47. Screening for Adolescent Idiopathic Scoliosis

RECOMMENDATION

There is insufficient evidence to recommend for or against routine screening of asymptomatic adolescents for idiopathic scoliosis. Clinicians should remain alert for large spinal curvatures when examining adolescents.

Burden of Suffering

Scoliosis, a lateral spinal curve of 11° or greater, affects an estimated 500,000 adults in the United States.[1] Idiopathic scoliosis accounts for about 65% of cases of structural scoliosis,[2,3] and a large proportion of these cases develop during adolescence. A lateral spinal curve of 11° or greater is present in about 2–3% of adolescents at the end of their growth period. Curves greater than 20° occur in less than 0.5% of adolescents.[4] The potential adverse effects of scoliosis include the progressive development of unpleasant cosmetic deformities, back pain, social and psychological problems during both childhood (e.g., poor self-image, social isolation) and adulthood[5] (e.g., limited job opportunities, lower marriage rate), and the financial costs of treatment.

There is little firm evidence that persons with idiopathic scoliosis are at significantly greater risk of experiencing back complaints than is the general population; most existing epidemiologic studies have lost a large proportion of patients to follow-up and lack adequate statistical power to detect a difference.[6,7] Data on the psychosocial effects of scoliosis and poor cosmesis are also limited. Long-term studies suggest a poor correlation between the location or magnitude of curves and the extent of psychosocial complaints.[6] A number of surveys and uncontrolled long-term studies of scoliosis patients have reported low marriage rates in women and high rates of unemployment, disability, and poor self-esteem,[9–12] but these studies lacked internal control groups, and many patients in the cohorts had spinal conditions other than adolescent idiopathic scoliosis. Persons with severe curves are at increased risk of restrictive pulmonary disease and increased mortality, but such curves are usually early-onset and at least 100–120° in magnitude.[6,12–15] Severe curves of this magnitude have become uncommon in the United States and generally occur only as a consequence of severe, early-onset infantile or juvenile scoliosis.[17]

Only a subset of curves detected through screening are destined to progress to a point of potential clinical significance. The probability that curves will progress more than 5° can vary from 5% to 90%, depending on the patient's age, sex, and skeletal maturity, and the pattern and magnitude of the curve.[18–21] Progression is less likely in older children with greater skeletal maturity and with smaller curves. Depending on the patient population, between 25% and 75% of curves detected on screening may remain unchanged, and 3–12% of curves may improve.[21–24] The reported probability that curves less than 19° will progress is 10% in girls between age 13 and 15 and 4% in children over age 15.[18,19] In curves that progress, one study found that the probability was 34% that the curves would progress more than 10°, 18% that they would progress more than 20°, and 8% that they would progress more than 30°.[21] Another study of patients with untreated curves found that 25% ceased progression before reaching 25° and that 12% ceased progression before reaching 29°.[22]

Accuracy of Screening Tests

The principal screening test for scoliosis is the physical examination of the back, which includes upright visual inspection of the back and the Adams forward-bending test.[25] Patients with abnormal findings on initial physical examination are often then referred for a more thorough physical examination. Some physicians also obtain a standing roentgenogram to measure the degree of curvature (e.g., Cobb angle). Roentgenographic findings serve as the reference standard for estimating the sensitivity and specificity of screening tests. The reported 95% confidence interval for intraobserver and interobserver variability in measuring the Cobb angle on radiographs is 3–5° and 6–7°, respectively.[26,27]

A relatively large proportion of children screened in schools are found to be "positive" on initial examination, but only some of these cases are ultimately found to have scoliosis. In studies of school screening, 11–35% of screened children were classified as positive and were referred for further evaluation; in one study, 37% of those referred for orthopedic evaluation were found to have no abnormality.[24,28] The sensitivity and specificity of the physical examination depend on the skills of the examiner and the degree of spinal curve being sought. In one study, public health nurses with special training in school screening were able to detect all children (sensitivity of 100%) with a Cobb angle greater than 20°. The specificity of the examination was 91%. The sensitivity and specificity of the examination in detecting curves greater than 10° were 73.9% and 77.8%, respectively.[29]

