

Early onset scoliosis: Can Best Practice Guidelines Be Provided in Europe?


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Abstract

Background: The goal of treatment in early onset scoliosis patients is to control the deformity while allowing chest and spinal growth. However, management, either conservative or surgical, remains controversial and require further clinical evidence. The aim of this study was to determine best practice guidelines for the management of early onset scoliosis patients in Europe.

Methods: A Delphi approach was used among European experts to determine consensus in: (1) identification of progression risk factors, (2) clinical and radiographic evaluation, (3) conservative treatment, and (4) surgical management. Idiopathic and non-idiopathic groups were analyzed separately. Statements were only included in the final agreed consensus if at least 80% of respondents agreed with the statement.

Results: A total of 22 pediatric orthopedic surgeons completed the first and second surveys, and 21 the third. Out of open-ended questions included in the initial Delphi, 75 statements for 59 multiple-choice questions in the idiopathic group and 47 statements for the 44 multiple-choice questions for the non-idiopathic group were formed for the second round. Of the total 122 statements, 50 (40.9%) reached consensus, and 72 did not. Among the 72 statements re-evaluated in Round 3, consensus was reached on 69 (96%).

Conclusion: Early onset scoliosis represents a rare, heterogeneous, but life-threatening condition often associated with insufficient relevant evidence. Consensus is difficult to achieve (39.3% of the 122 statements), especially in the non-idiopathic group. However, 48 guidelines (33 for idiopathic and 15 for non-idiopathic) were provided by European experts to improve early onset scoliosis management.

Level of Evidence: IV

Keywords: Early onset scoliosis, idiopathic, syndromic, neuromuscular, congenital, conservative management, growth-friendly management

Introduction

Early onset scoliosis (EOS) is defined as a spinal deformity before the age of 10 years, often associated with a high progression risk if left untreated. The main etiologies are idiopathic, syndromic, neuromuscular, and congenital.¹ EOS is a very heterogeneous condition in terms of magnitude, curve location, and progression, but also sagittal alignment. The goal of treatment, either conservative or surgical, is to control the deformity while allowing chest and spinal growth, with the least possible negative consequences on patients' quality of life.

Due to the rarity of this condition, it is often difficult to provide evidence-based treatment guidelines. The standard of care, therefore, greatly varies among but also within countries, and even between surgical departments. In addition,

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physicians have a tendency to modify their management over time based on the literature and their personal experience. As a matter of fact, Matsumoto et al. have recently shown that the initial enthusiasm that existed with growth-friendly surgical techniques has declined, and that preferences for conservative treatment have increased over the last 10 years.² The spectrum of management strategies ranges from observation to casting, full or part-time bracing, and numerous growth-friendly surgical techniques (distraction-based, growth guidance, or compression-based). However, all these procedures remain associated with high rates of complications and unplanned returns to the operating room.³ Therefore, early spinal fusion is also an option in children with limited growth potential, such as some syndromic etiologies (skeletal dysplasia for example), or with severe neurologic co-morbidity (cerebral palsy).^{4,5}

When no high-quality clinical studies are available (or are even practical to perform), consensus recommendations by specialist organizations are a constructive way forward. The Delphi method is an interactive way for a panel of experts to reach consensus.^{6–8} It has the advantage that many individuals can contribute anonymously, without the process being dominated by a few individuals who may have particularly strong opinions on a topic. The aim of this study was, therefore, to provide best practice guidelines for EOS (idiopathic and non-idiopathic) from a group of European experts.

Materials and methods

The European Paediatric Orthopaedic Society (EPOS) spine study group identified the need to provide best practice guidelines for EOS management. Therefore, a three-round Delphi process was used to try to obtain consensus among 22 European experts in pediatric spine, representing 11 countries. The experts included 16 members of the EPOS Spine study group, and 6 non EPOS-members, identified and selected by vote by the study group members. All experts were highly specialized in pediatric deformity care, with more than 15 years of practice, and worked in academic environments (20 different departments) with access to the most advanced technologies. The steering committee consisted of three experienced pediatric orthopedic spine surgeons and a biostatistician expert on Delphi. The relevant literature was reviewed, and areas of controversy were established regarding (1) definition, (2) prognostic risk factors, (3) conservative treatment, (4) surgical management, (5) complications,

and (6) graduation. Surgeons were surveyed for current practices, presented with the results of the systematic review, and asked to vote anonymously for or against item inclusion during the three iterative rounds.

Delphi process

Round 1 questionnaire included open-ended questions (52 for idiopathic and 57 for non-idiopathic), and the respondents were asked to specify their treatment choices in different scenarios (Figure 1). Round 2 questionnaire included multiple-choice questions (MCQs; 59 for idiopathic and 44 for non-idiopathic) based on the data obtained in the first round. Questions were created as a “5-point Likert-type scale” (strongly agree, agree, neutral, disagree, strongly disagree) or had multiple answers. Threshold value for consensus was initially fixed as 80% at the second round. All the questions below this threshold value were asked again in the third round. In the third round, the response categories were given as “agree-neutral-disagree” in a three-point Likert-type scale, and previous answers from the group were visible. The classification of the consensus rates based on the responses received in the third round was as follows: $\geq 80\%$ consensus rate: strong agreement or disagreement, 70%–79% consensus rate: moderate agreement or disagreement, 50%–69% consensus rate: weak agreement or disagreement, and $< 50\%$ consensus rate: cannot be decided.

