## **Review Article**

Scheuermann's Kyphosis: Diagnosis, Management, and Selecting Fusion Levels

## Abstract

Scheuermann's kyphosis (SK) is a rigid structural deformity of the thoracic spine defined radiographically as three or more contiguous vertebrae with at least 5° of wedging anteriorly. Prevalence of the disease is thought to be between 0.4% and 10%. The true cause of SK remains unclear; however, various theories include growth irregularities, mechanical factors, genetic factors, and/or poor bone quality as the causes. Patients with mild disease (less than 70°) generally have a favorable prognosis with good clinical outcomes. Most patients with SK are successfully treated nonsurgically with observation, anti-inflammatory medications, and physical therapy. Surgical intervention is indicated in patients with greater than 70° to 75° thoracic curves, greater than 25° to 30° thoracolumbar curves, intractable pain, neurologic deficit, cardiopulmonary compromise, or poor cosmesis. Because of advances in posterior spinal instrumentation, surgery can typically be performed through a posterior-only approach. When surgical treatment is planned, appropriate selection of the upper- and lower-instrumented vertebrae is important to achieve a wellbalanced spine, preserve motion segments, and reduce the risk of junctional kyphosis.

**C** cheuermann's kyphosis (SK) is a **O**rigid spinal kyphosis affecting the mid-thoracic or thoracolumbar spine, which was first described by Holger Werfel Scheuermann in 1920.<sup>1,2</sup> The condition is associated with anterior wedging of the vertebrae, end plate irregularities, and Schmorl's nodes.<sup>3</sup> In 1964, Sorensen<sup>4</sup> was the first to define SK radiographically by the presence of at least three adjacent vertebrae wedged a minimum of 5°. Prevalence of the disease is thought to be between 0.4% and 10%,4,5 affecting men and women equally.<sup>6</sup> The etiology of SK remains unclear; however, multiple theories have been proposed. Scheuermann<sup>1</sup> believed that osteonecrosis of the vertebral ring apophysis resulted in longitudinal growth arrest of the anterior vertebral body, thus causing a wedging of the vertebrae. Schmorl postulated that disk material herniated through the vertebral end plates lead to loss of disk height, vertebral body wedging, and node formation.<sup>3</sup> Both these early theories have since been called into question. Some authors noted a familial predilection for the disease, including a high rate of heritability and an autosomal dominant pattern, suggesting possible genetic causes.<sup>4</sup> Growth hormone abnormalities have also been implicated as a causative factor; however, the true cause of the disease remains unknown.

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## Classification

SK can be classified into two distinct groups: typical and atypical. In the more common typical SK, the apex of the deformity is usually in the mid-thoracic spine (T7-T9) and Sorensen's criteria (greater than three adjacent vertebrae wedged  $5^{\circ}$  or more) are met. In atypical SK, the apex of the deformity is frequently in the thoracolumbar or lumbar spine. Classic radiographic findings such as disk space narrowing, end plate changes, and Schmorl's nodes are still present, but Sorensen's criteria are not always met.<sup>2</sup>

### **Natural History**

The natural history of SK tends to be benign. In patients with a smaller degree of kyphosis (less than  $60^{\circ}$ ), good clinical outcomes can be expected.<sup>6</sup> Murray et al<sup>6</sup> followed 67 patients for 32 years. For patients with curves less than 85°, the authors found no differences in the number of days absent from work because of back pain, extent that the pain interfered with activities of daily living, self-consciousness, self-esteem, or level of recreational activities. Also, the patients reported little preoccupation with their physical appearance. Patients with SK had normal or above normal pulmonary function overall. Only in patients with severe kyphosis of greater than 100° was pulmonary function affected negatively. Patients with SK did report more spine tenderness and limited thoracic extension on physical examination. They

also pursued jobs that required less strenuous physical activity. Ristolainen et al7 investigated 19 untreated patients with mild thoracic SK, with a mean follow-up of 46 years. The authors found that the degree of radiographic deformity increased only slightly during longterm follow-up. Over the course of the study, the mean thoracic kyphosis increased from 46° (range,  $25^{\circ}$  to  $78^{\circ}$ ) at baseline to  $60^{\circ}$  (range,  $34^{\circ}$  to  $82^{\circ}$ ) (*P* < 0.001), and the mean of the vertebrae wedge angle increased from 8.8° to 9.9° at final follow-up (P = 0.046). No correlation existed between the extent of kyphosis progression and the function at final follow-up. In severe kyphosis, neurologic complications have been reported in a small number of patients.8

