
Cervicomedullary compression in young patients with achondroplasia: Value of comprehensive neurologic and respiratory evaluation

We studied prospectively 26 young patients with achondroplasia to test two hypotheses: that respiratory problems may be the result of occult spinal cord compression, and that achondroplastic patients with cord compression might have occult respiratory abnormalities. Respiratory abnormalities were present in 85%, the majority caused by a primary problem of the pulmonary system, such as small thoracic cage or obstructed airway. Three patients had hypoxemia, recurrent cyanotic spells, and episodes of respiratory distress explainable only by cervicomedullary cord compression; in each patient, respiratory problems were alleviated by decompressive surgery. Another six patients with cervicomedullary compression had, in addition, at least one primary pulmonary cause of respiratory problems. After decompressive surgery the respiratory problems improved in three and were unchanged in three. Reconstructed sagittal CT images proved the most sensitive technique for detecting cranio-cervical stenosis as a cause of cervicomedullary cord compression, although some degree of stenosis was present in nearly all of the patients. (J PEDIATR 1987;110:522-30)

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Achondroplasia is an autosomal dominant skeletal dysplasia primarily of endochondral bone. Dysplasia of the cranial base and upper cervical spine may cause stenosis of the neural canal and compression of the distal medulla and upper cervical spinal cord at the craniovertebral junction. Quadriplegia may result at any age.¹ Cervicomedullary cord compression also has been identified as one cause of respiratory compromise, apnea, and sudden infant death.²⁻⁴

The surmised pathophysiology is compression of the medullary respiratory center, denervation of respiratory tract muscles, or both. Neurologic signs, such as upper motor neuron findings, have been subtle or absent in many

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CT	Computed tomography
SEP	Somatosensory evoked potentials

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documented cases of cord compression in infancy and childhood.³⁻⁵ Because hypotonia in infancy and delayed motor development are thought to be characteristic of achondroplasia,⁶ determining when these signs point to a

neurologic abnormality is problematic. Some infants with achondroplasia who died suddenly and unexpectedly have had respiratory or neurologic symptoms; others have not.^{3-5,7} In either event, necropsy has usually revealed cervicomedullary compression and medullary gliosis.^{3-5,7} Some infants who died had pulmonary artery hyperplasia, right ventricular hypertrophy, or both, without congenital cardiac abnormalities. These pathologic changes suggest that pulmonary hypertension, perhaps the result of occult respiratory abnormalities, existed before death.^{5,7}

Because both restrictive and obstructive respiratory disease occur in achondroplasia, the former caused by a constricted thoracic cage and the latter by bony encroachment of the upper airways,^{2,8-13} interpretation of respiratory problems in children with achondroplasia is difficult. Furthermore, few patients have had systematic evaluations that might indicate how to disentangle the multiple potential causes of respiratory compromise.

We encountered several young patients with cervicomedullary compression whose respiratory abnormalities could not otherwise be explained. Cervicomedullary compression presented a clinical challenge: the manifestations were often subtle, but failure to diagnose and treat could result in sudden death. Our experiences led to these hypotheses: (1) infants and children with achondroplasia with respiratory problems may have occult cervicomedullary compression, and (2) some patients with cervicomedullary compression have occult respiratory problems. We undertook this clinical study to evaluate children with achondroplasia prospectively and comprehensively. Because signs and symptoms of cervicomedullary compression may be non-specific, and primary and neurogenic causes of respiratory disorders may coexist, we evaluated patients for both neurologic and respiratory abnormalities.

METHODS

Patient population. Thirteen boys and 13 girls with achondroplasia, aged 4 months to 6 years, were enrolled. They represented 42% of these patients seen in our clinics between December 1982 and November 1984. Ten were referred, because of symptoms, from outside our institution; 16 were regular clinic patients. Of the 36 nonparticipating patients, the parents of six were approached, but declined participation, and 30 were not informed of this study by their clinic physician.

All but one patient had parents unaffected by achondroplasia. At the time of admission to the study, 12 had respiratory symptoms alone, three had neurologic symptoms alone, four had both, and seven had neither. None of the nonparticipants had major respiratory symptoms noted in their medical records.

