

## Neck Disorders In Little People

By Dr. George S. Bassett, M.D.

A variety of neck problems may occur in patients with dwarfing conditions because of alterations in the anatomy of the cervical spine. The cervical spine is made up of seven vertebrae, separated by discs ("shock absorbers"), joints, and held together by various ligaments. The cervical spine connects the skull to the rest of the body and is constructed so that a tremendous range of motion is possible. Each vertebra is made up of a block of bone in front ("vertebral body") and an arch of bone behind through which the spinal cord runs. The spinal cord exits the bottom of the skull through a hole called the foramen magnum and then passes through the cervical vertebra down the rest of the spine. Neck problems may occur at three different levels of the cervical spine depending upon the type of dwarfing condition: the foramen magnum, the upper cervical spine and the lower cervical spine.

The growth disturbance responsible for the changes seen in achondroplasia also affects the hole at the bottom of the skull (foramen magnum) through which a portion of the brain and spinal cord pass. In some infants with achondroplasia, the foramen magnum may be too small ("stenosis"). A variety of problems may occur if there is not enough room for the brainstem and spinal cord at the level of the foramen magnum. These include muscle weakness ("hypotonia") or complete paralysis; breathing problems including breath-holding ("apnea") or blue spells ("cyanosis"); swallowing difficulties or other feeding problems; or sudden infant death syndrome ("SIDS"). These problems would most commonly become evident during the first two or three years of life. The diagnosis of foramen magnum stenosis may be made by a combination of a physical examination, electrical tests ("SSEPS"), and special X-rays (MRI). Treatment is based upon the severity of the symptoms and findings. The foramen magnum will grow with time so observation alone may be all that is required in most children. However, the symptoms in some children may be of such severity that surgical enlargement ("decompression") of the foramen magnum is required even at a young age. This is a major surgical procedure and generally should not be performed unless there is significant evidence of cord compromise.

Problems of the upper two cervical vertebrae may occur in little people of all ages with spondyloepiphyseal dysplasia (SED), pseudoachondroplasia, chondrodysplasia punctata (Conradi's), metaphyseal chondrodysplasia (Schmid or McKusick types), or metatropic dysplasia. The first cervical vertebra (C1) is ring-shaped whereas the second vertebra (C2) normally has a block of bone in front shaped like a "post" (odontoid process) projecting toward the skull around which the ring of C1 rotates. The spinal cord passes behind this post (odontoid process) as it courses through the ring of C1. The back half of C2 is shaped like all other vertebrae with an arch of bone to protect the spinal cord. In those dwarfing conditions listed above, the odontoid process may be too small allowing for too much motion ("instability") when the head is moved forwards or backwards. This excessive motion may be further increased if the ligaments holding C1 and C2 together are too loose. This is called C1-C2 instability or atlantoaxial instability. Since the spinal

cord runs behind the odontoid through the ring of CI, injury to the cord may occur if there is excessive motion.

Symptoms of upper cervical instability may include muscle weakness, paralysis, numbness, electrical sensations ("paresthesias"), problems with walking, loss of bowel or bladder control, breathing difficulties, or "black-out" spells. However, many patients do not have any symptoms associated with their cervical spine instability ("asymptomatic"). However, these patients are at "high risk" for spinal cord injury while participating in sports, or if they should fall or become involved in a motor vehicle accident. The diagnosis may be confirmed by cervical spine X-rays taken with the neck in full flexion and full extension. If too much motion is present, these two vertebrae should be surgically fused even if there are no symptoms so that inadvertent injury to the spinal cord does not occur. If symptoms are present, surgery must be seriously considered.

Abnormalities of the lower cervical spine often occur in diastrophic dysplasia, campomelic dysplasia and Larsen's syndrome. In these conditions, the neck may lose its normal contour ("lordosis") because of underdevelopment of the third or fourth cervical vertebrae anteriorly. The body of the vertebrae may not be completely square in front, but rather triangularly shaped. In addition, the normal ring of the vertebrae may be deficient behind the spinal cord ("posteriorly") and may be associated with loose ligaments. This deficiency of bone anteriorly combined with loose ligaments posteriorly may allow the involved vertebra to slide forward on the next lower normal vertebrae and create a large angular deformity of the spine ("kyphosis"). This kyphosis may also cause injury to the spinal cord and lead to weakness or complete paralysis. Early detection is the key and will require a careful history and physical, as well as cervical spine X-rays. A neck brace may be initially used in younger children with mild deformities of the cervical spine in the hope that the deficient vertebral body will grow larger and stabilize the kyphosis providing that there is no evidence of instability or spinal cord compromise. However, surgical stabilization ("fusion") is strongly advised for little people with progressive deformities, instability, or signs of spinal cord compromise.

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