#### Nonsurgical Treatment

In contrast to idiopathic scoliosis, in which most surgeons agree on threshold ranges for surgical intervention, clinical equipoise still exists on when to intervene in SK. Conventionally, most clinicians have treated curves less than 50° to 80° nonsurgically.9-11 Hard indications for surgical intervention are still being investigated in the literature.<sup>9</sup> There is a paucity of data to support bracing treatment of SK, and the effect of bracing treatment on the natural history and progression of disease remains unknown.12,13 In addition, all available studies investigating bracing treatment in SK have been small, retrospective, and limited to level IV evidence. Criteria

for bracing treatment include smaller and more flexible curves (ie, curves less than  $55^{\circ}$  to  $80^{\circ}$ , with passive correction of 40% or more). Some authors supported bracing treatment in immature patients, with the goal of vertebral body remodeling.<sup>13,14</sup> Brace wear is recommended for 16 to 23 hours per day until apical wedging is corrected. Riddle et al<sup>14</sup> reported that initial bracing treatment can achieve an almost 50% reduction in kyphosis in many patients, but some loss of correction occurred after termination of brace treatment. Another paper by Sachs et al reported on 120 patients with SK treated with a Milwaukee brace. Of the patients who were compliant with brace wear, 76 patients displayed improvement in their kyphosis, 10 were unchanged, and 24 demonstrated some worsening. Seven of the 24 patients who worsened went on to require surgical intervention.

Regardless of brace treatment, a formal exercise program that emphasizes thoracic extensor strengthening and endurance can be helpful. Weiss et al reported on long-term results of physical therapy, osteopathy, manual therapy, an exercise program, and psychological therapy for a group of 351 patients. At the end of the treatment regimen, male and female patients both reported a statistically significant reduction in pain.<sup>15</sup>

## Surgical Management

Indications for surgical intervention include progressive kyphosis despite brace compliance, neurologic deficit, persistent pain, or notable deformity in a skeletally mature individual.

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Measurements of 55° to 80° of kyphosis have been used as a threshold for surgical intervention; however, a hard curve magnitude threshold is not currently supported in the literature.9,10 Polly et al9 and the Spinal Deformity Study Group recently reviewed outcomes of 150 patients from various centers with SK treated surgically and nonsurgically. Their results confirmed the notable variability in treatment decisions among various surgeons. Mean curve magnitude in the surgical population was 73° versus 70° in the nonsurgical group. Maximal Cobb angle was not found to be a notable predictor for surgical intervention or patient-reported outcome scores. The authors did note that surgically managed patients were older, had more pain, had lower selfappearance scores, and had higher body mass index than those treated nonsurgically. Overall, the authors concluded that pain and patient dissatisfaction played important role in determining when to proceed with surgical intervention.

# Surgical Approach and Technique

The surgical management of SK has evolved greatly over time, particularly with the advent of segmental spinal instrumentation and ultimately pedicle screw (PS)-based posterioronly approaches. Harrington compression instrumentation was initially used in the management of SK. Bradford et al<sup>5</sup> reported on 22 patients treated with this technique; the authors observed a notable loss of correction postoperatively. The reason for failure was likely because of the posterior-based tension-sided fusion and lack of anterior column support. The authors ultimately advocated for a combined staged AP approach to minimize this complication. In their next series, using anterior discectomies and interbody fusions through thoracotomy, only 5 of 24 patients experienced loss of correction of more than 10°.<sup>11</sup> Luque and Cotrel-dubousset instrumentation was also implemented historically in the management of SK. However, particularly high rates of junctional kyphosis plagued these techniques.<sup>12,16</sup>

Herndon et al<sup>17</sup> studied the results of a combined approach by evaluating 13 patients who underwent anterior release and fusion, followed by posterior fusion. The authors reported an average correction of 51° and that 12 of 13 patients had good pain relief. Lowe and Kasten<sup>16</sup> similarly noted good results in patients who underwent staged AP spinal fusion. Yang et al<sup>18</sup> showed 16 patients who underwent a similar AP approach to have had adequate initial correction and good maintenance of correction at follow-up. In one of the largest retrospective reviews of surgically treated SK, Lonner et al<sup>10</sup> noted a markedly higher overall complication rate with combined AP surgery compared with a posterior-only approach (23.8% versus 5.5%). Most of these complications were approach related (such as pneumothorax and pulmonary effusions) or related to the magnitude of the procedure; patients who underwent combined approaches had markedly greater preoperative Cobb angles.

