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GRAND ROUNDS

Slow correction of severe spastic hyperlordosis in an adult by means of magnetically expandable rods

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Abstract

Background We describe a case of severe and progressive lumbar hyperlordosis (160°) in a 28-year-old female university student with cerebral palsy. Her main complaints were abdominal wall pain and increasing inability to sit in her custom wheelchair.
Method When deciding on our opinion about the most promising treatment strategy, we contemplated slow continued correction by means of percutaneously expandable magnetic rods (MAGEC) after the index surgery as a key component of a satisfactory correction in this severe and rigid curve. After an initial radical release and partial correction, a release and correction procedure was required for the bilateral hip flexion contracture. A final in situ posterior fusion was performed as a second spinal procedure, once the desired final correction at 66° of lumbar lordosis was achieved.
Result Three years after the completion of surgery, the patient has a stable clinical and radiological result as well as a solid posterior fusion on CT.
Conclusion This is the first case published in which percutaneous magnetic distraction was successfully used in an adult patient.

Keywords Cerebral palsy · Hyperlordosis · Spasticity · Magnetically controlled growing rods · Slow deformity correction · Viscoelastic

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Case presentation

A 28-year-old female university student with cerebral palsy presented to our spine clinics with slowly progressive abdominal wall pain and increasing difficulty of sitting in her customized wheelchair.

According to her history, pain and a subjective curve increase had been noted over the previous 2–3 years and various attempts with bracing and with botulinum toxin

Fig. 1 Lateral photograph in the supine position at the time of surgery (under general anaesthesia, bottom image) and taken 12 years before (top image)

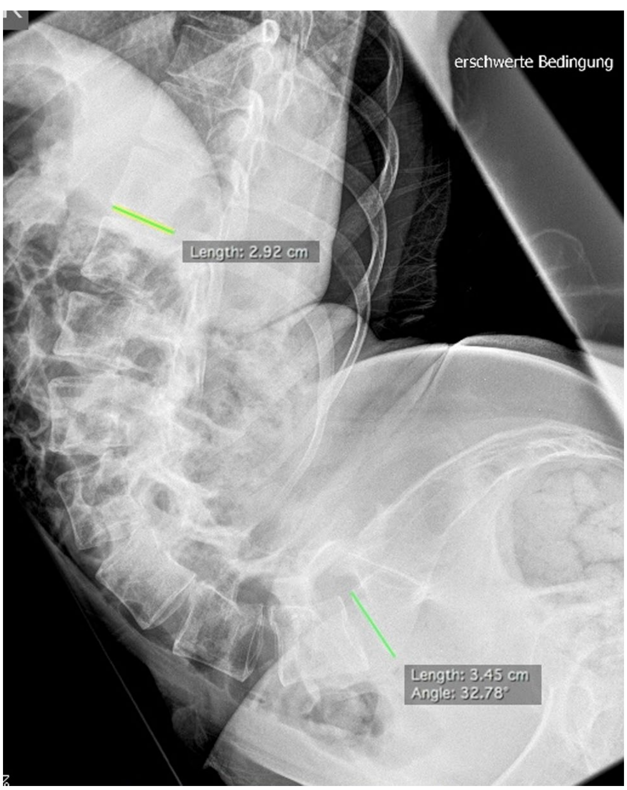

Fig. 2 Lateral radiograph of the lumbar spine acquired in a supine position 6 months prior to the presentation. At this point, the lordosis angle measured only 150°