The positive predictive value (PPV) of visual inspection and the forward-bending test varies with the degree of curvature by which a "true positive" is defined, the prevalence of scoliosis in the screened population,

and the skills of the examiners. The magnitude of the PPV is inversely related to the degree of curvature used to define scoliosis, since the prevalence of small curves is greater than large curves. In an Australian study, the PPV was 78% for curves greater than 5° in a population with an estimated prevalence for this degree of curvature of 3%.[30] In another study, the PPV was 54% for curves greater than 10° (prevalence of 2%) and 24% for curves greater than 20° (prevalence of 1%). A Canadian study involving specially trained school nurses reported a PPV of 18% in detecting curves greater than 10° (prevalence of 1.7%) and a PPV of 4% in detecting curves greater than 20° (prevalence of 0.3%).[28]

Other scoliosis screening tests include the inclinometer[31,32] and Moire topography. The inclinometer has a reported sensitivity of 96–98%, specificity of 29–68%, and reliability coefficients of 0.86–0.97 in detecting a Cobb angle of 20° or more.[33] In some studies, Moire topography correlates poorly with the Cobb angle.[34] A study that combined Moire topography with the forward bending test found that Moire topography had a sensitivity of 95% and the forward bending test had a sensitivity of 46% in detecting curves of 10° or greater. The calculated PPV of the test was 29% (study prevalence of 4%).[35]

There is limited information about the value of repeated screening of children who have previously tested negative for scoliosis. Although the probability of false-positive results would be increased by such a practice, repeat screening could potentially detect cases in older adolescents that escaped detection in early puberty or that developed into significant curves after screening was performed. There are few data that confirm these benefits. In one study, 43% of the cases that were detected on screening during tenth grade had previously tested negative 2–3 years earlier.[30]

Effectiveness of Early Detection

Direct evidence of the effectiveness of scoliosis screening would require controlled prospective studies demonstrating that persons who receive screening experience better outcomes than those who are not screened. No such studies have ever been published, although there is some evidence that patients with advanced curves may be more likely to fail treatment (to progress further or undergo surgery) than patients with smaller curves.[36]

The effectiveness of screening has been inferred from temporal studies that compared outcomes in local communities before and after the institution of large screening programs. These studies reported an increase in the number of referrals to local scoliosis clinics, the proportion of curves detected by screening, and the use of braces; they also reported a decrease in the mean age of referred cases, mean curve size, number of curves progressing to 40°, proportion of cases requiring treatment, and the rate of

spine fusions.[37–40] However, most of the studies provided limited information about the comparability of the "before" and "after" groups, making it difficult to determine whether the time trends were due to screening or to other temporal factors.

The rationale behind screening is the assumption that the early detection of curves permits prompt initiation of conservative therapeutic measures that may prevent progression of the curves and thereby avoid the complications of advanced scoliosis. The principal forms of conservative treatment for curves detected through screening include spinal orthoses (braces), electrical stimulation, and exercise therapy. Surgery may also be recommended for cases detected through screening, and it is argued that early surgery for large curves may produce better outcomes than surgery performed at later ages.

Braces are generally effective in providing immediate correction of curves; initial standing roentgenograms often demonstrate a 50–60% correction in the curve.[41] The effectiveness of braces in preventing progression is less certain. There have been no published controlled prospective studies establishing the effectiveness of brace treatment. A multicenter prospective controlled trial of brace therapy has recently been completed,[42] but the results were not published as of this writing. Most existing evidence regarding the effectiveness of brace therapy comes from uncontrolled case series reports. Early series with limited follow-up reported corrections in lateral curvature of as much as 50%. Although gradual loss of correction over the course of treatment was noted, follow-up 1–2 years after discontinuing brace treatment revealed significant improvement over pre-brace values in a large proportion of patients.[43] Mean rates of curve progression in braced patients were lower than rates expected from natural history data.[44] Long-term studies (more than 5 years of follow-up) have since demonstrated that the early post-treatment correction observed in these reports was often temporary. A gradual loss of correction was noted in the years following brace treatment, with mean overall improvement in such studies averaging 0–4° compared with pre-brace values.[45–47]