Each patient was admitted to the Pediatric Clinical

Research Unit for multidisciplinary evaluation, and informed consent was obtained. The protocol was approved by the institutional review board.

Respiratory evaluation. Parents were interviewed about the health and medical history of their child; respiratory and neurologic symptoms were elicited systematically with a standardized questionnaire. Each child was examined, and arterial blood gas measurements while breathing room air, electrocardiogram, cross-sectional and M-mode echocardiogram, chest radiograph, and overnight multichannel polysomnography were performed.

Neurologic evaluation. A detailed neurologic examination was performed on each child by a neurologist who was unaware of the history or laboratory findings. Axial CT of the head and upper cervical spine, reconstructed CT images of the craniocervical junction in sagittal planes, brainstem auditory evoked potentials, and upper limb somatosensory evoked potentials were also performed. Both CT and evoked potentials (except for the two SEP conducted intraoperatively) were interpreted by one of the investigators (H.W. or R.W.M.) who was unaware of the history or the results of the examination of the patient. Neurologic state was considered abnormal if one of the following was present: hyperreflexia, sustained clonus, hypertonia, paresis, asymmetry of movement or strength, or abnormal plantar response. Hypotonia was considered an unreliable sign in view of the frequent joint hyperextensibility in achondroplasia.

Axial CT of the head and neck was performed by conventional methods using a Somatom DR3 Scanner (Siemens Corp., Iselin, N.J.). For multiplanar reconstructions, axial sections 2 or 4 mm thick were obtained at 0-degree gantry angulation, beginning at the C2-C3 level and continuing craniad to the brainstem, with the patient in neutral position. All scans were evaluated by the same neuroradiologist (H.W.). Ventricular size was compared with cortical mantle.¹⁴ The foramen magnum was designated as small if the anteroposterior or transverse diameter, measured on axial CT scans, was >3 SD smaller than the mean for age for children without achondroplasia,¹⁵ and irregularities from the normal oval shape were noted.¹⁶ In addition we assessed, in each patient, configuration of the bony craniocervical junction, shape of the medulla and spinal cord, subarachnoid space surrounding the spinal cord, and spatial relationship between the odontoid process and the craniocervical junction.

Evoked potentials were performed with a Nicolet CA 1000 (Nicolet Instruments, Madison, Wis.) with magnetic disk storage (Fig. 1). Brainstem auditory evoked potentials were elicited by rarefaction clicks at intensities 50, 70, and 80 dB above hearing threshold with 35 dB contralateral masking. The active electrode was placed over the vertex

