



**The High Risk Infant
Central Nervous System Dysfunction**

A Training Curriculum

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**For the
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Training Program**

**University of Pittsburgh
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316 & 936: The High Risk Infant: Central Nervous System Dysfunction

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An Overview of the Curriculum

Rationale

As Foster Parents, Adoptive Parents, and Child Welfare Professionals continue to care for medically complex children, a better understanding of various disease entities becomes increasingly significant. Caregivers need to be educated on conditions related to alterations in the central nervous system in order to better care for their medically complex child. Caregivers must also keep abreast on the current management and home care techniques which are available for their medically complex child.

Learning Objectives

- Define conditions leading to a disturbance in the central nervous system.
- Identify those infants who are at greatest risk for developing neurological problems.
- Understand the physiology of various neurological conditions.
- List appropriate interventions for the care of infants with a disturbance in the central nervous system.
- Understand the home care involved with infants who have a disturbance in the central nervous system.
- List current management for the care of infants who are experiencing central nervous system dysfunction.

Competency to be Addressed in Curriculum

316-1. The Child Welfare Professional knows health and medical conditions which can affect the well being of children and families or which can contribute to or result from child abuse/neglect.

Workshop Training Time

3 hours

Target Audiences

Child Welfare Professionals, Family Preservation Workers, Foster Caregivers, and Adoptive Parents.

Expectations of Trainer

This curriculum has been developed to be delivered by a trainer knowledgeable in the medical, developmental, and psychosocial needs of the high risk infant have an awareness of community resources available to meet the needs of this diagnostic group, and have experience with disease management in the home setting. The trainer should have a strong medical background and have a minimum of a Bachelor's Degree. The trainer should have some knowledge in child welfare practice, specifically in direct services to children and families. The trainer should have considerable experience in training workshops and should possess excellent group facilitation skills. The trainer must have knowledge and experience in diversity awareness so that special attention can be afforded to the provision of culturally congruent healthcare.

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An Overview of the Curriculum (continued)

Equipment Needed

Specific materials needed to conduct the training are listed for each section of the curriculum. All sections require overhead projector and flip chart.

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The High Risk Infant Central Nervous System Dysfunction Content & Schedule of Activities for Three Hour Curriculum

0.5 Hours	INTRODUCTION AND OPENING ACTIVITIES
Section I 1.0 Hour	SPINA BIFIDA/MYELOMENINGOCELE Pathophysiology Clinical Manifestations Therapeutic Management Family Teaching Home Care
Section II 1.0 Hour	HYDROCEPHALUS Pathophysiology Clinical Manifestations Therapeutic Management Family Teaching Home Care
Section III 0.5 Hours	EVALUATION AND CLOSURE Transfer of Learning Activity

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Dear Trainer –

This curriculum is an extension of “The Infant at Risk” segment of “Meeting the Needs of Medically Complex Children”. You may want to review that curriculum, as well as applicable section of the “Accessing Services Directory” prior to teaching this segment on Central Nervous System Dysfunction.

Caring for a child experiencing Central Nervous System Dysfunction can be a sometimes frightening and overwhelming experience for those involved. It is our hope that the education information provided in this curriculum will offer some assistance to those caregivers.

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Preface

This curriculum was designed as an introductory segment on Central Nervous System Dysfunction and provision of care to infants with this diagnosis. It must be understood that as research and medicine evolve, so do the treatments and educational materials, with the advancement of technology. Workers, parents, and trainers should continue to remain updated on new information to assist in managing Central Nervous System Dysfunction.

The trainer should explain to the audience that he/she will be purposely using both medical and non-medical terminology in order to familiarize participants with verbiage they may see on the child's medical record or hear during healthcare provider appointments.

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Incorporating Transfer of Learning Into This Curriculum

In order for this training to be deemed effective, there must be a mechanism present to promote transfer of learning from the training session atmosphere to the care environment surrounding the child with Central Nervous System Dysfunction.

You will find a “What Have I Learned?” activity at the conclusion of this curriculum to promote this transfer of learning. Take the time to review learning objectives before training begins to facilitated mutual expectations of outcomes.

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INTRODUCTION AND OPENING ACTIVITIES

Rationale: Participant interaction and group trust promote a positive environment for learning.

Learning Objectives: Participants will be able to:
Introduce themselves to other participants.
Identify their training needs.
Describe the course and competency.

