IDIOPATHIC SCOLIOSIS

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ABSTRACT

Idiopathic scoliosis (IS) is a lifetime condition and is defined as a structural, lateral rotated curvature of the spine of >10° on standing coronal plane radiographs. It should be distinguished from other causes of scoliosis. It can be classified as infantile, juvenile, and adolescent according to age. As a rule of thumb, about 80% of all curves are idiopathic, right convex thoracic, and present in otherwise healthy girls at the beginning of puberty. A family member most commonly detects scoliosis. The structural asymmetry of the spine is best observed by asking the patient to bend forward. IS is often seen in more than one member of a family, but the aetiology remains unknown. Multiple genes are likely to be involved with incomplete penetrance and variable expressivity. Early detection by screening allows for monitoring curve progression and timely initiation of bracing, but school screening is controversial and practises vary worldwide. Most patients have minor scoliosis and treatment is generally not recommended for patients with curves <20°, but in many European countries clinicians recommend physiotherapy (scoliosis specific exercises) for smaller curves. The indication for bracing is a progressive curve of 25-45° in a growing child. Its effectiveness has been debated, but in a large recent randomised study, the number of teenagers with high-risk curves who progressed to the threshold of surgery was significantly reduced. Surgery is recommended for patients with curves >45°. Scoliosis surgery was not successful until the introduction of Harrington's instrumentation in the 1960s. Modern instrumentation has evolved from the Cotrel-Dubousset system in the 1980s, and a variety of methods are available today. Although scoliosis may be a burden, long-term studies suggest that a good quality of life is maintained in most patients.

Keywords: School screening, bracing, surgery, genetics, quality of life, physiotherapy.

INTRODUCTION

Idiopathic scoliosis (IS) affects 2-3% of children and usually presents in adolescence. It is a life-long condition and is defined as a structural, lateral rotated curvature (Cobb angle) of the spine of >10° on standing coronal plane radiographs. Adolescent IS usually arises in otherwise healthy children in the growth spurt at the beginning of puberty. The spinal curve is most commonly detected by the family and not by healthcare providers and it is difficult to determine the exact age of onset.[1,2] Age at presentation is therefore more accurate.

Most patients with IS have minor curves and are not recommended any treatment.[3,4] About 90% of those treated are girls.[5,6] IS can be classified as infantile (<age 4 years), juvenile (age 5-9 years), and adolescent (10 years or older).[7] Comprehensive reviews on adolescent IS have been published.[2,8] The aim of the present review is to briefly describe the current knowledge about aetiology, natural course, screening and diagnosis, and outline the evidence of physiotherapy, brace treatment, and surgery.

AETIOLOGY AND DIAGNOSIS

IS is often seen in multiple members of a family. One identical twin may have a large curve while the other has a small curve, which suggests that genetic, epigenetic, and environmental factors are involved. Studies of twins[9] have reported a concordance in 73% of monozygotic twins and

36% in dizygotic twins. Despite a number of genetic studies with different study designs, the aetiology remains unknown. Different methods of inheritance have been reported, but no single locus has been identified.[10] Multiple genes are likely to be involved with incomplete penetrance and variable expressivity. Candidate gene analyses[11-14] have not found associations for connective tissue genes, but for vitamin D and oestrogen receptor genes. Polymorphism of the oestrogen gene has shown association both for the probability of having scoliosis and for curve progression.[13,14] The hypothesis that an abnormality of the paravertebral muscles contributes to the development of IS has not been confirmed.[8] Magnetic resonance imaging (MRI) studies[15] indicate that the growth of the vertebral bodies is disproportionate compared to age matched controls, but the mechanisms involved are poorly understood.

Scoliosis has been a recognised condition for centuries. Structural scoliosis must be discriminated from functional scoliosis that may be caused by leg length discrepancy or back pain in a patient with disc herniation. A structural scoliosis is clinically suspected if it appears as a keel of a boat when examining the patient bending forward (Figure 1). The keel or gibbus is an expression of the rotation of the spine as structural scoliosis, and is of a three-dimensional (3D) deformity.

