Eur Spine J
DOI 10.1007/s00586-017-5113-8

GRAND ROUNDS

Tailor-made management of thoracic scoliosis with cervical hyperextension in muscular dystrophy

A. Angelliaume[1] · L. Harper[1] · A. Lalioui[1] · A. Delgove[1] · Y. Lefèvre[1]

Received: 21 March 2017 / Accepted: 25 April 2017

Abstract

Purpose We report the case of a 13-year-old boy managed for fixed cervical hyperextension due to congenital muscular dystrophy with partial merosin deficiency. He presented a right decompensated thoracic scoliosis (T6–L1 Cobb angle 72°) associated with cervical and lumbar lordosis. The spinal extension was accompanied by major flexion of the hip resulting in the trunk being bent forward. This posture caused daily severe back pain responsible for significant loss of quality of life. This led to the decision to perform surgery.
Methods Initially, the surgery was limited to the thoraco-lumbo-sacral area. An anterior release was done, followed by posterior T1-pelvis vertebral fusion using a modified Luque–Galveston technique. The correction achieved was satisfactory in the coronal plane, but the correction of the thoracic kyphosis was insufficient to compensate for the cervical hyperextension. Cervical spine was fixed at 52° of lordosis, and associated with a left 50° rotation and a right 45° inclination of the head. We performed a posterior and lateral release of the cervical muscles followed by positioning of the halo, itself connected to a made-for-measure thoracic corset. A daily adjustment of the threaded rods was done daily for 3 months to correct the cervical position. Then, we performed a spinal fusion without instrumentation, by posterior articular abrasion and grafting from the occiput to T1. Following that, the halo-corset was kept in place for 4 months.
Results At the end of 8 month treatment, the clinical result was satisfactory with a balanced spine both face on, and sideways, allowing for comfortable painless positioning. At 5 year follow-up, he showed stable spinal fusion without any loss of correction.
Conclusion There is no gold standard treatment for cervical hyperextension, but approaches have to be tailor-made to the patient's needs and the team's experience.

Keywords Muscular dystrophy · Neuromuscular scoliosis · Cervical hyperextension · Surgery

Case presentation

We report the case of a 13-year-old boy managed for fixed cervical hyperextension due to congenital muscular dystrophy with partial merosin deficiency. He was 140 cm tall and weighed 29 kilos (Body mass index = 14.8 kg/m^2). He was unable to walk and needed

✉ A. Angelliaume
angelliaume.audrey@gmail.com

[1] Department of Pediatric Orthopaedics, Pellegrin University Hospital, Place Amélie Raba-Léon, 33076 Bordeaux, France

Published online: 07 June 2017

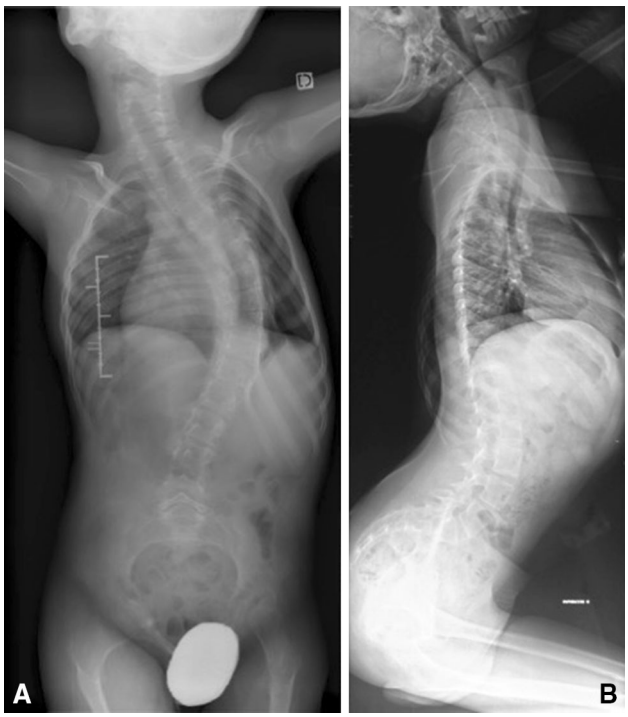

Fig. 1 Preoperative radiography. **a** Right thoracic scoliosis with Cobb's angle at 70°. **b** Spine hyperextension with lumbar hyperlordosis, thoracic hypokyphosis, and cervical lordosis