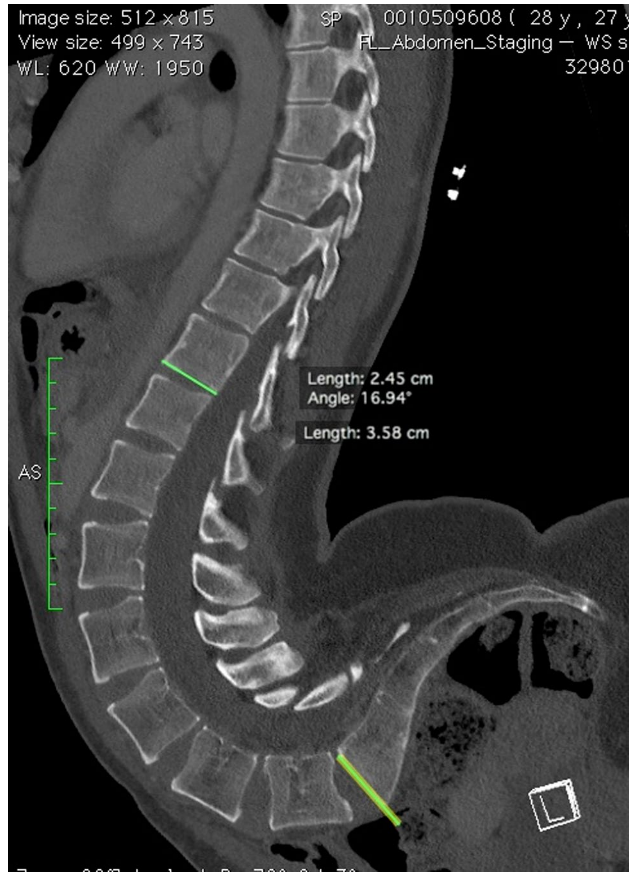

Fig. 3 Sagittal reconstruction from a CT scan acquired a few weeks prior to surgery. The lordosis angle now measures above 160°

injections had been unsuccessful in halting the progression of the deformity.

Her motivation to seek treatment was both the pain and the increasing impairment of her university studies by the limitation in being able to sit.

Clinically, there were an apparent severe hyperlordosis, flexion-adduction deformities of both hips and a visual pulsation of the skin below the umbilicus (video 1), which was also the region where the abdominal wall pain was mostly located.

A firm resistance was palpable directly under the skin in the proximity of the pulsation.

Personal photographs taken 12 years before showed a much less pronounced lumbar lordosis than upon presentation (Fig. 1). Radiographs, CT and MRI gave evidence to a lordosis angle of 160° without any major coronal deformity (Figs. 2, 3, 4). Angio-CT and duplex ultrasound demonstrated that the aortic bifurcation, the confluens of the iliac veins as well as the stomach and possibly the duodenum were stretched out in front of the spine and pressing against the abdominal wall (Figs. 5, 6, video 2).

Initially a primary anterior approach as described by several authors [1, 2] appeared tempting on biomechanical grounds because of the tall, distracted discs. Resecting these would have led to significant anterior column shortening and mobilization, allowing for at least a good partial correction. Once the Angio-CT and the duplex ultrasound exam were performed, however, we did not pursue this avenue further because of the in our opinion prohibitively high risk of vascular and abdominal complications.

When considering a primary posterior approach, we were aware that even a radical soft tissue release as well as a complete mobilization of the spine would probably not permit a sufficient correction in a single-stage procedure. On accepting that a minimum of two posterior surgeries would be necessary, we considered to attempt a radical release with a slow correction by means of percutaneously expandable magnetic rods and discussed the off-label application of that paediatric spine technology with the patient. We obtained a fully informed consent and agreed to decide intraoperatively, and depending on the correction that would be attainable during that procedure whether or not a continued percutaneous correction after the initial surgery should be performed.

In the first surgery, a radical posterior soft tissue release was performed, consisting of a stepwise release of the erector spinae muscles from the posterior iliac crest, intermittent partial incisions of the lumbar and the muscle fasciae at multiple levels as well as partial transections of the erector spinae muscles at multiple levels. At the same time, a pelvic base, consisting of bilateral S1 and iliac screws, was constructed as well as a thoracic base consisting of bilateral T12 and T9 pedicle screws and sublaminar bands at T10 and T11. A temporary rod was anchored by means of laminar hooks between a lumbosacral and a thoracic crossbar for slow and stepwise distraction, while the release procedure continued with the spine: here all lumbar and lumbosacral facet joints were released, the interspinous ligaments were completely resected and several ankylotic laminae were osteotomized as required and separated, much like with Ponte osteotomies. Eventually, all lumbar segments could be mobilized, but despite all efforts and under severe soft tissue tension in the sagittal plane, we had to accept that clearly less than 90° of lordosis could be achieved.