Time: 0.5 Hours

Methods: Presentation by trainer. Group discussion.

Materials: Name tents (large index cards or heavy stock paper), markers, 3x5 index cards
Overhead #1 – Agenda, Learning Objectives, and Competency

Activities: Activity #1 – Name tents

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Trainer Notes

INTRODUCTION AND OPENING ACTIVITIES

Trainer Note: Name tent materials and 3x5 cards may be given to participants when they arrive or placed on the participants' tables prior to the start of training.

- Introduction of the trainer and welcome participants to the training.
- Review specifics of Competency-Based Training:
 - 15-minute rule
 - Sign-in sheet
 - Evaluation form completion and submission
 - Availability of continuing education units
 - Review "housekeeping rules" pertaining to specific training site.

Trainer Note: Use Overhead #1 here. "Agenda, Learning Objectives, and Competency"

- Review agenda, learning objectives, and competency.

Activity #1 – Name Tents and Question Cards

Step 1 – Instruct participants to write their name in the center of their paper name tent with a marker. They should also note if they are caseworker, foster parent, or adoptive parent.

Step 2 – Trainer should go around the room and have each participant give their name and why they are attending the training session (i.e. foster parent for child with central nervous system dysfunction, caseworker for child with central nervous system dysfunction, etc.)

Step 3 – Trainer should explain that 3x5 index cards are for writing questions participants may have. These questions will be addressed throughout the training as time permits.

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SPINA BIFIDA / MYELOMENINGOCELE

Rationale: Child welfare professionals, family preservation staff, foster care providers, and adoptive parents must have better understanding of Spina Bifida/Myelodysplasia if they are to be participants in the care of an infant with this disorder.

Learning Objectives: Participants will be able to:
Describe Spina Bifida and Myelomeningocele and be aware of care interventions appropriate for a child with this complication.

Time: 1.0 Hour

Methods: The trainer presents the content in lecturette form using overhead transparencies as reinforcement.

Materials:
Overhead #2 – Spinal Cord Segments
Overhead #3 – Midline Defects
Handout #1 – Latex Allergy in Children with SB
Handout #2 – Folic Acid for the Prevention of Neural Tube Defects
Handout #3 – Case Study #1
Handout #4 – Case Study #2
Handout #5 – Case Study #3
Handout #6 – Case Study #4

Activities: Activity #1 – Small group activity of case study scenarios

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SPINA BIFIDA / MYELODYSPLASIA

Myelodysplasia refers broadly to any malformations of the spinal canal and cord. Midline defects involving failure of the bony spine to close are called Spina Bifida (SB), the most common defect of the central nervous system. SB is categorized into two types: Spina Bifida Occulta and Spina Bifida Cystica.

Trainer Note: Use Overhead #2 here. “Spinal Cord Segments”

Spina Bifida Occulta refers to a defect that is not visible externally. It occurs most commonly over the lumbar or sacral area such as L-5 and S-1. Routine x-ray exams indicate that the disorder may be in as many as 10% to 30% of the general population, however, it may not be apparent unless there is an associated skin, cutaneous manifestation, or neuromuscular disturbance. Because of this, caregivers should be alert to any lower spine changes. If a child has an attached spinal cord, the child may seem perfectly normal as an infant. The only sign may be hair or pigmentation over the site of attachment. As the child grows, the cord will pull and stretch and curvature of spine and loss of sensation and paralysis will occur unless surgically repaired. Once nerve damage occurs, it can't be repaired.

Superficial skin indications include either a skin depression or dimple, which may also mark the outlet of a sinus tract that extends to the subarachnoid space (part of the spine); a port wine stain; dark tufts of hair; and a soft, subcutaneous lymphoma or fatty tissue. These signs may be absent, appear as a single entity, or be present in combination.

If the Spina Bifida Occulta is associated with neurological involvement, it is known as occult spinal dysraphism. Here, the neurological involvement consists of progressive or static changes in the gait with foot weakness, foot deformity, problems with the bowel and bladder. Manifestations may not be evident until the child walks or begins to become toilet trained.

Trainer Note: Use Overhead #3 here. “Midline Defects”

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Spina Bifida Cystica refers to actually a visible defect with an external sac-like protrusion. The two major forms of Spina Bifida Cystica are:

Meningocele – Encases meninges (part of the brain) and the spinal fluid but no neural (nerve) elements. Meningocele is not associated with neurologic defects which do occur in varying, often serious degrees in Myelomeningocele.

