. Neurosurgery was one of these domains and a general summary of current information was provided based upon the key issues of hydrocephalus, the Chiari malformation, tethered spinal cord, tethering in adults, syringomyelia, neurosurgical causes of scoliosis and the reliability of tests for neurologic function.

Specific Recommendations for further investigation centered upon 5 topics:

1. Assessment and treatment of hydrocephalus in patients with SB.
2. The role of tethered cord release in the long-term management of patients with SB.
3. The long term neurological priorities, challenges, complications and treatments for adults with SB
4. Identification of neurologic deterioration from HCP, shunt malfunction, TCS, Chiari and syringomyelia in SB.
5. The role of neurosurgical causes of scoliosis in people with SB.

In the intervening 14 years there have been over 600 papers published on Neurosurgical topics in Spina Bifida. The Centers for Disease Control has sponsored and coordinated a National Spina Bifida Patient Registry. A variety of North American Neurosurgery meetings in which Neurosurgical issues related to Spina Bifida were considered have occurred annually including the American Society of Pediatric Neurosurgery, the Pediatric Section of the AANS/CNS, the International Society of Pediatric Neurosurgery and the Society for Research in Hydrocephalus and Spina Bifida (SRHSB). In addition, there have been a number of regional and invitational meeting within the Pediatric Neurosurgery community to address issues related to care for patients with SB.

Several major developments were unforeseen and have significantly impacted the Neurosurgical issues in children with SB. The first of these was the 2006 publication of MOMs Trial in the New England Journal of Medicine. This prospective randomized, multi-center trial demonstrated significant improvements in spinal level, need for shunting and decreased incidence of a symptomatic Chiari malformation in a cohort of infants with SB who were treated with in utero MMC closure (IUMMC). These findings were exciting developments and remain amongst the most actively discussed issues surrounding Neurosurgical care of infants with SB. Some controversies and many unknowns exist surrounding this important intervention and IUMMC will remain at the forefront of Pediatric Neurosurgical discussions related to SB for the foreseeable future.
The second major development was the development of Endoscopic Third Ventriculostomy with Choroid Plexus Coagulation (ETV-CPC) which was developed and popularized by Benjamin Warf in Africa. In seminal publications in 2004-2008 Warf demonstrated that ETV-CPC was effective in treating children with HCP from a variety of different causes without a shunt. A follow up publication by Stone and Warf documented similar success in SB related HCP in a North American cohort. These studies prompted consideration for utilization of this technique as a preferred means of treating hydrocephalus. The Hydrocephalus Research Network (HCRN) is currently conducting a randomized trial evaluating the effectiveness of this mode of treatment for hydrocephalus from a variety of different etiologies.

Another controversial development since the publication of the “Green Book” was the development of the “Xiao Procedure”. This procedure was developed and popularized by Dr. Xiao in China and reportedly resulted in improved bladder control for patients with Spina Bifida by surgically re-connecting dorsal motor roots to ventral ones in the sacral region. This reportedly with time resulted in trainable reflexes that would initiate volitional voiding with cutaneous stimulation of the leg, flank or abdomen. Despite initial enthusiasm it failed to show consistent performance and was ultimately shown of no benefit in a controlled North American trial.

In response to suggestions from the SBA-Professional Advisory Committee the CDC sponsored and developed the National Spina Bifida Patient Registry (NSBPR) which is a large patient registry for patients with SB. This prospective multi-center data base has gone through three consecutive versions which have progressively, sequentially increased the amount of data present. Over 15 centers and 6000 patients have been entered and there is sufficient information that early data analysis proposals are being considered by the CDC for papers studying basic questions pertaining to epidemiology, basic treatment and patient profile in Spina Bifida in North America.

Finally, there has been persistent though inconsistent Neurosurgical interest in transition for patients with Spina Bifida. A limited number of small cohort studies have addressed this issue and it continues to receive attention at meetings and conferences but little systematic study nor prioritization.

Against this background the Neurosurgical Working Group for the SBA Guidelines Initiative provides the following outline of issues and questions to drive the next iteration of guidelines for Neurosurgical Care of the patient with Spina Bifida in 2017.