Figure 1: A structural right convex thoracic scoliosis. The lateral deviation is shown in the left image while the rotation of the spine is shown as a keel of a boat (gibbus) by bending forward in the right image.

Other rare causes of scoliosis should be excluded, such as vertebral malformation, neuromuscular disorders, and syndrome scoliosis. The diagnosis is confirmed if a standing coronal radiograph shows a curve >10°. Several classification systems have been used for the description and development of curves. Most primary curves are right sided thoracic, but primary curves may, by example, be double shaped, thoraco-lumbar, or lumbar. The IS is usually s-shaped with one major and two compensatory curves, while neuromuscular scoliosis is c-shaped. Patients with rare conditions such as Rett syndrome may have either c-shaped or s-shaped curves.[16]

SCHOOL SCREENING

Early detection by screening allows for monitoring curve progression and timely initiation of bracing but school screening is controversial and practices vary worldwide.[17,18] Some studies have supported screening whilst others have discouraged routine screening. Currently most international scoliosis societies support and recommend screening.[19,20] While opponents of screening mainly cite the increased costs and lack of effectiveness of the programmes,[21] discontinuation has led to late detection and more surgeries in various countries.[1,22]

School screening is recommended at the onset of puberty, usually twice, at the ages of 11 and 13 years. Since girls are considered to be at higher risk compared to boys, screening in girls only may be more preferable. Screening can be performed by community nurses or physical therapists and provided in connection with other routinely contacts with the school's healthcare system. The examination includes the forward bending test with the use of a scoliometer to measure spinal rotation. School screening is easy to perform by trained examiners and takes an average of 9 minutes per child.[23] Children with rotation >7° are recommended a standing conventional X-ray examination, and those with a major curve >20° are referred for specialist examination.

The most comprehensive longitudinal school screening was performed in 115,190 children in Hong Kong.[24] Of these, 3,158 received X-rays, 264 were braced, 10 had surgery, and 29 had brace and surgery. Comparatively, in Norway which currently has no screening and has age cohorts of about 60,000, in 2012 there were 51 children who had brace treatment and 71 had surgery. Experiences from a clinical trial[5] on the effectiveness of bracing

suggest that patients and their parents prefer bracing to no treatment. The best way to allow for timely bracing may be to reintroduce screening. The procedure itself is cheap but it may not be cost-effective unless implemented in countries which have high surgical rates.

NATURAL HISTORY

The history of completely untreated IS is unknown.[2] Studies[25,26] that are cited when the natural history is described, may have included braced patients or have follow-up rates <50%. One study[26] that followed patients for 50 years reported that the level of work and disability did not appear different from controls. Although back pain was experienced more often, this was not clearly associated with curve size. Self-image was lower than in controls.

TREATMENT GUIDELINES

Treatment is generally not recommended for patients with curves <20°, but in many European countries clinicians recommend physiotherapy for smaller and moderate curves in addition to brace treatment.[27]

As scoliosis does not increase in a number of patients, progression to a curve size >25° in a growing child is usually required before bracing is recommended. Skeletal maturity is estimated from images indicating the epiphyseal growth plate at the iliac crest, and is classified as Risser Grade 0-5. Indications for bracing include Stages 0-2 and in some cases Stage 3 if other signs of puberty are not present. Bracing is usually maintained until Risser Stage 4-5 or about 2 years after menarche in girls, and until Risser Grade 5 in boys. Risk factors for curve progression are debated, but young onset age, flexible curves, or thoracic or thoracolumbar primary curves tend to develop faster. Surgery is recommended in the growing child with curve size >45-50°. In addition, other factors such as vertebral rotation and curve localisation are considered in the evaluation for surgery.

