THE UVMMC SPINA BIFIDA COLLABORATIVE CARE PROGRAM

Working Together Conference, August 23, 2016
Allison Fortuna, RN
Ida McNamara, RN

OBJECTIVES

• Review the anatomy and physiology associated with a diagnosis of spina bifida and the impact it has on various systems within the body

• Describe interventions, both surgical and medical that are used to treat the individual with spina bifida.

• Discuss the importance of the school nurse role with the care team.

• Review interventions to support the individual with spina bifida at school

SPINA BIFIDA

• Neural tube defect, the most common congenital central nervous system anomaly

• Characterized by a cleft in the vertebral column, with a corresponding defect in the skin, so that the meninges and spinal cord are exposed

• Failure of the neural tube to close prior to the reopening of the neurocele allows the cranial vesicles to collapse, is the mechanism for the development of the Chiari II malformation

• Prenatal diagnosis is accomplished by maternal screening of serum alpha fetoprotein (AFP) levels and the diagnosis is established by ultrasound
Myelomeningocele in a newborn infant

MRI showing acquired hydrocephalus

SPINA BIFIDA

- Incidence varies substantially across populations in patterns that follow nutritional geographic and ethnic factors
- Risk factors include: folic acid deficiency, family history, pregestational maternal diabetes, prenatal exposure to anticonvulsants such as valproic acid or carbamazepine, lower vitamin B12 levels during pregnancy, maternal obesity, maternal hyperthermia, maternal diarrhea, gestational diabetes
SPINA BIFIDA

• Prenatal counseling: discussion with the parents includes the natural history of myelomeningocele and the prenatal management decisions
• Discussion also includes postnatal management choices including surgical closure of the defect and possible need for ventriculoperitoneal shunt placement along with evaluations of the kidneys, bladder, bowel and treatment of any orthopedic abnormalities

SPINA BIFIDA PRENATAL SURGERY

• Can arrest leakage of spinal fluid from the back and might therefore prevent or reverse herniation of the hindbrain (Chiari II malformation) and hydrocephalus
• Is only performed at specialized centers that have the special expertise, multidisciplinary teams, and facilities to provide the intensive care required
• The Management of Myelomeningocele Study (MOMS) was a randomized controlled trial that evaluated the safety and efficacy of fetal surgery

SPINA BIFIDA

• Once born there is a very thorough assessment of the lesion
• Assessment of the level of the lesion will help in determining the potential for mobility
• Evaluation also includes associated abnormalities of other systems, particularly bladder and bowel
ONGOING MANAGEMENT

- Neurological complications: shunt malfunction, tethered cord
- Bladder management
- Bowel management
- Orthopedic concerns

NEUROLOGICAL MONITORING

- Shunt malfunction
- Tethered cord
- Chiari II malformation
- Seizures
- Cognitive development

MRI of tethered cord and lipoma
BLADDER MANAGEMENT
• Neurogenic bladder
• Clean intermittent catheterization
• Urinary Tract Infections
• Surgical considerations

BOWEL MANAGEMENT
• Innervation of the bowel and anus are affected, leading to dysmotility and poor sphincter control
• Goals of bowel management are to achieve continence by prompting regular elimination of stool and to avoid fecal impaction

ORTHOPEDIC CONCERNS
• Main goals are to maximize function and mobility and minimize deformity and pain
• Abnormalities are caused by unbalanced muscle action around joints, paralysis, and decreased sensation in lower extremities
• Increased risk for wound infection, pathologic fractures, nonunion after orthopedic surgery, as well as latex allergy and skin breakdown
• Rapid development of a problem may be caused by an acute neurologic pathology
Spina Bifida Collaborative Care Program (SBCCP)

- UVM Children’s Hospital Spina Bifida Collaborative Care Program began September 2015
- Includes:
  - Pediatric Nephrology
  - Pediatric Neurosurgery
  - Pediatric Urology
  - Pediatric Physiatry
  - Developmental Pediatrics
  - Certified Prosthetist & Orthotist
  - Social Work
  - Parent/Family Advisor
  - Vermont Family Network
  - Child Life

www.sbagreaterne.org

Goals of SBCCP at UVM Children's Hospital

- Goals:
  1) Improved care coordination for patients & families
  2) Improved communication within care team
  3) Providing opportunities for families to connect and meet children with similar medical needs
  4) Encouraging families to actively participate in care plan
  5) Continue to evolve clinic with feedback from patients & families