We then constructed a hinged construct with two 5.5-mm standard MAGEC rods (Ellipse Technologies, Aliso

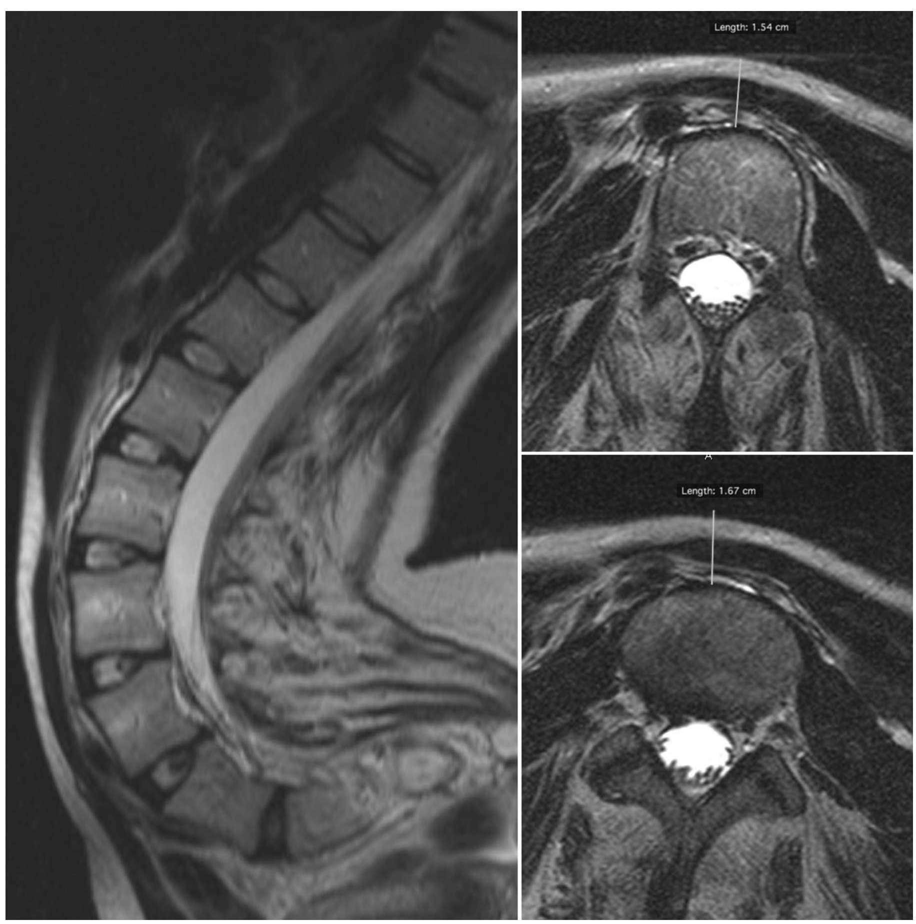

Fig. 4 Preoperative MRI (T2 sequences in sagittal and transverse orientations). The cauda equina appears normal; the lumbar discs are hyperdistracted and strongly wedged with a straight anterior annulus and with the posterior annulus being mostly inapparent. At the apex, the anterior vertebral cortex is measured as being 16 mm below the skin