Myelomeningocele – Clinically the term Spina Bifida is used to refer to Myelomeningocele. Contains meninges, spinal fluid, and nerves. Develops during the first 28 days of pregnancy when the neural tube fails to close and fuse at some point along its length. It is detected at birth, accounts for 90% of spinal cord lesions, and may be located at any point along the spinal column. Usually, the sac is encased in a fine membrane that is prone to tears through which cerebral spinal fluid can leak. In other instances the sac may be covered by dura, which is part of the brain lining, meninges, or skin, in which instances there is rapid and spontaneous growth of tissue.

The largest number of myelomeningoceles are found at the lumbar or lumbosacral area. The location and magnitude of the defect determine the nature and extent of neurological impairment. When the defect is located below the second lumbar vertebrae, the nerves are involved giving rise to certain symptoms such as flaccidness, areflexic, or partial paralysis of the lower extremities, and varying degrees of sensory deficit.

The anomaly most frequently associated with myelomeningocele is hydrocephalus. 90-95% of children with SB have hydrocephalus. Although present at birth, hydrocephalus may not be apparent until shortly thereafter. Careful monitoring of the head circumference, fontanel tension, and ventricle size by head ultrasound can indicate its presence. Hydrocephalus can occur because the neural tube defect itself disrupts the flow of the cerebral spinal fluid.

***Pathophysiology**

The pathophysiology of SB is best understood when it is related to the normal formative stages of the nervous system.

The primary defect in neural tube malformations is believed by most authorities to be a failure of neural tube closure. However, there is also evidence to indicate that the defects are a result of splitting of the already closed neural tube as a result of abnormal

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increase in cerebral spinal fluid during the first trimester. The incidence of neural tub malformations in the United States is about 1 in 1,000. The world's highest rate of Spina Bifida occurs in the Irish. In the United States, the rate is highest in Appalacia and Mexican immigrants. African Americans have a decreased risk for neural tube malformations. Incidence rates vary by season with the greatest risk occurring when conception takes place in a month associated with high incidence of viral infections. The incidence decreases in all ethnic groups as the socioeconomic status improves.

***Clinical Manifestations**

The manifestations of SB vary widely according to the degree of spinal defect. The defect is readily apparent on inspection. The degree of neurological dysfunction is directly related to the anatomical level of defect and, thus, the nerves involved.

Symptoms include both upper and lower motor impairment, which can be determined by observing the infant's response to either a pin prick over their legs or trunk. Defective nerve supply to bladder and bowel may cause problems with sphincter control. Sometimes the damage to the muscles of the lower extremities will also produce joint deformities while the infant is still in utero. These are primarily problems with extension contractures, club feet, kyphosis (curving of the spine), scoliosis, as well as hip dislocations.

Diagnostic Evaluation

The diagnosis is made on the basis of clinical manifestations and on examination of the meningele sac. Diagnostic measures to evaluate the brain and spinal cord include MRI, ultrasound, CT scan. Laboratory examinations are used primarily to determine the causative organisms and the major complications of the myelomingeles which include meningitis and urinary tract infections.

Trainer Note: Discussion of prenatal detection may be left out unless questioned by participants.

Prenatal Detection

It is possible to determine the presence of some major prenatal

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open neural tube defects. Ultrasound scanning of the uterus and elevated concentrations of what's known as alpha-fetal protein (AFP), a fetal specific gamma-1 globulin, in the amniotic fluid may indicate the presence of an anencephaly or myelomeningocele. There are no prenatal correction procedures for SB nor can the degree of damage be predicted. Currently, involved families are faced with the ethical dilemma of ending the pregnancy early or anticipating the birth of a medically complex child.

***Therapeutic Management**

Management of the child who has Spina Bifida/ Myelomeningocele requires a multi-disciplinary approach involving neurologists, neurosurgeons, pediatricians, urologists, orthopedists, physical therapists, occupational therapists, social workers, caseworkers, and caregivers.

The care planning efforts of this team is focused primarily on:

1. The defect itself, as well as problems associated with the defect such as hydrocephalus, paralysis, orthopedic difficulties, and urinary/genital abnormalities.
2. Possible acquired problems such as meningitis, hypoxia, and hemorrhage.
3. Other system abnormalities such as heart or gastrointestinal.

