CASE REPORT

Halogravity traction in the preoperative treatment of scoliosis in twins with Marfan syndrome

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SUMMARY

We report on the influence of the duration of halogravity traction for achieving curve correction in monozygotic twins with Marfan syndrome who underwent posterior spinal fusion. Review of the medical charts and standard radiograph analysis of twin girls treated at our department was performed. Halogravity traction with a four-pin skull construct was applied for 3 weeks in twin A and for 2 weeks in twin B with a maximum of 20% body weight used. Both were on a 24-hours-day halogravity traction regime. Achieved thoracic curve correction after halogravity traction was 31% in twin A and 18% in twin B. Although less curve correction after traction was achieved in twin B, this had no significant implications on final postoperative curve correction. Halogravity traction can be a useful tool in the preoperative treatment of scoliosis in patients with Marfan syndrome if applied for 3 weeks. In order to avoid complications, we propose that lower weights be used with a starting weight of 1.5 kg increased by 1 kg daily until 20% body weight is reached.

BACKGROUND

Scoliosis is a common feature in Marfan syndrome, and observed in up to 62% of affected patients;^{1–6} it is refractory to bracing, with only a 17% rate of success.⁷ Operative treatment is followed by higher perioperative risks, instrumentation-related complications and more reoperations compared with adolescent idiopathic scoliosis.^{8–10} More levels of surgical correction, more distal fusion and greater correction of sagittal balance are required, making the operation more demanding.^{8–9–11} Anterior release followed by posterior instrumentation was proposed in order to improve curve flexibility and enhance postoperative correction, but with reported 14% of respiratory complications.^{12–13} An alternative for anterior release is halogravity traction, a method of applying gradual, sustained traction in order to make surgery less complex, to improve safety and to maximise postoperative correction. It is well-documented in treatment of severe scoliosis and kyphoscoliosis.^{14–19} Although its application in patients with Marfan syndrome could also be beneficial, the literature is scarce. In addition, only Ogden and Southwick²⁰ reported of Marfan twins with scoliosis but without the use of halogravity traction. We present preoperative application of halogravity traction in twin girls with Marfan syndrome. This paper is intended to show influence of the duration of halogravity traction for achieving curve correction in monozygotic twins with Marfan syndrome.

CASE PRESENTATION

Eighteen-year-old white monozygotic twin girls presented at our department with leptosomic body type and marfanoid features, including long limbs and long thin fingers (twin A: height 176 cm, weight 43 kg, body mass index (BMI) 13.9 kg/m²; twin B: height 182 cm, weight 50 kg, BMI 15.1 kg/m²). The twins were diagnosed with Marfan syndrome in early childhood with mitral valve prolapse, moderate-to-severe pulmonary dysfunction and high myopia. Both developed severe and rigid scoliosis that was refractory to bracing, and had a rapid progression. Plain radiographs were obtained and revealed severe right thoracic curve in twin A (figure 1A). Twin B had severe left thoracic and right lumbar curves (figure 1B). Plain radiographs on bending showed only approximately 10% of correction in the thoracic curve (table 1).

In addition, MRI obtained of the twins showed dural ectasia in the lumbar region, a common occurrence in Marfan syndrome.

TREATMENT

At the age of 18 years, first twin A, and after 5 months twin B, were treated at our department.

The surgical plan was for halogravity traction for 3 weeks prior to the planned posterior fusion. Four skull pins were inserted in standard procedure with local anaesthesia, and halogravity traction began the next day. The starting weight was 1.5 kg, increased at a slow rate of 1 kg/day, as tolerated, to a maximum of 10 kg (20% of body weight). If the patient showed intolerance, the weight was

Figure 1 Plain radiograph of (A) twin A and (B) twin B.

