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The Core Curriculum

Myelodysplasia

Objectives

1. Define myelodysplasia, and list two separate clinical presentations of myelodysplasia
2. Describe the relationship of myelomeningocele with anencephaly
3. Describe the range of clinical severity in patients with myelomeningocele
4. Discuss the relationship of orthopaedic problems accompanying myelomeningocele to neurologic level
5. Discuss the prognosis for ambulation in patients with spina bifida
6. Define a dietary supplement of proven effectiveness in reducing birth rates of myelodysplasia
7. Describe the relationship of latex allergy and myelodysplasia
8. Describe the clinical presentation of spinal dysraphism without spina bifida aperta

Discussion points

1. Is upper limb function normal in patients with myelomeningocele?
2. Why is a multispecialty clinic setting preferred for management of children with myelomeningocele?

Discussion

The term myelodysplasia encompasses a wide spectrum of disruption of the structures formed from the neural tube. Failure of closure of the tube results in myelomeningocele at the caudal end or anencephaly at the cranial end. Several types of intracanal pathology (tethered cord, diastatomyelia, congenital kyphos) can result in neurologic deficits appearing later in childhood. Sacral dimpling or hair patches often, but not always, signal an underlying spinal dysraphism. Myelomeningocele (often simply called spina bifida) denotes an open neural tube at birth. This defect, usually fatal a half century ago, is now managed aggressively at birth by neurosurgeons with closure of the sac. Hydrocephalus often accompanies myelomeningocele, and can be aggravated by sac closure, thus shunting procedures are also routinely performed in infancy. Arnold-Chiari malformations can become problematic at essentially any age. The variable levels of motor and sensory dysfunction are generally recorded by level of sensation, many children will have some reflex activity of muscles distal to their sensory level. Subsequent functional level is obviously linked to the neurologic level. Asher and colleagues demonstrated 20 years ago that sustained ambulation was dependent on at least some hamstring function, although many children with greater levels of paralysis will ambulate for some period during childhood. A variety of orthopaedic problems accompany myelomeningocele. Patients with sacral levels of paralysis can develop problems secondary to insensate feet and foot deformity. Knee flexion and external tibial

torsion are common in patients with lumbar level paralysis. Hip dislocation can accompany any level, but is most common when the medial hamstrings and quadriceps are innervated, but the abductors and extensors are not. A continuing dialogue is available in the orthopaedic literature about the advisability of attempts to reduce dislocated hips in patients with myelodysplasia, present thought can probably be best characterized in saying that unilateral dislocations in ambulatory patients are better reduced, but that benefit to reduction in any other setting has not been established. A wide variety of spinal pathology accompanies spina bifida, requiring complex management and decision making. To add further complexity, upper limb function is usually not normal, and hydrocephalus may affect proprioception and balance even though sensation is present. Other factors, such as obesity, which is relatively common and difficult to control in older children with myelomeningocele, also affect functional status.

Latex allergy is almost universal in children with spina bifida who have been exposed, and avoidance of exposure to natural rubber latex is now advised for all children with myelomeningocele. There may be a genetic predisposition in this population to latex allergy.

Administration of folate to women of childbearing age has been shown to markedly reduce the incidence of spina bifida; a genetic focus for metabolism of folic acid has recently been identified. Women with defective folate metabolism have increased serum levels of homocystine which are reduced by ingestion of folate. Obese mothers also have an increased rate of children with spina bifida.

Management of the child with myelomeningocele is complex, involving pediatric, neurosurgical, urologic, and orthopaedic follow-up throughout the entire childhood and adolescence. A recent outcome study concluded that parental hope had more influence on the quality of life than neurologic deficit, indicating the influence of so-called nonmedical factors play a great role in outcome. Thus, management of children with myelomeningocele is best provided in settings with the ability to coordinate multidisciplinary care.