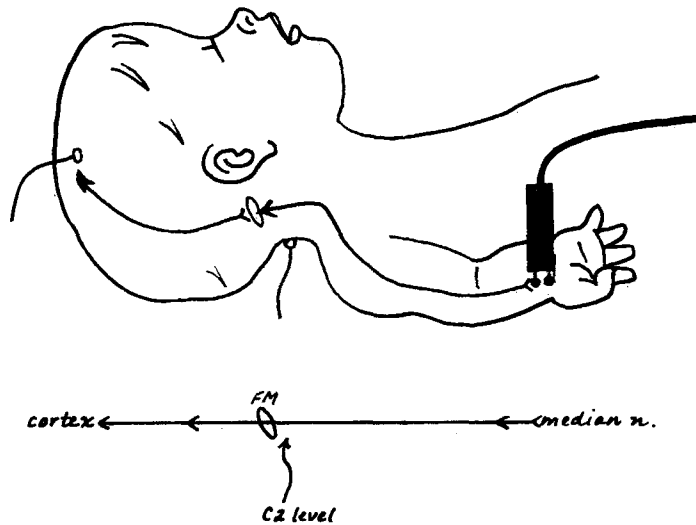


Fig. 1. Determination of somatosensory evoked potentials. Gold-plated silver cup electrodes were used, and impedance was maintained at <2000 ohms by dermal abrasion. Scalp electrodes were placed over contralateral cortex at C3' and C4' (international 10-20 system) and over second cervical vertebra (SC2), each referenced to midline forehead (Fpz). Each median nerve was stimulated transcutaneously at wrist. For each determination, 512 or 1024 square wave pulses of 200 msec duration were delivered at 5.9/sec (bandpass 5 to 5000 Hz), and responses were computer averaged. Recordings were considered adequate only if visible thumb twitch and reproducible responses over both sensory cortex and SC2 were obtained. Repeat determinations were separated by 2 to 3 minutes without stimulation. Latency of initial negative response at SC2, latency of initial negative wave over contralateral sensory cortex (N1), and amplitude of N1 were determined using cursor mode of computer. Interpretation of somatosensory evoked potentials was based on quality of N1 response (amplitude and shape) and central conduction time, defined as time delay between SC2 peak and N1 peak.²⁰ Because N1 peak intensity varies little with age,²¹ diminution or absence of response, in presence of adequate SC2 response, was considered abnormal. Central conduction time, an estimate of conduction from posterior column nuclei at C2 to sensory cortex, was chosen because of its theoretical independence of limb length. Central conduction times were considered abnormal when prolonged by at least 40% compared with expected values for age, which were extrapolated from studies of infants and adults.²⁰⁻²² FM, Foramen magnum.

(C2) and referenced to electrodes over each mastoid process. One thousand or 2000 stimuli were delivered at 11.9/sec (band pass 30 to 3000 Hz), and the responses were computer averaged. Replicate waves were obtained. Interpretation was based on the delay from wave I to wave V, because serous otitis, a common cause of hearing loss in achondroplasia, can result in a delay of auditory nerve input to wave I.

Definitions. Radiologic criteria for craniocervical stenosis were a small foramen magnum together with at least one of the following: deformity of the spinal cord, lack of subarachnoid space surrounding the cord, posteriorly placed odontoid process, or abnormal overgrowth of the occipital bone into the craniocervical junction (Fig. 2). In this study, cervicomedullary cord compression was defined as a chronic myelopathy. Thus craniocervical stenosis was defined radiologically, whereas cord compression was defined both anatomically and pathophysiologically; although suggested by clinical study, cord compression could be confirmed only at operation or autopsy.

RESULTS

Results for all patients, grouped according to initial complaints, are summarized in the Table.

History. Most (73%) patients had had respiratory symptoms (apnea, pneumonia, loud snoring, or dyspnea) in the preceding 6 months. Apneic or cyanotic episodes while awake occurred in seven (27%) patients without provocation, such as a recent feeding. Neurologic complications of achondroplasia, particularly cervicomedullary cord compression, had not been previously evaluated as a possible cause for any patient's respiratory symptoms.

Seven (27%) patients had previously known neurologic abnormalities, including five (two with quadriplegia) first diagnosed in our genetics clinics. One patient had had a ventriculoperitoneal shunt because of aqueductal stenosis, and progressive hydrocephalus was suspected in two patients because growth of their head circumference had crossed several percentiles on the reference charts for achondroplasia.¹⁷