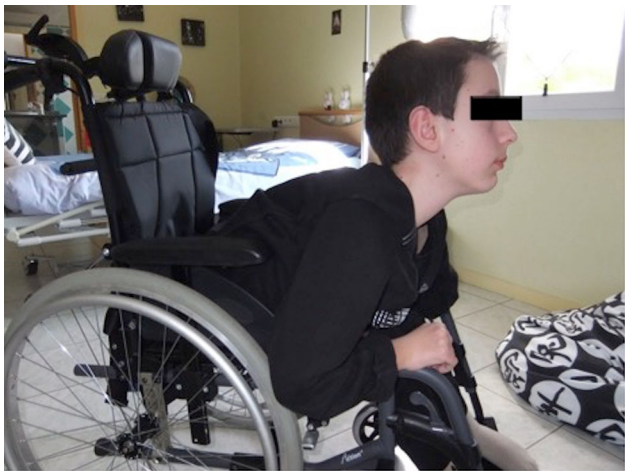

Fig. 2 Preoperative photo of our patient. Notice the cervical hyperextension and the hip hyperflexion with the trunk bent forward to look straight

help for mobilization, but had normal mental abilities. He presented a well-tolerated moderate restrictive lung disease and suffered from right decompensated thoracic scoliosis associated with cervical and lumbar lordosis. Before surgery, the Cobb's angle was 72° between T6 and L1, and the C7 plumb line showed a 2.7 cm frontal

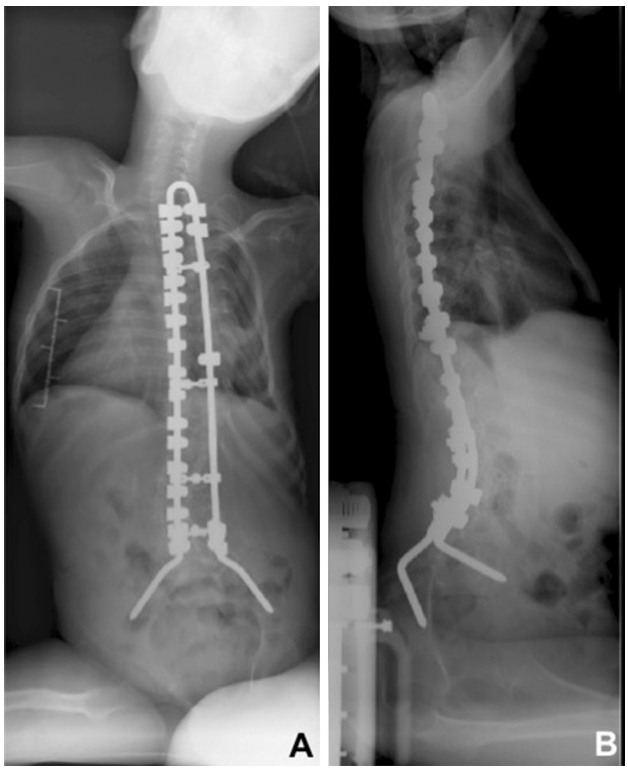

Fig. 3 Postoperative radiography and spine fusion with modified Luque–Galveston technique

displacement (Fig. 1). T1–T2 kyphosis was 9° and L1–L5 lordosis was 68° (Fig. 1). The spinal extension was accompanied by major flexion of the hip resulting in the trunk being bent forward (Fig. 2). He further accentuated this position, with his trunk practically horizontal, as this was the only way he could look straight ahead. This bent posture was his main problem, both because of the general appearance it gave him and because it caused daily severe back pain responsible for significant loss of quality of life (QoL). This led to the decision to perform surgery, initially limited to the thoraco-lumbo-sacral area. The first step consisted of an anterior release by thoracoscopy followed by posterior T1-pelvis vertebral fusion using a modified Luque–Galveston technique. The patient then presented an early (day 21) deep infection (pseudomonas aeruginosa and bacillus non cereus), requiring two redo surgeries for lavage and prolonged antibiotics.

The correction achieved was satisfactory in the coronal plane (postoperative Cobb's angle 27°, i.e. a 62% correction) but less so in the sagittal plane (T1T12 = 4° and L1L5 = 56°) (Fig. 3). Correction of the thoracic kyphosis was insufficient to compensate for the cervical hyperextension. Indeed, the cervical lordosis was fixed at 52°, associated with a left 50° rotation and a right 45°

inclination of the head. The objective at this stage was to correct the cervical hyperlordosis, rotation, and inclination, so that the patient could look straight when sitting (Fig. 4).