It is important to note that the response formats varied across the questions. While some questions were presented in an agree–disagree format, others required participants to select from predefined options. For the agree–disagree questions, the level of agreement was quantified and categorized as strong, moderate, or weak based on percentage thresholds. In contrast, for questions requiring a selection among categories, agreement was not applicable; instead, a selection rate was calculated.

The term clear categorization refers to these cases, where the distribution of responses indicated a consistent and unambiguous choice by the majority of participants.

Data collection and management

Questionnaires were submitted online to the respondents consecutively in each round. Study data was collected and managed using Research Electronic Data Capture

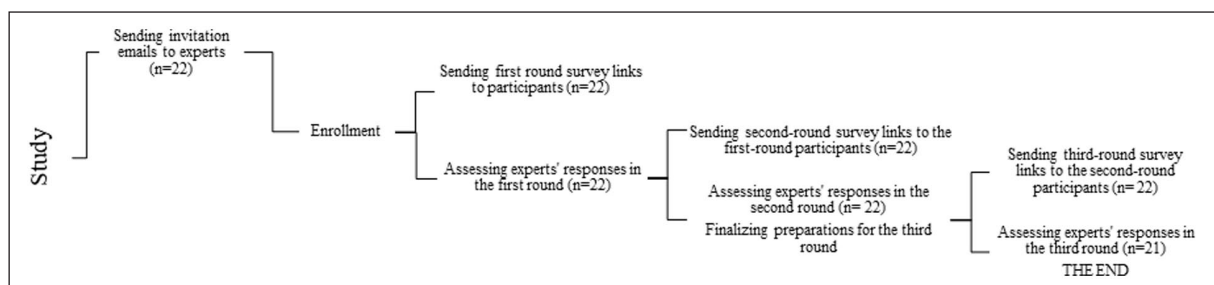


Figure 1. Flowchart on the Delphi method used in the current study.

Table 1. Statements regarding idiopathic EOS definition.

| Definition of idiopathic EOS | % of answers | Intensity of agreement/ disagreement | Round in which consensus established |
|--|--------------|--------------------------------------|--------------------------------------|
| The distinction between infantile and juvenile scoliosis is still relevant in idiopathic EOS | 80% agree | Strong | Third |
| MRI should be systematic before any surgery in the management of an idiopathic EOS patient without neurological symptoms | 86% agree | Strong | Second |
| Idiopathic EOS patients under 12 months-old represent a specific entity that should be managed differently than older patients | 91% agree | Strong | Second |
| Idiopathic EOS patients between 9 and 10 years of age (so called tweener) represent a specific entity that should be managed differently than younger patients | 80% agree | Strong | Third |

EOS: early onset scoliosis; MRI: magnetic resonance imaging.

Table 2. Statements regarding idiopathic EOS prognosis factors.

| Prognostic factors for idiopathic EOS | % of selection | Round in which consensus established |
|--|----------------|--------------------------------------|
| The most important demographic and/or clinical factors | | |
| Age at diagnosis | 85% agree | Third |
| Rib hump > 10° | 80% agree | Third |
| The most important radiological factors | | |
| RVAD >20 | 82% agree | Second |

EOS: early onset scoliosis; RVAD: rib vertebral angle difference.

Table 3. Statements regarding conservative treatment of idiopathic EOS.

| Conservative treatment of idiopathic EOS | % of answers | Intensity of agreement/ disagreement | Round in which consensus established |
|---|--------------|--------------------------------------|--------------------------------------|
| A 25° idiopathic EOS at first visit in a 1-year-old child should always be observed first | 80% agree | Strong | Third |
| A 25° is the uppermost limit of severity of the curve for observation in idiopathic EOS patients between 6 and 10 years | 90% agree | Strong | Third |
| The most preferred conservative method for the management of a progressive thoracic 60° idiopathic EOS before the age of 6 years (0–5 years of age) is “cast” | 82% agree | Strong | Second |

EOS: early onset scoliosis.

(REDCap) tools. REDCap is a secure, web-based application designed to support data capture for research studies, providing (a) an intuitive interface for validated data entry; (b) audit trails for tracking data manipulation and export procedures; (c) automated export procedures for seamless data downloads to common statistical packages; and (d) procedures for importing data from external sources.^{9,10}

Results

The questions that reached strong agreement or disagreement are summarized in Tables 1–8, but the answers from experts for each MCQ are reported in Tables 9 and 10.

Idiopathic EOS

Decisions were reached on 40 (53%) of the 75 statements for the 59 questions in the second round. Strong agreement was reached in 15 (38%) of these 40 statements, and clear categorizations were made in 25 (62%) of them. In 10 (29%) of the 35 statements remaining to the third round, a strong agreement was reached in the third round, clear categorizations were revealed in 10 (29%) statements, and weak and moderate agreement was obtained in 13 (37%) statements. At the end of the third round, no consensus was reached on 2 (5%) statements (Table 9).