Respiratory evaluation. Twenty-two (85%) patients had

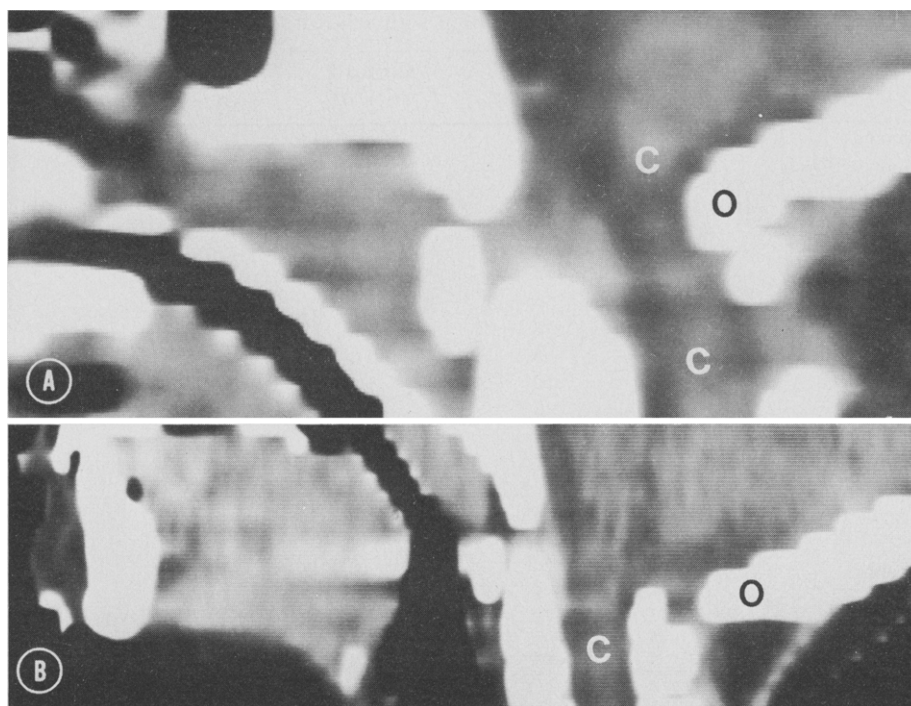


Fig. 2. Reconstructed midline sagittal CT images of craniocervical junction. **A**, Nine-month-old infant with achondroplasia. **B**, Twelve-month-old infant with no neurologic or skeletal abnormalities. Each image is oriented with face to left. Note in **A** that occipital bone (*O*) protrudes anteriorly and inferiorly and spinal cord (*C*) is thinned and distorted where it appears to abut bone at posterior foramen magnum.

respiratory abnormalities by our evaluations. All 16 patients with a small thorax¹⁸ had one or more abnormalities (hypoxemia, hypercapnea, pulmonary infiltrates, right ventricular hypertrophy on echocardiography, or right ventricular strain on ECG) suggesting a chronic respiratory disorder. However, three of 16 with small chests had hypoxemia during obstructive sleep apnea only. Six patients with respiratory abnormalities did not have small chests: four had obstructive sleep apnea, one had respiratory insufficiency while awake, and one had both. Respiratory insufficiency, defined as hypoxia with or without hypercarbia, was thus found in 15 (58%) patients, including 13 with and two without thoracic restriction.

Obstructive sleep apnea occurred in nine patients. Central sleep apnea, defined by the absence of respiratory effort, was found in two patients with obstruction; neither patient had apnea while awake. Among seven patients with waking apnea or cyanotic episodes, only one had sleep apnea (obstructive) documented on polysomnography. Two patients had cyanotic episodes associated with no respiratory effort witnessed by medical personnel, but none of these episodes was recorded on the polysomnogram.

Neurologic examination. In 11 (42%) patients, four of

whom had no symptoms, results of neurologic examinations were abnormal. Findings were confined to the lower extremities in five of the 11: three patients had hyperreflexia but normal strength, and two patients had flaccid paraparesis without hyperreflexia. Both upper and lower extremity signs were found in six of the 11: two had spastic quadriparesis, three had hemiparesis, and one had monoparesis and ankle clonus.

Computed tomography. In 25 of 26 patients, the foramen magnum was small in either transverse diameter alone (five patients) or both transverse and anteroposterior diameters (Fig. 3). Craniocervical stenosis was present in 15 patients. Ten others had a small foramen magnum but no other feature of stenosis. In reconstructed sagittal images, constriction of the craniocervical space was most often associated with abnormalities of the posterolateral portion of the occipital bone. In 14 patients with stenosis, the squama of the occiput extended anteriorly, forming an abnormal shelf of bone (termed "horizontalization"¹⁹). Ten of the 14 also had thickening of the posterior rim of the foramen magnum, which constricted the area further; in four the posterior rim protruded inferiorly and extended anterior to the posterior arch of the atlas (Fig. 2). Less

Table. Results of initial evaluations in 26 infants and children with achondroplasia