Initial care of the newborn involves prevention of infection, neurologic assessment including observation for associated anomalies and the emotional impact of this abnormality on the caregivers/family.

Most authorities believe that early closure, within the first 24-72 hours after birth, offers the most favorable outcome; especially with regard to morbidity and mortality from serious infection. Early closure, preferably within the first 12 hours, not only prevents local infection and trauma to the exposed tissue, but it also avoids stretching of other nerve roots which may occur as the meningeole sac expands during the first 24 hours after birth. This prevents further motor impairment.

One of the main considerations needs to be that of orthopedic problems. According to most orthopedic specialists,

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musculoskeletal problems that will effect later locomotion should be evaluated early and treatment should be instituted without delay. Neurological assessment will determine the level of the lesion, recognition of spasticity and progressive paralysis, the potential for deformity and functional expectations for the child.

Orthopedic management includes prevention of contractures, correction of the deformity, prevention of skin breakdown, and obtaining the best possible locomotor function.

Physical therapy and orthopedic management of children with myelomeningocele is considered to be a continuous process to achieve the maximum level of function. A variety of devices are available to provide mobility to children with spinal cord lesions, including lightweight braces, custom built wheelchairs, individually designed walkers.

Corrective procedures/surgery when appropriate are best done at an early age so that the child will not lag significantly behind classmates in developmental progress.

Management of Genitourinary Function

Myelomeningocele is one of the most common causes of neurological bladder dysfunction among children. In infants, the goal of treatment is to preserve renal function. In older children, the goal is to achieve urinary continence. Urinary incontinence, a chronic often debilitating problem, commonly arises from the dysfunctional bladder. In addition, neuropathic bladder dysfunction may cause the child to be prone to infection or other urinary problems.

The characteristics of bladder dysfunction in children vary according to the level of the lesion and the influence of bony growth and development of the spine. During infancy, urinary incontinence is physiological and normal. Ongoing urological monitoring and rapid management of complications are essential for adequate growth and maturation. As the infant grows, other methods will need to be investigated to achieve continence. Examples may be surgical procedures, catheterizations, medications, etc.

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Bowel Control

Some degree of bowel continence can usually be achieved in most children with a myelomeningocele with diet modification, regular toilet habits, and prevention of constipation and impaction. It is frequently a lengthy process. Fiber supplements, laxatives, suppositories and/or enemas aid in producing regular evacuation.

Latex Allergies

Trainer Note: Distribute Handout #1. “Latex Allergy in Children with Spina Bifida”

Latex allergy was identified as being a serious health hazard when a report linked intraoperative anaphylaxis with latex in children with SB. These children are at high risk for developing latex allergies because of repeated exposure to latex products during surgery and from numerous bladder catheterizations.

Allergic reactions range from sneezing and watering eyes to wheezing and rashes and anaphylactic shock. More severe reactions tend to occur when latex comes in contact with mucous membranes, wet skin, and the blood stream, and in individuals with atopic disorders.

It is important to note the Casein, a milk protein, is sometimes added to rubber. Therefore, individuals with a severe allergy to milk may also show signs of an apparent latex reaction.

The most important goals are prevention of latex allergies and the identification of children with a known hypersensitivity. The incidence of latex allergies in children with SB ranges from an estimated 18% to 60%. Allergy testing with latex extracts may be performed with skin prick tests, however, skin testing can cause allergic reaction. Avoiding latex products is the most important intervention. Sometimes cloth can be placed between the latex item and skin. Children with a latex allergy should carry identification, i.e. medic alert bracelet.

Trainer Note: If time permits, ask participants who have cared for a child with Spina Bifida to share experiences with the group.

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Prognosis

The prognosis for a child with a myelomeningocele depends on the neurological deficit present at birth, including motor ability and bladder innervation and the presence of associated cerebral defects. Early surgical repair of the spinal defect, antibiotic therapy to reduce the incidence of meningitis, prevention of urinary system dysfunction, and correction of hydrocephalus have significantly increased the survival rate.

Trainer Note: Distribute Handout #2. “Folic Acid for the Prevention of Neural Tube Defects”

With the widespread use of folic acid supplementation during the childbearing years, the incidence of SB should be decreased dramatically. Whether folic acid will lessen the severity of the defect in infants born with SB, despite supplementation, is still unknown.