Table 4. Statements regarding surgical treatment of idiopathic EOS.

| Surgical management of idiopathic EOS | % of answers | Intensity of agreement/ disagreement | Round in which consensus established |
|--|--------------|--------------------------------------|--------------------------------------|
| TGR should still be considered as a surgical option for progressive idiopathic EOS | 86% agree | Strong | Second |
| Growth-friendly surgery should not be performed in patients with closed TRC | 80% agree | Strong | Third |
| There is still a place for 4.5 diameter rods in the surgical management of progressive idiopathic EOS | 82% agree | Strong | Second |
| Preoperative halo-gravity traction is recommended for idiopathic EOS patients with stiff curves >75° | 91% agree | Strong | Second |
| Single-rod constructs are no longer the first-choice option in the surgical management of progressive idiopathic EOS | 86% agree | Strong | Second |
| Thoracic hyperkyphosis can influence the type of growth-friendly surgical technique recommended in idiopathic EOS? | 95% agree | Strong | Second |
| MCGR lengthening should be performed at least 3 times a year (3- to 4-month intervals) | 82% agree | Strong | Second |
| TGR lengthening should be performed more than once a year | 85% agree | Strong | Third |
| Radiographs (T1T12 and T1S1 lengths) are the best method to monitor growth gain after lengthening in idiopathic EOS | 86% agree | Strong | Second |
| I believe in the law of diminishing return in growth-friendly surgical techniques | 95% agree | Strong | Second |
| The law of diminishing return exists with both MCGR and TGR growth-friendly surgical techniques | 90% agree | Strong | Second |
| Spine-to-spine (spinal) constructs are superior to ribs to spine (extraspinal) constructs | 86% agree | Strong | Second |

TGR: traditional growing rods; TRC: triradiate cartilages; EOS: early onset scoliosis; MCGR: magnetically controlled growing rods.

Table 5. Statements regarding complications and graduation for idiopathic EOS.

| Complications | % of answers | Intensity of agreement/ disagreement | Round in which consensus established |
|---|---------------|--------------------------------------|--------------------------------------|
| Asymptomatic PJK occurring during the growth-friendly surgical management of an idiopathic EOS child should only be addressed at final fusion in the absence of skin problem or anchor failure. | 86% agree | Strong | Second |
| Graduation | | | |
| Final fusion is not always necessary at the end of growth in idiopathic EOS | 80% agree | Strong | Third |
| MCGR can be left in place (and observed) at the end of growth in idiopathic EOS | 100% disagree | Strong | Third |
| Implant removal followed by observation is a reasonable option after growth-friendly surgery in idiopathic EOS | 91% disagree | Strong | Second |
| 10 years of age is the lowermost age in a girl to consider early final fusion in case of lengthening failure of growth-friendly techniques | 85% agree | Strong | Third |

PJK: proximal junctional kyphosis; EOS: early onset scoliosis; MCGR: magnetically controlled growing rods.

Non-idiopathic EOS

Decisions were reached in the second round for 10 (21%) of the 47 statements for the 44 questions. Strong agreement was reached in 4 (40%) of these 10 statements and clear categorizations were made in 6 (60%) of them. Out of the 37 remaining statements to the third round, 9 (24%) obtained strong agreement, 1 (3%) had clear categorization, 26 (70%) had weak and moderate agreement, and 1 (3%) had no consensus after the last round (Table 10).

Discussion

The Delphi process led to a strong agreement for 44% of the statements for idiopathic EOS and only 31.9% in the non-idiopathic subgroup. EOS is a rare condition representing less than 10% of all pediatric spine patients, and its treatment is centralized into academic medical centers in Europe, as the risk of complications is significant and requires multiple specialties.

Table 6. Progression risk factors and additional imaging for non-idiopathic EOS patients.

| Prognostic factors | % of answers | Intensity of agreement/ disagreement | Round in which consensus established |
|---|--------------|---|---|
| The most important diagnosis which associates with progressive non-idiopathic EOS before the age of 6 years | | | |
| Hemivertebra with contralateral bar | 91% | Strong | Second |
| Imaging | | | |
| Full spinal magnetic resonance images are always indicated with non-idiopathic EOS >20° at diagnosis | 95% agree | Strong | Third |
| Children with NFI should undergo cervical and/or spinal radiographs before the age of 6 years to detect pathologies early | 84% agree | Strong | Third |
| Cervical spine flexion-extension radiographs should always be taken before growth-friendly management of non-idiopathic EOS in children with skeletal dysplasia | 95% agree | Strong | Third |

EOS: early onset scoliosis.

Table 7. Non-idiopathic EOS-specific statements regarding conservative treatment.

| Conservative treatment | % of answers | Intensity of agreement/ disagreement | Round in which consensus established |
|--|--------------|---|---|
| It is usually indicated to only observe a 25° non-idiopathic EOS in the age group before the age of 5 years | 84% agree | Strong | Third |
| 60° would be the uppermost degree of non-idiopathic EOS which you regard as an indication for casting under general anesthesia in the conservative treatment of progressive non-idiopathic EOS in the age group between 2 and 5 years of age | 84% agree | Strong | Third |
| It is always indicated to treat conservatively a 40° non-idiopathic EOS (excluding congenital) in the age group before the age of 3 years | 86% agree | Strong | Second |
| Continuing with thoracolumbosacral orthosis is better than immediate cessation following a successful casting treatment (curve below 30° at the end of casting period) in the conservative treatment of progressive non-idiopathic EOS before the age of 5 years | 95% agree | Strong | Second |

EOS: early onset scoliosis.