Physiotherapy

Physiotherapy is favoured in many European countries as first-line treatment of small curves and for those with a low risk of progression. Different methods are available and scoliosis-specific exercises are described as different from physiotherapy in general.[28] A recent Cochrane review[29] reported that there is a low quality of evidence that scoliosis-specific exercises may be more effective than electro stimulation, traction, and postural training, but the authors suggest that more research is warranted. A previous systematic review[30] included more studies of low quality and concluded that scoliosis-specific exercises reduced progression rate and brace prescription in patients in early puberty. At present there is little knowledge about the advantages of scoliosis-specific exercises as compared to participation in regular sports or as an adjunct to bracing. Physiotherapy is commonly used in all patients with scoliosis, both in a long-term follow-up of middle-aged previously braced patients, and of younger operated patients; around 30% reported to have undertaken physiotherapy in the last year.[31-33] The use of physiotherapy is also much debated, and scoliosis surgeons often claim that it is not indicated. High-quality studies examining indications and effectiveness in terms of curve reduction and health-related quality of life (HRQoL) are warranted.

Bracing

Bracing has been used for centuries, the aim is to stop curve progression to avoid surgery. Many different braces are available for the treatment of scoliosis in the growing child or teenager.[34] Principally these can be divided into three types: rigid day and night braces, rigid night braces, and dynamic braces. Advocates of the two latter types argue that these are as effective as rigid day and night braces and more user friendly, but this is not documented in controlled studies. The rigid brace is custom-made from a pre-shaped model and fitted by orthopaedic engineers. A Cochrane review[35] found only one controlled study examining the efficacy of bracing. This study[36] reported that about 25% progressed with brace treatment and 55% without.

The effectiveness of bracing was documented in a large recent randomised study.[5] Bracing significantly decreased the progression of high-risk curves to the threshold of surgery in patients with adolescent IS. Longer hours of brace wear were associated with greater benefit. For the average patient the wearing of a rigid brace did not reduce HRQoL. Patients were recommended to wear the brace for 20 hours daily. The curve reduction in a compliant girl is shown in Figure 2.

Compliance was examined by thermosensors and indicates that patients with brace wear >13 hours