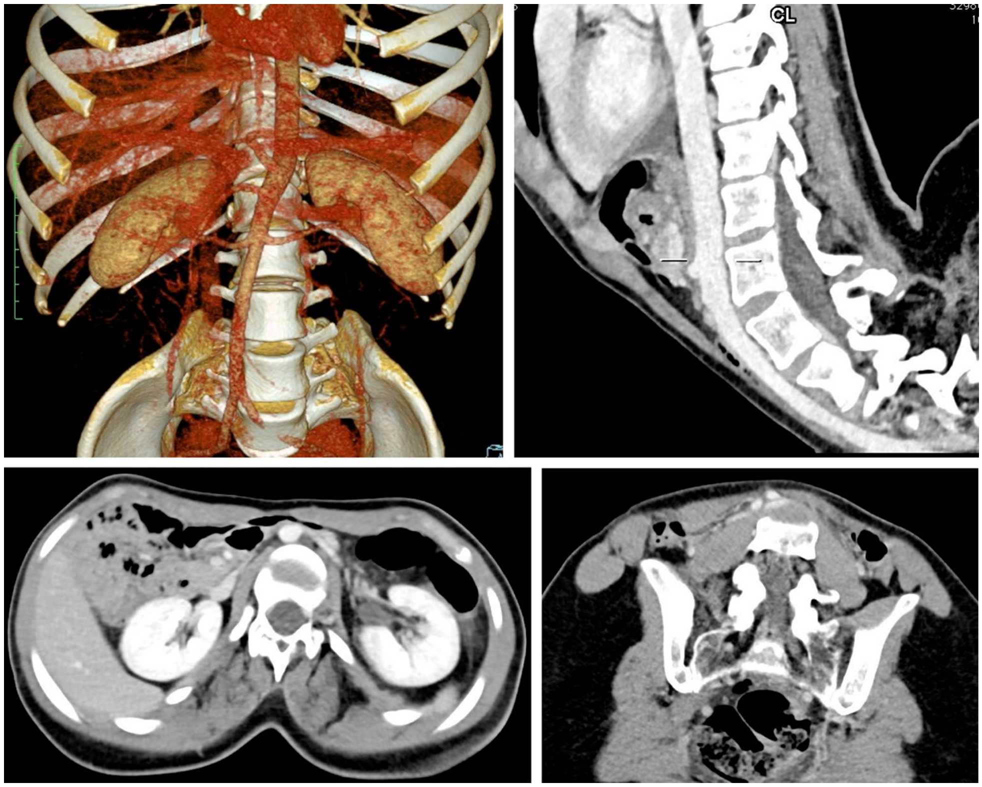

Fig. 5 Images from an Angio-CT that was performed to help decide on the best surgical strategy. Top left: 3D reconstruction. Top right: sagittal plane view, showing aorta and a gas-filled abdominal organ (stomach or duodenum) stretched out over the apex of the deformity and tenting the abdominal wall. Bottom left: cross section at the level of the renal arteries. Bottom right: cross section at the level of the aortic bifurcation

Viejo, CA, USA; local reseller: Orthovative GmbH, Gmund, Germany) with the motors positioned subcutaneously and with the rods connected to the crossbars by laminar hooks to maintain articulation. The hooks were secured with FibreWire® ligatures to prevent them from accidental disengagement from the crossbars.

Probably because of the high intraabdominal pressure on the operating table caused by the deformity and particularly the pressure on the liver, the patient suffered transient liver failure and a subsequent systemic inflammatory response syndrome (SIRS), from which she fortunately recovered within a few days. After complete wound healing which was uneventful and with the patient more frequently sitting, a radiograph showed some further spontaneous improvement to a lordosis angle of 85° (Fig. 7). Over the following 3 months, the rods were expanded on six occasions and always until the maximum tension of the system was reached, as indicated by an audible and palpable clicking of the proprietary magnetic actuator that controls the magnetic motor within the rod itself [3]. As the deformity slowly corrected further, the actuator was ultimately distracted by a total of 16 mm on the right and 12 mm on the left side. With the hip flexion deformities increasingly becoming a problem because of the spinal deformity correction, she then required a hip release and a rectus-psoas transfer to straighten her hip joints.

After she had fully recovered from the intercurrent hip surgery and after further distractions, we assessed her sagittal plane balance when standing and walking with help and decided to finalize the correction at a lordosis angle of 66° to maintain a sufficiently positive SVA (Fig. 8). This was done by performing a standard posterior instrumented fusion from T9 to the pelvis in exactly the achieved position and without any corrective tension brought onto the implants. The explanted MAGEC rods were visually intact and without any surrounding metallosis; there were no implant-related complications. During the subsequent 2 years of the follow-up, there was no appreciable loss of correction in the lumbar spine but a reduction in thoracic kyphosis from 36° to 26° (Table 1). This could potentially be interpreted as a reciprocal change in the unfused portion of the thoracic spine in response to the lumbar correction. There are limitations to the precision and the comparability of measurements of the mobile spine between supine studies (CT) and sitting

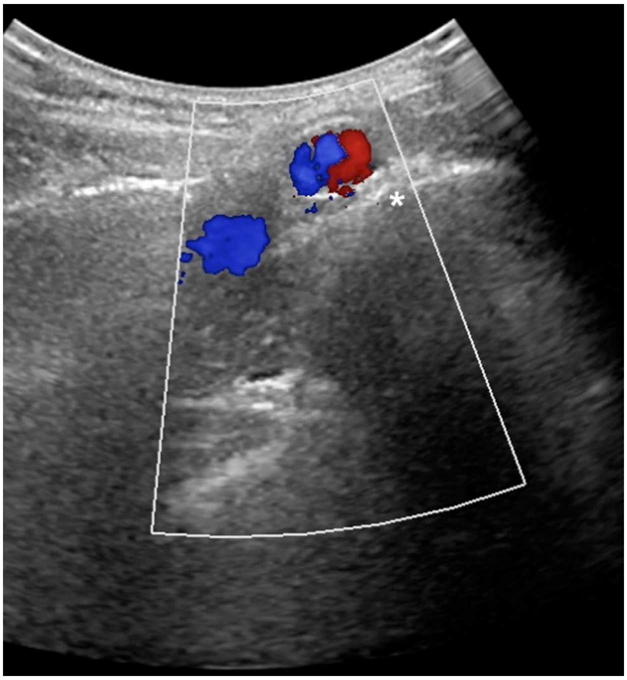

Fig. 6 Color-coded duplex ultrasound in cross-sectional orientation at the apex of the hyperlordosis. The asterisk indicates the location of the anterior vertebral cortex. The dual flow signals (red for aortic bifurcation, blue for iliac vein confluens) can be appreciated. The large blue signal to the right belongs to the right renal vein