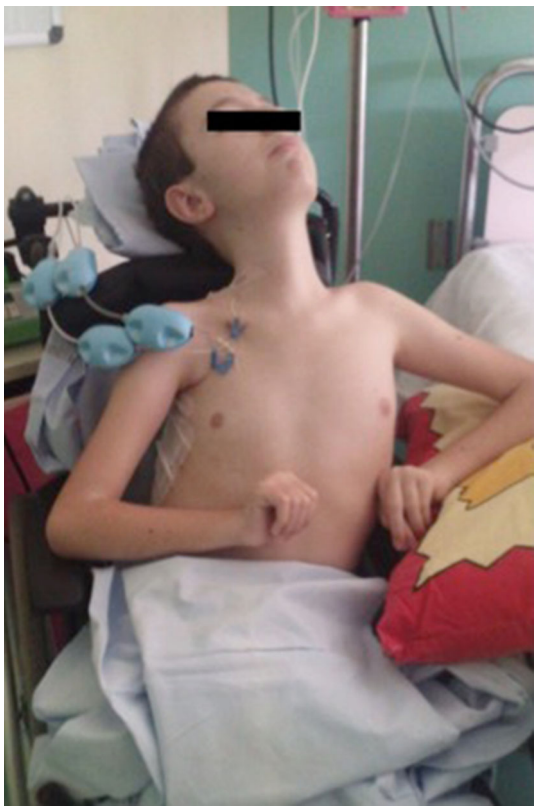

Fig. 4 Postoperative photo of our patient who could not look straight when sitting

Procedure

We decided to correct the cervical hyperextension progressively using an adjustable cranial halo ring fixed to a thoracic corset following the same principles used when external fixators are used for limb deformities.

The first step consisted of a posterior and lateral release of the cervical muscles (freeing of the muscles and their insertions) followed by positioning of the halo, itself connected to a made-for-measure thoracic corset (operating time 130 min and operative bleeding less than 300 cc). Because of the shape of the cranial halo ring, we had to put it back to front to position it correctly (Fig. 5).

Extended release of the cervical muscles only allowed for limited correction of the abnormal cervical position. We then decided to progressively correct the deformation by modifying the position of the halo relative to the corset using threaded rods, as used for limb fixators. This allowed us to apply pressure in the sagittal, coronal, and transversal planes. The contraption consisted of two vertical posterior rods (right and left), which could be used to force cervical flexion using posterior distraction. An anterior left rod, oblique towards the bottom and the front (contraction), and a right anterior rod, oblique towards the top and the front (distraction) allowed us to correct the rotation and inclination of the head. Adjustments of the threaded rods did not exceed 1 mm per day to avoid pain. Limb motility was assessed several times a day and pain was monitored every 3 h. We pursued this management for 3 months, with several interruptions because of difficulties in tolerance of the corset.

At the end of the correction period, the cervical spine was correctly centered in profile. Inclination and rotation were neutral and clinical evaluation confirmed that the patient

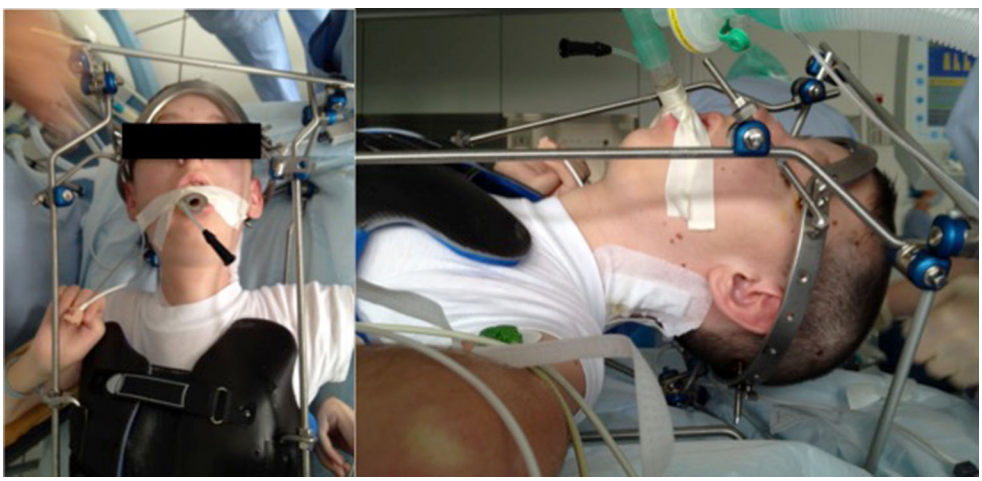

Fig. 5 Photography with halo relative to the corset, with threaded rods, as used for limb fixators, to correct the cervical position