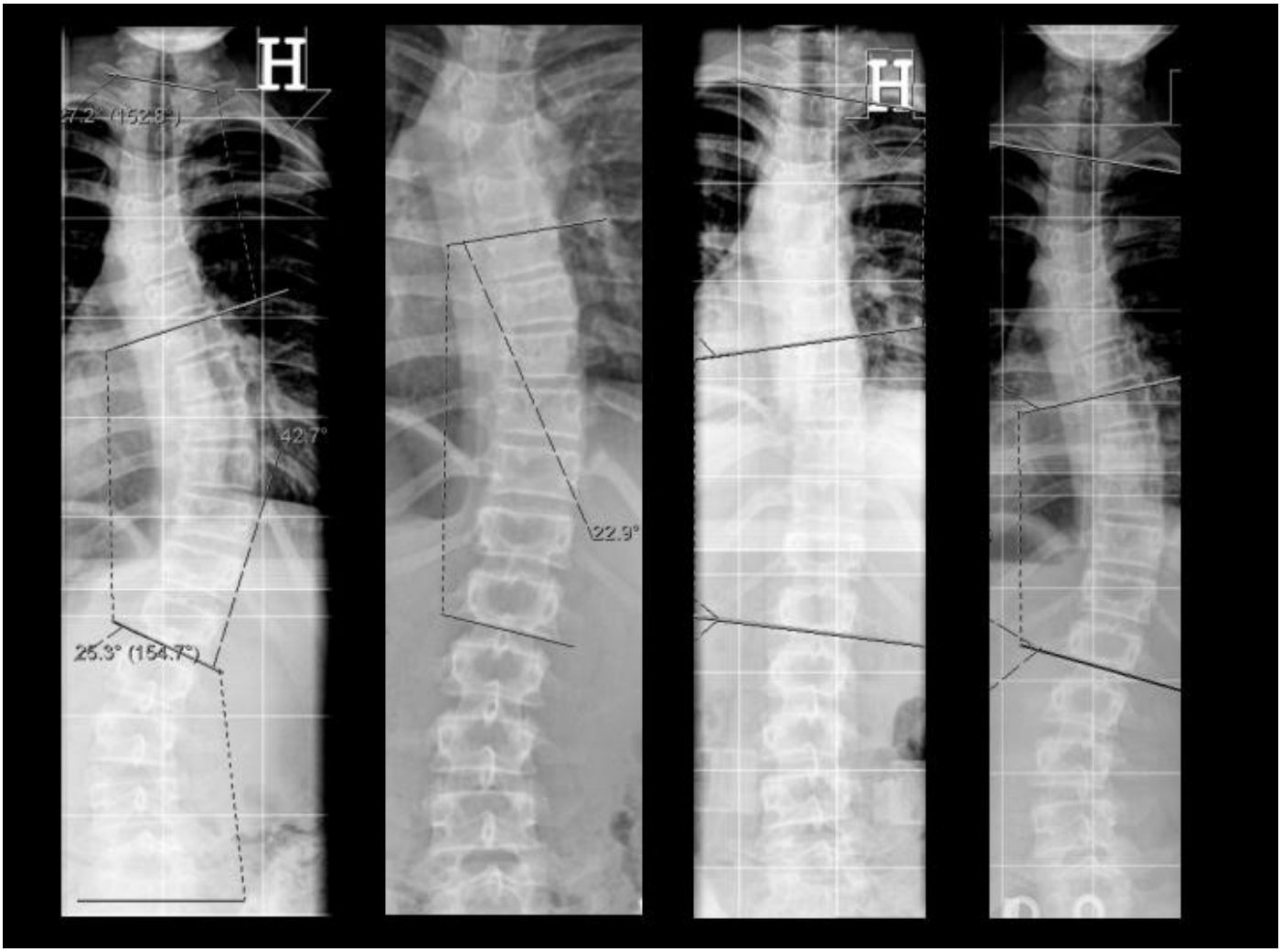

Figure 2: Radiographs showing a right convex primary curve of 42.7° in a girl with adolescent idiopathic scoliosis who started bracing at age 12 years. She had no signs of growth maturity, Risser was 0, and she had not had menarche. She was highly compliant and used the brace about 20 hours daily. The curve was flexible before bracing and was reduced to 22.9° lying prone, to 15.6° in a rigid brace (indicating good brace fit), and the primary curve was 32.0° at brace weaning. She participated in sports 1-2 hours daily.

had a success rate of >90%, while those with brace wear <6 hours had a success rate of 42%. This is in agreement with the results of a large longitudinal cohort study with >20 years follow-up.[6] In this study there were 284 compliers and 71 non-compliers. 68 of the compliers and 41 of the non-compliers had progressed >6° at long-term (OR: 5.8 [95% confidence interval 3.3-10.2]) and 17 versus 10 had surgery (OR: 8.6 [3.7 to 19.9]). The curve progression in a girl who initially used the brace as prescribed but later ended treatment is shown in Figure 3.

Surgery

The aims of surgery are 3D curve correction and improved appearance by balancing the trunk. Scoliosis surgery is major surgery in an otherwise healthy child and it is an overall goal to keep short-term and long-term complications to a minimum. The mortality rate is 1.3 in 1,000 operations and spinal cord injury is reported in about 0.5% of operations.[37,38] Improved preoperative and intra-operative neurophysiologic monitoring and blood salvage procedures contribute to safer surgery. Late infections are usually caused by skin (acne) bacteria and are suspected by onset of pain or signs of a fistula or skin abscess and usually indicate removal of the inserted instruments. Other causes of reoperations are instrument failure or pain. Reoperations are conducted in 5-23% of the patients.[31,39,40] Scoliosis surgery is expensive and total costs are number two of all surgeries in children and adolescents in the USA, next to appendicitis.[5]

