

# Efficacy of adaptive seating system for the management of neuromuscular scoliosis in a young adult with cerebral cavernous malformation: A case report

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## Abstract

Cerebral cavernous malformations (CCMs) are vascular malformations causing seizures and cerebral hemorrhages. Neurological and functional profile can differ from each individual, and the management requires high complexity. The aim of the present case report was to explore the management of neuromuscular scoliosis in a young adult with CCM, comparing the use of a scoliotic brace with a customized adaptive seating system (AdSS). X-ray examinations were performed in three different situations: sitting in a flat bench without postural support, sitting in a flat bench with scoliotic brace, and sitting in an AdSS. For the analysis of the AdSS, an experimental radiolucent device was used. Differences in spine alignment and postural support were found. The best option to care was the use of the scoliotic brace. Nevertheless, the use of a customized AdSS offers a good solution for the management of neuromuscular scoliosis, with a difference of 10 degrees with respect to the scoliotic brace. Position was maintained at 3-month follow-up, with a reduction of the overhang of the spine. The use of an AdSS represents a valid attempt for the management of neuromuscular scoliosis in people with CCM. This approach can be considered when the patient suffers the scoliotic brace and/or when the brace interferes with breathing or other clinical conditions.

## Keywords

X-ray, posture, cerebral cavernous malformations, spine, case report

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## Backgrounds

Cerebral cavernous malformations (CCMs) are vascular malformations causing seizures and cerebral hemorrhages.<sup>1</sup> CCM may cause serious neurological symptoms because of severe bleeding or pressure on the brain or nerves.<sup>2</sup> The majority of affected patients carry a sporadic lesion, but 10%–40% may have a positive family history or carry multiple CCMs, suggesting a genetically determined cerebral cavernomatosis.<sup>3</sup> CCM is a rare disease, and neurological and functional profile can differ from each individuals, and the management requires high complexity. The present case report describes the management of severe neuromuscular scoliosis in a young adult with CCM. Neuromuscular scoliosis may occur in association with multiple diagnoses, and surgical management is often the only option other than supportive care.<sup>4</sup> Nonoperative treatment cannot control the deformity but may be used while significant spinal growth remains. However, because of many factors (e.g., health care costs, clinical conditions, or simply the refusal consent to treatment), for many people, a conservative approach is the only option to care. However, bracing can be

ineffective because of reduced tolerance of brace wear, skin irritation, and respiratory problems, as well as the aggressive nature of the deformity; it is important that the potential advantages, disadvantages, and expected outcomes of bracing are clearly understood by the patients, their family, and carers if it is used.<sup>5,6</sup> This study reports the first investigation for the management of neuromuscular scoliosis in a young adult with CCM, comparing the efficacy of scoliotic brace vs. the use of an adaptive seating system (AdSS) by using a new x-ray-based methodology.

## Methods

### Patient information

The present case report describes the case of a patient suffering from neurological sequelae of truncal bleeding causing by CCM. From 1998 to 2003, he underwent different surgery procedures in posterior cranial fossa. He displayed symptoms associated with hemiplegia, gait, and balance disorders. From 2005 to 2018, more neurological deficits occurred with bilateral spastic hemiplegia, corneal lesion in the right eye, and breathing and feeding difficulties. A percutaneous endoscopic gastrostomy was done, and a mechanical ventilation was placed. In 2020, he was admitted to the emergency department for fever and respiratory difficulty. Next, thorax X-Ray was performed; the examination showed right apical perillary thickening. Furthermore, a brain computed tomography highlighted hyperdensity in the right peduncular compatible with vascular anomaly type cavernous angioma, with phenomena of intralesional bleeding. Therefore, in September, he underwent surgery to evacuate the hemorrhagic area located on the right lateral

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**Table 1.** Functional classification.

