Instructional Lecture: Paediatrics

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Treatment strategies for early-onset scoliosis

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- Early-onset scoliosis (EOS) is defined as a spinal deformity occurring before the age of ten years.
- Untreated EOS or early spinal fusion resulting in a short spine is associated with increased mortality and cardiopulmonary compromise.
- EOS may progress rapidly, and therefore prompt clinical diagnosis and referral to a paediatric orthopaedic or spine unit is necessary.
- Casting under general anaesthesia can be effective and may prevent or delay the need for surgery in curves of less than 60°.
- 'Growing' rods (traditional or magnetically-controlled) represent the standard surgical treatment in progressive curves of 45° or greater.
- Children with congenital scoliosis associated with fused ribs benefit from surgery with a vertical titanium prosthetic rib.
- Surgery with growth-friendly instrumentation is associated with a high risk of complications.

Keywords: early-onset scoliosis; casting; growing rods; complications; vertical prosthetic titanium rib

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Introduction

Early-onset scoliosis (EOS) is defined as a spinal deformity occurring before ten years of age.[1,2] Untreated EOS or early spinal fusion resulting in a short spine is associated with increased mortality and cardiopulmonary compromise.[3-5] Since EOS is a heterogeneous condition, a uniformly accepted classification has been proposed.[1] This includes age, aetiology (congenital, neuromuscular, syndromic and idiopathic), major curve, kyphosis and progression modifier. Surgery is indicated for progressive deformities.[4,6,7] EOS may progress rapidly and, therefore, prompt clinical diagnosis and referral to a paediatric orthopaedic unit is necessary (Figs 1a, 1b and 1c).[1,7]

Growth of the spine

Truncal height will increase by 350% and weight twenty-fold from birth to adulthood.[8-11] In addition to 2D growth, volumetric growth occurs: at birth the volume of the thorax is 6.7% of the final volume and the volume of lumbar vertebrae will be multiplied by six from the age of five years to skeletal maturity (Fig. 2).[9] The growth of the spine, thoracic cage and lungs are closely associated with each other.[8-11] Disturbance within spinal or thoracic cage growth will adversely affect growth of the lungs. Severe scoliosis and early spinal fusion negatively affect the growth of the spine.[5]

Most vertebrae have at least three growth zones.[8-10] The pattern of posterior arch growth is linked to the presence of the neural stem and differs from vertebral body growth, which more or less resembles the growth of long bones.

Ossification of the vertebral bodies starts at the third month of intra-uterine life.[12] Three primary ossification centres are present within each vertebra, except for C1, C2 and the sacrum. Ossification first appears in the lower thoracic and upper lumbar spine and radiates from there in both cranial and caudal directions.[12]

The skeleton has two rapid growth periods – from birth to five years and during puberty.[8-11] At birth, the standing height of the neonate is about 30% of the final height. The spine makes up to 60% of the sitting height, whereas the head represents 20% and the pelvis the remaining 20%.[9,10] The length of the spine will nearly triple between birth and adulthood. The T1-S1 segment measures about 19 cm at birth, 28 cm at the age of five years and 45 cm at skeletal maturity (Fig. 2). This segment represents 49% of the sitting height and 64% of the length of the spine. During the first five years of life, its rate of growth is > 2 cm per year, 0.9 cm between the ages of five and ten years and 1.8 cm during puberty.[9] The thoracic spine (T1-T12) is about 11 cm long at birth, 18 cm at five years of age and 22 cm at ten years, and will reach a length of 28 cm in boys and 26 cm in girls at maturity.[10]

The length of the thoracic spine is critical for normal lung development. The final length of the thoracic spine is