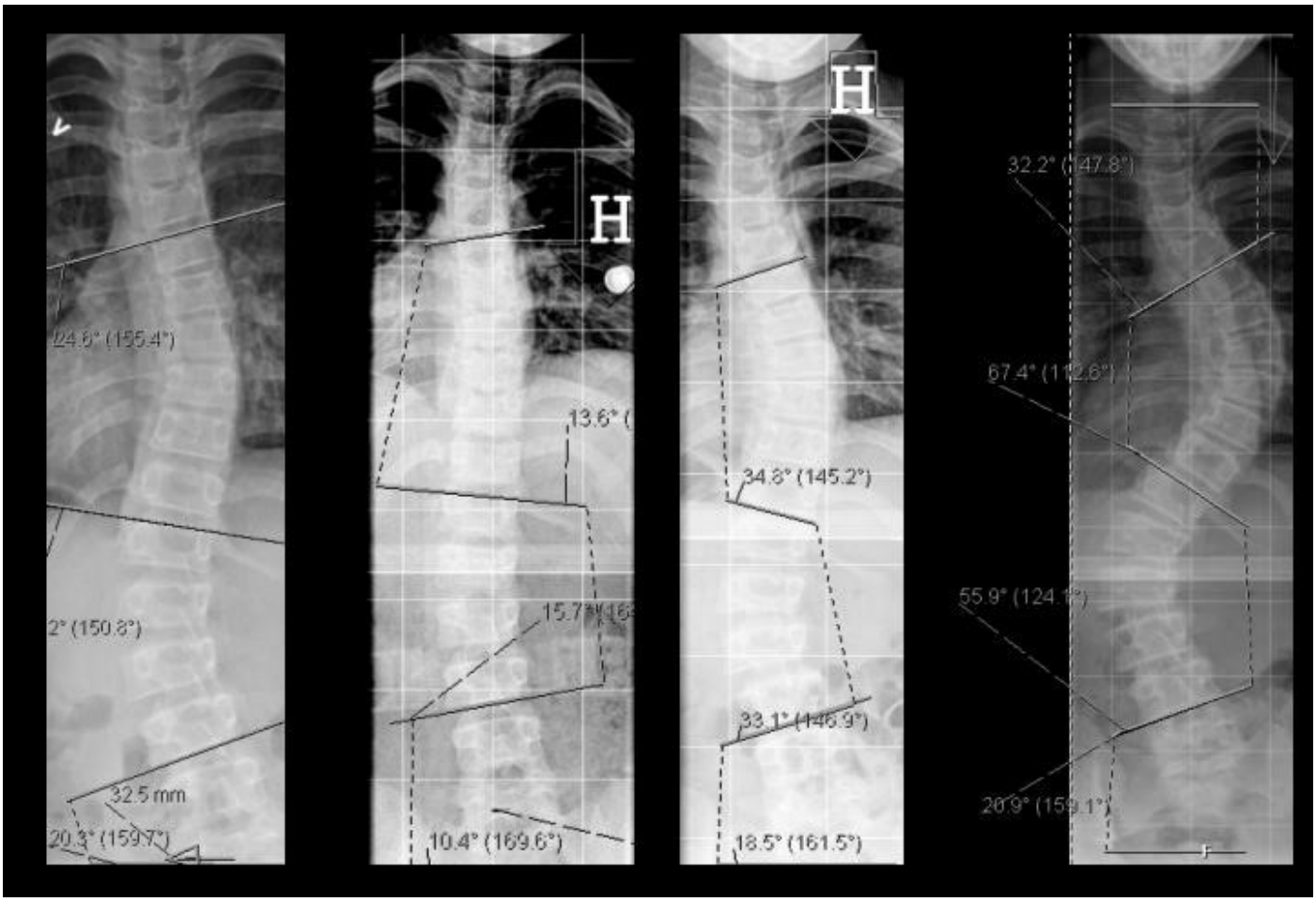

Figure 3: Radiographs show juvenile idiopathic scoliosis in a girl. She started bracing at age 9 years, the double shaped curve was 24.6° and 29.2°, respectively. Curves were flexible and were reduced to 13.6° and 15.7° in a rigid brace. She started to become non-compliant at age 11 years, curves increased to 34.8° and 33.1°. She stopped to use the brace at 12 years of age and did not attend any appointment for 2 years and her scoliosis had increased to 67.4° and 55.9° at 14 years of age, respectively, and she was referred for surgery.