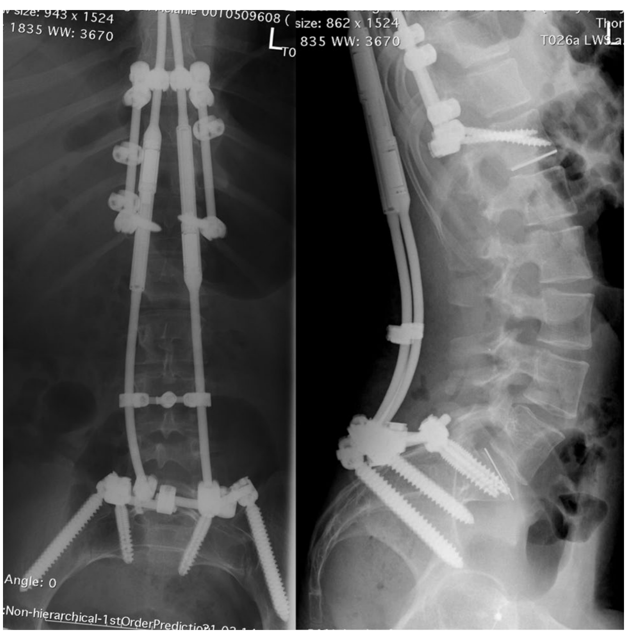

Fig. 7 AP and lateral sitting radiographs of the lumbar spine after recovery from SIRS and after mobilization for several weeks. The lordosis angle LEP T12—UEP S1 is 85°. It can be seen from the AP image how the 2 interconnected magnetic rods are hinged by means of laminar hooks on a 5.5-mm crossbar at the thoracic anchor and on a cross connector at the pelvic anchor

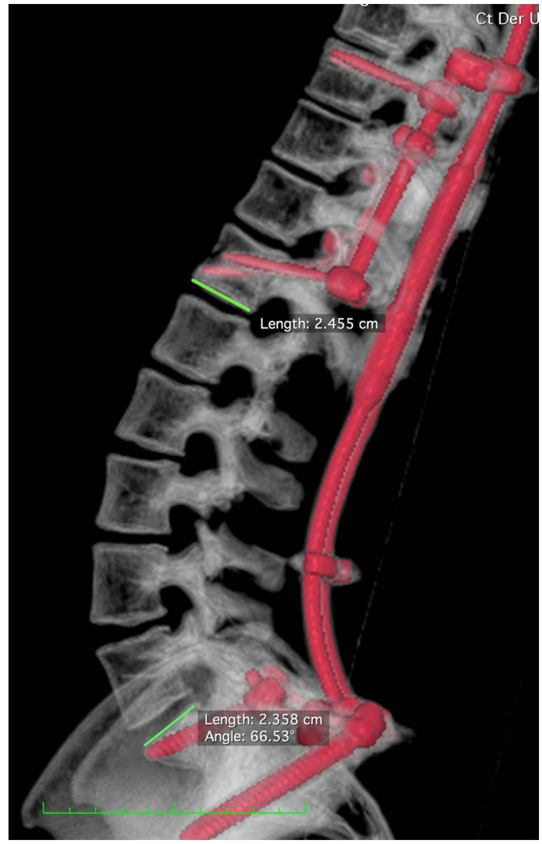

Fig. 8 3D reconstruction from a CT scan at the end of the distraction phase. In comparison with the preop images, the disc spaces have been reduced in volume, the segmental lordotic angles have reduced and the posterior annuli have been stretched. The slow shortening of the spine over time not only permitted further angular correction but also a significant translation of the lordosis apex backwards towards the instrumentation

Table 1 Development of lumbar lordosis and thoracic kyphosis from immediately preop to final follow-up

	Preop (°)	After initial surgery (°)	Prior to final fusion (°)	Final FU (°)
Lumbar lordosis[a]	160	85	66	68
Thoracic kyphosis	36	36	33	26