In 39 well-matched patients, Lee and Lenke compared a combined AP versus a posterior-only approach using PS fixation with rods. The authors found that the posterior-only surgery provided superior results with regard to surgical time, blood loss, and overall complication rate (0.00% versus 38%). Equivalent results in terms of kyphosis correction and SRS-30 outcome measures were reported. Koptan et al<sup>19</sup> also reported on PS-only techniques; the authors compared a PS-only approach with a hybrid construct using hooks, PSs, and sublaminar wires. Although this was a relatively small series of 33 subjects, they found that patients treated with PS only had greater percentage correction, less loss of correction, and statistically significant decrease in estimated blood loss and surgical time.

In a more recent report, Koller et al<sup>20</sup> investigated a combined AP versus a posterior-only approach in 92 well-matched patients. The authors primarily investigated radiographic parameters and found that AP and posterior-only approaches both averaged similar degrees of final correction. The authors concluded that satisfactory clinical results can be obtained with either procedure; however, in select patients with severe deformity, posteriorthree-column osteotomies based may be required to achieve adequate correction if the surgeon chooses a posterior-only approach. Therefore, the morbidity of these osteotomies should be balanced against the potential complications associated with anterior releases. Ponte and Shufflebarger also investigated a posterior-only surgical approach. The authors argued against the need for anterior releases and that successful outcomes can be obtained with a posterior-only approach using primarily PS anchors and posterior column osteotomies. Seventeen patients were treated with the procedure, with a mean correction of 9.3° per osteotomy site and an overall correction of 49%. They reported only two minor complications.<sup>21</sup>

In our practice, multilevel posterior column osteotomies (Schwab type 2 or Ponte Osteotomies)<sup>22</sup> generally are used across the apex of the deformity to improve spinal flexibility before correction; these osteotomies can afford approximately 5° to 10° of correction per level.<sup>23</sup> For sharp angular and more rigid curves, advanced three-column osteotomy



Case 1: 18-year-old male patient with thoracic kyphosis that is accentuated by bending forward.

techniques with higher grades of resection may be required; these osteotomies involve resecting the pedicles (Pedicle Subtraction Osteotomy, Schwab types 3 to 4) and/or parts of the vertebral body along with the adjacent disks (Vertebral Column Resection, Schwab types 5 to 6).<sup>20</sup> The sagittal deformity angular ratio (DAR) is the maximum kyphotic angle divided by the number of vertebral levels involved.24 The DAR can be used to differentiate a sharp angular kyphosis from a smooth kyphosis that is spread over multiple levels and can help determine the potential need for advanced osteotomies. A patient with a high DAR may be better served with a three-column spine shortening

osteotomy than with posterior column osteotomies. The DAR also strongly correlates with the risk of intraoperative spinal cord monitoring alerts, with up to 75% incidence of a motor-evoked potential alert for a sagittal DAR greater than  $22^{\circ}.^{24}$ 

In terms of implant choice, an overall lack of consensus exists in the literature. Number and type of fixation points, rod material and its diameter, and the most appropriate corrective techniques are all areas of debate. Some authors recommend a minimum of eight fixation points above and below the apex of the kyphotic deformity.<sup>19</sup> We recommend cobalt chromium or stainless steel rods measuring at least 6.0 mm in diameter. Smaller, less rigid rods have a tendency to loose correction over time in patients with kyphosis. However, in general, the use of larger and stiffer implants should be balanced against the potential risk of junctional kyphosis.