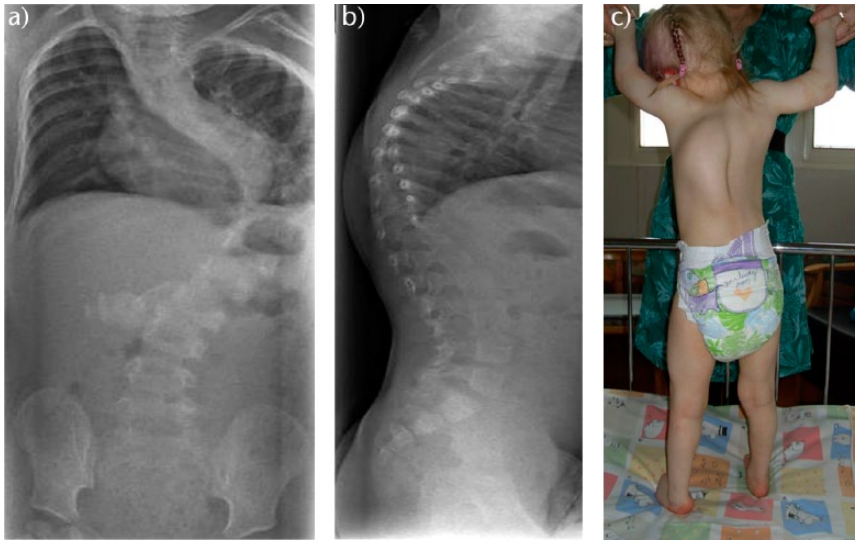

Fig. 1 Two-year-old girl with severe thoracic early-onset scoliosis: a) and b) standing posteroanterior and lateral spinal radiographs; c) clinical photograph demonstrating major rib hump and deformity.

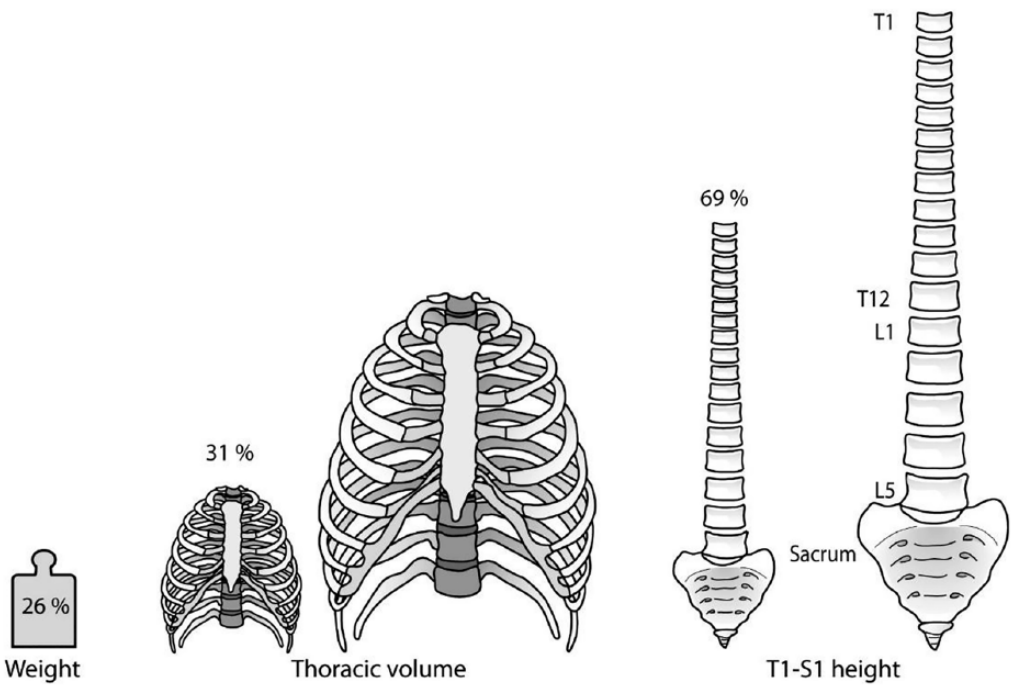

Fig. 2 Relative size of thorax and spine in a 5-year-old as compared with an adult. Reproduced from Helenius, I.J. (2011) 'Normal and abnormal growth of spine',[11] with permission from Springer.

