Occult Respiratory Dysfunction in a Craniovertebral Anomaly*

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Craniovertebral anomalies are developmental disorders that affect the skeleton and the enclosed neuraxis at the junction of the cranium and cervical spine. The two fundamental types of malformations result from faulty metameric segmentation or from dysraphic anomalies in the median sagittal plane. The associated clinical syndromes may be caused by any one of several factors, among which are bony deformities that produce compression or traction upon the neuraxis, intrinsic malformations of the nervous system, and disturbed circulation of cerebrospinal fluid.2

The effect of these lesions on respiratory function has received scant attention in the literature. The hazards accompanying operative intervention were first appreciated by List8 and were described secondarily by Mullan and Raimondi.19 The flexed position of the head, which is of utmost importance for proper surgical exposure, may stretch already compromised neural tissues and so produce changes in respiration which, if not corrected, may result in apnea and death. Pathological confirmation of respiratory peril was described by Bharucha and Dastur1 who, in 20 operations, reported four operative mortalities from ventilatory failure. Necropsy in two patients demonstrated areas of softening and hemorrhage in the cervicomedullary region. In a later communication,4 a similar danger was associated with decompression of atlanto-axial dislocations.

Case Report

A 50-year-old woman was admitted with a complaint of occipital pain of 2 years duration. The pain was increased by coughing and sneezing; and, associated with these maneuvers, she experienced momentary periods of apnea and tetraparesis. She noted dizziness when she extended the neck. She had increasing difficulty in gait, as well as clumsy movements and numbness of her arms and hands. She denied bowel or bladder dysfunction.

Examination. She was a short, dysmorphic woman with a short neck; otherwise, the general systemic examination was unremarkable. She was alert and oriented, the cranial nerves were intact, except for absent gag reflexes. Motor examination demonstrated a spastic, ataxic gait, worse on the right, but no loss in strength on either side. Coordination was impaired in all four extremities. Deep tendon reflexes were moderately hyperactive bilaterally, as was the jaw jerk. Hoffman and Babinski signs were present bilaterally. On sensory testing, vibration and position sense were impaired in both legs, but temperature, pinprick, and light touch were appreciated normally. Routine laboratory data were unremarkable.

Roentgenologic Findings. The chest x-ray was normal. Plain films (Fig. 1) of the cervical spine demonstrated abnormalities of the neural arches of C-1 and C-2. The anterior element of the atlas was fused with the base of the skull. In addition, platybasia was present with basilar invagination. There was minimal mobility of the dens in relation to the anterior arch of the atlas on flexion and extension. The diameter of the foramen magnum, from dens to posterior lip, measured 18 mm. Pantopaque myelography (Figs. 2 and 3) demonstrated widening of the spinal cord shadow in both the anteroposterior and lateral views beginning at the third cervical vertebra. The cisterna magna was small, and the fourth ventricle was in normal position. Further examination of the upper cervical spinal cord was carried out with air (Fig. 4). After the introduction of
FIG. 1. X-ray films of the lateral cervical spine. *Left:* Fusion of the arches of C-1 and C-2 is seen (*white arrow*) with minimal basilar invagination (*black arrow*). *Right:* Fusion of the anterior arch of the atlas with the base of the skull is seen (*white arrow*). The distance from the posterior lip of the foramen magnum to the dens is only 18 mm.

FIG. 2. Myelograms with the patient in the prone position. *Left:* The spinal cord is abnormally wide above the C-3 level. *Right:* There is encroachment on the anterior subarachnoid space.