	Group 1 (n = 12)	Group 2 (n = 3)	Group 3 (n = 4)	Group 4 (n = 7)	Total (n = 26)
Respiratory symptoms					
Apnea or cyanotic episodes	4	1	2	0	7
Loud snoring	8	1	2	4	15
Recurrent pneumonia	4	1	3	0	8
Respiratory system evaluation					
Small chest circumference	8	3	3	2	16
Lung infiltrates	5	1	2	2	10
Right ventricular strain	8	1	2	2*	13
Hypoxemia while awake	5	0	4	1	10
Hypoxemia while asleep	10†	3	3	1†	17
Hypercapnea	6	1	0	0	7
Sleep apnea, obstructive	5	1	1	2	9
Sleep apnea, central	1	0	0	1	2
Neurologic examination					
Pyramidal signs	2	3	3	0	8
Paresis	2	2	3	1	8
Computed tomography					
↑ Lateral ventricles	7	1	2‡	3	13
↑ Extracerebral CSF	3	2	2	1	8
Small foramen magnum	12	3	4	6	25
Craniocervical stenosis	7§	3	2§	1	13
Absent posterior subarachnoid space	3	3	3	0	9
Abnormal somatosensory evoked potentials	2	3	2	0	7

Patients are grouped by type of complaint on entry: group 1 had respiratory symptoms; group 2 had neurologic symptoms; group 3 had both respiratory and neurologic symptoms; group 4 had neither.

*One patient had 3 mm cardiac interventricular septal defect.

†Not determined for one patient.

‡Additional patient had shunted aqueductal stenosis.

§Metrizamide CT myelography demonstrated stenosis and cord atrophy not documented on initial CT in one additional patient.

||In one additional patient, SEP was not done prospectively, but was performed immediately prior to operative decompression, with abnormal results.

commonly, dysplasia of the odontoid process caused constriction of the craniocervical space anteriorly; three patients had posterior displacement of the odontoid, and one had an osteophyte-like projection from the odontoid protruding cranially into the foramen magnum.

Two types of spinal cord deformity were seen. In reconstructed sagittal images, 11 patients had an hourglass deformity characteristic of spinal cord atrophy; in four of these patients the cord was sigmoid. Axial CT scans at the level of the atlas in patients with hourglass deformity showed the spinal cord as a flattened oval (Fig. 3). Of nine patients with an overhanging posterior rim of the foramen magnum, eight had an hourglass deformity. The second type of deformity, found in four patients, was posterolateral indentation of the cord by bony irregularities of the foramen magnum.

In six patients with an hourglass deformity of the cord, a thin rim of cerebrospinal fluid encircled the cord despite a narrowed craniocervical space. The other nine patients with anatomic stenosis lacked a posterior subarachnoid space, either at the foramen or caudally at the posterior arch of the atlas.

Thirteen patients (not including the patient with a ventriculoperitoneal shunt) had enlarged lateral ventricles for age, but none of the 13 had either progressive dilation or symptoms or signs of hydrocephalus. In addition, seven of those with ventricular dilation and the patient with a shunt had collections of excess cerebrospinal fluid over the frontal cortex.

Evoked potentials. The central components of brainstem auditory evoked potentials were normal in all patients. Nine patients had abnormal SEP; 17 had wave forms and central conduction times within the age-related normal guidelines.²⁰⁻²² In two patients, abnormal SEP were documented intraoperatively. Preoperative studies in the other seven yielded abnormal findings: five lacked cortical responses either unilaterally or bilaterally, two had flattened cortical responses with poor waveform, and two had extreme prolongation of the central conduction time. Abnormal SEP occurred exclusively among the patients with craniocervical stenosis and lack of posterior subarachnoid space on CT scans.

Results of initial evaluation. In nine patients, cervicomedullary cord compression was suspected. Somatosensory