| Classification                               | Level                                                                                             |
|----------------------------------------------|---------------------------------------------------------------------------------------------------|
| Gross Motor Function Classification System   | Level 5—Transported in a manual wheelchair                                                        |
| Manual Ability Classification System         | Level 5—Does not handle objects and has severely limited ability to perform even simple actions   |
| Communication Function Classification System | Level 4—The person is not always consistent at communicating with familiar communication partners |
| Eating and Drinking Classification System    | Level 5—Unable to eat or drink safely—tube feeding may be considered to provide nutrition         |

profile of the bulb and for the complete removal of three additional cavernomas located in the cerebellar vermis and contiguous left cerebellar hemispheric site. The patient was an adolescent boy aged 23 years and 8 months and had a corneal lesion in the right eye, double spastic hemiparesis, chronic respiratory failure with mechanical ventilation, left convex neurogenic scoliosis (128 degrees), gastroesophageal reflux disease, and percutaneous endoscopic gastrostomy. For neuromotor disorders, he attended physical therapy and respiratory physiotherapy. For the management of the severe neuromuscular scoliosis, the medical doctors team suggested the use of a scoliosis brace. However, family reports difficulty in using and low compliance.

### Clinical findings

To have a comprehensive overview of the functional profile, the research group decided to use classification systems generally used for cerebral palsy population because of the lack of specific classification systems and/or assessment tools for rare diseases. Therefore, we used the Gross Motor Function Classification System,<sup>7</sup> the Manual Ability Classification System,<sup>8</sup> the Communication Function Classification System,<sup>9</sup> and the Eating and Drinking Classification System.<sup>10</sup> All these classification systems are hierarchy ordinated into 5 levels: the first one describes the best functioning while the fifth describes the worse. These are not assessment tools but allow classifying of functioning in different domains. The patient's functioning profile for each classification system is presented in Table 1.

To describe posture in seating position, the Italian version of the Seated Postural Control Measure [11] was used. The boy was seated in an AdSS composing of a tilt-in-space AdSS, with customized backrest and cushion, using pelvic belt, medial and lateral thigh supports, lateral thoracic support, and posterior head support. The frontal evaluation showed a severe right pelvic obliquity with trunk lateral shift and moderate inclination of the left scapular girdle. On the sagittal plane, the trunk is maintained in extension with thoracic hyperlordosis of the spine and anterior tilt of the pelvis. On the transverse plane, there was evidence of severe left pelvic rotation and moderate right scapular rotation. Right lateral shift of the head, with rotation and hyperextension.

## Results

### Diagnostic assessment and therapeutic intervention

To investigate the best solution for the management of the neuromuscular scoliosis, the research group decided to compare the use of scoliotic brace with a customized AdSS. The assessment

was performed according to biomechanical perspective investigated with an x-ray examination. Therefore, anteroposterior trunk x-ray was made in three different situations:

1. In sitting on a flat bench without brace and/or support;
2. In sitting on a flat bench with the scoliotic brace; and
3. In sitting on the experimental device reproducing the AdSS used by the young boy.

To measure the efficacy of the AdSS, an experimental device completely radiolucent was used. Standards for wheelchair fabrication were followed, collaborating with an orthopedic company accredited with the National Health System. The experimental device presents easily adjustable parameters for anthropometric



**Figure 1.** Experimental device used for x-ray examination. The experimental device is considered an evaluation tool for simulating different postural configurations. In fact, it presents easily adjustable parameters for anthropometric measures and applying different postural components and supports is possible. Over the seat and the back, it is possible to apply a customized seating system or a commercial one.

measures (adjustable length, width, and depth for both the seat and the back) and to apply different postural components (e.g., trunk and hand supports) as well as different postural support (e.g., pelvic belt and anterior trunk support). Materials used for the prototypal fabrication of the seat and the back were completely radiolucent (polymethyl methacrylate). Over these planar units, it was possible to apply the customized AdSS used by the young boy and to perform an x-ray examination. Figure 1 illustrates the experimental device.

The radiological examination was performed in the frontal projection. The study of the spine showed left convex scoliotic deviation with angular values of 128 degrees in sitting position, 112 degrees when sitting on AdSS, and 102 degrees when sitting with a scoliotic brace (limit vertebrae D3-L4). X-ray examinations are reported in Figure 2.

According to these findings and to family perspective—that refer poor tolerance of the scoliotic brace and breathing difficulties—the research group suggested using only the AdSS for the management of neuromuscular scoliosis. This is because although the scoliotic brace was shown to be more effective, it could not be used for longer periods. By contrast, the customized AdSS, despite less support, offered greater tolerability and use over time. Considering the particular case, the research group decided to follow up the boy for a short period when using only the AdSS.