The absence of internal controls in most bracing studies limits inferences about the independent effects of braces on outcomes. Some investigators have relied on historical control groups to infer effectiveness. A recent review of over 1000 braced patients, for example, concluded that braces altered the natural history of the disease because treatment failures were significantly less common in this series than was observed in a 1984 study by the same authors.[48] A retrospective review that did include a control group of matched, untreated patients reported that braced patients had a lower rate of curve progression and a higher rate of curve regression than untreated patients.[49] The differences were not statistically significant, but the study may have lacked adequate sample size to detect a difference.

Another controlled study of similar design reported no statistically significant differences in any parameter of curve progression but also had a small sample size.[50]

Outcome measures in most bracing studies relate only to curve correction and provide little information on health outcomes (e.g., back pain, patient feelings about their appearance, psychosocial impact). Available evidence is limited to an uncontrolled study, which found that braced patients noted an improvement in back "surface shape," as determined by a computerized photogrammetric surface mapping procedure.[51] Compliance problems limit the effectiveness of brace treatment.[52] Braces are generally recommended to be worn for 23 hours/day, a program that is often difficult for adolescents to follow and that influences compliance.[53] One study reported that only 15% of patients were highly compliant and that patients wore their braces an average of 65% of the recommended time.[54] Another study reported that complaints were uncommon among adolescents who wore braces.[55]

Lateral electrical surface stimulation (LESS), in which surface electrodes are applied to the skin nightly for at least 8 hours until skeletal maturity is attained,[56] has only been evaluated in uncontrolled case series reports. Although early case series reports found low rates of progression (0–5%) in patients who received LESS,[57,58] subsequent studies found that 18–56% of patients progressed more than 10°.[59,60] A chart review of patients who had completed treatment with LESS and were fully compliant found that over two thirds of curves progressed at least 5°; 50% of the patients required fusion or ended treatment with a curve greater than 40°.[61]

Exercises have been advocated as prophylactic therapy to prevent the need for more extensive treatment (e.g., braces) and as adjunctive therapy to enhance the effectiveness of braces.[62] Scientific evidence to support either use of exercise therapy is limited. Exercise alone has historically demonstrated poor effectiveness in preventing curve progression,[62,63] although there have been few published studies in this area. A study of a school-based exercise program for adolescents with scoliosis found that curve progression after 1 year was not significantly different between the study group and a matched control group.[63] Supporting evidence includes a small randomized controlled trial (grade I evidence) of adolescents wearing a cast, which showed that exercise was more effective than traction in improving curves on lateral bending;[64] an uncontrolled cohort study that showed improved vital capacity in hospitalized scoliosis patients who received physiotherapy;[65] and an uncontrolled case series report, which found that some braced patients who performed a thoracic flexion exercise had reduced vertebral rotation and thoracic curves after exercise.[66] The study lacked controls, follow-up, and an assessment of clinical outcomes.

Surgery is generally not considered unless significant progression has occurred. Few clinical trials have compared surgery with no surgery to assess its efficacy; case series reports provide the largest body of evidence. In these studies, Harrington instrumentation and other surgical techniques appear to be effective in correcting scoliotic curves in the frontal plane—Cobb angles are corrected by 40–70%[67–71]—but thoracic hypokyphosis, deviations in axial rotation, and lordosis are often not corrected.[72] Reduced lumbar lordosis ("flat-back" deformity)[73] and "crankshaft" deformities (in skeletally immature patients with posterior arthrodeses)[74,75] can develop over time, although modifications in devices and techniques have reduced the risk of these complications. A small improvement in pulmonary function has also been reported.[76] Cotrel-Dubousset instrumentation appears to achieve correction in the frontal plane while maintaining normal sagittal contour, and some correction of axial rotation with improved cosmesis has also been reported.[77] Spinal decompensation due to torsional changes and spinal cord damage are potential complications of Cotrel-Dubousset instrumentation.[78,79] Another limitation to surgical techniques is loss of fixation, which can result in partial or total loss of correction. There is an estimated 10–25% loss of correction from Harrington instrumentation, but the risk may be lower in patients who are immobilized by a cast or brace.[80,81] Loss of correction appears to be uncommon with Cotrel-Dubousset instrumentation (loss of correction less than 2%), and the latter does not require immobilization.[82]