[a]After the first surgery, the inflection point between the lumbar lordosis and the thoracic kyphosis was altered, so that the preop lordosis was measured from the lower end plate of T11 to the upper end plate of S1, whereas all later lordosis measurements are from the lower end plate of T12 to the upper end plate of S1. Thoracic kyphosis was measured from the upper end plate of T4 to the lower end plate of T11

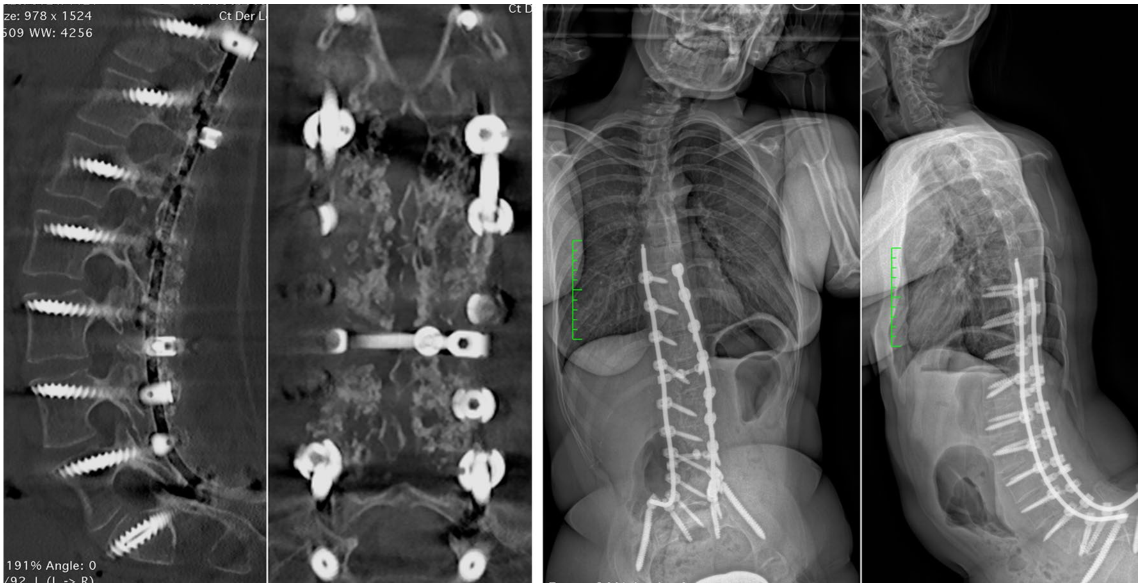

Fig. 9 **a** Exemplary CT reconstructions in the sagittal plane and in the coronal plane at 2 years after completion surgery. A solid posterior fusion mass, an unchanged correction and a stable construct without implant loosening or failure can be seen. **b** Sitting EOS full spine scan at 2 years after surgery. The patient is leaning forward against a holding bar for support. Despite these positional influences, an unchanged spinal situation and an intact construct are visible

radiographs, while on the other hand the fused lumbar spine is less susceptible to these.

Outcome, follow-up

The clinical and radiological results have remained stable over 3 years with a solid posterior fusion and a stable construct demonstrated by means of CT at 2 years after the final surgery (Fig. 9). The patient has obtained significant benefit from the procedures with the abdominal pain completely resolved and with full and unrestricted sitting capacity as well as newly gained limited standing and walking capacity with aids. Overall, her functional score improved from Gross Motor Function Classification System (GMFCS) level IV preoperatively to an intermediate level III–IV.

Hyperlordosis is an uncommon condition to begin with, and it has rarely been described in the context of cerebral palsy. Most of the earlier publications mention hyperlordosis as a sequela of dorsal rhizotomy, a procedure that has been abandoned since [4–8]. Song et al. [1] described a case with 120° of hyperlordosis that also had associated hip flexion contracture. In this case and in a multi-month effort, the authors first performed femoral extension osteotomies and spinal extensor tenotomies, which initially worsened the lordosis to 150°. The actual spinal correction was a staged anterior discectomy and fusion, followed by traction and then a posterior Luque–Galveston extension and instrumentation, resulting in a correction to 63°. Lipton et al. [9] published a series of spastic hyperkyphoses and hyperlordosis with a mean lordosis angle of 92° corrected to a mean of 44° by means of single-stage unit rod instrumentation [9]. Vialle et al. [2] reported on a series of five patients with milder lordosis curves that were corrected from between 90° and 120° of hyperlordosis to between 52° and 90° in either a single-stage posterior procedure (2 cases) or a staged antero-posterior procedure (3 cases) [2]. These latter three cases had an anterior discectomy and fusion a few days prior to the posterior correction and instrumented fusion. Karampalis and Tsirikos [10] published their series of 13 patients with lordosis angles ranging from 80° to 150°. By single-stage posterior correction and fusion, these were corrected to between 42° and 85°. The authors mentioned that preop increased lumbar lordosis and sacral slope were associated with higher complication rates that in their series included excessive blood loss, chest infections and superior mesentery artery syndrome.

