Fact Sheet

Primary Care Needs of Children with Hydrocephalus

Children with hydrocephalus may have life-long special health needs. These needs may alter their primary care. It is important that care givers understand hydrocephalus in order to provide primary health care to these children and their families.

Clinical Manifestations at Time of Diagnosis

Although the signs and symptoms of hydrocephalus may be somewhat varied by the specific cause of the condition, there are common clinical manifestation associated with increased intracranial pressure. If the accumulation of excessive cerebrospinal fluid (CSF) occurs slowly, the infant or young child may be asymptomatic until the hydrocephalus is quite advanced. Significant dilation of the ventricle may occur before abnormal head growth is apparent. Full or distended fontanels, frontal bossing, prominent scalp veins, vomiting, irritability, and even opisthotonic posturing may be observed before dramatic changes are noted in head circumference.

In the older child with fused cranial sutures, the development of hydrocephalus may result in the non-specific symptoms of headache, nausea, vomiting and personality changes, including irritability, lethargy and loss of interest in normal daily activities. Spasticity or ataxia of the lower extremities and urinary incontinence may occur. These children frequently complain of vision problems as increased intracranial pressures on the second, third or sixth cranial nerves result in extraocular muscular paresis and papilledema. Alterations in growth, sexual development and fluid and electrolyte imbalance may occur if increased pressure occurs at the site of the hypothalamus.

Implantation of a shunting device is usually the treatment of choice.

Associated Problems

Intellectual

Intellectual function is difficult to predict early in the disease process. The cause of hydrocephalus appears to be the most important determining factor of intellectual function. Uncomplicated hydrocephalus has a better cognitive prognosis than hydrocephalus associated with brain injury. In recent studies two-thirds of children with hydrocephalus had normal or borderline normal intelligence. In children with Intelligence Quotient (IQ) scores above 70, performance IQ scores are lower than full-scale and verbal IQ's. This discrepancy indicates a need for preschool and school counseling and testing to identify areas of learning disability.

Visual

Visual abnormalities are often found at the time of diagnosis or during episodes of shunt malfunction. Increased intracranial pressure results in optic nerve pressure, limited upward gaze, extraocular muscle paresis and papilledema.

Even though the shunt is functioning and hydrocephalus is controlled, these children commonly have vision problems. Gaze and movement problems such as strabismus, astigmatism, nystagmus and amblyopia are found in approximately 25-33% of children with hydrocephalus. Refractive and accommodation error are found in approximately the same percentage of children but not necessarily in the same children. Abnormalities in vision are associated with a lower performance scores but not with lower verbal intelligence scores.

Motor Disabilities

As many as 75% of children with hydrocephalus will have some form of motor disability. These disabilities vary from severe paraplegia to mild imbalance or weakness. The severity of the motor deficit is most often diagnosis-related; children with conditions such as porencephaly, Dandy-Walker malformation and the meningomyelocele have more serious motor defects than children with simple congenital hydrocephalus.

Hydrocephalus also affects fine motor control. Kinesthetic-proprioceptive abilities of the hands are often affected negatively, and this, coupled with impaired bimanual manipulation and frequent visual deficits, may make it difficult for the child with hydrocephalus to perform well on time-limited, nonverbal intelligence tests.

Seizures

Seizures in infancy are not uncommon at the time of initial diagnosis of hydrocephalus because of increased intracranial pressure. About 20% of infants with hydrocephalus continue to have seizures after the first year of life. These seizures may be simple or complex and usually can be well managed with standard anticonvulsant therapy. Acquired hydrocephalus is more often associated with seizure activity caused by the underlying reason for the development of hydrocephalus (i.e. brain tumor, CNS trauma or infection). These seizures may be more difficult to control and are usually more focal in origin.

Primary Care Management

Growth & Development

Both precocious puberty and short stature have been reported in children with hydrocephalus. Sexual development before the age of 8 in girls and 10 in boys is considered precocious and warrants further diagnostic study. Heights below the 5th percentile, if not compatible with family stature, indicate growth retardation. Treatment is available for both of these conditions, and children should be referred to an endocrinologist if symptoms occur.

In children suspected of having hydrocephalus, or who are known to have hydrocephalus, head circumference should be measured by experienced personnel. Until the cranial sutures are completely fused, which can be delayed in these children, growth of head size is a major diagnostic tool in evaluating the child's condition. Once a shunt has been placed, head circumference may decrease 1 to 2 cm as the pressure is relieved. After this initial decrease, the head should grow only in proportion of the child's body. The significance of head size measurements in the child who has a shunt cannot be overestimated, and daily measurements may be necessary when evaluating the shunt-dependent infant for possible shunt malfunction.