References

1. Alman BA, Bhandari M, Wright JG. Function of dislocated hips in children with lower level spina bifida. *Journal of Bone & Joint Surgery - British Volume* 1996;78(2):294-8.
2. Asher M, Olson J. Factors affecting the ambulatory status of patients with spina bifida cystica. *J Bone Joint Surg (Am)* 1983;65:350.
3. Crandall RC, Birkebak RC, Winter RB. The role of hip location and dislocation in the functional status of the myelodysplastic patient. A review of 100 patients. *Orthopedics (Thorofare, NJ)* 1989;12(5):675-84.
4. Duffy CM, Hill AE, Cosgrove AP, Corry IS, Mollan RA, Graham HK. Three-dimensional gait analysis in spina bifida. *Journal of Pediatric Orthopedics* 1996;16(6):786-91.
5. Fraser RK, Menelaus MB. The management of tibial torsion in patients with spina bifida. *Journal of Bone & Joint Surgery - British Volume* 1993;75(3):495-7.
6. Fraser RK, Bourke HM, Broughton NS, Menelaus MB. Unilateral dislocation of the hip in spina bifida. A long-term follow-up. *Journal of Bone & Joint Surgery - British Volume* 1995;77(4):615-9.

7. Frawley PA, Broughton NS, Menelaus MB. Anterior release for fixed flexion deformity of the hip in spina bifida. *Journal of Bone & Joint Surgery - British Volume* 1996;78(2):299-302.
8. Frawley PA, Broughton NS, Menelaus MB. Incidence and type of hindfoot deformities in patients with low-level spina bifida. *Journal of Pediatric Orthopedics* 1998;18(3):312-3.
9. Furderer S, Hopf C, Schwarz M, Voth D. Orthopedic and neurosurgical treatment of severe kyphosis in myelomeningocele. *Neurosurgical Review* 1999;22(1):45-9.
10. Heeg M, Broughton NS, Menelaus MB. Bilateral dislocation of the hip in spina bifida: a long-term follow-up study. *Journal of Pediatric Orthopedics* 1998;18(4):434-6.
11. Hullin MG, Robb JE, Loudon IR. Ankle-foot orthosis function in low-level myelomeningocele. *Journal of Pediatric Orthopedics* 1992;12(4):518-21.
12. Kirpalani HM, Parkin PC, Willan AR, Fehlings DL, Rosenbaum PL, King D, et al. Quality of life in spina bifida: importance of parental hope. *Archives of Disease in Childhood* 2000;83(4):293-7.
13. Mita K, Akataki K, Itoh K, Ono Y, Ishida N, Oki T. Assessment of obesity of children with spina bifida. *Developmental Medicine & Child Neurology* 1993;35(4):305-11.
14. Muen WJ, Bannister CM. Hand function in subjects with spina bifida. *European Journal of Pediatric Surgery* 1997;7(Suppl 1):18-22.
15. Niggemann B, Breiteneder H. Latex allergy in children. *International Archives of Allergy & Immunology* 2000;121(2):98-107.
16. Ou CY, Stevenson RE, Brown VK, Schwartz CE, Allen WP, Khoury MJ, et al. 5,10 Methylene tetrahydrofolate reductase genetic polymorphism as a risk factor for neural tube defects. *American Journal of Medical Genetics* 1996;63(4):610-4.
17. Rodgers WB, Frim DM, Emans JB. Surgery of the spine in myelodysplasia. An overview. *Clinical Orthopaedics & Related Research* 1997(338):19-35.
18. Sarwark JF, Weber DT, Gabrieli AP, McLone DG, Dias L. Tethered cord syndrome in low motor level children with myelomeningocele. *Pediatric Neurosurgery* 1996;25(6):295-301.
19. Sarwark JF. Kyphosis deformity in myelomeningocele. *Orthopedic Clinics of North America* 1999;30(3):451-5, viii-ix.
20. Shaw GM, Todoroff K, Finnell RH, Lammer EJ. Spina bifida phenotypes in infants or fetuses of obese mothers. *Teratology* 2000;61(5):376-81.
21. Szepfalusi Z, Seidl R, Bernert G, Dietrich W, Spitzauer S, Urbanek R. Latex sensitization in spina bifida appears disease-associated [see comments]. *Journal of Pediatrics* 1999;134(3):344-8.
22. Williams EN, Broughton NS, Menelaus MB. Age-related walking in children with spina bifida. *Developmental Medicine & Child Neurology* 1999;41(7):446-9.