Table 8. Non-idiopathic EOS-specific statements regarding surgical treatment, complications, and graduation.

| Surgical treatment | % of answers | Intensity of agreement/ disagreement | Round in which consensus established |
|--|--------------|---|---|
| Traditional growing rods are my preferred method in the growth-friendly management of non-idiopathic EOS needing follow-up for spinal cord malformation | 89% agree | Strong | Third |
| Intraoperative navigation is useful in the surgical treatment of EOS | 82% agree | Strong | Second |
| Bilateral rods are always indicated in the growth-friendly management of progressive non-idiopathic EOS to prevent rod fractures | 86% agree | Strong | Second |
| Complications | | | |
| A revision surgery is always indicated on a 6-year-old non-idiopathic EOS child with T1–T12 length of 180mm and failure to lengthen MCGR bilaterally | 84% agree | Strong | Third |
| An asymptomatic non-idiopathic EOS child treated using growth friendly surgery with proximal junctional kyphosis of 25° should undergo an extension of the instrumentation | 95% disagree | Strong | Third |
| Graduation | | | |
| Final fusion is not indicated in well corrected non-idiopathic EOS treated using traditional growing rods | 84% agree | Strong | Third |
| Final fusion is my preferred method in the surgical management of a 60° non-idiopathic, non-neuromuscular scoliosis at 10 years of age | 84% agree | Strong | Third |

EOS: early onset scoliosis; MCGR: magnetically controlled growing rods.

Table 9. Results on the 75 statements related to idiopathic EOS management.

| Definition | % of answers | Intensity of agreement/ disagreement | Round in which consensus is established |
|---|---------------------------|--|--|
| The definition of idiopathic EOS (under 10 years of age) is sufficient to define the subgroup of patients. | 55% agree | Weak | Third |
| The distinction between infantile and juvenile scoliosis is still relevant in idiopathic EOS. | 80% agree | Strong | Third |
| MRI should be performed within the first 6 months after initial diagnosis in the follow-up of an idiopathic EOS patient without neurological symptoms. | 55% agree | Weak | Third |
| MRI should be systematic before any surgery in the management of an idiopathic EOS patient without neurological symptoms. | 86% agree | Strong | Second |
| The C-EOS classification should be used in routine in the follow-up of idiopathic EOS patients. | 45% agree/ 5% disagree | Cannot be decided | Third |
| Idiopathic EOS patients under 12 months-old represent a specific entity that should be managed differently than older patients. | 91% agree | Strong | Second |
| Idiopathic EOS patients between 9 and 10 years of age (so called tweener) represent a specific entity that should be managed differently than younger patients. | 80% agree | Strong | Third |
| Prognostic factors | | | |
| | % of selection | Round in which consensus is established | |
| The most important demographic and/or clinical factors | | | |
| Age at diagnosis | 85% | Third | |
| Rib hump > 10° | 80% | Third | |
| Trunk imbalance | 55% | Third | |
| Thoracic lordosis | 65% | Third | |
| The most important radiological factors | | | |
| RVAD > 20 | 82% | Second | |
| Initial Cobb > 30° | 68% | Second | |
| Apical vertebral rotation | 64% | Second | |
| The most relevant clinical parameters to take into account to choose a management method (observation, conservative or surgery) | | | |
| Age | 82% | Second | |
| Trunk imbalance | 55% | Second | |
| Family compliance and conditions of life (including child's behavior) | 50% | Second | |
| The most relevant radiological parameters to take into account to choose a management method (observation, conservative or surgery) | | | |
| Cobb angle | 82% | Second | |
| Progression rate | 73% | Second | |
| Skeletal maturity | 55% | Second | |

(continued)

Table 9. (continued)