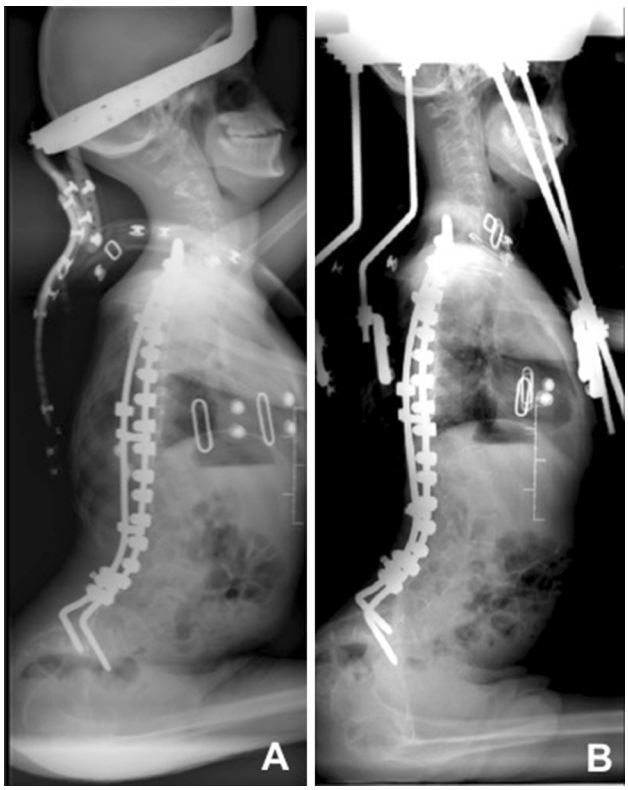

Fig. 6 Radiography, lateral view of the whole spine (see the correction of cervical hyperlordosis). **a** Day zero of the halo installation. **b** 6 months later

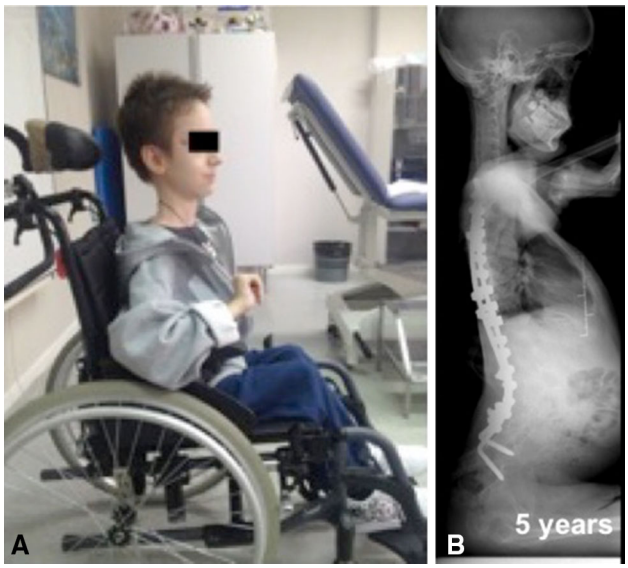

Fig. 7 Patient 5 years after the first surgery. **a** Sitting in comfortable position looking forward. **b** Solid spinal fusion with correct position of the whole spine

could look straight ahead with a horizontal view (Fig. 6). He then underwent spinal fusion without instrumentation, by posterior articular abrasion and grafting from the occiput to T1. Following that, the halo-corset was kept in place for 4 months, so as to obtain sufficient fusion, which was evaluated by X-rays. After a total of 7 months, the halo-corset was removed and replaced by a foam collar for one month.

During the correction phase there were no changes in motility or sensitivity.

All in all, at the end of 8 months treatment, the clinical result was satisfactory with a balanced spine both face on, and sideways, allowing for comfortable painless positioning (Fig. 7). At 5 year follow-up, he showed stable spinal fusion without any loss of correction (Fig. 8).