# Level Selection and Junctional Kyphosis

No matter the technique implemented, junctional kyphosis has been a major issue for surgeons treating SK and has been reported in multiple series.<sup>11,25,26</sup> Reinhardt and Bassett reported a 40% rate of junctional loss of correction between the fused and unfused segments. Lowe and Kasten<sup>16</sup> also noted an

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incidence between 20% and 30%. In their series, proximal junctional kyphosis (PJK) was found to be related to obtaining greater than 50% correction at the time of surgery. The authors also noted that overcorrection and PJK can be avoided if the postoperative kyphosis remains greater than 40°. More recently, Lonner et al reviewed 78 patients treated with either combined AP or posterior-only spinal fusions. The authors noted a 32% rate of PJK and a 5.1% rate of distal junctional kyphosis (DJK).<sup>10</sup> Junctional kyphosis was defined as a Cobb measurement of greater than 10° between the fused and unfused segments. Other authors reported DJK in up to 28% of patients.<sup>27</sup> In Lonner's report, a larger magnitude of kyphosis both before surgery and at final follow-up tended to be associated with the development of PIK. PIK was also found to be directly correlated with the magnitude of pelvic incidence, that is, the greater the pelvic incidence for the individual, the higher the magnitude of PJK. Importantly, patients who were fused at or proximal to the Cobb end vertebra were markedly less likely to develop PJK.

Figure 2

Various techniques have been recommended to avoid junctional problems. Proximally, a general consensus exists in recommending the inclusion of the proximal end vertebra in the construct.<sup>27</sup> However, some surgeons argue that the construct should extend to one neutral vertebra above the end Cobb vertebra. Careful analysis of preoperative radiographs and diligent surgical planning are critical to ensuring the surgeon is selecting the appropriate level. It is important to obtain highquality upright and hyperextension radiographs, and close attention should be given to the upper end of the intended construct.

In addition to appropriate level selection, other techniques have been



Case 1: Radiographs showing thoracic kyphosis measuring 90°. L2 is the sagittal stable vertebra as it is the most proximal vertebra touched by the posterior sacral vertical line (red line).

used to avoid PJK. It is paramount to ensure appropriate maintenance of the facet joints, soft tissue, and ligamentous structures between the fused and unfused segments. It is important to preserve the interspinous ligament between the fused and unfused segments. Denis et al<sup>27</sup> pointed to disruption of the junctional ligamentum flavum (in addition to other factors) as a possible cause of PIK in their series. Some have advocated for a "soft landing" with hooks or wires at the top of the PS construct or ligament augmentation with tape or allograft. Biomechanical studies also confirmed a more gradual transition to normal motion when hooks, ligament augmentation, or tethers are used.<sup>28</sup> Hassanzadeh et al<sup>29</sup> compared hooks with PSs at the upper instrumented vertebra (UIV) in adults treated with long posterior spinal fusion. PJK did not develop in any of the patients treated with hooks versus 29.6% of patients in the PS group, and patients with hooks were noted to have markedly



Case 1: Postoperative radiographs showing instrumentation from T2 to L2, with kyphosis measuring  $50^{\circ}$ .

higher SRS-22 functional scores at final follow-up. In adolescent idiopathic scoliosis (AIS), using hooks at the top of the construct has been shown to reduce the rate of PJK as well.<sup>30</sup> However, in Lonner's SK series, proximal anchor type (screw versus hook) had no effect on the development of PJK.<sup>10</sup> Last, the use of transition rods may potentially transmit stress more gradually between the fused and unfused segments.<sup>31</sup> Both Koller et al and Lonner et al noted a relationship between global spinopelvic morphology and PJK.<sup>10,32</sup> Their data showed that among patients in whom junctional kyphosis had developed, increasing pelvic incidence correlated with increasing magnitude of PJK.<sup>15</sup> In addition, Lowe and Kasten<sup>16</sup> found that patients with SK tend to be in negative sagittal balance, and this may become further negative with surgery, particularly if the patient is overcorrected, thus predisposing them to junctional kyphosis. Although further research is needed in this area, restoration of proper sagittal alignment and global spinopelvic alignment is paramount to ensuring good clinical outcomes. Overcorrection of the kyphotic deformity should be avoided in general, and particularly in the setting of high pelvic incidence; the unfused spinal segments may not be able to adjust for postoperative changes to achieve appropriate overall alignment.<sup>32</sup>

