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Case Study

Scoliosis in adulthood—a case with untreated early onset scoliosis presenting at the age of 76 years

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Abstract. [Purpose] Untreated early-onset scoliosis may eventually progress to more than 90° after growth, cause severe health problems, and increase chance of mortality. Therefore, surgical intervention is often indicated prior to the development of a life-threatening deformity. This case report aims to reveal how a 76-year-old male patient with curves exceeding 110° is functioning with minimal difficulty. [Subject and Methods] The patient, who has never had surgical intervention for scoliosis, can perform his everyday activities. His curves were 111° thoracic and 118° lumbar when he presented at the author's office in January 2015. [Results] The patient reported that he rarely needs a physician and participates in endurance sports like jogging. Despite this, the patient recognizes his restrictive ventilation disorder (shortness of breath) when he is inactive. The patient complained of shortness of breath and cosmetic concerns. [Conclusion] The current guidelines indicate that early-onset scoliosis should be operated at a young age; however, this protocol is not supported by high-quality evidence. Notably, patients with curvatures exceeding 100° after puberty may have a reasonable quality of life when they lead an active life with regular participation in endurance sports and physical rehabilitation.

Key words: Early onset scoliosis, Endurance sports, Physical rehabilitation

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INTRODUCTION

Patients with untreated adolescent idiopathic scoliosis (AIS) rarely progress to a degree of curvature which may cause a significant impairment in general health¹). Aging patients with AIS generally function well in a 50 year follow-up of untreated scoliosis²). However, early-onset scoliosis (EOS), which typically exceeds 90° after growth when left untreated³), can lead to severe health problems (mainly cor pulmonale) and an increased chance of mortality^{4, 5}). Therefore, surgical intervention is often indicated for these cases before a life-threatening deformity develops⁶).

A case series⁶⁾ and several long-term case reports^{7–11)} have demonstrated that while patients who have had spinal fusion surgery may function well later on, they regularly need re-operations. Few publications have shown untreated cases in aging patients and curvatures exceeding $100^{\circ 1, 4, 5}$. The present case report aims to demonstrate how an untreated 76-year-old male patient with 118° lumbar/111° thoracic scoliosis is functioning.

SUBJECT AND METHODS

A 76-year-old man with severe curvatures of 111° thoracic and 118° lumbar presented at the office of the author in January 2015 (Fig. 1). His scoliosis was first observed before he was 4 years old. The patient reported having no brace treatment or surgery over the course of his lifetime and rarely needing a physician. He participated in regular sports activities and was treated by a physiotherapist in Germany for his scoliosis. His office visit marked the fourth time in his life he has had radio-

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graphs. He stays active by participating in endurance sports like jogging but recognizes his restrictive ventilation disorder (shortness of breath) when he is inactive. The patient never considered scoliosis surgery. His only complaints are shortness of breath and cosmetic concerns.

RESULTS

The sole reason for the patient's visit was to inquire whether his exercises and sports are appropriate for his condition. The author examined the patient clinically and suggested a radiograph to determine the appropriate advice. The patient initially did not want to undergo radiography, but consented after the author explained its necessity in the absence of any of the patient's previous radiographs. The radiograph revealed a well-balanced double major curve pattern. The angle of trunk rotation at presentation exceeded 30° on both curves. The patient's forced vital capacity (VC) is 1.4 liters—less than 50% of the predicted value, when measured according to his current height. When estimating the patient's expected height without scoliosis, his VC is less than 30% of the predicted value. The diagnosis on the record (Fig. 2) was "severe restrictive ventilation disorder." Dyspnea is evident while talking as he draws breath sharply in between his sentences.

DISCUSSION

Winter and Lonstein⁶ reported a case of long-term follow-up after early surgery for pediatric spinal deformity. Their case series includes patients with scoliosis of different etiologies. Their non-operative cases show a poor outcome. One patient, with a decompensated single thoracic curve, suffered a collapsing spine and early death due to severe cor pulmonale.