Standard early infant developmental screening assessment tools used in primary care practice, such as the Denver Developmental Screening Tool, may be of little help in assessing infants with hydrocephalus. It is important for the practitioner to interpret developmental findings in light of other clinical observations

to assist the parents in developing reasonable expectations for their infant. Some motor delays can be expected during infancy and childhood, given that approximately 75% of children with hydrocephalus have some form or motor disability. The primary care provider must document carefully motor skill acquisition because a loss of skill may indicate shunt malfunction or progression of the primary cause of hydrocephalus. This applies to older school-aged children as well. Ataxia, slurred speech, lack of progression in school, or incontinence also may indicate a deterioration of neurological status and the need for further evaluation.

Often children with hydrocephalus will benefit from infant stimulation programs or physical therapy, and the practitioner must be familiar with the programs available in the family's community to help them identify programs that would be most beneficial for their child.

Immunizations

Diphtheria, Tetanus, Pertussis (DPT). Pertussis can pose a special problem in infants with hydrocephalus. Because children, with a history of seizures, are at increased risk of seizures after pertussis vaccination deferral of the DPT immunization may be prudent until neurologic stability is ascertained. The risk of contracting pertussis is low but because neurologically impaired children may be at an increased risk or morbidity and mortality from illness caused by Bordetella pertussis, immunization is not absolutely contraindicated, and in some patients (such as those with well-controlled seizures, corrected hydrocephalus or cerebral palsy) will be indicated. The primary care provider will need to weigh the risk of disease versus the risk of side effects of the vaccination. In these difficult situations, consultation with the child's neurosurgeon or neurologist may be advisable to help assess the child's potential for having seizures.

Measles. Measles vaccine also has been implicated in post-vaccination seizures, with a higher incidence of this occurring in infants and children who have a history of convulsions. It is not believed these post-vaccine seizures produce permanent neurologic damage and the high ongoing risk of natural measles with its high morbidity rate justifies measles immunization in children with a personal of convulsions.

Haemophilus influenza type B. Conjugated polysaccharide-diphtheria Haemophilus influenza type B (HIB) vaccination is recommended at 18 months for all children. The use of HIB vaccine is variable but generally this vaccine is given less often than other recommended vaccinations. Because of the increased risk of HIB CNS infections in children with shunts, children with hydrocephalus who have shunts should definitely receive the new conjugated vaccine as recommended. Children who have a history of documented HIB disease before 2 years of age may not produce adequate antibodies to prevent a second infection and therefore should be immunized.

Other immunizations. Vaccination for polio, mumps and rubella should be given as routinely scheduled.

Screenings:

Visual Screening:

Because of the high incidence of visual defects in children with hydrocephalus, special attention should be paid to visual screening. The Hirschberg test and funduscopic examinations should be performed during each office visit and the result carefully documented. At about six months of age, the child should be referred to a pediatric ophthalmologist for a thorough examination. Yearly examinations should be scheduled thereafter.

Hearing Screening:

In addition to routine office screening for hearing acuity, infants with a history of CNS infection or antibiotic treatment with amino-glycosides should undergo auditory-evoked response testing. Subsequent shunt malfunctions or CNS infections require reassessment of hearing, and periodic evaluation by an audiologist is recommended.

Dental Screening:

Routine dental care is advised. If the child is receiving phenytoin for seizure control, more frequent dental care may be needed because of the possibility of hyperplasia of the gums. If the child has a ventriculoatrial shunt, prophylaxis with penicillin is recommended for all dental work including adjustment of braces to prevent bacterial endocarditis.

Special Family Concerns

Parents of children with hydrocephalus constantly worry about continued shunt function. With every malfunction there is a need for surgery and the perceived threat of brain damage. This constant worry and the daily responsibilities and stress of caring for a child who may have multiple medical problems is very difficult for families. Financial strain caused by numerous medical visits or surgical procedures may deplete a family's financial reserve, and private insurance may not be obtainable unless it is offered through a large group employment policy. Concerns about the child's ability to be self-supporting and independent in the future are also an issue for parents as the child grows into adolescence.

Depending on the child's condition, the number of specialists and community resources used, and the family's strengths or abilities, the parents may need assistance from the practitioner in case management. Coordination and communication between specialists, practitioners, family and school personnel can often become complex and overwhelming to parents. The practitioner can function as a central clearing house to help the parents understand the advised medical treatments, to follow-up on necessary referrals and to identify priorities for that family and child.

For additional resources about hydrocephalus and information about the services of the Hydrocephalus Association, please contact:

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