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**Standards
of Care
Outcome
Measures**

***for Children With
Myelodysplasia
(Spina Bifida)***

Developed By:
The Myelodysplasia Standards Committee
of the Bureau for Children With Medical Handicaps

Standards of Care for Children with Myelodysplasia (Spina Bifida)

Definition	A midline defect of the neural tube, the regional vertebral arches and overlying skin.
Incidence	Approximately one out of 1,000 live births in the United States
Goals	These standards are prepared to assure that children with myelodysplasia achieve maximum habilitation throughout life.
Care Needs	The care needs of the child should be addressed in a family-centered, community-based, coordinated and culturally competent manner. The care needs include medical, surgical, nutritional, psychosocial, educational, vocational, recreational and habilitation needs.
Team Management	<p>The care needs will be met by a coordinated team of pediatric specialists who are prepared by education and experience to care for the child with myelodysplasia. The team should consist of a pediatrician, neurosurgeon, urologist, orthopedist, clinical nurse specialist or nurse practitioner, service coordinator, social worker and dietitian. Other professionals with extensive pediatric experience including, but not limited to, an orthotist, physiatrist, physical therapist, occupational therapist, psychologist, ophthalmologist, audiologist, dentist and early childhood educator may be available for consultation.</p> <p>Parents of children with myelodysplasia, or persons with myelodysplasia, should actively participate as part of the coordinated team. The plan of care is developed with attention to the preferences and strengths of the child/adolescent and his/her family. The team discusses the child's prognosis with the family and provides them with ongoing emotional support. The team pediatrician keeps the primary care physician informed of the child's follow-up care.</p>
Referral	Referral should be made at the time of diagnosis of the spinal defect. Prenatal diagnosis is preferred. Referral should be made to a child care center approved by the Bureau for Children with Medical Handicaps, in which the coordinated team functions. It is emphasized that, while a coordinated team includes a pediatrician to manage the range of services required by children having this congenital lesion, the child requires ongoing primary pediatric care.
Prenatal Care	Referral should be made at the time of diagnosis of the spinal defect to a child care center approved by the Bureau for Children with Medical Handicaps, in which the coordinated team functions. Upon prenatal diagnosis, the coordinated team and genetics center should work cooperatively to assure that appropriate information, including medical, genetic and preventive information, is provided to the family.

First days of life

- (1) The initial contact includes assessing the neurological deficit and determining evidence of hydrocephalus. The team neurosurgeon should be involved in the neonate's care within the first 24 hours of life for repair of the cele, as well as during assessment and treatment of the hydrocephalus, if present.
- (2) Following the initial assessment, the significance, implications and need for early team management of the congenital defect are explained to the infant's family by the neurosurgeon, pediatrician, social worker and nurse. Printed materials about the disability, as well as verbal information, are provided to the family.
- (3) Surgical repair of the myelomeningocele is made early, preferably within the first 24 hours of life. Contraindications include a continuous skin coverage of the lesion or complications which make immediate surgical repair inadvisable. In these cases, medical care will be administered.
- (4) A comprehensive evaluation of the infant must be achieved by the coordinated team during the initial hospitalization. Every effort is made to involve both parents in decision making, even though the mother may be hospitalized in a separate location.
- (5) After resolution of the neurosurgical problems, other team members evaluate the infant and institute appropriate treatment prior to the infant's discharge.
- (6) Urologic evaluation should be obtained as soon as neurosurgical problems are resolved and the infant is stable.
- (7) The managing physician prepares a summary of the hospital course and ongoing plan of treatment for the primary care physician and the Bureau for Children with Medical Handicaps, if an application has been made for BCMH services.
- (8) The following subjects should be discussed with parents prior to discharge and repeated during follow-up care: genetic aspects, services of the specialized clinic, follow-up appointments, medical complications (e.g., shunt malfunction, urinary tract infections), anticipated development, anticipated interventions and availability of financial and community support, including parent groups. Printed materials on these topics should also be provided to the family.
- (9) Service coordination by the team service coordinator and local public health nurse service coordinator should be initiated and provided on an ongoing basis.
- (10) Latex precautions are instituted to decrease sensitization and avoid adverse exposure. The family receives information on latex sensitivity and precautions to take.
- (11) The team assists the family in screening all available financial and community program support.

First years of life

- (1) If there are no specific problems, follow-up evaluation by the coordinated team is made at intervals not greater than every three months during the first year of life and every four months for the second year of life.
- (2) Records are maintained including a flow sheet of pertinent data on the neurological, urological and orthopaedic status and developmental functioning of the child. This information is communicated to the child's primary care physician.
- (3) The neurosurgeon is responsible for follow-up care of the shunts and necessary lengthening, revision, etc. The neurosurgeon should also participate in any subsequent surgery or management of problems affecting the central nervous system.
- (4) The urologist is responsible for periodic evaluation of the urinary tract with the goal of preservation of renal function.
- (5) Aiming for early mobility, the orthopaedist performs regular assessments of the child's motor function and recommends treatment such as correction of joint deformity, dislocation and scoliosis.
- (6) The team pediatrician gives the family ongoing explanation of the child's condition, provides coordination of specialty care, and maintains continuity of care. The pediatrician assesses (or refers for assessment of) the child's intelligence and motor and sensory function.
- (7) Nutritional assessment is provided on an ongoing basis to ensure adequate nutrient intake and appropriate weight for height.
- (8) Other team members actively participate in the ongoing follow-up care of the child.
- (9) Primary pediatric care is given by the child's primary care physician.
- (10) Families are informed of the services of the local early intervention program.
- (11) Special emphasis is given to the development of self-help skills and the establishment of the child's bowel/bladder continence.
- (12) The team assists the family in securing all available financial and community program support.