closely related to the lung volume obtained at skeletal maturity.[5] If the T1-T12 segment reaches the length of 18 cm (normal value at the age of five years) at maturity, a lung volume (vital capacity) of approximately 45% of normal is achieved, which is compatible with survival.[5] However, the T1-T12 segment should achieve the length of 22 cm (normal length at the age of ten years) to obtain normal lung volume at maturity.[5]

Indications for interventions

EOS can be treated with serial casting, bracing or surgery (see Fig. 3). Casting is indicated for progressive infantile scoliosis (diagnosed before the age of three years),[13] while surgery is typically recommended when the Cobb angle progresses beyond 50° in the setting of failed conservative management and documented progression.[4,6,7] Progressive

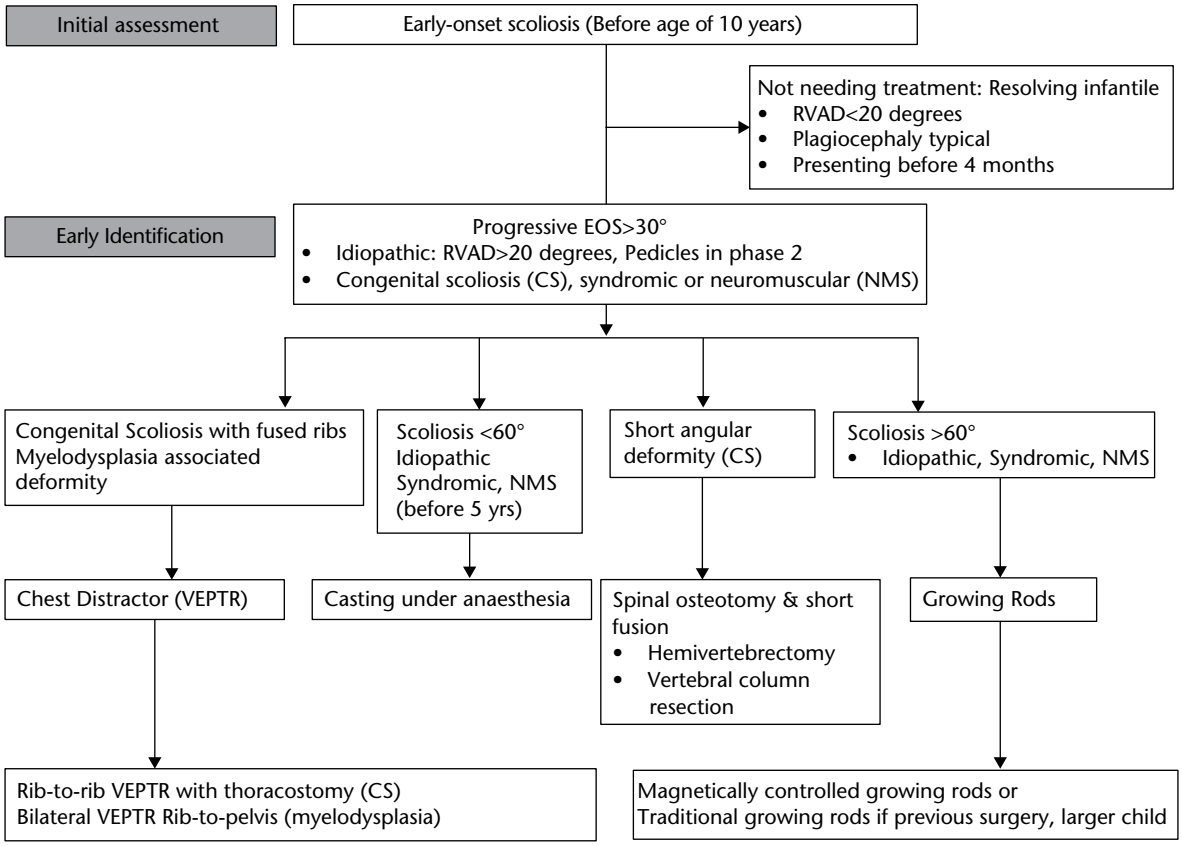

Fig. 3 EOS treatment flowchart.