Until recently, the most appropriate lowest instrumented vertebra (LIV) was thought to be the level below the first lordotic disk (FLD).16,33 The FLD is defined as the most proximal thoracolumbar or lumbar disk below the level of the kyphosis with  $\geq 5^{\circ}$  of anterior opening. Because of hyperlordosis in the lumbar spine in patients with SK, determining the FLD can sometimes be difficult, thus leading to fusion short of the true FLD and subsequent DJK. Cho et al<sup>34</sup> reported on using the sagittal stable vertebra (SSV) instead of the FLD. The stable vertebra is defined as the most proximal touched vertebra by the posterior sacral vertical line. The authors noted a markedly decreased incidence of DJK (4% versus 71%) compared with patients whose constructs ended at the FLD (which typically is more proximal than the SSV). However, it is important to note that patients in this series were treated with hook instrumentation at the LIV, which may be an independent risk factor for DJK.<sup>27</sup> This may explain the high incidence of DJK in their patients fused to the FLD. In an attempt to avoid screw pullout and subsequent DJK, many surgeons now opt for longer PSs at the LIV to obtain maximum vertebral body purchase and fixation into the anterior column.

Using a PS-only posterior construct, Kim et al<sup>35</sup> investigated the incidence of DJK in patients who were fused at or below the SSV versus above the SSV. Patients who were fused at or below the SSV were found to have markedly greater lordotic disk angles below the LIV and lower revision surgery rates for DJK (5% versus 36.3%). The authors concluded that the SSV method may reduce complications secondary to DJK, such as screw pullout, loss of fixation, and/or sagittal decompensation, but at the expense of incorporating additional motion segments, which can be worrisome, especially in a relatively young patient population. Conversely, Yanik et al<sup>36</sup> found no difference in the incidence of DJK in patients based on LIV choice. Patients who were instrumented to the SSV and the FLD had similar radiographic measurements and clinical outcome scores at most recent follow-up. The authors concluded that it is not necessary to extend the fusion down to the SSV, thus sparing motion levels.

Based on our experience, we recommend using the SSV concept for distal level selection. If the selected SSV is just barely touched by the sacral vertical line, the adjacent disk space should be evaluated further. If the proximal disk space is lordotic, the "barely touched" SSV is still a safe choice. After the distal fusion level is selected, we recommend maintaining symmetry of the construct overall. The fusion should extend roughly the same extent from the apex proximally and distally, with some consideration for adding one additional proximal fusion level to ensure that the proximal end vertebra is also included in the fusion construct. For example, if the SSV is L2 and the apex of the kyphosis on upright radiograph is T8, the construct should start at the level of T2.37

# **Complications**

In a retrospective review, 693 surgically treated patients with SK were evaluated

through the Scoliosis Research Society morbidity and mortality database.<sup>38</sup> The investigators reported an overall complication rate of 14%. Complications were markedly more common in adult patients. The overall incidences





Case 2: Radiographs showing thoracic kyphosis measuring  $82^{\circ}$ . L1 is just touched by the posterior sacral vertical line (red line).

of complications associated with the posterior, anterior, and same-day AP procedures were 14.8%, 4.1%, and 16.9%, respectively. These data are in stark contrast to the paper by Lee et al<sup>39</sup> in which the posterior spinal fusion-alone group displayed a mark-edly lower complication rate compared with the patients in combined AP approach (0.00% versus 38%). The most common complication in the Scoliosis Research Society (SRS) study was wound infection at 3.8%. Acute neurologic complication rate was

1.9%, which included four documented spinal cord injuries (0.6%), one of which was complete (0.15%). Four patients (0.6%) died of surgical complications, including sepsis, pulmonary embolism, and cardiorespiratory failure. More recently, Lonner et al<sup>40</sup> reviewed data from the Harms Study Group. The authors reported a 16.3% major complication rate in SK, with a revision surgery rate of 14.4%. Again, the most common complication was wound infection (10.3%), followed by instrumentation-related

complication (3.1%) and neurologic complications (2.1%).