All of these previously published cases have in common that these were either adolescents or young adults below the age of 19 years, whereas in our case, the patient was older and the deformity rather rigid as a function of age and the partial spontaneous ankyloses. We therefore realized that a significant amount of release would be necessary, if any meaningful correction was to be achieved.

Table 2 Measurements and calculations from a CT scan that was performed about a month prior to surgery and a CT scan that was performed 3 months after surgery (which is not yet the full final correction)

	11.3.14	29.07.14	Diff	% of pre	% Delta
Lordosis LEP T11—UEP S1 (°)	159	82	− 77	52	− 48
Length along anterior vertebral cortex (mm)					
T12–S1	221	195	− 26	88	− 12
Length along posterior vertebral cortex (mm)					
T12–S1	148	153	5	103	3
Length sublaminar line (mm)					
T12–S1	118	150	32	127	27
Length anterior annulus (mm)					
T12–L1	1.1	0.9	− 0.2	83	− 17
L1–2	1.0	0.8	− 0.1	87	− 13
L2–3	0.9	0.8	− 0.1	87	− 13
L3–4	1.1	0.8	− 0.3	72	− 28
L4–5	1.3	1.0	− 0.3	77	− 23
L5–S1	1.2	1.1	− 0.1	91	− 9
Sum T12–S1	***6.6***	***5.5***	***− 1.2***	***83***	***− 17***
Length posterior annulus (mm)					
T12–L1	0.2	0.3	0.1	166	66
L1–2	0.2	0.3	0.1	179	79
L2–3	0.2	0.3	0.1	146	46
L3–4	0.2	0.5	0.3	243	143
L4–5	0.2	0.4	0.2	202	102
L5–S1	0.3	0.3	0.1	124	24
Sum T12–S1	***1.2***	***2.1***	***0.9***	***177***	***77***
Lordotic angle disc space (°)					
T12–L1	18.9	10.0	− 8.9	53	− 47
L1–2	19.0	9.4	− 9.5	50	− 50
L2–3	17.4	10.6	− 6.8	61	− 39
L3–4	19.8	4.2	− 15.5	21	− 79
L4–5	22.4	10.2	− 12.2	46	− 54
L5–S1	19.7	12.1	− 7.6	61	− 39
Sum T12–S1	***117.1***	***56.6***	***− 60.5***	***49***	***− 51***
Surface area mid-sagittal disc space (cm^2)					
T12–L1	1.80	1.44	− 0.36	80	− 20
L1–2	1.70	1.48	− 0.22	87	− 13
L2–3	1.77	1.40	− 0.37	79	− 21
L3–4	2.02	1.63	− 0.39	81	− 19
L4–5	2.55	1.79	− 0.75	70	− 30
L5–S1	2.44	1.90	− 0.54	78	− 22
Sum T12–S1	***12.27***	***9.65***	***− 2.63***	***79***	***− 21***

The measurements were taken as displayed in Fig. 10. As can be seen, the global lordosis angle measured from the lower end plate T11 to the superior end plate S1 changed from 159° to 82°, a reduction of 48%. When summarizing the individual disc space angular changes, the means was a reduction of 51%, closely matching the global measurement. Interestingly, the mean length increase of the posterior annulus was 77%, whereas the mean length decrease of the anterior annulus was only 17%. Intervertebral disc volume changes significantly as measured on the central sagittal cross-sectional surface area with a slice thickness of 2 mm. The largest change was seen at the L4–5 disc with a 30% volume decrease with neighbouring discs reducing their volume by between 13 and 22%. It appears from these measurements that the complex correction observed under slow distraction and increased axial loading of the previously unloaded lumbar spine is a function of both, a volume reduction of the intervertebral discs and predominantly a stretching of the previously compressed posterior annulus