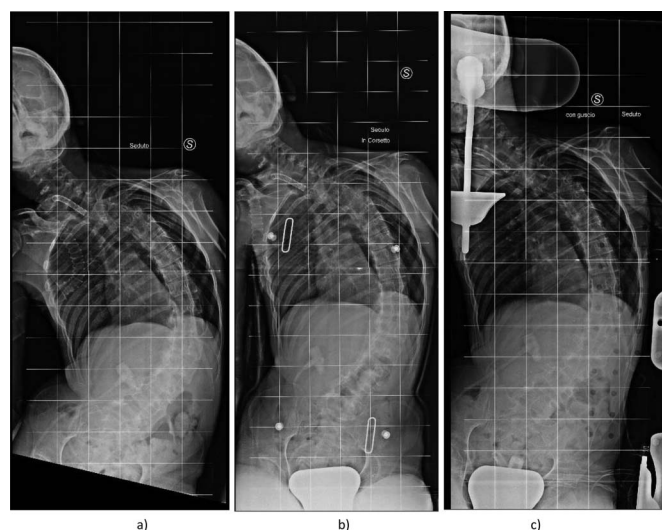
### Follow-up and outcomes

A new radiological examination after the 3-month follow-up was performed. During this period, the boy used only the AdSS. A second anteroposterior radiological examination was performed in sitting position using the AdSS. The study of the spine showed left convex scoliotic deviation with angular values of 112 degrees. Compared with the previous examination, radiology technicians confirm the stability over time, observing a reduction of the right overhang of the spine. X-ray examination at 3-month follow-up is reported in Figure 3.

### Discussion

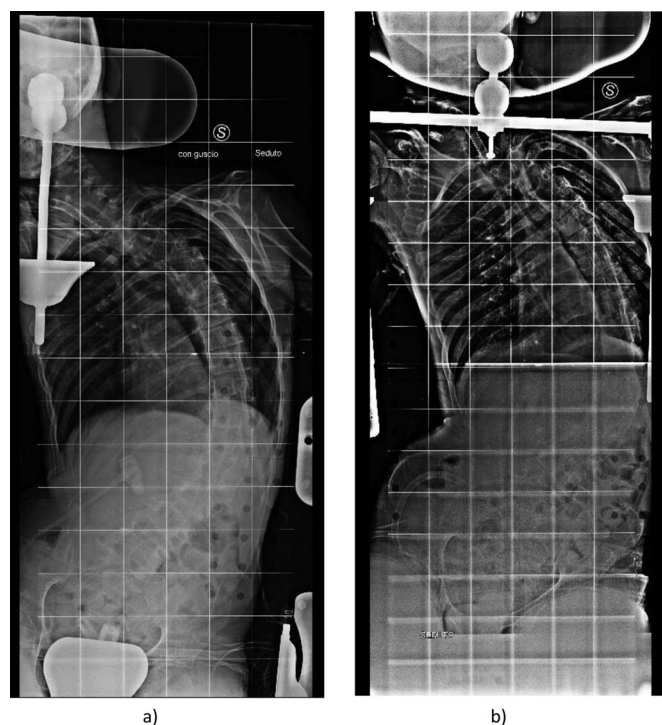
Spinal deformity cannot be attributed to trunk muscle weakness alone; indeed, in certain pathologies, trunk muscle hypertonia rather than paralysis is primarily implicated.<sup>12</sup> In certain pathologies inducing severe muscular deficiency, orthopedic treatment should be very early: It begins with passive bracing, but in most cases, conservative treatment of neuromuscular spinal deformity is only an interim measure awaiting surgery.<sup>12</sup> However, bracing can help to manage the spinal deformity by providing postural support, maintaining sitting position, improving muscle balance, and augmenting function while permitting further growth to occur before surgical intervention.<sup>13,14</sup> Furthermore, bracing may limit complications such as impaired pulmonary development, and therefore, reduce the risks associated with any future surgery.<sup>12</sup> However, in the present case report, there was a poor tolerance of the scoliotic brace. So, the research group decided to investigate how an AdSS can effectively contribute in the management of neuromuscular scoliosis.

