ABSTRACT. Objective. The purpose of this study was to determine the prevalence of kyphosis in a Turner syndrome (TS) population.

Methods. Standing lateral thoracic spine and standing anterior-posterior (A-P) scoliosis radiographs were obtained on all girls with TS between the ages of 5 and 18 years seen in a TS clinic between July 2000 and March 2001. Medical histories were reviewed, and a pediatric orthopedic surgeon evaluated the radiographs of each patient (N = 25). Excessive kyphosis was defined as an A-P curvature > 40°, vertebral wedging as any A-P deformity > 5° at an individual vertebral body, and scoliosis as a lateral curvature > 10°.

Results. Fifteen (60%) of 25 patients were found to have abnormal radiographic findings: 10 (40%) of 25 with excessive kyphosis, 10 (40%) of 25 with vertebral wedging, and 5 (20%) of 25 with scoliosis. Forty-eight percent of the girls had both excessive kyphosis and/or vertebral body wedging. Two girls had kyphosis ≥ 55°, and 5 had scoliosis ≥ 25°. Girls with excessive kyphosis and/or vertebral body wedging were older (13.6 ± 3.9 years vs 10.6 ± 2.8 years).

Conclusions. The prevalence of excessive kyphosis and vertebral body wedging seems to be increased in girls with TS and corresponds with advancing age. Routine radiologic surveillance may facilitate detection of developing deformities so that treatment with a brace can be considered to prevent or slow the process. Pediatrics 2002;109(6). URL: http://www.pediatrics.org/cgi/content/full/109/6/e93; Turner syndrome, Scheuermann’s disease, kyphosis, scoliosis, skeletal, orthopedics, radiography, spine.

ABBREVIATIONS. TS, Turner syndrome; A-P, anterior-posterior; BMI, body mass index; GH, growth hormone.

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older (13.6 ± 3.9 vs 10.6 ± 2.8 years) than those who did not. All girls older than 14 years of age (N = 8) had excessive kyphosis/wedging (Table 1). Seventy-five percent (9/12) of the girls with excessive kyphosis/wedging had a 45,X karyotype versus 46% (6/13) who did not.

Logistic regression revealed that chronological age alone was predictive of the presence of excessive kyphosis/wedging (P = .053). By stepwise linear regression analysis, chronological age was also predictive of the degrees of kyphosis (P = .032; r^2 = 0.24). No other variables (karyotype, BMI percentile for age, presence of scoliosis, use and duration of GH, oxandrolone, and estrogen treatments) were predictive of the presence of excessive kyphosis/wedging or degree of kyphosis (Table 2).

**DISCUSSION**

Our study suggests that the prevalence of vertebral wedging and excessive kyphosis in the TS population is high. Irregular vertebral epiphyseal growth rings and anterior vertebral wedging in TS were first reported in the Polish medical literature in the late 1950s. In 1968, additional reports appeared in the American and German orthopedic literature. In these studies, 22 of 39 subjects with TS ages 12 to 50 years (mean age: 27 years) and 11 of 22 subjects with TS older than 12 years of age, respectively, were reported to have abnormalities of the vertebral epiphyseal rings (“Scheuermann-like disease”). In the latter study, 5 of the patients were reported to have kyphosis. More recent orthopedic reviews of TS state that abnormalities of the vertebral epiphyseal rings rarely cause significant kyphosis or pain, and general reviews of TS rarely mention vertebral wedging or kyphosis at all.

In the present study, 48% of girls with TS in a population ranging in age from 5 to 18 had kyphosis >40°, a prevalence markedly increased compared with the reported prevalence of 3% in a general population of school-aged children. The cause and natural history of structural kyphosis remain controversial, even in the general population. However, most researchers agree that mechanical factors, osteoporosis, and the adolescent growth spurt are major determinants. In TS, the cause of kyphosis seems to be multifactorial and is likely to include the above factors, an intrinsic bone defect, and hormonally induced growth spurts.

Individuals with TS have bone growth abnormalities that result in short stature, abnormal body proportions, growth plate and epiphyseal irregularities, abnormal fusion of bones, variably delayed maturation of bones, and osteopenia. These skeletal disturbances are attributable, at least in part, to haploinsufficiency of pseudoautosomal genes on the X chromosome, such as SHOX, a transcription factor that is expressed in the developing limbs, the first

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<th>Patient Number</th>
<th>Age (Years)</th>
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<th>Scoliosis (Degrees)</th>
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* Scoliosis treated with brace, resulting in a decreased kyphotic curvature.
† Spinal abnormality treated with surgery.