Bold italics indicates the sums of the individual segments

This is the first such case published where slow percutaneous distraction was employed. We used this secondary distraction as an addition to the initial release, and the overall correction was successful and was aided in its effect by a progressive reduction in disc height under increasingly physiological loading conditions. The articulating and in that sense “dynamic” construct facilitated such loading in contrast to what would have been the case with a rigid pedicle screw construct. We observed that the slow adjustment of the intervertebral discs under loading conditions led to a progressive angular correction (lordosis angle) as well as to a translation of the apex of the lumbar lordosis in posterior direction—closer to the gravity line. An analysis of the preoperative CT and a CT obtained at 3 months postoperatively shows that over that period, the lumbar disc spaces reduced their volume, changed angles and reduced anterior annulus height while increasing posterior annulus height (Table 2, Fig. 10).

We feel that the concept of slow, controlled distraction deserves renewed thought also in severe adult spinal deformity. While currently this is an expensive and time-consuming strategy, it is safe because the distraction happens very slowly and in an awake patient. It also takes full advantage of the intervertebral discs’ viscoelastic properties and their capacity to slowly shed fluid under increased loading, effects which cannot be harnessed during an instantaneous correction that would be performed within just a few hours. Overall, it seems much less aggressive and potentially also more powerful with regards to the final result, because of the ability to optimally judge the appropriate endpoint of a correction and because the final construct does not need to be under corrective tension as would be the case in a single-step correction.

Fig. 10 Illustrations of the measurements for Table 2. Left partial image shows the lengths along the anterior and the posterior vertebral cortices as well as of the sublaminar line. Right partial image shows how lordotic angles, annular lengths and mid-sagittal cross-sectional surfaces of the discs were measured

Compliance with ethical standards

Conflict of interest No author has a conflict of interest in relation to this work.

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Informed consent The patient concerned has given her full and informed consent to this publication.

References

1. Song EW, Lenke LG, Schoenecker PL (2000) Isolated thoracolumbar and lumbar hyperlordosis in a patient with cerebral palsy. J Spinal Disord 13:455–460
2. Vialle R, Khouri N, Guillaumat M (2006) Lumbar hyperlordosis in cerebral palsy: anatomic analysis and surgical strategy for correction. Childs Nerv Syst 22:704–709. https://doi.org/10.1007/s00381-005-0011-5
3. Cheung JP, Cahill P, Yaszay B, Akbarnia BA, Cheung KM (2015) Special article: update on the magnetically controlled growing rod: tips and pitfalls. J Orthop Surg (Hong Kong) 23:383–390. https://doi.org/10.1177/230949901502300327
4. Crawford K, Karol LA, Herring JA (1996) Severe lumbar lordosis after dorsal rhizotomy. J Pediatr Orthop 16:336–339
5. Steinbok P, Hicdonmez T, Sawatzky B, Beauchamp R, Wickenheiser D (2005) Spinal deformities after selective dorsal rhizotomy for spastic cerebral palsy. J Neurosurg 102:363–373. https://doi.org/10.3171/ped.2005.102.4.0363
6. Johnson MB, Goldstein L, Thomas SS, Piatt J, Aiona M, Sussman M (2004) Spinal deformity after selective dorsal rhizotomy in ambulatory patients with cerebral palsy. J Pediatr Orthop 24:529–536
7. Golan JD, Hall JA, O'Gorman G, Poulin C, Benaroch TE, Cantin MA, Farmer JP (2007) Spinal deformities following selective dorsal rhizotomy. J Neurosurg 106:441–449. https://doi.org/10.3171/ped.2007.106.6.441
8. Peter JC, Hoffman EB, Arens LJ (1993) Spondylolysis and spondylolisthesis after five-level lumbosacral laminectomy for selective posterior rhizotomy in cerebral palsy. Childs Nerv Syst 9:285–287 **(discussion 287–288)**
9. Lipton GE, Letonoff EJ, Dabney KW, Miller F, McCarthy HC (2003) Correction of sagittal plane spinal deformities with unit rod instrumentation in children with cerebral palsy. J Bone Jt Surg Am 85-A:2349–2357
10. Karampalis C, Tsirikos AI (2014) The surgical treatment of lordoscoliosis and hyperlordosis in patients with quadriplegic cerebral palsy. Bone Joint J 96-B:800–806. https://doi.org/10.1302/0301-620x.96b6.33020