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Operative treatment of severe scoliosis in osteogenesis imperfecta: results of 20 patients after halo traction and posterior spondylodesis with instrumentation

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Abstract Correction and stabilisation of the scoliotic spine in osteogenesis imperfecta is difficult. The optimal technique has yet to be determined, since no large series in which a single procedure has been carried out by a single surgeon using a single protocol has yet been described. The charts of 20 patients with osteogenesis imperfecta who had undergone halo gravity traction (HGT) and a posterior spondylodesis with Cotrel-Dubousset (n = 18) or Harrington (n = 2) instrumentation were reviewed. No correction was made at the time of the surgical spondylodesis. The average follow-up was 4.8 years (range 2–10.5 years). The preoperative traction improved the Cobb angle of the scoliosis by 32% (from a mean of 78.5° to a mean of 53.3°) and improved the kyphosis by 24% (from a mean of 56.0° to mean of 42.5°). This correction deteriorated slightly at final follow-up, for both the scoliosis and the kyphosis (mean 57.6° and 44.4° respectively). Few complications were encountered during the HGT period. In 16 cases no complications occurred during the follow-up period. Ambulation and functional ability were upgraded for 7 of 20 patients.

Key words Osteogenesis imperfecta · Scoliosis · Kyphosis · Traction · Spinal fusion

Introduction

Osteogenesis imperfecta (OI) is a heritable connective tissue disease characterised by bone fragility and ligamentous laxity. Scoliosis, blue sclerae, dentinogenesis imperfecta, basilar impression and early onset hearing loss are the main characteristics [17]. Sillence et al. [15] classified OI into four groups based on genetic criteria, clinical symptoms and radiological findings.

The management of scoliosis in OI is a difficult task. The scoliosis usually progresses until severe kyphoscoliosis is present in adulthood [2, 12, 13]. Besides the age-related progression, previous studies have reported a strong correlation between scoliosis and the more severe types of OI [2, 7]. Progressive scoliosis and concomitant chest deformity can be painful and will interfere with self-care. Mobility and vital functions may also decrease [19]. Therefore, correcting the scoliosis, or at least arresting its progression, is important in patients with this overall disabling disease.

Progression of the spinal curvature is hard to restrain, because bracing deforms the chest and ribs probably without any influence on the scoliotic curvature [1, 11, 14, 18]. On the other hand, surgery of the spine in OI is hazardous [16] and difficult due to the extreme bone fragility. Therefore, Benson et al. [2] recommended spinal fusion at a young age with or without instrumentation. Norimatsu et al. [13] argued against correction of spinal curvature in severe scoliosis, as the holding power of bone in OI is very limited.

Gitelis et al. reported a case where partial correction of the spine was obtained using a halo gravity traction (HGT) device preoperatively, which seemed to benefit the final outcome of reduction [10]. The purpose of our study is to evaluate retrospectively the results of HGT and spinal fusion with instrumentation in 20 consecutive patients with OI and severe scoliosis.

Materials and methods

The study included 20 consecutive patients with osteogenesis imperfecta who were treated between 1977 and 1995. Sixteen were operated at the Centre Helio Marin in Roscoff and four at the Hôpital des Enfants Malades in Paris, by one senior author (G.F.), according to a standard protocol. All records were analysed retrospectively. Almost all patients are still seen at the outpatient clinic on a yearly basis.

The patients were classified according to Sillence [15]. Characteristics of the patients are listed in Table 1.

Functional ability was scored according to Bleck [3]. The indication for spinal surgery was pain and/or progression of scoliosis interfering with functional ability. Anteroposterior and lateral roentgenograms of the total spine were made before, bi-weekly during halo traction and postoperatively, while the patient was sitting or standing, depending on their functional capacity. Roentgenograms at the most recent follow-up were also examined by an independent observer (G.J.M.J.). Curves of scoliosis and kyphosis were measured according to the Cobb method in a standardised way [6]. Any kyphosis at the thoracolumbar junction was considered pathologic; a severe kyphosis in the thoracic region was not seen.

All patients underwent the same procedure. The halo was placed in a subequatorial fashion with six to ten pins. At the beginning of our experience we noticed migration of the halo when six pins only were used. We therefore changed to using ten pins. The patients were then mobilised in a standing halo frame. Weights were gradually increased to a maximum of half the bodyweight of the patient. Close clinical observation of neurological symptoms was maintained during the entire procedure.

When no further correction of the spine could be measured on successive roentgenograms, the operation was performed.