Scoliosis surgery was revolutionised by Harrington's instrumentation in the 1960s.[41] Modern instrumentation has evolved from the Cotrel-Dubousset system in the 1980s.[42] Combinations of wires, hooks, pedicle screws, and long rods are used to conduct modern 3D correction. On average, the major curve is corrected by about 60% of its original size and the instrumentation includes about ten segments. The use of segmental pedicle screws in the thoracic spine has been introduced to improve better fixation.[43] No conclusive evidence exists about advantages in outcome, such as improved HRQoL, including better self-image. Interestingly, a previous study[44] reported no difference in long-term outcome after Harrington's instrumentation compared with Cotrel-Dubousset posterior instrumentation at 10-years follow-up. We examined 86 patients at 10-years follow-up after operative treatment with Cotrel-Dubousset.[31] The average primary curve was reduced from 56° to 19° (Figure 4), five patients had implants removed, and 79% of the patients considered their back function as excellent or good. Despite this, 45% reported to have consulted a physician or received physiotherapy the last year before the 10-year follow-up.

Anterior instrumentation is used mainly for isolated thoracolumbar and lumbar curves. The main advantage is the reduced number of fusion levels. A small study[45] with 17-years follow-up reported good correction, no infection, <10% reoperations, and good scoliosis specific HRQoL.

Casts are rarely used post-operatively today. Patients are usually hospitalised for about 10 days,