***Family Teaching/Home Care**

As soon as the parents and caregivers are able to cope with the infant’s conditions, they are encouraged to become active participants in the care. They need to learn how to continue at home the care that has been initiated in the hospital such as positioning of the infant, feeding the infant, skin care, and range of motion exercises when appropriate. Parents are taught clean urinary catheterization technique when prescribed. The family needs to know the signs and symptoms of complications and how to reach assistance when needed.

As the child grows and develops, parents need guidance to encourage and stimulate the infant to accomplish age appropriate developmental tasks within the limits imposed by the disabilities. Referral to county Early Intervention is extremely helpful to caregivers. Upper limb movements can be stimulated early by placing the infant on the floor in what’s known as a prone position, or on their tummies, with toys within reach. Activities that encourage body consciousness, such as rolling over and pulling to a sitting position, are encouraged at appropriate times. Creeping and crawling, even in a limited way, help the child to explore the environment.

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Parents may need help to modify appliances and activities normally expected of a growing child. For example, if the infant is a paraplegic, he or she should be encouraged to use arms and shoulders as much as possible. When the infant is sitting in an infant seat, high chair, or feeding table, the hips will need to be supported and foot rests provided. Hard soled shoes will need to be worn to maintain the feet in correct alignment and to protect the insensitive feet from trauma.

It is important for the family to understand the nature of the sensory deficit in a child with a spinal defect. The child will be insensitive to pressure or other sources of tissue injury. Therefore, the family must be alert to hot or cold items that could cause thermal injury to tissue and to inspect the skin regularly for signs of pressure, especially over bony prominences.

Because of sensory impairment, the child is unaware of bladder discomfort, therefore, signs of urinary tract infections may be easily overlooked. Urinary tract infection is often considered when the child becomes ill.

Assistance with preparing the child with regard to school includes their special needs and disabilities, and will require help providing a better initial adjustment to broader social experiences.

Numerous organizations and agencies are able to offer assistance and support to children and families. The Spina Bifida Association of America provides services and support for families of children with spinal lesions.

Activity – Individual Case Studies

Trainer Note: Divide the participants into four groups. Each group will be given one of the scenarios outlined below. After approximately 10 minutes of group discussion, a spokesperson from each group will share with the other participants their thoughts and interventions for the case study.

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Case Study #1 (Handout #3)

You are a foster parent and have just received a 1-week-old infant for emergency placement. You are aware that the birth mother of this infant was a Mexican immigrant and had little or no prenatal care. As you are changing the infant's diaper, you notice a small dimple at the base of the infant's spine. Upon closer inspection, you observe several tufts of hair protruding from those dimple areas. What, if any, suspicions would you have regarding this infant? What would you do regarding those suspicions?

Trainer Note:

- Mother is in high risk group for having child with neural tube defect.
- Dimple and hair tufts may indicate Spina Bifida occulta.
- Encourage pediatric follow-up to caseworker.

Case Study #2 (Handout #4)

You are the caseworker involved with a family who has a child with Spina Bifida. The family has just relocated to this area from another state and are feeling overwhelmed with the child's care. What are some of the interventions and/or referrals that you would suggest?

Trainer Note:

- County Early Intervention
- Home Care
- Special Kids Network
- Spina Bifida Support Groups

Case Study #3 (Handout #5)

You are the foster parents of a 1-month-old child with Spina Bifida. The myelomeningocele has been surgically repaired, and the suture line is healed. The infant does not appear to be feeling well. He is feeding poorly and his skin seems warm. He does not seem to have any symptoms of a cold, such as runny nose or congestion. He is passing urine, however, the diapers do not seem quite as saturated, and the color appears a little more yellow than usual. What would you suspect? What would you do?

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Trainer Note:

- Signs of infection – warm skin, feeding poorly, not feeling well.
- Urine concentrated.
- Spina Bifida children prone to urinary infections.
- Contact pediatrician.

Case Study #4 (Handout #6)

You are the caseworker involved in the care of a child with Spina Bifida. The child has just recently started a school-aged day care program. In preparation for this activity, concentrated efforts have been made to maintain a mechanism for urinary continence. Currently, the child is being straight cathed at regular intervals. Recently, it has been reported that the child has been crying and appears very uncomfortable during straight cath procedures. The caregivers are noting rashes on the child's legs. What might you suspect? Based on that suspicion, what would your interventions be?

