Congenital kyphosis

Objectives
1. Describe the embryology and formation of congenital kyphosis
2. Describe the natural history of congenital kyphosis, with emphasis on the differences from congenital scoliosis
3. Discuss differences in treatment that would be indicated depending on etiology (defect of segmentation or formation) and age of patient

Discussion points
1. What is the reason for the increased risk of paraplegia with congenital kyphosis?
2. What is segmental spinal dysgenesis?
3. What is the role of posterior fusion for congenital kyphosis? Is it different than for congenital scoliosis?

Discussion
Congenital kyphosis is less common than congenital scoliosis, and the natural history is less favorable. The embryology is the same as that for congenital scoliosis (discussed in more depth under that topic) but the failure of formation (more frequent) or segmentation is in the anterior vertebral body. Anterolateral vertebral body deficits can cause kyphoscoliosis. If congenital kyphosis is allowed to progress untreated, paraplegia typically follows during the adolescent growth spurt. Treatment, therefore, is directed toward identification in the young child and prevention of progression. In children < age 3-5 with an unsegmented posterior hemivertebra, posterior fusion often, but not always results in an improvement of the kyphosis. If a failure of segmentation is present, posterior fusion can arrest progression, but will not result in improvement. In older children, more demanding anterior correction and posterior instrumentation are necessary. In the untreated spine, the cord becomes compressed over the kyphotic deformity, with vascular embarassment of the cord. Decompression and stabilization have a better chance of success if the paraplegia is of recent onset and incomplete.

There is no role for bracing in the treatment of congenital kyphosis. An ominous subset of congenital kyphosis is segmental spinal dysgenesis, which is accompanied by narrowing of the canal and has such a poor prognosis that surgical intervention is indicated as soon as the diagnosis is made.
References


