



# POSNA

## The Core Curriculum

### **Congenital scoliosis**

#### **Objectives**

1. Describe the embryology of the spine from the notochordal stage until complete vertebral segmentation
2. List the common types of congenital scoliosis described by McMaster and the prognosis of each one
3. Discuss the incidence and nature of other anomalies accompanying congenital scoliosis
4. Discuss the role of bracing for congenital scoliosis
5. Describe the types and locations of congenital scoliosis most likely to progress, and appropriate age to consider surgical intervention

#### **Discussion points**

1. What kind of workup for other anomalies should be performed on an infant with congenital scoliosis?
2. What are the indications for imaging of the spinal cord in a patient with congenital scoliosis?
3. What are the indications for imaging of the spinal cord in a patient with congenital scoliosis?
4. Why is there such a high rate of other anomalies accompanying congenital scoliosis?
5. What types of surgical procedures are indicated for treatment of congenital scoliosis? When would excision be indicated? Posterior fusion? Anterior and posterior fusion?

#### **Discussion**

A general knowledge of spinal embryology is necessary to understand the pathoanatomy and rationale for treatment of congenital scoliosis. As the neural tube is formed during the third week of the embryonal period, cells of the paraxial mesoderm coalesce to form the 42 to 44 pairs of somites. Somites give rise to the sclerotome, myotome, and dermatome. For congenital scoliosis, we are primarily concerned with the sclerotome, but the relationship of the spinal nerves which originate from the neural tube toward the center of the sclerotome, and the segmental arteries which grew between the sclerotomes are also important. In what is called a metameric shift, the cranial portion of each sclerotome combines with the caudal part of the adjacent superior sclerotome. The segmental arteries thus pass around the middle of the newly formed vertebral body (composed of the adjacent hemisclerotomes of two separate sclerotomes), and the spinal nerves pass between the vertebral bodies. It is not difficult to see the combinations of spinal anomalies which could result from a failure of orderly resegmentation of the sclerotomes. The process involves segmentation and re-formation, thus patterns of congenital scoliosis can be regarded as failures of segmentation

(unsegmented hemivertebrae of unsegmented bar) or of formation (hemivertebrae). The frequent association of kidney anomalies with congenital scoliosis can be explained by the fact that the lateral portion of the same paraxial mesodermal cells that form the somites forms the mesonephros. Other anomalies are frequently present with congenital scoliosis. Intraspinous anomalies have been found in 30%. Reduced pulmonary function has been noted in more severe thoracic curves.

The prognosis of any given deformity is a product of the disruption of symmetrical growth. A hemivertebra which has growth potential may not change in its configuration during growth. A hemivertebra which is accompanied by a failure of separation on the contralateral side is subject to rapid deformation. The natural history of congenital scoliosis has been studied in depth, especially by McMaster, and his work is the basis of our present treatment approach. Presently, he recommends treatment in the first year of life for unsegmented bars accompanied by hemivertebra in the midthoracic, thoracolumbar, or lumbar spines. If progression is recognized early, convex anterior and posterior epiphyseodesis is effective, but this must be performed early in life (before age 3-5, depending on the deformity) to be effective. Posterior procedures alone are not presently regarded as reliable for congenital scoliosis. For established deformity, hemivertebra excision has been reliable. Hemivertebrae at L5-S1 routinely require excision if a balanced spine is to be achieved.

The role of bracing is limited, but for control of compensatory curves and maintenance of coronal balance, bracing occasionally has a role.

## References

1. Bradford DS, Boachie-Adjei O. One-stage anterior and posterior hemivertebral resection and arthrodesis for congenital scoliosis. *Journal of Bone & Joint Surgery - American Volume* 1990;72(4):536-40.
2. Callahan BC, Georgopoulos G, Eilert RE. Hemivertebral excision for congenital scoliosis. *Journal of Pediatric Orthopedics* 1997;17(1):96-9.
3. Day GA, Upadhyay SS, Ho EK, Leong JC, Ip M. Pulmonary functions in congenital scoliosis. *Spine* 1994;19(9):1027-31.
4. Holte DC, Winter RB, Lonstein JE, Denis F. Excision of hemivertebrae and wedge resection in the treatment of congenital scoliosis. *Journal of Bone & Joint Surgery - American Volume* 1995;77(2):159-71.
5. Keller PM, Lindseth RE, DeRosa GP. Progressive congenital scoliosis treatment using a transpedicular anterior and posterior convex hemiepiphyseodesis and hemiarthrodesis. A preliminary report. *Spine* 1994;19(17):1933-9.
6. King JD, Lowery GL. Results of lumbar hemivertebral excision for congenital scoliosis. *Spine* 1991;16(7):778-82.
7. King AG, MacEwen GD, Bose WJ. Transpedicular convex anterior hemiepiphyseodesis and posterior arthrodesis for progressive congenital scoliosis. *Spine* 1992;17(8 Suppl):S291-4.
8. Lazar RD, Hall JE. Simultaneous anterior and posterior hemivertebra excision. *Clinical Orthopaedics & Related Research* 1999(364):76-84.

9. Loder RT, Urquhart A, Steen H, Graziano G, Hensinger RN, Schlesinger A, et al. Variability in Cobb angle measurements in children with congenital scoliosis [see comments]. *Journal of Bone & Joint Surgery - British Volume* 1995;77(5):768-70.
10. Lonstein JE. Congenital spine deformities: scoliosis, kyphosis, and lordosis. *Orthopedic Clinics of North America* 1999;30(3):387-405, viii.
11. McMaster MJ, Ohtsuka K. The natural history of congenital scoliosis. A study of two hundred and fifty-one patients. *Journal of Bone & Joint Surgery - American Volume* 1982;64(8):1128-47.
12. McMaster MJ. Congenital scoliosis caused by a unilateral failure of vertebral segmentation with contralateral hemivertebrae. *Spine* 1998;23(9):998-1005.
13. Prahinski JR, Polly DW, Jr., McHale KA, Ellenbogen RG. Occult intraspinal anomalies in congenital scoliosis. *Journal of Pediatric Orthopedics* 2000;20(1):59-63.
14. Shahcheraghi GH, Hobbi MH. Patterns and progression in congenital scoliosis. *Journal of Pediatric Orthopedics* 1999;19(6):766-75.
15. Thompson AG, Marks DS, Sayampanathan SR, Piggott H. Long-term results of combined anterior and posterior convex epiphysiodesis for congenital scoliosis due to hemivertebrae. *Spine* 1995;20(12):1380-5.
16. Winter RB, Lonstein JE, Denis F, Sta-Ana de la Rosa H. Convex growth arrest for progressive congenital scoliosis due to hemivertebrae. *Journal of Pediatric Orthopedics* 1988;8(6):633-8.
17. Winter RB, Lonstein JE. Congenital scoliosis with posterior spinal arthrodesis T2-L3 at age 3 years with 41-year follow-up. A case report. *Spine* 1999;24(2):194-7.