Trainer Note:

- Spina Bifida child has frequent exposure to latex.
- Pain during catheterization could be reaction to latex catheter.
- Rash on legs could be due to caregiver's latex gloves.
- Alert pediatrician.
- Test for allergy.
- Get latex-free equipment.
- Identification alert for child.

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HYDROCEPHALUS

Rationale: Child welfare professionals, family preservation staff, foster care providers, and adoptive parents must have better understanding of hydrocephalus if they are to be participants in the care of an infant with this disorder.

Learning Objectives: Participants will be able to:
Explain what is hydrocephalus and understand appropriate care interventions for an infant with this disorder.

Time: 1 Hour

Methods: The trainer presents the content in lecturette form using flipcharts and overhead transparencies as reinforcement.

Materials: Overhead #4 – Ventricles of the Brain
Overhead #5 – Ventriculoperitoneal Shunt
Handout #7 – Vignette #1
Handout #8 – Vignette #2
Handout #9 – Vignette #3
Handout #10 – Vignette #4

Activities: Small group activity

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HYDROCEPHALUS

Trainer Notes

Hydrocephalus means “water head” and is defined as an abnormal accumulation of cerebrospinal fluid (CSF) within the ventricles of the brain. Hydrocephalus may occur as frequently as 1 in every 500 children born. It results from a congenital or acquired condition.

Congenital hydrocephalus is usually a result of a mal-development, intrauterine infection, or Spina Bifida. Acquired hydrocephalus can be caused by infection, neoplasm, hemorrhage, or trauma.

***Pathophysiology**

To appreciate the condition, an understanding of the dynamics of cerebral spinal fluid and the relationship between various structures that make up the ventricles in the subarachnoid spaces is necessary.

Trainer Note: Use Overhead #4 here. “Ventricles of the Brain”

The two mechanisms by which cerebral spinal fluid is formed includes the secretion by the choroid plexuses and the lymphatic-like drainage by the extra cellular fluid of the brain. Cerebral spinal fluid circulates throughout the ventricular system and is then absorbed within the subarachnoid spaces by a mechanism that is not entirely clear.

The causes of hydrocephalus are varied. The results is either impaired absorption of the cerebral spinal fluid from the subarachnoid space or obstruction to the flow of the cerebral spinal fluid through the ventricular system. Rarely, a tumor of the choroid plexuses causes increased cerebral spinal secretion.

Any imbalance of secretion in absorption causes increased accumulation of cerebral spinal fluid in the ventricles, which then become dilated and compress the brain substance against surrounding rigid bony cranium. When this occurs before the fusion of the cranial sutures, it provides an enlargement of the skull, as well as a dilation of the ventricles. In children under the age of 10-12 years of age, previously closed suture lines,

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especially with the sagittal suture, may become diastatic or opened.

***Clinical Manifestations**

The two factors that influence the clinical picture in hydrocephalus is the acuity of the onset and the presence of pre-existing structural lesions.

In infancy, before closure of the cranial sutures, head enlargement is the predominant sign. Whereas in older infants and children, the lesion responsible for the hydrocephalus produce neurological signs through pressure on adjacent structures.

In infants with hydrocephalus, the head grows at an abnormal rate. The first signs may be bulging fontanel without head enlargement. The anterior fontanel is tense, often bulging, and you will be unable to palpate the pulse of the heart through the anterior fontanel. Scalp veins are dilated and markedly so when the infant cries. With the increase in the inner cranium volume, the bones of the skull will become very thin and the sutures will become palpable.

There may be frontal enlargement or bossing with depressed eyes, and the eyes may be rotated downward producing what is known as the classic “setting sun” sign in which the sclera may be visible above the pupil. Pupils are sluggish with unequal response to light.

Trainer Note: Draw example of “setting sun” eyes on the flipchart.

The infant is often very irritable and lethargic. He or she may feed poorly and may display changes in level of consciousness. They may demonstrate lower extremity spasticity. The infant often cries when picked up or rocked, and quiets only when allowed to lie still.

Early infantile reflexes persist, and normally expected responses may not appear. If hydrocephalus is allowed to progress, Development of lower brain stem function is disrupted as manifested by difficulty in sucking and feeding and often times a very shrill, brief, high-pitched cry. Eventually, the skull becomes enormous and the cortex is destroyed.

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If the hydrocephalus is rapidly progressing, the infant may display vomiting, somnolence, seizures, and cardiopulmonary distress. Severely effected infants usually do not survive the neonatal period.