Patients were placed prone, with the HGT in situ, using pillows to make their position comfortable. A posterior spinal fusion with Cotrel-Dubousset paediatric (n = 18) or Harrington (n = 2) instrumentation was performed. No attempt was made to correct the spinal curvature during operation. The rods were prebent according to the curvature. Severe osteopoenia of all patients compelled a bone allograft. All bone was harvested from the iliac crest of a close relative, usually one of the parents, within 24 h prior to the intervention. The graft bone was not treated in any way, nor was HLA typing performed. It was kept sterile in a freezer at 70 °C below zero.

Postoperatively, the halo frame was worn for some additional weeks together with a plaster cast, which was worn for 1.5–3 months. Hereafter a body jacket was worn for between 0.5 and 1.5 years, depending on the roentgenograms. Patients remained hospitalised in the rehabilitation centre while in the halo frame.

Mean Cobb angle and standard deviations of scoliosis and kyphosis were calculated before treatment, after HGT, after surgical intervention and at final follow-up. A paired t-test was used to analyse the results of traction and surgical intervention.

Table 1 Characteristics of the patients, presented chronologically, by date of operation (*OI* osteogenesis imperfecta, *CD* Cotrel-Dubousset instrumentation, *HT* Harrington instrumentation)

No	OI	Sex	Weight (kg)	Traction period		Operation		
				Days	Complications	Age (Yr+Mo)	Typ	Complications
1	I	F	30	160	Halo pin infection	14 + 5	CD	–
2	III	F	25	125	Nystagmus and hyperreflexia	14 + 10	HT	–
3	III	F	21	145	–	13 + 10	CD	–
4	III	F	25	100	–	17 + 3	HT	Loss of correction after 4 years
5	IV	F	27	60	–	14 + 4	CD	–
6	IV	F	32	95	–	12	CD	–
7	III	F	23	85	–	13 + 11	CD	–
8	III	F	24	165	–	15 + 8	CD	Pseudarthrosis and rod fracture
9	III	F	18	135	–	11 + 10	CD	–
10	III	M	15	75	–	12 + 9	CD	Loss of correction after 3.3 years
11	III	M	18	87	–	15 + 2	CD	Loss of correction after 1 year
12	III	M	17	71	–	12 + 3	CD	Fracture intra-op
13	III	F	15	63	–	14 + 7	CD	–
14	III	M	27	120	–	14 + 3	CD	–
15	III	F	16	57	–	13 + 3	CD	–
16	III	M	35	71	Migration of halo	17 + 7	CD	–
17	III	F	29	64	–	15	CD	Fracture intra-op
18	I	F	18	42	–	11 + 10	CD	–
19	III	M	35	38	–	14	CD	–
20	III	F	34	41	–	14 + 4	CD	–

A *t*-test was used to analyse the influence of age (median 14 years) on scoliosis, kyphosis, HGT and surgical intervention (α = 0.05). *P*-value <0.05 was considered significant. In double major curves only the largest curve was analysed, as the sample of minor curves was too small for statistical analysis.

Results

The mean period of follow-up was 58 months (range 24–126 months). Mean age at operation was 14 years (range 11.8–17.6 years). Six of the patients were male, 14 female.

The traction period lasted a mean of 90 days (range 38–165 days). The average traction weight was 10 kg (range 5–16 kg), and corresponded to a mean 43% of bodyweight. This construction had a striking effect in enabling some patients, who had previously been non-ambulatory, to walk.

Roentgenograms of the total spine were made before, bi-weekly during halo traction and postoperatively (Fig. 1).

