TREATMENT OF KYPHOSIS AND LUMBAR STENOSIS IN ACHONDROPLASIA

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KYPHOSIS (1-5)

Kyphosis is fairly common in achondroplasia; 30 percent of the early thoracolumbar kyphoses persist. A third of these or approximately 10 percent of achondroplasts have a progressive kyphosis. Some control of the kyphosis early is possible orthotically using an underarm orthosis or TLSO (thoraco-lumbo-sacral-orthosis). This improves the clinical appearance and may have some impact on the lumbosacral hyperlordosis. There are no long term longitudinal studies as yet to document stabilization of the spine in the corrected position. The important question is whether there is a decrease in the 30 percent residual kyphoses or 10 percent progressive kyphoses in adulthood in treated patients. Of additional concern is the respiratory effects of some of these restrictive devices on the pulmonary function in children with small thoraces and thus pre-existing potential respiratory dysfunction.

Surgery for kyphosis in achondroplasia is indicated in the following situations. Any acute angular thoracolumbar kyphosis needs to be treated surgically. Orthotic treatment is ineffectual in these acute angular structural kyphoses. Any progressive kyphosis should be treated surgically. This can either be kyphosis that increases during growth or more commonly an increase that occurs following a laminectomy for neurological compression. At the time of the laminectomy, if there is a pre-existing kyphotic deformity, fusion should occur at the time of the decompression, thus preventing subsequent progression of the kyphosis. Significant kyphotic deformities over 60-70 degrees in children require stabilization.

For these kyphotic deformities, as with any kyphotic deformity, a two-stage combined anterior and posterior approach is necessary (6). Anteriorly, via a thoracolumbar approach, the anterior longitudinal ligament is divided and the intervertebral discs are excised. The excised rib is cut into small pieces and used for an interbody fusion, being augmented by autologous iliac bone or an allograft of banked bone when additional bone graft is necessary (Fig. 1). If there is an angular
Fig. 1. D.M. presented at 31+5 after having had a laminectomy at another center for symptomatic stenosis. He presented with back pain and progressive kyphosis and a lateral radiograph showed a 58 degree long kyphosis (A). The kyphosis was treated with anterior disc excision and interbody fusion, with correction to 16 degrees on the day of surgery (B). Postoperatively he was cast for 10 months spending the first 6 months supine. He was braced for an additional 6 months. At this time the fusion was solid and the kyphosis measured 28 degrees (C).