The signs and symptoms in early to late childhood are caused by an increase in the inner cranial pressure. Most commonly these symptoms include headache upon awakening with improvement following emesis or vomiting or upright posture, swelling of the optic nerve, strabismus, and difficulty walking.

As with infants, the child will become irritable, lethargic, apathetic, often times confused, and often time incoherent in their speech.

Diagnostic Evaluation

In infancy, the diagnosis of hydrocephalus is based on the head circumference that crosses one or more gridlines on the measurement chart within a period of 2-4 weeks and associated neurological signs that are present and progressive. However, other diagnostic studies are needed to localize the site of the cerebral spinal obstruction. Routine daily head circumference measurements are carried out on infants with myelomeningoceles who have experienced a hemorrhage or intercranial infections.

In evaluation of a preterm infant, especially adapted head circumference charts are consulted to distinguish abnormal head growth from rapid head growth that often takes place normally. The primary diagnostic tool for detecting hydrocephalus are the Computed Tomography (CT) scan and Magnetic Resonance Imaging (MRI). Sedation is often required since the child must remain absolutely still for an accurate picture. Diagnostic evaluation in children who have symptoms of hydrocephalus after infancy is similar to that employed in those with a suspected intercranial tumor.

***Therapeutic Management**

The treatment of hydrocephalus is directed toward:

1. Relief of the hydrocephalus
2. Treatment of complications
3. Management of problems related to the effect of the disorder on psychomotor development

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The treatment is, with few exceptions, surgical. Medical therapy has been largely disappointing. Many newborn infants with progressive cranial enlargement secondary to inter cranial hemorrhage demonstrate spontaneous stabilization resolution.

Serial lumbar punctures and medications have been used with varying degrees of success. The administration of Acetazolamide and Isorbide or Lasix is somewhat beneficial in decreasing the production of cerebral spinal fluid in selected cases of slowly progressive disease. The medications reduce the inter cranial pressure until spontaneous arrest of hydrocephalus takes place or as a temporary measure when surgery is contraindicated.

Surgical Treatment

Trainer Note: Use Overhead #5 here.
“Ventriculoperitoneal shunt”

Improved techniques have established surgical treatment as the therapy of choice in most cases of hydrocephalus. This is accomplished by direct removal of the obstruction such as a tumor or, in rare instances, of fluid overproduction by electric coagulation of the choroid plexus. However, most children do require a shunt procedure that provides drainage of the cerebral spinal fluid from the brain ventricles to another compartment, usually the peritoneum.

The preferred procedure is the ventroperitoneal shunt, especially in neonates and young children. There is greater allowance for excess tubing, which minimizes the number of revisions needed as the child grows.

Since it requires repeated lengthening, the ventroatrial, or VA shunt, is reserved for older children who have attained most of their symatic growth and children with abdominal pathology. The VA shunt is contraindicated in children with cardiopulmonary disease or elevated cerebral spinal fluid protein.

Trainer Note: Ask participants who have cared for a child with an AP shunt to share experiences with the group.

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Complications

The major complications of VP shunts are infection and malfunction. All shunts are subjected to mechanical difficulty such as kinking, plugging, separation, or migration of the tubing. If malfunction occurs, the child will demonstrate signs and symptoms of increased intracranial pressure as outlined earlier. Malfunction is most of caused by mechanical obstruction either within the ventricles from particulate matter, such as tissue or exudate, or at the distal end from thrombosis or displacement as a result of growth.

The most serious complication, shunt infection, can occur at any time. The period of greatest risk is 1-2 months following placement. In the case of infection, one would see redness, swelling, tenderness at the shunt site and/or fever.

Prognosis

The prognosis of children with treated hydrocephalus depends largely on the rate at which hydrocephalus develops, the duration of raised intracranial pressure, the frequency of complications, and the cause of the hydrocephalus. For example, malignant tumors may have a high morbidity regardless of other complicating factors. Untreated hydrocephalus has a 60% mortality rate caused by the disorder or intercurrent illnesses.

Surgically treated hydrocephalus with continued neurological and medical management has a survival rate of about 80% with the highest incidence of mortality occurring within the first year of treatment.

Hydrocephalus in association with myelomeningocele has a less favorable prognosis. In some children, irreversible damage may have been produced by the hydrocephalus or from the original infection.