and non-progressive infantile scoliosis are typically differentiated mainly by using the rib-vertebra angle difference (RVAD).[13] A RVAD of 20° or more is typical of progressive infantile scoliosis, while most resolving curves show a RVAD < 20°. A definitive hallmark of progressive infantile scoliosis is the apical rib head in phase two. In this stage, the shadow of the head of the rib overlaps the corresponding vertebral body.[13]

Surgery can be performed with traditional 'growing' rods (TGR) requiring repeated surgical distractions typically performed every six months,[4,6,7] with Shilla (Medtronic International, Memphis, USA) and other growth guidance systems,[14] with a vertical expandable prosthetic titanium rib (VEPTR) (Fig. 4)[15,16] or with magnetically-controlled growing rods (MCGR) (Figs 5a and 5b)[17-20] which allow the spine to grow until skeletal maturity when final spinal fusion may be performed.[21,22]

Casting

Early casting has been shown to prevent progression and even correct EOS with long-lasting stable outcome in otherwise healthy children.[23] In young children casting is performed under general anaesthesia on a Cotrel frame. Each jacket is worn for eight to 16 weeks to allow for the rapid growth period of the spine and the trunk. In the landmark paper by Mehta,[23] 136 children under the age of four years with progressive infantile scoliosis (scoliosis diagnosed before the age of 3 years) were treated with casting. In 94 children with early referral (mean age 1 year 7 months) and with a mean Cobb angle of 32° (11° to 65°), the scoliosis resolved by a mean age of 3.5 years. They needed no further treatment and went on to lead a normal life. In contrast, in 42 children with late referral (mean age 2.5 years) with a mean Cobb angle of 52° (23° to 92°), casting could not reverse the deformity. In all, 15 of these children (36%) underwent spinal fusion.

'Growing' rods

Surgical treatment using TGRs or MCGRs has been the standard for EOS following several outcome studies documenting long-term follow-up after growth with subsequent definitive final fusion or just observation following the final lengthening.[4,6,7,17-22] TGR is a non-fusion technique, which requires repeated surgical lengthenings and is associated with a high risk of surgical complications.[24,25] These include deep surgical site infection, rod fractures and failure of proximal fixation. MCGRs (Figs 5a and 5b) represent a new distraction-based spinal instrumentation

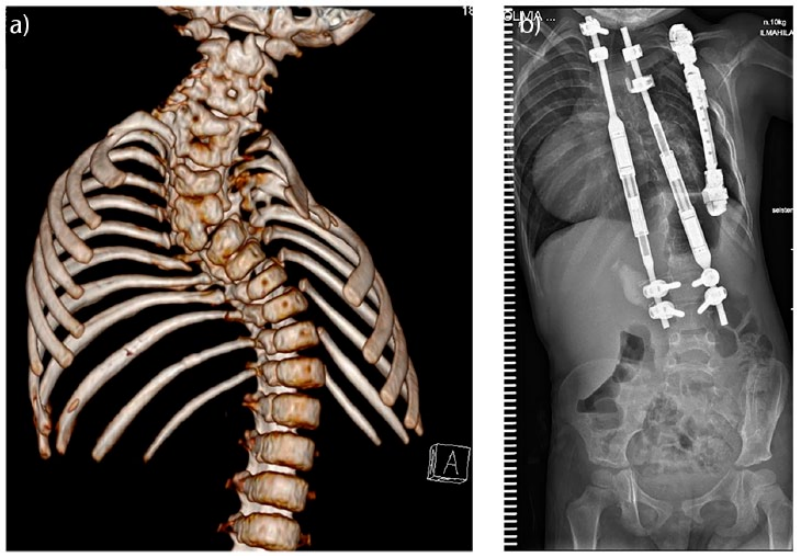

Fig. 4 a) 3D CT format demonstrating congenital scoliosis with fused ribs (spondylocostal dysostosis) at the age of one year; b) three-year postoperative standing radiograph after staged thoracostomy and rib-to-rib vertical titanium prosthetic rib instrumentation (at the age of one year) and magnetically controlled growing rod surgery at the age of 1.5 years.