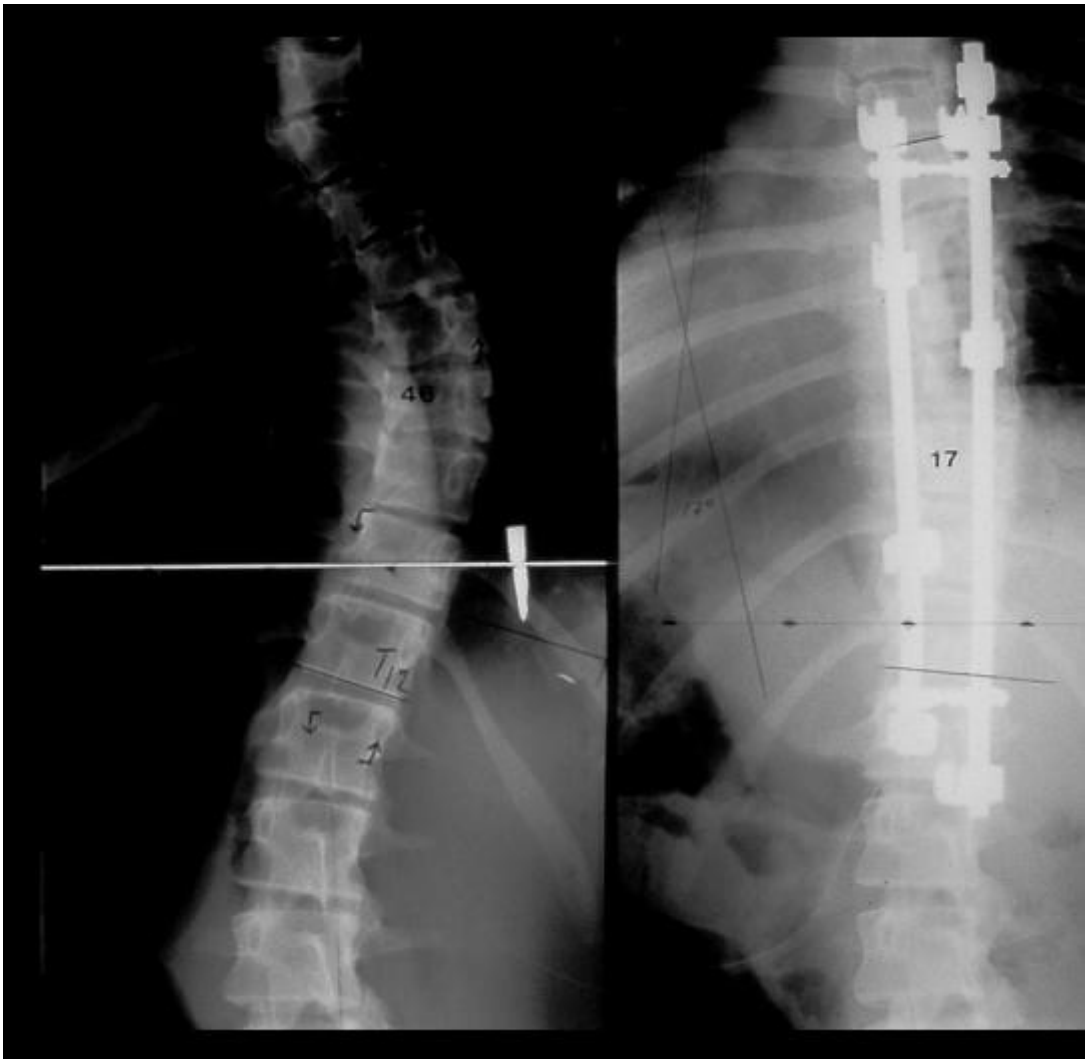

Figure 4: Radiographs that show idiopathic scoliosis in a girl aged 14 years before and after surgical treatment using Cotrel-Dubousset instrumentation.

and thereafter stays at home for another 10 days before the general condition is acceptable for attendance of regular school classes.

HEALTH AND HRQOL

Dyspnoea is associated with curves >80°.[2] Pulmonary function was improved after brace and surgery at 25 years follow-up.[46] Pregnancy, childbearing, and delivery experience of braced and operated patients are comparable to controls.[32,33,47] Spinal mobility is decreased after bracing and surgery, but less after modern instrumentation compared with Harrington's instrumentation.[44,48] Muscle endurance was reduced in one study after bracing and surgery, but muscular strength tests were comparable to controls in another study.[44,48]

Pain is reported more often by operated scoliosis patients than controls without scoliosis.[40,49] Also, a considerable number of patients consulted a physician or had physiotherapy the year before long-term follow-up both after bracing and surgery.[31-33] Average scores of self-image were slightly decreased in both braced and operated patients at long-term follow-up.[31,31-33] QoL was not reduced after bracing and comparable to controls in patients in a recently published clinical trial.[5]

HRQoL is measured by various scoliosis-specific questionnaires. Different self-report outcomes are available. The most commonly used is the Scoliosis Research Society questionnaire, which assess five domains (physical function, pain, self-image, mental health, and patient satisfaction).[50] This questionnaire is validated for use in many different countries. For scoliosis patients this questionnaire is more accurate and valid compared with a generic questionnaire such as the EQ-5D.[51] Reporting of

scoliosis-specific HRQoL in scoliosis patients is influenced by co-morbidity.[52]

CONCLUSION

The aim of the present review is to briefly describe the current knowledge about aetiology, natural course, screening and diagnosis, and outline the evidence of physiotherapy, brace treatment, and surgery. Despite a number of genetic studies with different study designs, the aetiology remains unknown. The natural course is difficult to outline in the long-term because many patients have been braced. Available evidence suggests that QoL in most patients is, in the long-term, beneficial. The indication for school screening is to detect patients who benefit from bracing. The cost-benefit of screening is debated. Scoliosis is idiopathic in most patients, but curve patterns and other causes should be carefully evaluated to exclude other causes. The evidence of physiotherapy for small curves is sparse, while a recent milestone trial documented the effectiveness of bracing. Surgical methods for operating on large curves are continuously revised and should preferably be evaluated by high-quality studies before implementation.

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