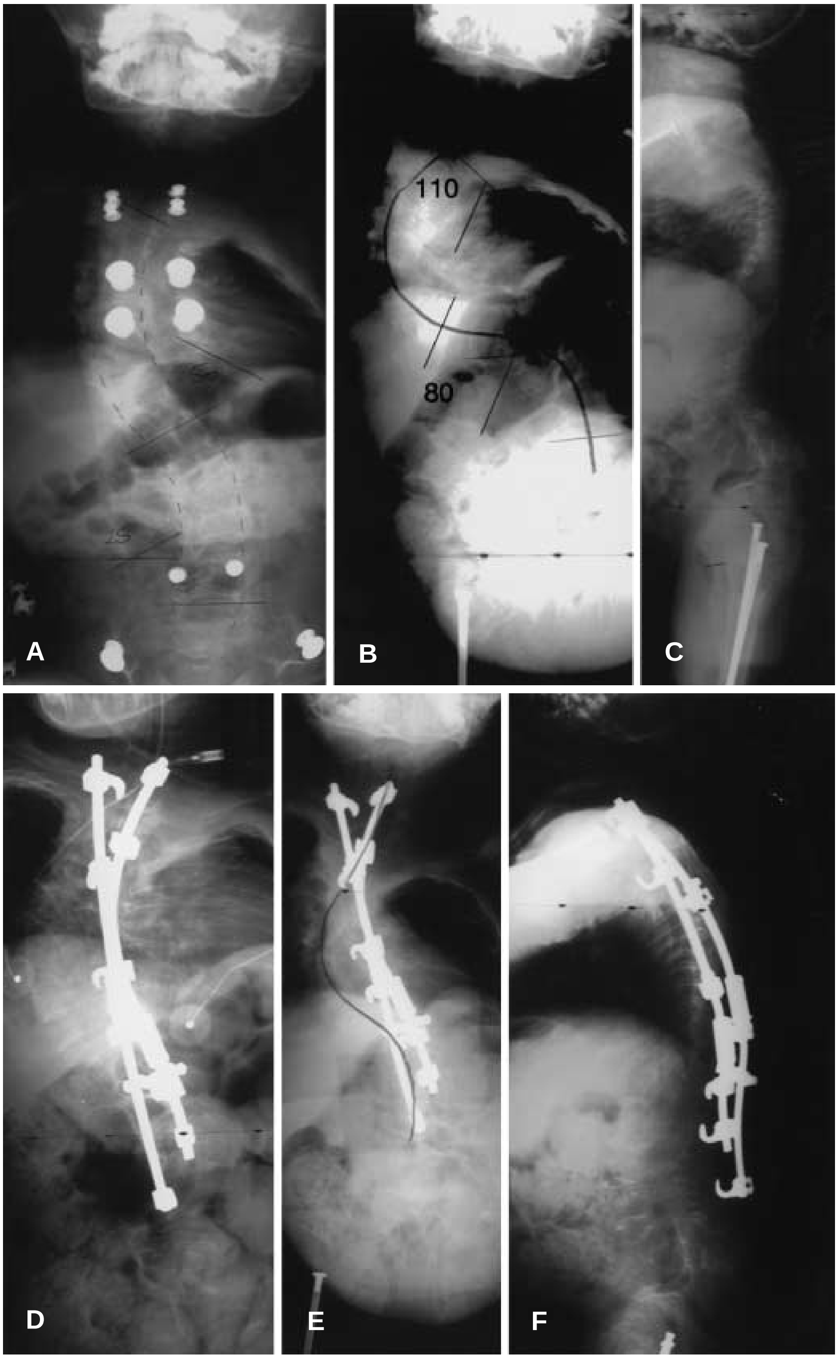

Fig. 1 **A** The spine of a 6-year-old boy suffering osteogenesis imperfecta (OI) type III. No operation was performed at this stage. **B, C** Anteroposterior (AP) and lateral roentgenograms at the age of 12, just prior to a traction period of 2 months. The Cobb angle of the major curve measured 110°, and of the minor curve 40°. A severe kyphosis was present, with a Cobb angle of 70°. **D** Postoperative roentgenogram. The Cobb angle of the major curve is 55° and of the minor curve 35°. **E, F** Two years after surgery the AP and lateral roentgenograms show that the alignment of the spine is maintained without loss of correction despite the pull-out of the distal hook at the time of the surgery due to the fracture of the lamina

Table 2 Details of scoliosis and kyphosis in 20 patients with OI operated after halo gravity traction (*DM* double major, *KS* kyphoscoliosis, *TL* thoracolumbar, – missing data, / no kyphosis)[a]