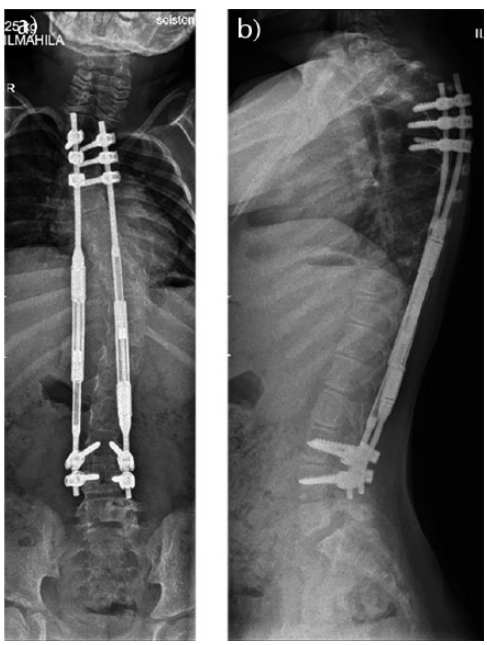

Fig. 5 The same patient as shown in Fig. 1. a) standing posteroanterior and lateral radiograph at the age of nine years; b) five years after conversion from traditional 'growing' rods to magnetically controlled growing rods.

for EOS, which allows non-surgical, outpatient construct lengthenings with possibly a reduced risk of deep surgical site infection.[17-20]

Both TGRs and MCGRs are used in a submuscular, dual-rod fashion.[4,7,17-19,]Typical spinal fixation involves upper thoracic pedicle screws or laminar hooks and mid-lumbar pedicle screws.[7] Growing rods provide correction of the spinal deformity using indirect methods: distraction on the concave side and cantilevering on the convexity of the curve.[4,6,7] Severe scoliosis (defined as scoliosis > 90°) remains difficult to correct using growing rod surgery.[26,27] Thus, coronal curve correction was typically lower (43% in this study) for severe EOS than when using segmental pedicle screw instrumentation in children undergoing surgery for adolescent idiopathic scoliosis (> 60%).[4,6,7,27]

When successful, the initial 'growing' rod surgery typically provides approximately half of the spinal length increase and the periodic lengthenings provide the remaining half in children with EOS treated with growing rods.[4,6,7]

In a multicentre study, Akbarnia et al[4] evaluated 23 patients (seven idiopathic, three congenital, 13 secondary) who were operated on using TGRs with a minimum two-year follow-up. All patients underwent lengthening of the implants every six months. The average number of lengthenings was 6.6 and this resulted in growth of 4.6 cm or 1.2 cm/year. Patients with congenital scoliosis received significantly less length during the initial procedure while lengthening produced similar growth. Distraction of the spine with growing rods may stimulate growth of the spine, since growth of 1.2 cm per year exceeds that of the normal spine. The 'Growing' Spine Study Group[28] evaluated the T1-S1 gain over repeated surgical lengthenings. A decrease of T1-S1 gain from 10 mm at the first lengthening to 6 mm at the seventh lengthening occurred, but some gain occurred even after multiple lengthenings (a type of 'law of diminishing returns').

Bess et al[24] observed at least one complication in 81 of 140 children (58%) treated using a TGR during a

minimum five-year follow-up. Older age at surgery and dual submuscular growing rods decreased the risk of complications, whilst every surgical procedure increased this risk by 24%.[10]