| Conservative treatment | % of answers | Intensity of agreement/ disagreement | Round in which consensus is established |
|--|---|---|--|
| A 25° idiopathic EOS at first visit in a 1-year-old child should always be observed first. | 80% agree | Strong | Third |
| A 25° is the uppermost limit of severity of the curve for observation in idiopathic EOS patients under 6 years old (0 to 5 years of age) | 55% | Third | |
| A 25° is the uppermost limit of severity of the curve for observation in idiopathic EOS patients between 6 and 10 years. | 90% | Third | |
| Serial casting should always be the first option in a progressive thoracic idiopathic EOS before the age of 6 (0–5 years of age). | 75% agree | Moderate | Third |
| The most preferred conservative method for the management of a progressive thoracic 25° idiopathic EOS before the age of 6 years (0–5 years of age) “cast or Full-time brace (Thoraco-lumbo-sacral-orthosis, TLSO).” | 32% | Second | |
| The most preferred conservative method for the management of a progressive thoracic 60° idiopathic EOS before the age of 6 years (0–5 years of age) is “cast.” | 82% | Second | |
| The most preferred conservative method for the management of a progressive thoracic 25° idiopathic EOS between 6 and 10 years old is “Full-time brace (TLSO).” | 59% | Second | |
| Select your most preferred conservative method for the management of a progressive thoracic 60° idiopathic EOS between 6 and 10 years old. | For this question “Cast, Full-time brace (TLSO), Full-time Milwaukee, Night-time brace” were the response categories, but participants (36%) think that none of these options is the most preferred conservative method for the management of a progressive thoracic 60° idiopathic EOS between 6 years and 10 years old. | | |
| A 60° is the uppermost limit of severity of the curve for conservative treatment in idiopathic EOS patients under 6 years old (0–5 years of age). | 70% | Third | |
| 50° or 70° are the uppermost limits of severity of the curve for conservative treatment in idiopathic EOS patients between 6 and 10 years old. | 35% | Third | |
| There is a role for night-time only orthosis in the primary conservative treatment of idiopathic EOS before 6 years old (0–5 years of age). | 70% disagree | Moderate | Third |
| There is a role for night-time only orthosis in the primary conservative treatment of progressive thoracic EOS before 6 years old (0–5 years of age). | 65% disagree | Weak | Third |
| There is a role for night-time only orthosis in the primary conservative treatment of progressive thoracic EOS between 6 and 10 years old. | 70% | Third | |
| 6 years olds your uppermost age limit for casting an idiopathic EOS. | 100% disagree | Strong | Second |
| Neuromonitoring is necessary during idiopathic EOS casting. | 80% agree | Strong | Third |
| General anesthesia is necessary during idiopathic EOS casting. | 45% | Third | |
| The minimum length of a serial casting program to treat a 3-year-old idiopathic EOS is 3 months. | 85% agree | Strong | Third |
| Full-time brace is the best option to continue conservative treatment after a successful casting period (curve <30° after casting)? | 75% agree | Moderate | Third |
| Family and patient reports are sufficient to monitor brace compliance in idiopathic EOS. | | | |
| Surgical technology | | | |
| MCGR represent to date the best surgical option for progressive idiopathic EOS. | 60% agree | Weak | Third |
| TGR should still be considered as a surgical option for progressive idiopathic EOS. | 86% agree | Strong | Second |
| Access to intraoperative navigation is necessary to optimize the surgical management of idiopathic EOS. | 55% disagree | Weak | Third |

(continued)

Table 9. (continued)

| Surgical management | 60% agree | Weak | Third |
|---|---------------|----------|--------|
| The uppermost age limit to perform growth-friendly surgery in progressive idiopathic EOS is 10 years old. | 60% agree | Weak | Third |
| Growth-friendly surgery should not be performed in patients with closed TRC. | 80% agree | Strong | Third |
| Indications for growth-friendly surgery in idiopathic EOS should remain limited before the age of 6 (0–5 years old). | 75% disagree | Moderate | Third |
| Indications for growth-friendly surgery in idiopathic EOS should remain limited under the weight of 15 kgs (0–19 kgs). | 75% disagree | Moderate | Third |
| There is still a place for 4.5 diameter rods in the surgical management of progressive idiopathic EOS. | 82% agree | Strong | Second |
| Preoperative halo-gravity traction is recommended for idiopathic EOS patients with stiff curves > 75° | 91% agree | Strong | Second |
| "MCGR" is the most preferred management strategy for a progressive 60° idiopathic curve in 8- to 10-year-old girl who failed conservative treatment. | 45% | Second | |
| "Observation until early final fusion" is the most preferred management strategy for a progressive 60° idiopathic curve in a 9- to 11-year-old boy who failed conservative treatment | 32% | Second | |
| The most relevant demographic and clinical parameters to discuss with caregivers before choosing a growth-friendly technique in progressive idiopathic EOS. | 82% | Second | |
| Growth expectation | 55% | Second | |
| Weight and nutritional status | 50% | Second | |
| Curve flexibility | 95% agree | Strong | Second |
| Thoracic hyperkyphosis can influence the type of growth-friendly surgical technique recommended in idiopathic EOS! | 82% agree | Strong | Second |
| MCGR lengthenings should be performed at least 3 times a year (3- to 4-month intervals). | 85% agree | Strong | Third |
| TGR lengthenings should be performed more than once a year. | 86% agree | Strong | Second |
| Radiographs (T1T2 and T1S1 lengths) are the best method to monitor growth gain after lengthenings in idiopathic EOS. | 95% agree | Strong | Second |
| I believe in the law of diminishing return in growth-friendly surgical techniques. | 90% agree | Strong | Second |
| The law of diminishing return exists with both MCGR and TGR growth-friendly surgical techniques. | 86% agree | Strong | Second |
| Spine-to-spine (spinal) constructs are superior to ribs to spine (extraspinal) constructs for the surgical management of idiopathic EOS. | 90% agree | Strong | Second |
| The most appropriate constructs to secure the proximal anchor fixation of 2 growth-friendly rods in idiopathic EOS. | 86% agree | Strong | Second |
| 3 pairs of pedicle screws (3 levels) | 36% | Second | |
| 2 pairs of pedicle screws (2 levels) | 27% | Second | |
| Single-rod constructs are no longer the first-choice option in the surgical management of progressive idiopathic EOS. | 86% agree | Strong | Second |
| The most common expected complications in the surgical management of idiopathic EOS. | | | |
| Failure to lengthen/autofusion | 73% | Second | |
| Proximal anchor dislodgement | 68% | Second | |
| Sagittal iatrogenic malalignment (PJK, curves flattening) | 64% | Second | |
| Complications | | | |
| The best option for revision of a 6-year-old idiopathic EOS child with T1–T12 length of 180 mm and failure to lengthen MCGR is transition to TGR. | 75% agree | Moderate | Third |
| Asymptomatic PJK occurring during the growth-friendly surgical management of an idiopathic EOS child should only be addressed at final fusion in the absence of skin problem or anchor failure. | 86% agree | Strong | Second |
| Graduation | | | |
| Final fusion is not always necessary at the end of growth in idiopathic EOS. | 80% agree | Strong | Third |
| MCGR can be left in place (and observed) at the end of growth in idiopathic EOS. | 100% disagree | Strong | Third |
| Implant removal followed by observation is a reasonable option after growth-friendly surgery in idiopathic EOS. | 91% disagree | Strong | Second |
| 10 years of age is the lowermost age in a girl to consider early final fusion in case of lengthening failure of growth-friendly techniques. | 85% agree | Strong | Third |
| 10 years of age is the lowermost age in a boy to consider early final fusion in case of lengthening failure of growth-friendly techniques. | 70% agree | Moderate | Third |
| The main reasons not to perform final fusion in an idiopathic EOS patient treated with growth-friendly technique. | | | |
| Acceptable residual deformity (balanced and < 40°) without symptoms | 77% | Second | |
| Wide auto fusion | 64% | Second | |
| Patient and family satisfied with final deformity | 59% | Second | |