No.	Area	Pattern	Scoliosis Cobb angle				Kyphosis Cobb angle			
			Pre HGT	Preop.	Postop.	Final follow-up	Pre HGT	Preop.	Postop.	Final follow-up
1	T3-L2	DM + KS	110° 50°	55° 30°	55° 30°	60° 30°	55°	–	–	25°
2	T4-L4	DM + KS	85° 75°	43° 53°	47° 58°	60° 55°	35°	21°	22°	20°
3	T4-L3	TL + KS	70°	40°	36°	40°	55°	35°	32°	35°
4	T4-L3	TL+KS	95°	65°	60°	74°	55°	45°	45°	54°
5	T4-L3	TL + KS	79°	40°	38°	40°	55°	35°	32°	40°
6	T3-L5	TL	56°	40°	40°	45°	/	/	/	/
7	T1-L2	DM	70° 75°	– –	43° 40°	45° 45°	/	/	/	/
8	T3-L5	DM + KS	81° 55°	45° 40°	54° 40°	85° 50°	55°	45°	45°	48°
9	T2-L5	TL + KS	60°	45°	35°	48°	90°	60°	60°	60°
10	T1-L1	TL + KS	85°	75°	75°	82°	75°	75°	75°	75°
11	T3-L2	TL + KS	85°	42°	55°	55°	75°	47°	36°	67°
12	T4-L4	DM + KS	80° 40°	65° 35°	65° 35°	65° 35°	70°	63°	56°	60°
13	Th3-L5	TL	80°	55°	55°	60°	/	/	/	/
14	T4-L4	DM + KS	55° 37°	– –	32° 20°	44° 25°	46°	–	16°	30°
15	T3-L5	TL + KS	83°	67°	85°	80°	70°	65°	70°	75°
16	T3-L5	DM	130° 90°	82° 41°	55° 50°	70° 60°	/	/	/	/
17	T3-L3	DM + KS	78° 88°	48° 40°	42° 33°	48° 30°	28°	28°	35°	35°
18	C7-L5	TL + KS	77°	66°	67°	65°	39°	22°	22°	25°
19	T3-L4	DM + KS	63° 65°	42° 39°	43° 33°	55° 40°	48°	25°	31°	27°
20	T6-L5	DM + KS	60° 60°	45° 27°	29° 25°	30° 25°	45°	29°	36°	35°
Mean Cobb angle			78.5	53.3	50.6	57.6	56.0	42.5	40.9	44.4

[a] In double curves, the upper line relates to the upper curve and the lower line to the lower curve

Before treatment, the average Cobb angle of the scoliosis was 78° (range 55°–130°) (Table 2). This decreased to 53° before surgery, 51° after surgery and 58° at the last follow-up. Mean correction achieved by traction was 25°, which is a 32% correction ($P < 0.001$). At final follow-up, the mean gain had subsided to 19° (25% correction) compared to the curve angle before traction ($P < 0.001$).

The Cobb angle of kyphosis decreased from 56° (range 28°–90°) before HGT, to 42.5° before surgery, and was 44° at final follow-up. Mean correction of kyphosis, comparing the Cobb angle before HGT with that at final follow-up, was 12° (21%) ($P = 0.001$). Here, too, a slight loss of correction occurred after operation. No age-related significant differences in correction of scoliosis or kyphosis were observed in the treatment period.

Complications of the treatment were divided into those occurring during the traction period and those occurring afterwards. At first, only six pins were used for fixation of the HGT. After migration of one halo frame, eight or ten pins were used, and no further cases of migration occurred. One deep wound infection appeared at one of the halo pins, without any sequela. One patient developed a nystagmus and hyperreflexia of the legs, which disappeared after reducing traction weight.

In two cases, a lamina broke during the operation and consequently the hook lost its anchorage and was replaced at an adjacent level. In three cases, failure of instrumentation was noted several years postoperatively (1.0, 3.3 and 4.0 years, respectively) and there was a partial loss of correction. In another patient, one rod broke after 2 years and the other after 3 years, resulting in a complete loss of reduction (patient 8).

In almost all our patients, a reduction of kyphoscoliosis of the spine was achieved, which can result in better

Table 3 Level of ambulation according to Bleck [3] of patients with osteogenesis imperfecta before and after posterior spondylodesis

	Before operation	After operation
Wheelchair	10	5
Exercise walker	2	4
Household walker	5	8
Neighbourhood walker	3	3
Community walker	–	–

sitting balance. Seven out of 20 patients upgraded their ambulation level (Table 3). Four patients upgraded their ambulation level from “wheelchair bound” to “exercise walker”; two patients became “household walker” from “exercise walker”. One patient even upgraded his ambulation level from “wheelchair” to “household walker”. No patient decreased their functional ability.

Discussion

Implant pull-out is the major complication in OI, due to the poor bone quality. Gitelis et al. [10], in their case report, and Yong-Hing and MacEwen [20] described fractures of a pedicle while correcting the spinal curve. Therefore, we performed an in situ spondylodesis with instrumentation after correction by HGT. Our strict postoperative policy of immobilising the spine in a body jacket for 7.5–18 months will further have minimised failure of the instrumentation. Despite these precautions, three hooks pulled out several years after the surgical intervention. On the other hand, the breakage of rods observed 2 and 3 years after, as a result of pseudarthrosis, indicates force can be taken by segmental instrumentation embedded in sufficient bone stock. Retrospectively, the immobilisation of the spine of this very active patient was insufficient, and a pseudarthrosis developed.