**TABLE 1.** Spinal Abnormalities by Age

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<tr>
<th>Patient Number</th>
<th>Age (Years)</th>
<th>BMI Percentile</th>
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SD indicates standard deviation.

<table>
<thead>
<tr>
<th>Variable</th>
<th>Girls With Kyphosis/Wedging Mean ± SD (n = 12)</th>
<th>Girls Without Kyphosis/Wedging Mean ± SD (n = 13)</th>
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</thead>
<tbody>
<tr>
<td>Age (y)</td>
<td>13.6 ± 3.9</td>
<td>10.6 ± 2.8</td>
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<tr>
<td>BMI for age (%)</td>
<td>83.4 ± 17.3</td>
<td>72.5 ± 26.5</td>
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<tr>
<td>Duration of estrogen (y)</td>
<td>1.0 ± 1.3</td>
<td>0.4 ± 0.1</td>
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<tr>
<td>Duration of GH (y)</td>
<td>4.14 ± 3.6</td>
<td>3.3 ± 2.2</td>
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<tr>
<td>Duration of oxandrolone (y)</td>
<td>1.4 ± 1.5</td>
<td>0.4 ± 1.0</td>
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2 of 5 KYPHOSIS AND TURNER SYNDROME

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and second pharyngeal arches, and in bone marrow.\textsuperscript{14}

In TS, body proportions of the torso are abnormal, because there is a greater relative reduction in body height than in body width.\textsuperscript{15} Developmental abnormalities of individual bones that may affect the spine include hypoplasia of cervical vertebrae, coalition of vertebrae and hemivertebrae, although these were not found in our study population. One might also hypothesize that variability in rate of bone matura-
tion, a phenomenon well documented for bones in the hand,\textsuperscript{16} may also occur in the spine and lead to abnormal mechanical stresses. Finally, osteoporosis is more common in individuals with TS.\textsuperscript{12} Even in the prepubertal years, many girls have a coarse trabecular bone pattern and decreased bone deposition at primarily cortical sites. During adolescence, there is increased bone turnover with osteopenia predominantly at trabecular sites such as the vertebrae.\textsuperscript{17} Abnormally low levels of estrogens secondary to gonadal dysgenesis and delays in estrogen replacement therapy no doubt contribute to this increased bone turnover state. Abnormalities of bone content and structure can alter the strength and mechanics of the spine, predisposing individuals to kyphosis as well as scoliosis.

Although most girls with TS undergo a blunted adolescent growth spurt, growth velocity is increased with GH therapy. Although one might predict that kyphosis would progress with increased growth-enhancing therapies, GH might have a protective effect by increasing bone mineral density.\textsuperscript{17,18}

In the present study, girls with structural kyphosis and vertebral wedging were older (average age: 13.6 years) than those without these abnormalities (average age: 10.6 years). However, this study was unable to determine the role of age and hormonal therapies in the development of kyphosis because of the small number of subjects, most of whom had received GH with or without estrogen and oxandrolone.

Early detection and treatment of kyphosis are important. Possible sequelae of excessive kyphosis include progressive deformity, back pain, and embarrassment about physical appearance. In extreme cases, impairment of neurologic and cardiopulmonary function can occur. Although treatment recommendations vary, bracing is generally recommended for progressive kyphosis $\geq 50^\circ$ in a skeletally immature patient.\textsuperscript{19} Surgical correction is considered for larger curves, particularly if the deformity is expected to worsen with continued growth.

Fig 1. Lateral views of an adolescent with TS and excessive kyphosis.
CONCLUSION

The prevalence of excessive kyphosis and vertebral body wedging seems to be increased in TS and to correlate with advancing age in childhood. Larger, longitudinal studies are needed to determine the relative roles that intrinsic bone defects, obesity, osteopenia, growth-promoting agents, and sex steroid therapy play in the development of excessive kyphosis. The phenotype (short neck, stocky build) of many girls with TS may hinder the clinical diagnosis of kyphosis. Routine radiologic surveillance may be indicated to aid in its early detection and allow for timely implementation of bracing. Although such radiographs seem to be most informative during adolescence, optimal timing of radiologic surveillance has yet to be established.

ACKNOWLEDGMENTS

We thank Dr Ali S. Calikoglu for his insightful comments and editing of this manuscript, and Lauren Lindblad, BS, and Paul W. Stewart, PhD, for statistical expertise.

REFERENCES


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