School Nurse Role on Care Team

- Obtaining student care plan info prior to start of school
  - Awareness of baseline
  - Involved in child’s IEP & 504 Plan team
  - Determine training needs of staff
  - Emergency plan, knowing when to alert family and care team
- Awareness of associated secondary medical conditions:
  - Brain abnormalities & symptoms of shunt malfunction
  - Bowel & bladder dysfunction
  - Mobility & skin integrity challenges
  - Chronic Pain
- Knowledge of interventions to improve quality of life
- Provide support of these interventions in the school setting
- View each child as an individual
Brain Abnormalities & Their Application in the School Environment

- Brain abnormalities associated with Spina Bifida affect the child’s ability to learn and be independent
  - Disgenesis of Corpus Callosum
  - Cortical thinning of posterior brain
  - Cortical thickening in anterior brain
- Shunt malfunction symptoms to assess for:
  - Personality changes
  - Headache
  - Vomiting
  - Lethargy & confusion
  - School performance deterioration
  - Swelling/redness along shunt tract
  - Seizures

Bladder Dysfunction Interventions

- Bladder dysfunction depends on level of lesion
- Often bladder nerves damaged, child can’t feel when bladder is full nor empty bladder (neurogenic bladder)
- Interventions:
  - Clean intermittent catheterization (CIC) q2-6 hours
    - Requires extra time & safe place
  - Mitrofanoff: continent stoma, typically cath q3-4 hours through abdomen or belly button
  - Bladder augmentation: Surgical reconstruction to relieve urinary incontinence & prevent kidney damage caused by urine back-up
- Promoting urinary continence at school improves quality of life allowing children to focus on learning

Mitrofanoff Catheterization

www.mitrofanoffsupport.org.uk/mitrofanoff-procedure/
Bowel Dysfunction Interventions

- Due to nerve damage, many children with SB are unable to feel urge to have bowel movement nor control their bowel movements.
- Interventions:
  - Bowel regulation and continence can be accomplished with scheduled bowel regime.
  - Antegrade continence enema (ACE) is a common procedure, child will do an enema through continent stoma to empty bowels, usually at night.
- In case of an accident, children should always have a change of clothes available.
  - Handle discretely and sensitively as accidents are out of child’s control.

Antegrade Continence Enema (ACE)

Mobility & Ambulation Challenges and Interventions

- The higher the lesion, the greater level of paralysis
  - S1-S5
  - L1-L5
  - T2-T12
- Associated orthopedic issues include: scoliosis, kyphosis & tethered spinal cord
- Interventions:
  - Be aware of supplies needed for mobility & level of assistance needed to operate.
  - Careful evaluation of skin breakdown for children w/ braces & in wheelchairs.
  - Circulation exercises to prevent pressure ulcers & infection.
  - Educate importance of safety in children with loss of sensation.
- Importance of inclusion in activities to promote development and self-confidence.
Importance of Active Participation in Play

- Studies have found children with disabilities value being “where the action is”
  - But have lower rates of active participation specifically in recess & playground activities
- “This pattern of participation might therefore reflect the cognitive functional profile of children with SB...that they are successful in everything that is guided but have more problems acting on their own initiative.”
  (Peny-Dahlstrand et al., 2013)

Latex Allergy

- Individuals with SB are at high risk for latex allergy
  - Current theory
  - Most practice latex precautions to prevent allergy development
- Symptoms of allergic reaction to latex:
  - Itching, hives, rash, skin redness, sneezing, runny nose, itchy watery eyes, horse throat, cough, wheezing, shortness of breath
  - Anaphylactic response
- Students sensitive or allergic to latex may also be allergic to bananas, avocados, chestnuts, kiwi & papaya
- How to Prevent Reactions in the School Environment
  - Educate staff, survey school environment, remove and replace
Living with Spina Bifida Video

- The Spina Bifida Experience-Centers for Disease Control & Prevention

REFERENCES

- Orthopedic issues in myelomeningocele (spina bifida).  UpToDate 2016
- Overview of the management of myelomeningocele (spina bifida).  UpToDate 2016
- Pathophysiology and clinical manifestations of myelomeningocele (spina bifida).  UpToDate 2016