Few controlled studies have evaluated surgery in terms of clinical outcomes, such as back pain and functional status. Although spinal curves and axial rotations are influenced by surgery, they do not correlate well with the incidence of back pain or other symptoms.[83] Studies that have demonstrated effects on clinical outcomes have suffered from design limitations. An uncontrolled retrospective study of patients who underwent spinal fusion found that complaints of low back pain were lower than reported rates in the general population and in scoliotic patients who do not receive fusion.[84] This study did not include internal controls, and it was performed in the years before spinal instrumentation was introduced. Similarly, a review of 32 patients who underwent fusion reported that the preoperative prevalence of poor self-image (38%), uncomfortable sexual intercourse (35%), and frequent or constant back pain (53%) had decreased to zero when surveyed 24–50 months after surgery.[85] This study also lacked a control group. A retrospective cohort study found that surgically treated patients were less likely than nonsurgically treated patients to report pain and were more likely to be performing manual work.[86] The study and comparison groups were not selected randomly and there were important differences between groups in preoperative characteristics. A survey found that patients who had undergone Harrington instrumenta-

tion differed significantly from persons without scoliosis in terms of employment, activity levels, and complaints of back pain.[87] The control group did not consist of persons with scoliosis who did not receive surgery, and thus it is unclear whether observed differences were due to scoliosis or to the effects of surgery.

The adverse effects of screening itself are generally minor, but follow-up testing of abnormal findings may incur anxiety, inconvenience, work and school absenteeism for return visits, financial costs for visits and radiographic tests, and radiation exposure from follow-up roentgenograms (although roentgenograms are not routinely ordered on all follow-up evaluations and, when obtained, radiation exposure can be reduced by modern imaging and shielding techniques). Confirmed or suspected scoliosis may affect future health insurance and work eligibility. These postulated adverse effects have not been proven in controlled studies. Treatment may also incur adverse effects from follow-up visits (e.g., inconvenience, absenteeism, radiation exposure) and from treatment itself. Brace wear, for example, may produce skin irritation, disturbed sleep, restrictions on physical and recreational activities, and difficulty in finding clothes, but studies confirming these effects are lacking. Studies have shown an association between brace wear and adverse psychological effects, diminished self-esteem, and disturbed peer relationships.[88,89]

The potential adverse effects of surgery can include the general risks of surgery, such as anesthesia risks, pain, and postoperative complications (e.g., bleeding, infection, pulmonary embolism), although these have been reduced by modern surgical and anesthetic techniques.[90] The overall risk of spinal cord damage is about 1–3%,[71,91] but rates are thought to be lower in uncomplicated surgery or when somatosensory evoked potential spinal cord monitoring is performed.[92] Fusion at certain ages during adolescence may affect the longitudinal growth of the spine.[93] Hook dislodgement and laminar fracture are possible. Other adverse effects of surgery include financial costs, inconvenience and lost productivity associated with hospitalization and convalescence, and external immobilization with casts or braces, which may be required for a period of months after surgery. Potential long-term complications occur generally in adults and include the development of pain caudad to the level of fusion, bursitis, pseudo-arthrosis, kyphotic deformities, and loss of normal lumbar lordosis.[91,94] Often these complications require further surgery during adulthood.