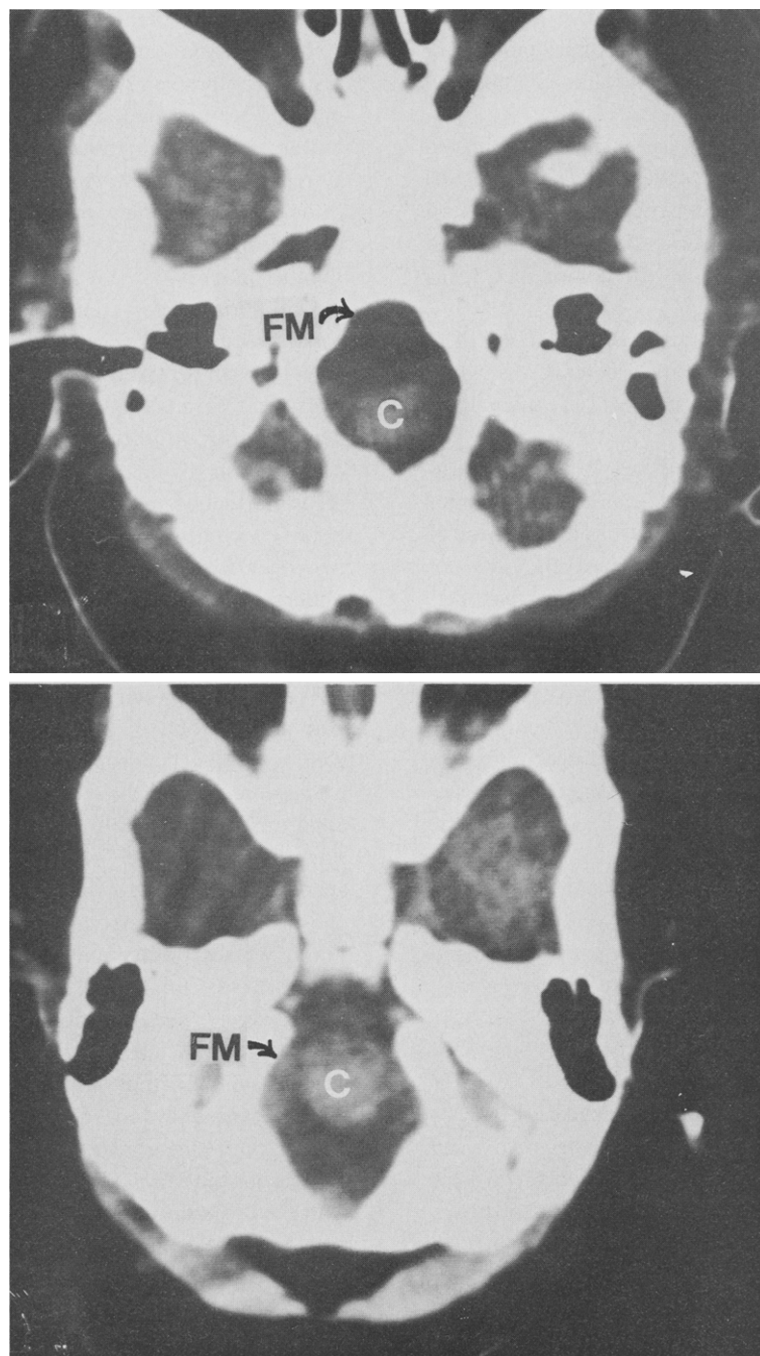


Fig. 3. Unenhanced axial CT images through foramen magnum. **A**, Fourteen-month-old infant with achondroplasia. **B**, Twelve-month-old infant with no neurologic or skeletal abnormalities, shown for comparison. Each image is oriented with face to top. Craniocervical stenosis is suggested in patient by small foramen magnum (*FM*) and narrowness of subarachnoid space posterior to spinal cord (*C*).

evoked potentials were abnormal in seven of these. In the other two, SEP studies were not performed because of severe respiratory problems in one and technical problems in the other; both had abnormal SEP intraoperatively.

Axial and reconstructed sagittal CT images without contrast enhancement showed craniocervical stenosis in seven. In the other two, CT-myelography showed stenosis and cord atrophy. All nine had respiratory problems, which in

three patients were not caused by obstructive sleep apnea, restrictive respiratory disease, or other primary pulmonary system disorders. One of these three also had no abnormal neurologic signs.

Management. All of the nine patients with suspected cervicomedullary compression underwent neurosurgical procedures. At operation, each patient had severe cord compression with atrophy. Five patients also required laminectomy of C1 and C2; in one the lamina of C3 was removed as well.