Yong-Hing and MacEwen [20] were the first to perform an extensive survey of the treatment of scoliosis in osteogenesis imperfecta. Of interest is the fact that treatment was performed by 51 orthopaedic surgeons in 14 countries. Of the 121 patients treated for scoliosis, 60 underwent spinal fusion; 55 had only posterior fusion, of whom 39 had instrumentation with Harrington rods and 16 had no instrumentation. Average length of follow-up was 3 years. The complication rate was high in patients who had undergone only posterior spinal fusion; 16 of 39 patients had one or more complications. The incidence of complications seemed to be related to the severity of kyphosis.

At operation, stabilisation had a higher priority than correction. The HGT made it possible to correct partially a severe scoliosis preoperatively, so no corrective force by the instrumentation was necessary. As OI is also characterised by ligamentous laxity [1], we assume that this accounts for the preoperative transformation of the scoliotic spine by HGT. The increase in the postoperative Cobb angle of scoliosis at latest follow-up, averaging 7°, is comparable with the 8° postoperative increase in the study of Benson et al. [1]. The mean postoperative Cobb angle of kyphosis deteriorated by 3° with our procedure. Although an anterior release may allow for more correction, in severe scoliosis it can increase morbidity [1].

Segmental spinal instrumentation offers the best distribution of forces in an osteoporotic spine, as in OI. Bradford prefers Luque rods and sublaminar wires, but at the moment no results of this technique have been reported, to the authors’ best knowledge [4]. Hanscom et al. recommended the use of segmental instrumentation and obtainment of minimal correction [11]. Unfortunately only three patients were reported with this technique, all with worsening curvatures at follow-up. The main goal is the distribution of forces along the spinal column. So nowadays it is our opinion that the use of segmental instrumentation by sublaminar wires and/or hooks is the best way to achieve this.

Yong-Hing and MacEwen described the use of methylmethacrylate cement (MMA) in four patients, but draws no conclusion about this technique [20]. Hanscom et al. described a patient who developed a pseudarthrosis stabilised with Harrington rods and MMA [11]. Although MMA increases hook purchase [18], it prevents development of fusion mass. As no corrective force was necessary by the method described, we preferred a spondylodesis performed by allograft bone laid around the rod and hooks. This gives a more biological fixation in time. Although no massive immune response was seen after the bone graft from a close relative, the same amount of bone stock can be obtained using freeze-dried, commercially available bone graft.

The use of HGT in scoliosis is still in dispute. Edgar et al. [8] concluded in 1982 that preoperative traction is only necessary in a severe and rigid scoliosis, and roentgenographic assessment during this traction period is useful. In this retrospective study of consecutive patients, HGT was chosen because of the particular problems associated with this disease. Due to the treatment protocol, no comparisons can be made with treatment without traction. Possibly, the newer correction and fixation techniques, such as Luque wires or Cotrel-Dubousset instrumentation, do make it possible to obtain correction without using preoperative traction, but at the moment no comparative data are available.

Mobilisation of the patient was possible a few days after surgery by using HGT. This is in sharp contrast to the patients reported by Yong-Hing and MacEwen [20], who were kept supine for an average of 2.9 months. Despite the long period of HGT, neither severe complications nor any severe discomfort of the patients was noted.

Potential benefits of correction and stabilisation of the spine include better sitting balance. Due to their laxity, a significant proportion of patients have a collapsing spine, which can be stabilised by spondylodesis. This was achieved in almost all our patients. Improvement of functional ability and ambulation level seldom occurs in severe OI patients after the first decade [5]. In these patients, a plateau phase in functional ability is frequently observed [9]. Therefore, we hypothesize that the higher level of functional ability in our patients is mainly due to spinal correction and stabilisation.

Being able to walk during the traction period also improved the general muscular condition of the patients. More importantly, it improved their mental status. In this period they achieved goals that had previously seemed unreachable.

Conclusion

The major goal for operative treatment is spinal correction and stabilisation, resulting in better self-care, mobility and sitting balance of the individual patient with OI. In our study we described an overall improved sitting balance and an increased functional ability in 7 of 20 patients after surgery. In 16 out of 20 cases no complications occurred during the follow-up period. Although partial loss of correction seems inevitable, operative stabilisation is possible.

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