Pre-school and early school years

- (1) Functional monitoring and appropriate treatment is given by the neurosurgeon, urologist and orthopaedist as needed. If there are no specific problems, follow-up evaluation is made by the coordinated team at intervals not greater than every six months.
- (2) Special emphasis is given to the development of self-help skills and the establishment of the child's bowel/bladder continence.
- (3) The coordinated team of professionals and parents are advocates for the child. In this capacity, they focus on the psychosocial development of the child and communicate with school personnel to assure appropriate educational placement.
- (4) Evaluation of independent mobility and encouragement of physical activity as appropriate.
- (5) Learning disabilities and attention deficit disorders are identified and remedial intervention and management is instituted.
- (6) Coordination with school providers, especially physical therapists and occupational therapists is essential at this time.
- (7) Pediatric care is provided by the appropriate primary care physician.
- (8) The team assists the family in securing all available financial and community program support.
- (9) Pediatric care is provided by the child's primary care physician.

Adolescence

- (1) The coordinated team evaluates the adolescent at semiannual intervals, or more frequently if necessary.
- (2) Members of the team, in consultation with school personnel, evaluate capacity for self care, mobility and further education and assist in arranging for the vocational assessment and placement of the young adult.
- (3) The team assists the adolescent and family in securing all available financial and community program support.
- (4) With the goal of integrating the adolescent into adult society, the team provides support, education and counseling relating to adult medical care, psychosocial, sexual and developmental concerns.

BCMHS Services

Requests for diagnostic and treatment services for children with myelomeningocele must be made by a physician provider who is a member of an approved coordinated team. Requests for service coordination are submitted by a team service coordinator.

Myelodysplasia Outcome Criteria

Outcomes

Process Measures

A. Quality Management

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| 1) The child's care for the myelodysplasia condition is managed by an established multidisciplinary myelodysplasia team. | 1 a) Referral made to the team for initial evaluation within the first few days of life.
b) The treatment plan is developed and implemented on the basis of team recommendations.
c) Team actively solicits family participation and collaboration in treatment plan.
d) Family has access to appropriate providers with each visit to the team.
e) Care coordination is provided by team with provision for local care whenever possible.
f) Complex diagnostic and surgical procedures are performed only in major centers.
g) Documentation of long and short term outcomes of the treatment plan.
h) Family has access by phone to team members at times other than regularly scheduled appointments. |
| 2) Service coordination is provided at both the team and local levels. | 2 a) The child has an identified team service coordinator and a local service coordinator (i.e. public health nurse).
b) There is documentation of a comprehensive service plan developed by the family, and the team and local service coordinators which addresses the medical, social, vocational, educational, nutritional and recreational needs of the child.
c) The family demonstrates an understanding of, and agreement with, the content of the plan.
d) The comprehensive service plan is updated at least twice yearly. |
| 3) Appropriate pediatric care is given throughout childhood. | 3 a) The child has an identified primary care physician who is an extended member of the team.
b) Physical exams and routine well child care are provided on a regular basis.
c) Immunizations are current.
d) Periodic monitoring for growth and development and related problems such as precocious puberty, attention deficit disorder, learning disabilities and obesity.
e) Mutual communication between the specialty team and primary care provider regarding the child's plan of care, progress, and special needs/problems.
f) Family demonstrates knowledge of child's routine health needs and treatment plan.
g) Child is screened for latex allergy and family receives instruction regarding latex precautions. |

4) Preparation for transition to adulthood is initiated at an appropriate age.

- 4 a) A transition plan is developed during the teenage years.
- b) The adolescent and family have input into the transition plan.
- c) Qualified adult health care providers are available to assume care at the appropriate time.
- d) The adolescent demonstrates the ability to self-direct care or obtain appropriate assistance.

B. Physiological

1) The child maintains adequate nutritional status.

- 1 a) Documentation of periodic nutritional assessment.
- b) Documentation of weight for height (length) ratio on National Center for Health Statistics (NCHS) growth grids.
- c) Documentation of skin fold measurements.

2) The child attains and maintains optimal neurological function.

- 2 a) Parent recognizes the signs and symptoms of changes related to neurological status and demonstrates knowledge of appropriate actions to take.
- b) Parent/child recognize the signs and symptoms of shunt malfunction and demonstrate knowledge of appropriate actions to take.
- c) Parent/child can identify actions and side effects of prescribed medications.
- d) Optimal seizure control achieved while maximizing function.

3) The child attains and maintains maximal level of skin integrity.