In 17 patients who did not undergo a neurosurgical procedure, treatment for respiratory problems was symptomatic (e.g., bronchodilators) or surgical (e.g., adenoidectomy or tracheostomy).

Follow-up. Twenty-two of 26 patients have been reevaluated 6 to 24 months after the initial evaluation. Eight of nine patients who had neurosurgery survived; the cause of death in one patient, 3 weeks postoperatively, was not determined, and autopsy was not allowed. Other postoperative complications included bacterial meningitis in three patients and obstructive hydrocephalus in one. One patient required a second decompressive procedure 1 year after the first because of reappearance of apnea, and 6 months later he was doing well. Five patients who had decompression had marked improvement in neurologic function, one had mild improvement by 6 months, and one had little improvement.

Despite marked neurologic improvement, obstructive sleep apnea worsened in two patients 1 year after spinal cord decompression; adenoidectomy in one patient and tracheostomy in the other improved the respiratory status. Two of the three patients with the most severe respiratory insufficiency preoperatively had no further respiratory problems; the other was unchanged.

Of 17 patients who did not have neurosurgery, 14 returned for follow-up. One had mild obstructive sleep apnea, eight had improved respiratory problems, two were unchanged, and three continued to have no abnormalities.

One patient who had craniocervical stenosis without evidence of cord compression on *initial* examination developed neurologic abnormalities. Cisternography subsequently showed, and surgical exploration confirmed, severe compression of the cord at the level of the foramen magnum.

DISCUSSION

Respiratory abnormalities and cervicomedullary cord compression. The few reports of clinical and postmortem findings in young patients with achondroplasia with respiratory problems suggest an association with cervicomedullary cord compression.^{1-5,7} In several patients, disorders of respiration were thought to be the earliest clinical manifes-

tations of a life-threatening abnormality of the neuraxis.^{3,4} Achondroplasia causes several possible mechanisms for respiratory problems, however, and rarely have reported patients been evaluated sufficiently to identify the contribution of neurogenic causes relative to primary pulmonary system abnormalities. Moreover, the frequencies of respiratory and neurologic problems in young patients with achondroplasia are unknown, although both are higher than in children without skeletal dysplasia.

Our serial observations support several conclusions. First, respiratory abnormalities are common and often severe in young patients with achondroplasia. Second, effective treatment is possible, but only after accurate determination of the causes of the problems. Third, in confirmation of our first hypothesis, some patients with respiratory abnormalities, but no clinical neurologic signs, have cervicomedullary compression. Finally, in support of our other hypothesis, some patients who have cervicomedullary compression, but no respiratory problems evident to their parents or pediatricians, have episodes of apnea or hypoxemia.

Frequency of respiratory and neurologic complications.

Our study group of 26 patients comprised 42% of all children with achondroplasia seen in our clinics. On initial evaluation, 85% of the study participants had respiratory abnormalities, and 35% had cervicomedullary compression. Even if neither problem occurred among the 36 achondroplastic children who were not studied, a minimum of 34% of our entire patient population younger than age 7 years had respiratory abnormalities and 14% had cervicomedullary compression. Although some bias in favor of more severely affected patients exists in clinics at a tertiary referral center, some of the patients with abnormalities were "asymptomatic" in the eyes of their parents or pediatricians, and were referred for other reasons. Whether the occurrence of other phenotypic manifestations that are reasons for referral, such as severe lower limb deformity or recurrent otitis, is related to respiratory or neurologic problems requires further investigation. In any event, the respiratory and neurologic abnormalities identified in our study group are likely more common in children with achondroplasia than was previously recognized.

Multiple causes of respiratory abnormalities in children with achondroplasia. The respiratory problems of most of the patients resulted from some primary abnormality of the pulmonary system, especially thoracic cage restriction or upper airway obstruction. However, three patients, each with cervicomedullary compression, had respiratory distress that could not be explained by a primary pulmonary cause. Our suspicion that the respiratory problems were neurogenic was confirmed in each case by marked improvement after surgery to decompress the cervicomedullary junction. The other six patients who had cervicom-

edullary compression also has respiratory abnormalities, but all had at least one primary pulmonary cause. Three of these patients had some improvement in respiration after neurosurgery, and three did not.