Several case reports^{7–11} conclude that early surgery is necessary to achieve the best possible results. However, nearly all of the patients required reoperation and some experienced disabling chronic pain. Alternatively, the patient discussed in this case report demonstrates that not all patients with EOS would choose scoliosis surgery retrospectively, even more than 70 years after the initial diagnosis.

Given the contrasting cases presented, stating that scoliosis surgery is lifesaving is definitively impossible⁷. Some patients with curvatures exceeding 100° after growth, such as this man, are living a reasonable life without surgery, although not without distinct challenges.

The current state of scoliosis literature largely ignores this patient population for various reasons. A lack of follow-up may occur between patients and their physicians, particularly those who resist surgical intervention. Patients who were recommended for surgery may also seek alternative treatments or no treatment at all. However, to date, parents of children diagnosed with EOS are less likely to refuse scoliosis treatment. The reason for this is that evidence now exists showing that scoliosis may potentially progress without intervention, especially in high-risk cases such as EOS¹). Furthermore, bracing can successfully halt or slow curve progression for patients and families not willing to consent to surgery¹²). Newly developed 3D braces supported by specific exercises have been shown to be highly corrective and can be used to successfully treat early-onset pediatric spinal deformities^{12, 13}. This computer-aided bracing technology, designed according to curve classification¹⁴), offers the possibility of improving EOS during the pubertal growth spurt, not just stopping curvature progression (Fig. 3).

At present, no evidence suggests that scoliosis surgery is superior to the natural history of the condition^{15–19)}. Moreover, the long-term effects of spinal fusion may lead to a high reoperation rate^{15, 20, 21)} and other long-term complications^{21, 22)}. When treating EOS and congenital scoliosis²³⁾, high-quality conservative treatment²⁴⁾ should be attempted to avoid spinal surgery. In light of new developments in the field of scoliosis research, only a small subset of patients with early-onset spinal deformities treated with bracing¹⁴⁾ may require scoliosis surgery.

Finally, the patient from this case report never consulted a spine surgeon during the course of his adult life. This implies that a subset of EOS patients who have declined surgery exists. As a result, these patients are not included in studies put forth by spine surgeons. In addition, we have to consider that patients suffering from adverse effects of spine surgery are often lost to follow-up²⁵⁾. This may cause a reasonable bias in publications on this topic.

A case report is of limited evidence. However, a phenomenon exists wherein untreated scoliosis patients with curvatures exceeding 100° may have a reasonable quality of life and would not choose to undergo surgery. More cases of untreated patients and curvatures exceeding 100° need to be found and published to gain more evidence.

In conclusion, the surgery practice for EOS is not supported by a high-quality research. Patients with curvatures exceeding 100° after puberty may have a reasonable quality of life when they lead an active life with regular participation in endurance sports and physical rehabilitation. In the absence of symptoms, some patients do not want surgery. Severe cases, as reported in literature, now have the potential to be managed with high-quality conservative treatment rather than immediate surgery.

Conflict of interest

HR Weiss is receiving financial support for attending symposia and receives royalties from Koob GmbH & Co KG. The company is held by the spouse of HR Weiss.



Fig. 1. *Left:* Clinical picture of the patient described in this case report, from the back. *Right:* X-ray taken at the time of the latest presentation in January 2015. The Cobb angles of the patient were 111° thoracic and 118° lumbar.



Fig. 3. *Left:* Early onset scoliosis patient with a curvature of 48° at the start of the pubertal growth spurt. She has been treated with a computer-aided design Chêneau-style Gensingen brace. *Right:* After brace weaning, she appears more balanced with a residual curvature of 24°.



Fig. 2. Patient's pulmonary function report (January 12, 2015). The report is in German and is used for docuentation purposes only. His forced vital capacity measured 1.4 liters, which is 47% of the predicted value. When measuring predicted value according to the patient's expected height without scoliosis, his forced vital capacity is less than 30% of the predicted value. The recorded diagnosis was "severe restrictive ventilation disorder."

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