## Intraoperative Neuromonitoring

neuromonitoring Intraoperative (IONM) has become increasingly prevalent in spinal deformity surgery over the past several decades, and multimodal monitoring should be implemented during surgical correction of SK. Cheh et al41 reported on loss of IONM signals in 42 pediatric patietns with kyphosis treated with posterior column osteotomies and fusion. Fourteen patients in their cohort carried a diagnosis of SK, of which five had a true positive loss of IONM signals. Signals returned in all five patients after partial or complete release of correction and optimization of blood pressure. No patient sustained a neurologic deficit. Kundnani et al42 reported a sensitivity of 100%, a specificity of 98.5%, and a positive predictive value of 85% when multimodality neuromonitoring was used for the surgical management of AIS. Buckwalter et al investigated IONM in a large cohort of patients with idiopathic scoliosis. They reported a positive IONM alert incidence of 3.6%, with 0.3% of patients waking up with a deficit. In another more recent paper, Samdani et al43 reviewed 676 patients with AIS treated with spinal fusion. Overall, 5.3% of their cohort experienced an IONM alert. Importantly, no three-column osteotomies were performed in this cohort. Overall, there is a paucity of data on spinal cord neuromonitoring in SK, and its specific use in the management of this disease is an area in need of further investigation.

# **Case Examples**

## Case 1

An 18-year-old male patient presented with severe kyphosis and

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moderate apical thoracic back pain. His physical examination revealed thoracic kyphosis that was accentuated by bending forward (Figure 1). Radiographs confirmed a diagnosis of thoracic Scheuermann's kyphosis measuring 90° from T3 to T12 with apex at T9 to T10 (Figure 2). Posterior spinal instrumentation and fusion from T2 to L2 was planned. Including the first distal lordotic disk in the fusion would lead to choosing L1 as the LIV. However, L2 was found to be the SSV (Figure 2) and was therefore selected as the LIV. While the upper end vertebra for the kyphosis was T3, the UIV was selected as T2 to include the proximal FLD in the construct. Multiple posterior column osteotomies were used intra-op from T6 to T12. After placing the PSs, 6.0 mm cobalt chrome alloy rods were engaged into the proximal screws and a cantilever maneuver was used to reduce the rods into the distal screws. Postoperative thoracic kyphosis measured 50° (Figures 3 and 4).

## Case 2

A 17-year-old male patient presented with gradually progressive thoracic kyphosis and mild back pain. Imaging revealed a Scheuermann's 82° kyphosis from T3 to T12 with a more proximal apex at T7 to T8 (Figure 5). Posterior spinal instrumentation and fusion from T2 to L1 was performed. The SSV, L1, was just barely touched by the posterior sacral vertical line. A closer look at the imaging revealed that the T12-L1 disk was lordotic; therefore, L1 was chosen as the appropriate LIV. The UIV was selected as T2 to maintain a symmetrical fusion from the apex and to include the proximal FLD in the construct. Posterior column osteotomies were performed from T5 to T11. The intraoperative correction maneuver performed was similar to that in case 1 to yield a postoperative kyphosis of  $45^{\circ}$  (Figure 6).



Case 2: Postoperative radiographs showing instrumentation from T2 to L1, with kyphosis measuring 45°.

#### Summary

Surgical intervention in SK is indicated in patients with deformity greater than  $70^{\circ}$  to  $75^{\circ}$ , intractable pain, neurologic deficit, cardiopulmonary compromise, or poor cosmesis. Thanks to advances in posterior spinal instrumentation, surgery is now typically able to be performed through a posterior-only approach. However, junctional kyphosis remains a challenging problem. It is paramount to ensure appropriate maintenance of facet joints and soft tissue and ligamentous structures between the fused and unfused segments. It is also recommended to limit correction to 50% or less of the original deformity. Although some controversy remains regarding fusion level selection, most surgeons agree that the construct should be symmetrical from the apex of the kyphosis and include the end Cobb vertebra proximally. Distally, some authors maintain that the LIV should be the level below the FLD, whereas others argue that the construct should extend further to the SSV. Instrumenting to the SSV may markedly reduce the incidence of distal junctional failure while typically incorporating just one

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additional motion segment into the fusion.

### References

*Evidence-based Medicine:* Levels of evidence are described in the table of contents. In this article, references 6, 9, 10, 19, 20, 26, 29, 30, 34-36, 39 are level III studies. References 1, 2, 5, 7, 8, 11, 14-16, 18, 21, 23-25, 27, 32, 33, 38, 40-42 are level IV studies. References 3, 17, 22 are level V expert opinion.

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