The use of MCGRs is the latest technique that allows non-invasive lengthening of the spine.[17-20] Typically, dual MCGRs are fixed submuscularly to the spine with pedicle screws or hooks to connect the proximal and the distal fixation of the rods. The MCGR contains a magnetically-driven lengthening mechanism. After the primary operation, lengthening can be done without anaesthesia with an external remote controller on an outpatient clinic basis. It has been suggested that, because there is no need for repeated surgeries, the risk of wound infections would be lower than with TGRs.[17-19] MCGRs have been shown to be a safe and effective surgical technique in patients undergoing primary EOS surgery.[17,18] However, patients converted from a TGR to MCGR seem to achieve less growth than children operated primarily with MCGRs.[19] In one study, 47% of children undergoing surgery with a MCGR for EOS have required an unplanned re-operation during a minimum two-year follow-up.[17] In severe EOS a period of preoperative halo traction is a useful adjunct, since it has been shown to reduce kyphosis more effectively than spinal release in EOS, which might therefore reduce stress on the spinal instrumentation.[31] In a recent study, preoperative halo traction was not associated with major complications in severe EOS.[26]

Complications associated with growing rod surgery include anchor failure, rod fracture, autofusion and increased risk of deep surgical site infection.[24,25] Growing rod surgery for severe EOS has also been associated with a relatively high risk of neurological deficits (5%).[26] The main causes of these deficits were: 1) correction and distraction of the spine during initial growing rod surgery; 2) pedicle screw pull-out during follow-up and; 3) difficulties in placing thoracic pedicle screws during revision surgery.[26]

VEPTR

The VEPTR implant has been designed primarily for the treatment of congenital scoliosis associated with fused ribs[15,16] (Figs 4a and 4b). Campbell et al[16] evaluated the outcomes of 27 patients with congenital scoliosis associated with fused ribs who underwent an opening-wedge thoracotomy and VEPTR implantation at the age of 3.2 years (mean follow-up 5.7 years). A total of 25 patients had at least one hemivertebra on the convexity and a unilateral bar on the concavity (mean length 4.2 vertebrae). The mean length of the thoracic spine was 11.7 cm preoperatively, 12.3 cm immediately after the index procedure and 15.7 cm at final follow-up, representing an increase of thoracic height of a mean of 0.7 cm per year (0.2 to 1.37). These findings of continued thoracic spinal growth have been confirmed by Emans et al.[30] On the other hand, rib-based instrumentation may increase the compliance of the rib cage, and thus decrease the functional lung volumes while increasing residual volume.[30] On the other hand, a subset of patients with early thoracic reconstruction using the VEPTR treatment have shown a complete resolution of pulmonary support. Final spinal deformity surgery has been reported to be difficult following VEPTR instrumentation due to calcification along the device and partial autofusion, as well as increased stiffness of the chest cage.[30]

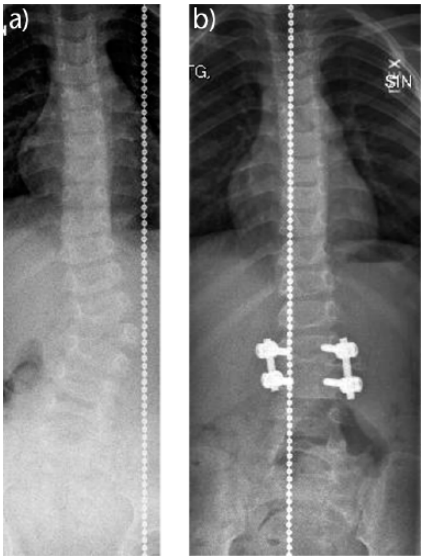

Fig. 6 a) Standing spinal radiograph demonstrating a fully segmented lumbar hemivertebra at the age of two years; b) standing radiograph two years after all posterior hemivertebrectomy and short 'single' level pedicle screw instrumentation.

Vertebral column resection

Vertebral column resection at an early age is indicated for short angular deformities, which are typical of congenital scoliosis or congenital kyphosis (Figs 6 and 7).[32-34] Hemivertebrectomy and short pedicle screw instrumentation for congenital scoliosis is the most common vertebral column resection before the age of ten years[32-34] (Figs 6a and 6b). Traditionally, hemi-vertebrectomy was performed using a combined anteroposterior approach, but currently most centres prefer a posterolateral approach.[33,34] The posterior approach results in a shorter operative time and less blood loss, but neural element manipulation and related transient deficits are more common than in the combined approach.[34] A combined anteroposterior approach may be needed at an early age when rigid fixation is not feasible and solid spinal fusion is warranted for an unstable condition, such as congenital dislocation of the spine (Figs 7a to 7e).