RYAD: rib vertebral angle difference; TRC: triradiate cartilages; PJK: proximal junctional kyphosis; MCGR: magnetically controlled growing rods; TGR: traditional growing rods; EOS: early onset scoliosis; MRI: magnetic resonance imaging.

Table 10. Results on the 47 statements related to non-idiopathic EOS management.

| Clinical, radiological and etiological risk factors for progression of non-idiopathic EOS | % of selection | Round in which consensus is established | |
|--|--|--|---|
| The most important diagnosis which associates with progressive non-idiopathic EOS before the age of 6 years. Hemivertebra with contralateral bar SMA | 91% 41% | Second Second | |
| The most important clinical findings which are associated with progression of non-idiopathic EOS. Significant rib hump (rotation) Young age of the patient | 64% 36% | Second Second | |
| The most important radiographic findings which are associated with progressive non-idiopathic EOS. Hemivertebra associated with contralateral bar Coronal Cobb angle > 50° | 73% 64% | Second Second | |
| Additional imaging for non-idiopathic EOS patients | % of answers | Intensity of agreement/disagreement | Round in which consensus is established |
| Full spinal magnetic resonance images are always indicated with non-idiopathic EOS > 20° at diagnosis. Children with CP GMFCSS should undergo spinal radiographs before school age to detect EOS early. Children with skeletal dysplasia should undergo neutral as well as flexion-extension cervical spine radiographs before the age of 6 years. Children with NFI should undergo cervical and/or spinal radiographs before the age of 6 years to detect pathologies early. Cervical spine flexion-extension radiographs should always be taken before growth-friendly management of non-idiopathic EOS in children with NFI. | 95% agree 68% agree 79% agree 84% agree 26% agree/ 32% disagree | Strong Weak Moderate Strong Cannot be decided | Third Third Third Third Third Third |
| Cervical spine flexion-extension radiographs should always be taken before growth-friendly management of non-idiopathic EOS in children with skeletal dysplasia. | 95% agree | Strong | Third |
| Conservative treatment | | | |
| It is usually indicated to only observe a 25° non-idiopathic EOS in the age group before the age of 5 years. Casting under general anesthesia is the first preference in the conservative treatment of progressive non-idiopathic EOS before the age of 5 years. 60° would be the uppermost degree of non-idiopathic EOS which you regard as an indication for casting under general anesthesia in the conservative treatment of progressive non-idiopathic EOS in the age group between 2 and 5 years of age. | 84% agree 79% agree 84% agree | Strong Moderate Strong | Third Third Third |
| It is always indicated to treat conservatively a 40° non-idiopathic EOS (excluding congenital) in the age group before the age of 3 years. Rigid thoracolumbosacral (TLSO) orthosis is the first preference in the conservative treatment of progressive non-idiopathic EOS. Casting under general anesthesia is always indicated in progressive congenital EOS (more than five levels) with a curve over 40° in the age group below 5 years of age. Casting under general anesthesia is my preferred method to treat progressive non-idiopathic EOS (excluding congenital) with a curve of 60° in the age group before the age of 5 years. 6 years is the uppermost age limit in the serial casting of progressive non-idiopathic EOS. Serial casting of progressive non-idiopathic EOS should last a minimum of 12 months. Continuing with thoracolumbosacral orthosis is better than immediate cessation following a successful casting treatment (curve below 30° at the end of casting period) in the conservative treatment of progressive non-idiopathic EOS before the age of 5 years. | 86% agree 53% disagree 63% disagree 79% agree 79% agree 58% disagree 95% agree | Strong Weak Weak Moderate Moderate Weak Strong | Second Third Third Third Third Third Second |