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Table 1 Results of halogravity traction and surgical correction in twins with Marfan syndrome

Curve	Twin A		Twin B	
	Th5–Th12	L1–L5	Th6–Th11	Th12–L5
Preoperative Cobb angle (°)	98	45	79	78
Bending Cobb angle				
Measured Cobb angle in bending (°)	85	25	73	50
Bending correction (%)	13	44	8	36
Achieved correction in halogravity traction				
Cobb angle at last day in halogravity traction (°)	68	38	65	67
Correction achieved (°)	30	7	14	11
Correction achieved (%)	31	16	18	14
Height increment	6 cm (176 cm preoperative)		4 cm (182 cm preoperative)	
Spinal instrumentation	Th5–Th12		Th6–L5	
Postoperative correction				
Postoperative Cobb angle (°)	49	25	42	43
Correction achieved (°)	49	20	37	35
Correction achieved (%)	50	44	47	45

increased by only 0.5 kg/day. The twins were both in a 24-hours-day halogravity traction regime in a halogravity traction bed, wheelchair and a walker frame. A physiotherapist had them perform exercises on a daily basis. Support by a psychologist was given before the initiation of the halogravity traction and, as needed, until the end of the treatment.

OUTCOME AND FOLLOW-UP

The halogravity traction was stopped after 2 weeks in twin B because of psychological discomfort and intolerance, which was not overcome despite daily support from the psychologist, while the planned 3-week regimen was completed by twin A. Twin A's thoracic curve was corrected by 31% (figure 2A), while twin B had less correction of 18% (figure 2B). Halogravity traction was

ceased in the operating room, under general anaesthesia, at the initiation of surgery in both twins. Posterior fusion with hybrid instrumentation was undertaken with the achieved curve correction of around 50% in both twins (figure 3A and B).

A satisfying outcome was retained after 2 years of follow-up, with a loss of thoracic correction of 3° in twin A and loss of lumbar correction of 4° in twin B.

DISCUSSION

A low success rate of 17% for brace treatment in Marfan scoliosis and a lower threshold for curve progression are inclined to favour surgery.⁷ The reluctance of the twins' family for surgical treatment postponed the operation until the twins reached the age of 18. There are no strict guidelines regarding duration of halogravity traction, but side effects, as high as 53%, tend to occur if 3 weeks of treatment are exceeded.^{15–19 21–24} Although presenting psychological discomfort, the halogravity traction was discontinued in twin B only after the 2 weeks of treatment were completed, after major correction was obtained, as shown

Figure 2 Radiograph from the last day in halogravity traction of (A) twin A, thoracic curve corrected to 31% and (B) twin B, achieved correction is 18% in the thoracic curve.

Figure 3 Postoperative radiograph of (A) twin A, thoracic curve corrected to 50% and (B) twin B with a correction of 47% achieved in the thoracic curve.

by Clark *et al.*²⁵ Feared complications, as documented in the literature,^{15 22 26–32} were not encountered, probably due to the steady and slow progression of weights of up to 25% of body weight, as reported by Yang,³¹ well-timed psychological support and a skilled team, which provided everyday nursing and hygiene. Lower weights were used because of the connective tissue abnormalities and ligament laxity evident in Marfan syndrome, which present a risk of cervical kyphosis development.³¹ As already mentioned, halogravity traction lasted only 2 weeks in twin B, which is the reason for less correction in her case. Anterior release was considered in both twins, but was rejected because of a relatively high respiratory complication rate^{12 13} as well as potentially disastrous vascular complications.³³ Although less curve correction after halogravity traction was achieved in twin B, this had no significant implications on the final post-operative curve correction.

Learning points

- We propose that halogravity traction in patients with Marfan syndrome start with a 1.5 kg weight, with a maximum daily increase of 1 kg or less, as tolerated, until 20% of body weight is reached.
- In the case of these twins, complications in halogravity traction were not encountered, probably because of the gradual increase to 20% of body weight.
- A satisfying outcome was achieved in both twins by means of applying up to 3 weeks of halogravity traction.
- Halogravity traction can be a safe and useful tool in preoperative treatment of patients with Marfan syndrome in the hands of a skilled and experienced team, and well-prepared patients.

Competing interests None declared.

Patient consent Obtained.

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