Treatment rationale and review of the literature

Partial merosin-deficient muscular dystrophy is a congenital muscular dystrophy (CMD), with an incidence between 4.5/10,000 and 8/100,000. Merosin deficit by mutation of the LAMA2 gene on chromosome 6q2 is responsible for 30–40% of cases. Clinical manifestations of the disease appear soon after birth, or during the early childhood, as a general muscular weakness, which mainly affects the girdles and trunk muscles. The motor handicap is severe and the patient's autonomy of movement is often limited to sitting. This, in turn, is often aggravated by orthopaedic complications such as: scoliosis with lumbar hyperlordosis caused by trunk muscle weakness, as well as flexion of the hips, knees, and elbows. Indeed, the muscles most affected by the disease are the cervical flexor muscles, the flexors of the trunk, and the proximal limb muscles, thus explaining the abnormal position. One key feature is the cervical hyperextension due to cervical flexor muscle weakness associated with contracture of the extensor muscles [1].

There are clear recommendations on the surgical management of neuromuscular scoliosis. These include meticulous pre-, per-, and postoperative multidisciplinary management [2, 3]. Surgery is performed if orthopaedic treatment is no longer possible either because of tolerance (pain, corset-related complications, or aggravation of the respiratory distress) or efficiency. The objectives include maintaining a sitting position without pain, improvement (or at least stabilization) of the restrictive lung disease [4, 5], facilitating care and hygiene, and decrease or prevention of back pain. Indeed, prolonged sitting in these patients often causes skin lesions and chronic back pain, which severely impairs QoL, leading to secondary depression, loss of energy and autonomy, and abnormal posture. Several studies have demonstrated good patient and family satisfaction following posterior vertebral fusion, even when the patients endured long and complicated postoperative courses [2, 4, 6]. There are few publications on the management of scoliosis associated with neuromuscular dystrophy except those concerning Duchenne dystrophy, and even those are fairly

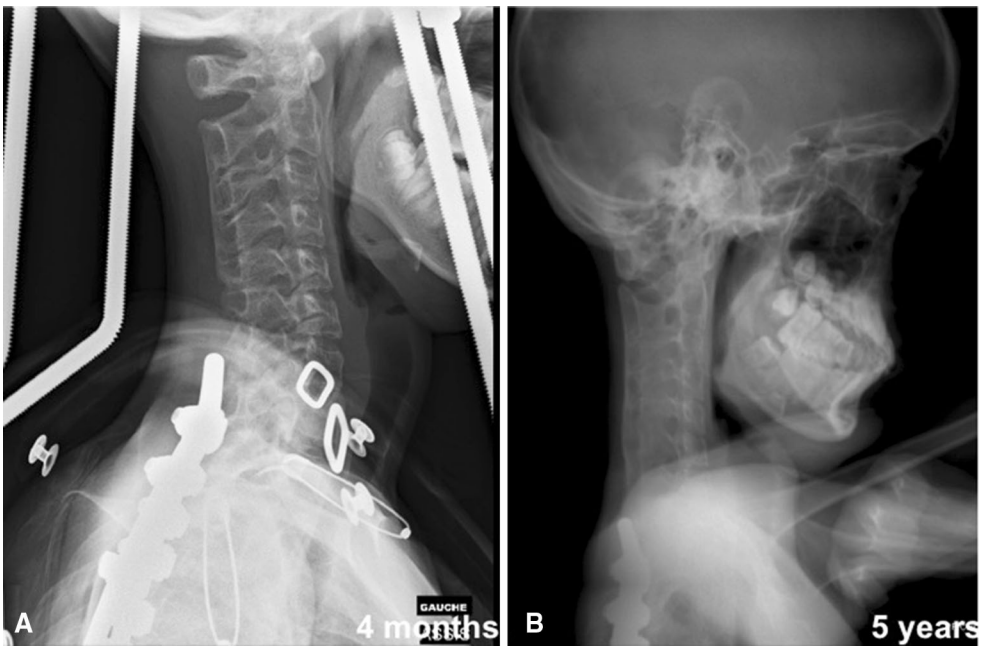

Fig. 8 Lateral view radiography of the cervical spine. **a** 4 months after beginning of treatment with halo. **b** Follow-up at 5 years, solid fusion in good position

ancient [2, 7]. Nevertheless, they do all agree on the general principles of management of these fragile patients, which are: short procedures and reasonable objectives so as to cause limited bleeding, as the latter directly influences the incidence of postoperative complications [8]. However, even when all the precautions have been taken, surgery in these patients is associated with a high risk of complication. In a series of 101 patients, Bentley et al. reported 130 complications [2], and Daher et al. report four major complications (2 cases of duodenal ischemia, one case of postoperative pseudarthrosis requiring redo-surgery and one case or severe respiratory distress rendering postoperative extubation impossible) in 11 patients suffering from non-Duchenne muscular dystrophy [7].