(continued)

Table 10. (continued)

| Surgical treatment | 82% agree | Strong | Second |
|---|--------------|----------|--------|
| Intraoperative navigation is useful in the surgical treatment of EOS. | 79% agree | Moderate | Third |
| The age of 24-months is the lowest age limit to perform any kind of orthopedic surgical intervention in progressive non-idiopathic EOS (excluding congenital). | 68% disagree | Weak | Third |
| Hemivertebrectomy can safely be performed before the age of 24 months for a fully segmented hemivertebra in the thoracolumbar area (T1–L5). | 58% agree | Weak | Third |
| MCGR is the primary surgical intervention in progressive non-idiopathic, non-congenital EOS. | 63% agree | Weak | Third |
| Preoperative halo-gravity traction is my preferred method in the presurgical growth-friendly management of an 80° non-idiopathic EOS associated with moderate thoracic kyphosis (<40°). | 68% agree | Weak | Third |
| Preoperative halo-gravity traction is always indicated in an 80° progressive non-idiopathic EOS associated with thoracic hyperkyphosis (>40°). | 74% agree | Moderate | Third |
| Anterior approach with releases and annulotomies is never indicated to facilitate curve correction of non-idiopathic EOS before the age of 10 years. | 89% agree | Strong | Third |
| TGR are my preferred method in the growth-friendly management of non-idiopathic EOS needing follow-up for spinal cord malformation. | 58% disagree | Weak | Third |
| TGR are my preferred method over MCGR in the surgical management of congenital scoliosis (more than five levels) with long curve types. | 53% disagree | Weak | Third |
| MCGR are indicated in the growth-friendly management of congenital scoliosis >5 levels. | 58% agree | Weak | Third |
| Rib-based instrumentation is never indicated in non-idiopathic EOS without rib pathologies. | 68% agree | Weak | Third |
| Thoracostomy (longitudinal rib osteotomies) and Vertical Expandable Prosthetic Titanium Rib (VEPTR) instrumentation is my preferred method to address congenital scoliosis with fused ribs. | 86% agree | Strong | Second |
| Bilateral rods are always indicated in the growth-friendly management of progressive non-idiopathic EOS to prevent rod fractures. | 53% agree | Weak | Third |
| Two to three pairs of pedicle screws are my preferred upper fixation method in the growth-friendly management of non-idiopathic EOS. | 63% disagree | Weak | Third |
| Two to three pairs of laminar, transverse or rib hooks are my preferred upper fixation method in the growth-friendly management of non-idiopathic EOS. | 53% agree | Weak | Third |
| Upper thoracic pedicle screws are associated with risk for neurologic deficits in the surgical treatment of non-idiopathic EOS. | | | |
| Complications | | | |
| A revision surgery is always indicated on a 6-year-old non-idiopathic EOS child with T1–T12 length of 180 mm and failure to lengthen MCGR bilaterally. | 84% agree | Strong | Third |
| Failure to lengthen MCGR on a 6-year-old with non-idiopathic EOS should be converted to TGR. | 68% agree | Weak | Third |
| Failure to lengthen MCGR on a 6-year-old with non-idiopathic EOS should be replaced to a new set of MCGR. | 79% disagree | Moderate | Third |
| An asymptomatic non-idiopathic EOS child treated using growth friendly surgery with proximal junctional kyphosis of 25° should undergo an extension of the instrumentation. | 95% disagree | Strong | Third |
| Anteroposterior instrumented spinal fusion is indicated to treat congenital scoliosis associated with segmentation defect (bony bar). | 79% disagree | Moderate | Third |
| Graduation | | | |
| Final fusion is indicated after growth-friendly management using MCGR even in well corrected non-idiopathic EOS. | 74% agree | Moderate | Third |
| Final fusion is not indicated in well corrected non-idiopathic EOS treated using TGR. | 84% agree | Strong | Third |
| Final fusion is my preferred method in the surgical management of a 70° neuromuscular EOS between the ages of 8 and 10 years. | 58% disagree | Weak | Third |
| Final fusion is my preferred method in the surgical management of a 60° non-idiopathic, non-neuromuscular scoliosis at 10 years of age. | 84% agree | Strong | Third |

EOS: early onset scoliosis; MCGR: magnetically controlled growing rods; TGR: traditional growing rods.

Idiopathic EOS

Twenty-seven guidelines with a high consensus rate ($\geq 80\%$) were provided by European experts for the management of idiopathic EOS. Four statements with strong agreement were found regarding the definition of EOS (Table 1), but experts recommended to consider patients under 1 year old and tweens as specific entities, which is currently not described in the EOS classification (C-EOS) reported by Williams et al.¹ Experts also agreed that magnetic resonance imaging was necessary before any surgical management, even without neurological symptoms, but not at initial diagnosis. Interestingly, the C-EOS was only moderately adopted by the European experts (45% of them used it in routine). Age and rotation (both clinical and radiological) were reported as the main prognostic parameters (Table 2). Radiologically, a rib vertebral angle difference $>20^\circ$ also reached strong agreement, while initial Cobb greater than 30° only reached a moderate one (68%).