Recommendations of Other Groups

The Scoliosis Research Society has recommended annual screening of all children aged 10–14 years.[95] The American Academy of Orthopedic Surgeons has recommended screening girls at ages 11 and 13 years and

screening boys once at age 13 or 14 years.[96] The American Academy of Pediatrics has recommended scoliosis screening with the forward bending test at routine health supervision visits at ages 10, 12, 14, and 16 years; this recommendation is under review.[97] The Bright Futures guidelines recommend noting the presence of scoliosis during the physical examination of adolescents and children ≥8 years of age.[98] The Canadian Task Force on the Periodic Health Examination concluded that there was insufficient evidence to make a recommendation.[99] Scoliosis screening is required by law in some states.[100]

Discussion

The clinical logic behind screening for adolescent idiopathic scoliosis is based on a series of critical assumptions. The logic assumes that screening tests are accurate and reliable in detecting curves, that early detection of curves results in improved health outcomes, and that effective treatment modalities are available for cases detected through screening. Implicit in this causal pathway are the assumptions that small curves detected through screening are likely to progress to curves of potential clinical significance, that scoliosis causes important health problems, and that the benefits of early detection outweigh the potential adverse effects of screening and treatment. Scientific evidence to support these assumptions is limited.

The principal screening test for scoliosis, the physical examination of the back, has variable sensitivity and specificity, depending on the skills of the examiner and the size of the curve being sought. The positive predictive value in typical screening settings is low, due to the low prevalence of clinically significant curves. There is little evidence about the incremental value of repeat screening in children with previously normal results.

There have been no controlled studies to demonstrate whether adolescents who are screened routinely for idiopathic scoliosis have better outcomes than those who are not screened. Decreased curve size and surgery rates have been observed in communities that have adopted aggressive screening programs, but it is unclear whether the changes were due to screening or to other temporal factors. Beyond temporary correction of curves, there is inadequate evidence that braces limit the natural progression of the disease. The effectiveness of LESS and exercise has not been demonstrated convincingly in currently available research. Surgery is effective in reducing, but not eliminating, the lateral scoliotic curve. The scoliotic curves for which surgery is recommended (e.g., documented progression beyond 40–50°) are more likely to be detected without screening.

The natural history of idiopathic scoliosis is such that most cases detected at screening will not require treatment because they will not

progress significantly. Indications for preventive treatment (e.g., braces) are therefore uncertain and can result in unnecessary treatment. Only a small proportion of adolescents with idiopathic scoliosis are currently considered candidates for treatment (e.g., those having progressive curves greater than 30°). Moreover, the burden of suffering associated with adolescent idiopathic scoliosis is uncertain. Cosmetic deformities and associated psychological and social effects have been difficult to evaluate in formal research. It is also unclear whether physical symptoms can be attributed to idiopathic scoliosis, except in severe cases. Finally, screening may result in mislabeling and the inconvenience, cost, and potential radiation exposure of follow-up evaluations. Both conservative treatment (e.g., braces) and surgery can be associated with medical, psychological, and social adverse effects.

In summary, there is insufficient evidence from clinical research that routine screening is effective in changing the outcome of adolescent idiopathic scoliosis. Limitations in the design of existing studies, however, also make it difficult to conclude that screening is ineffective or harmful. If screening for scoliosis is effective, discontinuation of school screening may have a disproportionate impact on disadvantaged adolescents. Adolescents who have access to primary care providers and to periodic health examinations have an opportunity outside the school setting to obtain back examinations. School screening may provide the only opportunity for back inspections of disadvantaged adolescents, including those from minority and low income families, who often lack access to such providers.

CLINICAL INTERVENTION

There is insufficient evidence to recommend for or against routine screening of asymptomatic adolescents for idiopathic scoliosis (“C” recommendation). The evidence does not support routine visits to clinicians for the specific purpose of scoliosis screening or for performing the examination at specific ages during adolescence. It is prudent for clinicians to include visual inspection of the back of adolescents when it is examined for other reasons. Additional specific inspection maneuvers to screen for scoliosis, such as the forward-bending test, are of unproven benefit.

Note: See the relevant background papers: U.S. Preventive Services Task Force. Screening for adolescent idiopathic scoliosis: policy statement. JAMA 1993;269:2664–2666; and U.S. Preventive Services Task Force. Screening for adolescent idiopathic scoliosis: review article. JAMA 1993;269:2667–2672.

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