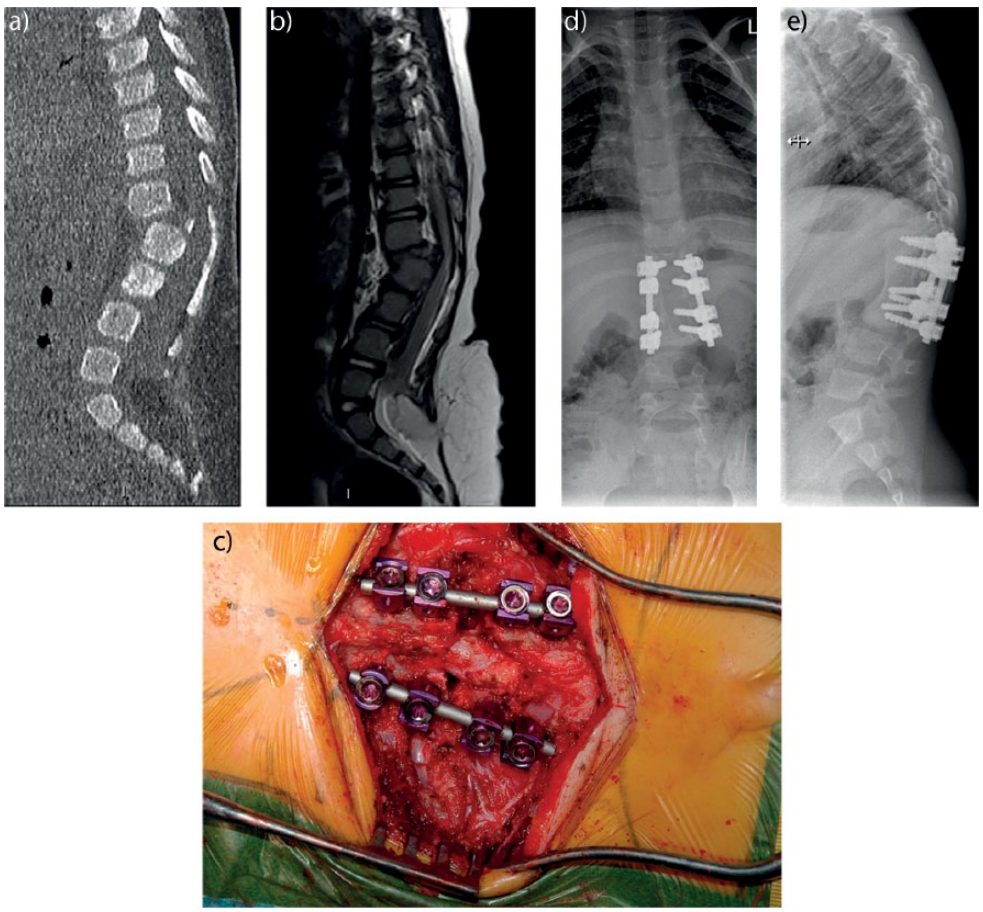

Fig. 7 a) Sagittal CT reformat on a nine-month-old boy with congenital dislocation of the spine (type A, small rudimentary hemivertebra at the dislocation); b) sagittal T2 MR image demonstrating cord compression and tethered cord with lipomeningocele; c) intraoperative photograph after posterolateral decompression of the cord via costotransversectomy; d) and e) standing posteroanterior and lateral radiographs three years after index surgery (anteroposterior spinal fusion) demonstrating solid spinal fusion.

Conclusions

EOS necessitates early diagnosis and prompt treatment to prevent severe and life-threatening cardiopulmonary compromise. Casting at an early phase may cure EOS, whilst more severe and progressive forms of EOS typically require surgery with 'growth-friendly' techniques, such as growing rods. The development of MCGRs reduces the need of repeated surgical measures and may reduce the risk of deep surgical site infection.

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