These results support the etiologic heterogeneity of respiratory abnormalities in infants and children with achondroplasia and emphasize the need for a thorough search for multiple causes, even in patients who have no initial complaints. Furthermore, some patients with severe cervicomedullary compression have their most impressive signs and symptoms in the respiratory system.

Evaluation of neurologic abnormalities. The neurologic examination was neither completely specific nor completely sensitive for cervicomedullary compression. When coupled with the potentially high frequency of cord compression in this population, the results of this study suggest that any young patient with achondroplasia should be evaluated for cord compression, even in the absence of abnormal symptoms or signs. What, then, is the most appropriate means of evaluation?

Somatosensory evoked potentials. SEP have been suggested as a sensitive early indicator of upper cord compression.²³ In our patients, abnormal SEP were present in all who had clinical signs, however mild, of cervicomedullary compression. The SEP were normal in all patients who were thought, because adequate subarachnoid space surrounded the cord, not to have persistent cervicomedullary compression. Thus the specificity of SEP in this situation is high. In several patients, on the other hand, SEP were normal despite a stenotic craniocervical junction on CT scan; none of these patients had abnormal neurologic signs and all had subarachnoid space around the hourglass spinal cord. It is unclear whether the presence of normal SEP despite a high likelihood of cord atrophy reflects a lack of sensitivity of SEP to subacute spinal cord damage, or whether the radiologic abnormalities are of little clinical consequence.

We found that SEP confirmed our clinical suspicion of cervicomedullary compression. This test might be most helpful when signs of myelopathy are equivocal and the SEP are abnormal. The meaning of a normal SEP in a patient who has nondiagnostic neurologic signs is unclear, as is the utility of serial SEP. Furthermore, because we chose to use a qualitative interpretation of SEP, either normal or abnormal, the diagnostic value and prognostic importance of subtle changes in wave form or conduction time are unclear, particularly in the absence of standards in young children with no neurologic problems.

Computed tomography. The reconstructed sagittal view showed, in addition to the reduced anteroposterior dimension of the foramen magnum, features of achondroplasia previously documented pathologically but heretofore underemphasized. The squamous portion of the occipital

bone was often "horizontalized" and extended anteriorly.²² The endochondrally derived lateral parts of the occiput formed a thickened and shortened posterolateral margin of the foramen magnum.^{22, 24, 25} Further, in some patients the foramen magnum was of adequate size, but posterior displacement of the odontoid effectively reduced the size of the craniocervical junction.²⁶

Routine lateral skull radiographs, cephalography, and axial CT scans are insufficiently sensitive techniques for detecting these bony abnormalities.^{14, 27} Magnetic resonance imaging can define spinal cord anatomy at the craniocervical junction in the transverse, sagittal, and coronal planes. Two problems, one technical and one logistic, prevent adoption of this technique. In the spin-echo pulse sequence T1 weighted images, the spinal fluid and ossified bone surrounding the thecal sac have similar signal intensity. In T2 weighted images, the spinal fluid and the spinal cord have almost identical signal intensities. Thus the presence or absence of a thin layer of spinal fluid between the cord and the occiput or vertebrae cannot be determined accurately. Moreover, close monitoring of ill or sedated patients while in the scanner is difficult.

Recommendations for initial evaluation. History and physical examination detect most, but not all, infants and children with achondroplasia who have cervicomedullary compression. Addition of an arterial blood gas measurement in the awake patient who is breathing room air, and polysomnography will detect those patients who have occult respiratory abnormalities that may or may not be neurogenic. Even in combination, these procedures will be nonspecific, and the patients identified as being at higher risk for cord compression should undergo CT with multiplanar reconstruction and SEP, in that order. In this way, the tests that are more costly and uncomfortable and that require sedation of the young patient can be reserved for those most likely to have serious neurologic compromise.

These young patients require comprehensive and detailed evaluations to detect and to manage abnormalities of the craniocervical junction. Close follow-up is needed for improving our understanding of this common complication of achondroplasia.

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