- 3 a) Parent/child can identify problems of decreased sensation.
- b) Parent/child can identify early signs and symptoms of skin breakdown, procedures to prevent skin breakdown and recommended treatment for skin lesions.
- c) Parent/child perform preventive measures such as daily skin checks, wearing non-restricting clothing, safety with hot and cold, wheelchair cushions, paddings, etc.

4) The child attains and maintains maximal level of urologic health, including upper tract functioning, bladder emptying and a socially acceptable degree of continence.

- 4 a) The child has urologic function tests within normal limits.
- b) Parent/child can identify early signs and symptoms of urinary tract problems and appropriate actions to take.
- c) Parent/child practice measures to prevent urinary tract problems by demonstrating ability to do clean intermittent catheterization and other urologic techniques as prescribed
- d) Parent/child can identify actions and side effects of prescribed medications.

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| 5) The child attains and maintains maximal level of bowel functioning, including physical and social aspects. | <ul style="list-style-type: none"> 5 a) Parent/child can identify early signs and symptoms of bowel problems and appropriate action to take. b) Parent/child demonstrate knowledge of recommended bowel program. c) Parent/child demonstrate knowledge of proper nutrition to support healthy and regular bowel evacuation. d) Parent/child can identify actions/side effects of prescribed medications. |
| 6) The child attains and maintains maximal level of spinal cord alignment and joint stability possible. | <ul style="list-style-type: none"> 6 a) Parent/child can identify signs and symptoms of spinal cord deformity and subsequent problems. b) Parent/child can identify recommended treatment plan. c) Parent/child demonstrate knowledge of safe use of adaptive equipment. |

C. Functional

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| 1) Child attains and maintains a maximal level of mobility. | <ul style="list-style-type: none"> 1 a) Parent/child can identify the treatment plan necessary to achieve mobility. b) Child can demonstrate typical exercise routine c) Obtains prescribed adaptive equipment, assistive devices and mobility aids. d) Parent/child can demonstrate knowledge of safe use of adaptive equipment, assistive devices and mobility aids. |
| 2) Child attains and maintains maximal motor skills. | <ul style="list-style-type: none"> 2 a) Parent/child demonstrate knowledge of treatment plan to promote development of motor skills. |
| 3) The child attains and maintains maximal self-help skills. | <ul style="list-style-type: none"> 3 a) Parent can identify the child's special safety needs related to the disability. b) Parent can identify the child's self-help needs within developmental guidelines. c) Parent teaches their child self-help skills (e.g. dressing, bathing, eating, toileting, etc.) d) Child can demonstrate skills compatible with highest educational level. |
| 4) Child obtains/maintains maximal understanding of sexual and reproductive capabilities. | <ul style="list-style-type: none"> 4 a) Child can identify implications of his/her neurosensory level as it pertains to sexual function. |

D. Psychosocial

- 1) Psychosocial response to handicapping condition is functioning level appropriate.
 - 1 a) Achievement of functioning level appropriate socialization skills.
 - b) Parent/child have information about support groups/peer groups.
 - c) Child demonstrates positive expressions of self-esteem.
 - d) Child participates in recreational activities.
- 2) Educational/vocational goals achieved.
 - 2 a) Appropriate neuropsychological testing is done.
 - b) Receives education in least restrictive environment with appropriate related services and supports.
 - c) Achieves employment/post secondary educational placement.

E. Family Satisfaction

- 1) Parent/child achieve satisfaction with outcomes of treatment.
 - 1 a) Parent/child have input into care plan.
 - b) Parent/child have access to choice of providers.
 - c) Parent/child demonstrate knowledge of myelodysplasia condition, complications, treatment plan, available resources, parenting skills and support opportunities.
 - d) Parent/child receive information about prevention and genetic counseling services available.
 - e) Parent/child identify cultural/religious beliefs and practices that need to be addressed in the delivery of care and services.

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The Myelodysplasia Standards Committee

Sonya Oppenheimer, M.D., Chairperson

Rosalind Batley, M.D.

Roberta Bauer, M.D.

Robert Bilenker, M.D.

Benedicta Enrile, M.D.

Gerald Erenberg, M.D.

Lorraine Fay, M.D.

Mary Jane Gerhardstein, M.S.W.

Jill Harris, P.T.

Deborah Hoffman

Meliss Klorer

Stephen Koff, M.D.

Edward Kosnik, M.D.

Kenneth Kropp, M.D.

Marlene Lutkenhoff, R.N., M.S.N.

Gloria Pina, R.N., C.P.N.A.

Mark Rayport, M.D., Ph.D.

Stephanie Reid

George Sich, Jr.

George Thompson, M.D.

Kurt Wegner, M.D.

BCMh Representatives:

James Bryant, M.D.

Karen Lane, R.N., M.A.

Kathie Myers, R.N., B.S.N.

Kathy Peppe, R.N., M.S.

Rosemary Feka, R.N.

Ann Weidenbenner, M.S., R.D., L.D.

Bureau for Children With Medical Handicaps

P.O. Box 1603

Columbus, Ohio 43216-1603

(614) 466-1700



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