***Family Support/Home Care**

The caregivers should have a basic understanding of cerebrospinal fluid (CSF) and their child's reason for hydrocephalus. They need to understand the functioning of their child's shunt. Although caregivers do not usually need to measure the child's head circumference every day, they should be shown to check fontanelles. (This should be done when infant is upright and quiet.) Because a child with hydrocephalus is at

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for increased risk seizures, caregivers must be taught how to recognize seizure activity and how to care for an infant/child during a seizure. Because children with hydrocephalus are prone to visual and hearing problems, regular screenings are especially important.

Caregivers must be instructed on how to recognize signs that indicate shunt malfunction or infection. Active children may have injuries such as a fall that can damage the shunt and the tubing may pull out of the distal insertion site or become disconnected during normal growth. The use of helmets is often suggested when the older child engages in activity which may result in falls.

The management of hydrocephalus in a child is a demanding task for both family and professionals, and helping the caregivers cope with a child is an important part of caregiving. The National Hydrocephalus Foundation provides information on the condition for families and assists interested groups in establishing local organizations.

Small Group Activity

Trainer Note: Divide participants into four groups. Each group will be given a vignette to discuss and share with other participants.

Vignette #1 (Handout #7)

You are a foster parent caring for a 4-year-old child with hydrocephalus. The child is very eager to play with and mimic the activities of his foster brother. The children are particularly fond of tricycle riding. What interventions/guidance can you provide to assist this child in being involved with normal play activity?

Trainer Note:

- Child should be encouraged normal play activity.
- Supervise activity.
- Have child fitted with protective helmet.
- Possible physical therapy consult for adaptive toys.

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Vignette #2 (Handout #8)

You are the caseworker involved in the care of an infant with hydrocephalus. The infant has just recently had an AP shunt placed. Since your last visit to this child one week ago, you seem to notice that the child's head appears larger, and she seems restless. What are some of the questions you would ask the caregivers regarding other symptoms? What would your suspicion be of a potential problem? What would your interventions be?

Trainer Note:

- Suspicious of blocked shunt.
- Irritable, poor feeding, lethargic, lower extremity spasticity, cries often, and vomiting may be other signs of increase in cerebrospinal fluid pressure.
- Call physician – take to emergency room.

Vignette #3 (Handout #9)

You are the adoptive parents of a child with hydrocephalus. You are concerned that this child may be treated “differently” by her classmates. What are interventions that you would suggest to assist in acceptance by peers, as well as understanding of the school?

Trainer Note:

- Explain condition to teachers.
- Style hair to make “shunt” less visible.
- Encourage play activity but provide safety helmet.

Vignette #4 (Handout #10)

You are the caseworker arranging emergency placement at 2 AM for an infant with hydrocephalus. What would be the key points that you would review with the foster family to provide them with the information necessary to survive the night until you can arrange for a next day visit for additional teaching?

Trainer Note:

- Signs & symptoms of blocked shunt.
- Name of physician and local emergency room.
- How to contact caseworker on call.

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EVALUATION AND CLOSURE **Transfer of Learning**

Rationale: This section is designed to assist participants in transferring information learned in training to their everyday practice, as well as to evaluate the trainer on his/her presentation.

Learning Objectives: Participants will:
Participate in the “What Have I Learned?” activity.
Complete and submit the program evaluation form.

Time: 0.5 Hours

Methods: Group activity.

Materials: Handout #11 – What Have I Learned?
Evaluation forms

Activities: Group discussion and completion of “What Have I Learned?”
Completion of evaluation forms

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EVALUATION AND CLOSURE

Trainer Note: Distribute Handout #11 . “What Have I Learned”

Have participants take about 10 minutes to complete the handout and then review it with the group at large.

***Transfer of Learning**

1. Why should a caregiver have a “dimple” lesion on their infant assessed by the pediatrician?
2. How would you feel if the agency caseworker called to inform you that a 2-week-old child with Spina Bifida was being placed in your home?
3. What are some of the key areas of involvement/referral for a child with hydrocephalus?

***Evaluation**

Trainer Note: Allow approximately 5-10 minutes for completion and collection of evaluation forms.

Before closing the training session, all participants must complete the evaluation forms and return to assist in future curriculum planning.

***Closure**

Thank participants for their attention and involvement during this very medically complex training session. Encourage their continued support and care of the medically complex child.

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