Correction of the cervical hyperextension is essential in these patients as it is directly related to QoL. However, as it is relatively rare, there is no consensus about its management in the literature. We found three reports of which two are from Giannini [9, 10]. He describes an original technique using posterior opening of the C2–C7 interspinous space. Spinal fusion is obtained using autologous bone grafts within the interspinal spaces. Postoperative immobilization is achieved using a collar-corset for 6 weeks, followed by a simple cervical collar for a further 6 weeks. At the end of the 12 week treatment, patients begin their reeducation by performing C1–C2 flexion, extension, and rotation. The long-term results obtained in the seven study patients are satisfactory with improved QoL and few complications. The spinal fusions remained solid and stable with time with a follow-up period of 10 years. Kose et al. report three cases of cervical hyperextension associated with Becker muscular dystrophy managed by anterior cervical osteotomy and anterior and posterior fusion [11]. Two patients were 16 and one was 21, and the osteotomy was performed at the C7–T1 level. The surgery was performed as a one-stage procedure with three separate steps and two changes in patient position. The first step through an anterior approach allowed for anterior release and osteotomy. The second step through a posterior approach allowed for further extensive release and instrumentation from C5 to T2. Reduction was achieved through external manipulation under spinal monitoring and fluoroscopic control. When the desired reduction was obtained, it was fixed using implants. A third anterior step stabilized the osteotomy using a plate. During each change in patient position, the neck was protected using a foam collar. Postoperative immobilization was maintained for 8 weeks using a foam collar. The only reported complications were C8 dermatome anesthesia, which disappeared completely with non-steroid inflammatory drugs and GABA-antagonist medication. 6-month postoperative results are satisfactory with correct restitution of sagittal balance, decrease in dysphagia in two patients, and clear increase in QoL, even if it is at the price of long and complicated surgery. The two techniques described are adapted to specific patients. In the report by Giannini, these are young patients with cervical hyperextensions which can be treated by one-step posterior reduction, whilst in the cases reported by Kose, these are adults with fixed deformities requiring a combined approach. Both series are fairly small with seven

and four patients. The techniques from the literature did not seem appropriated for our patient. His deformity was fixed and we feared that a simple non-instrumental posterior approach would be unsuccessful. Furthermore, he was at risk of infection as he presented a history of deep postoperative infection following his T1-pelvis fusion requiring two lavages. This is why we discarded the posterior instrumentation option. Our technique of progressive flexion of the cervical spine presented major advantages. It excluded the neurological risk associated with acute reduction of the hyperextension. Indeed, even if there are few complications reported in the two series mentioned above, Poulter et al. report three complications following complicated correction of hyperextension [12]. In one case, postoperative extubation was impossible leading to tracheotomy, with death of the patient 2 weeks later at home. Two patients suffered from medullar lesions; one C4 tetraplegia caused by medullar compression due to localized kyphosis and one case of transient paresthesia of the lower limbs also due to localized kyphosis.

To our knowledge, this is the first report of progressive correction of hyperextension in the literature.

We chose this approach for several reasons:

- Its global simplicity with short procedures in a fragile patient for whom the previous surgery had been complicated.
- Minimization of the postoperative infectious risk by lack of instrumentation.
- Minimization of the neurological and respiratory risks through the progressive nature of the correction with the possibility of going back a step in case of poor tolerance.
- Finally, the tailor-made approach was appealing as it allowed for correction in the three planes adapting the correction to the patient's tolerance and appreciation.

The principal difficulty of this method is the tolerance of the corset and the pressures it causes on the skin. This implies using an extremely well-adapted corset and all the protective pads possible. And more important even, correction must be applied slowly with pauses if necessary to allow for recovery of any skin lesions.

This method requires spreading the treatment over a long period, which in our case was done at hospital. This could, however, be done in day care. The patient's motivation for such a treatment is obviously essential, and we take the opportunity to commend our patient on his motivation and acceptance of the long hospitalization; this progressive correction required.

In conclusion, hyperextension is an essential aspect of scoliosis in congenital muscular dystrophies. They influence directly QoL. There is, for the moment, no gold standard treatment for these patients but approaches which have to be tailor-made to the patient's needs and the team's experience. Progressive correction using a halo-corset is an interesting alternative, offering excellent results with few risks. It does, however, rely on excellent patient motivation.

Compliance with ethical standards

Conflict of interest None of the authors has any potential conflict of interest.

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