Regarding conservative management, experts agreed that there was no rush to treat a 12-month patient with idiopathic EOS, and that 25° was the uppermost limit for observation between 6 and 10 years (Table 3). They also concluded that serial casting was the most appropriate treatment for a progressive 60° thoracic curve. The statement to always recommend cast as the first option in a progressive thoracic curve under 6 years old did not reach strong agreement (75%), as some experts preferred to use bracing (full or part-time) as initial management.

Experts agreed that growth-friendly surgery should not be performed in patients with closed triradiate cartilages, and that there is still a place for 4.5 diameter rods (Table 4). Europeans tended to be more surgically aggressive than their peers in the United States, especially in kids younger than 6 years old and under 20 kg (75%, moderate agreement).^{2,11,12} Preoperative halo-gravity traction is recommended for stiff curves $>75^\circ$, and growth expectation remains the most relevant demographic and clinical parameter to discuss with caregivers before choosing a growth-friendly technique. Thoracic hyperkyphosis can also influence the type of surgical technique. No consensus was found to consider magnetically controlled growing rods (MCGR) as the best surgical option, and the only final statement was that traditional growing rods (TGRs) should still be considered (primary surgery or revision). Only 45% of experts agreed that navigation was necessary during surgery to optimize outcomes, and robotics was not included in the survey. MCGR lengthening should be performed at least three times a year (3- to 4-month intervals), while TGR lengthening should be considered more than once a year. Radiographs (T1T12 and T1S1 lengths) are the best method to monitor growth gain, and the law of diminishing return exists with both MCGR and TGR, as previously reported.¹³ Spine-to-spine (spinal) constructs

are preferred to ribs to spine (extraspinal) ones, and single-rod constructs are no longer the first-choice option. Asymptomatic proximal junctional kyphosis (PJK) is frequently observed during treatment, but it should only be addressed at final fusion in the absence of skin problem or anchor failure (Table 5).

Graduation remains a hot topic, with many different opinions and strategies.^{14–16} However, European experts concluded that implant removal alone and leaving MCGR in place were not options to consider in idiopathic EOS, while early fusion and observing TGR were reasonable in some cases.

Non-idiopathic EOS

Only 15 statements (31.9%) reached a high consensus rate ($\geq 80\%$) in this heterogeneous and challenging subgroup. Hemivertebra with associated contralateral bar was identified as the situation with the highest progression risk. Full spinal magnetic resonance images should be obtained for all curves $>20^\circ$, and cervical instability should be ruled out if surgery is considered in patients with skeletal dysplasia or NF1 (Table 6). Early identification and casting can significantly delay the need for surgery.¹⁷ However, experts agreed that a 25° curve can only be observed first in children under 5 years old, while 40° curves require conservative management, and 60° curve represents the upper limit of casting (Table 7). A successful casting period should be followed by thoracolumbar bracing.

In case of surgery, bilateral rods are recommended to prevent rod fracture, and in opposition to the idiopathic group, navigation is considered helpful if available (Table 8). MCGR should not be considered when further spinal cord imaging is necessary during treatment, and radiological PJK is an expected and common phenomenon that does not require revision surgery in the absence of disabling symptoms. Failure to lengthen MCGR is another frequent complication, and revision (MCGR or TGR) is necessary if it happens in immature young children.¹⁸

Final fusion is not always indicated in well-corrected and pain-free non-idiopathic EOS treated with TGR, so further cost/benefit analysis should be performed in fragile patients who remain flexible (Table 8).^{19–21} In case of successful conservative management, early definitive fusion remains a reasonable alternative in 10-year-old non-neuromuscular patients with curves exceeding 60° , in order to avoid repeated surgeries.²²

Interestingly, the management of congenital scoliosis did not reach any consensus among the experts, reflecting the various options in this subgroup and the lack of clinical evidence to support decisions (Table 10). Even if the level of agreement was weak (63%), casting under general anesthesia was not recommended in this category, even in progressive curves exceeding 40° before 5 years of age.

Limitations

The limitations of this study are inherent to Delphi, such as the bias induced by the selection of participants, the lack of clarity on the meaning of “consensus,” and also the difficulty of generalizing the results to a wider population. However, even though the process was quite time-consuming and laborious, the number of drop-out remained very limited (1 at round 3).

In conclusion, defining best practice guidelines remains difficult among European experts in the field of EOS, especially for the non-idiopathic group, which is a very heterogeneous and challenging condition. A true expert involvement in the management of these uncommon and frequently very challenging conditions remains necessary. However, this Delphi study provided 48 consensus statements which would be a good guide for spinal surgeons who have less space for EOS in their daily practice. Seeing the differences between Europe and other regions of the world will also be a good starting point for developing common solutions in the future to improve patients' care.

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