In general, bracing has a positive influence on sitting function in children with neuromuscular scoliosis; the need of external support and the number of special adaptations of the AdSS decrease for



**Figure 2.** X-ray image examination. This figure reports the study of neuromuscular scoliosis in different situations: (a) sitting in a flat bench with a Cobb angle of 128 degrees, (b) sitting in a flat bench with scoliotic brace with a Cobb angle of 102 degrees, and (c) sitting in an adaptive seating system with a Cobb angle of 112 degrees.

many children when using the brace.<sup>15</sup> However, in the present case study, it was necessary to find alternative solutions because the boy did not tolerate the prolonged use of the brace. Despite our findings confirming that the scoliotic brace is more effective, it also reveals a good efficacy of the AdSS. In fact, the differences between two approaches are 10 degrees. In considering the high complexity



**Figure 3.** X-ray of sitting in an adaptive seating system at (a) T0 and (b) after 3 months. The study of the spine showed left convex scoliotic deviation with angular values of 112°. Compared with the previous examination (a), the 3-month follow-up x-ray examination (b) confirms the stability over the time, with a reduction of the right overhang of the spine.

for the management of neuromuscular scoliosis in CCM, as well as the patient's breathing difficulties, the clinicians have considered this difference acceptable. Furthermore, using a well-fitted AdSS improved postural alignment over time. The effects of AdSS on lateral spinal curvature was positive; as reported by Holmes and colleagues, the position of the lateral pads on a special seating system is important and, by the careful configuration of these supports, significant correction of a scoliosis can be obtained.<sup>16</sup> Furthermore, using the experimental radiolucent device allows measuring the column alignment when the boy used his own AdSS. The present investigation supports preliminary evidence on using AdSS for the management of neuromuscular scoliosis in CCM. This research also describes a new x-ray methodology to investigate how the AdSS can be effective for the management of neuromuscular scoliosis, investigating the relationship between load and body alignment in the real context.

### Patient perspective

The family reported a positive impact on daily management of the AdSS. In addition, having established that the AdSS actually contributed to a good postural support helped them reduce anxiety and stress levels, primarily because of poor adherence to orthotic treatment. The family was therefore highly satisfied of the proposed treatment.

### Informed consent

The family agreed to participate in this study, and informed consent was obtained from them. All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation and with the 1975 Declaration of Helsinki, as revised in 2008.

### Conclusion

Considering the high complexity for the management and clinical issues of CCM, particularly in such cases, using a well-fitted AdSS can be effective for the management of neuromuscular scoliosis.

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### Supplemental material

There is no supplemental material in this article.

### References

- Denier C, Labauge P, Brunereau L, et al. Clinical features of cerebral cavernous malformations patients with KRIT1 mutations. *Ann Neurol* 2004; 55: 213–220.
- Porter PJ, Willinsky RA, Harper W, et al. Cerebral cavernous malformations: natural history and prognosis after clinical deterioration with or without hemorrhage. *J Neurosurg* 1997; 87: 190–197.
- Labauge P, Denier C, Bergametti F, et al. Genetics of cavernous angiomas. *Lancet Neurol* 2007; 6: 237–244.
- Murphy RF and Mooney JF. Current concepts in neuromuscular scoliosis. *Curr Rev Musculoskelet Med* 2019; 12: 220–227.
- Roberts SB and Tsirikos AI. Factors influencing the evaluation and management of neuromuscular scoliosis: a review of the literature. *J Back Musculoskelet Rehabil* 2016; 29: 613–623.
- Miller AMD, Temple TMD and Miller FMD. Impact of orthoses on the rate of scoliosis progression in children with cerebral palsy. *J Pediatr Orthop* 1996; 16: 332–335.
- Palisano R, Rosenbaum P, Walter S, et al. Gross motor function classification system for cerebral palsy. *Dev Med Child Neurol* 1997; 39: 214–223.
- Eliasson AC, Krumlinde-Sundholm L, Rösblad B, et al. The Manual Ability Classification System (MACS) for children with cerebral palsy: scale development and evidence of validity and reliability. *Dev Med Child Neurol* 2006; 48: 549–554.
- Hidecker MJ, Paneth N, Rosenbaum PL, et al. Developing and validating the communication function classification system for individuals with cerebral palsy. *Dev Med Child Neurol* 2011; 53: 704–710.
- Sellers D, Mandy A, Pennington L, et al. Development and reliability of a system to classify the eating and drinking ability of people with cerebral palsy. *Dev Med Child Neurol* 2014; 56: 245–251.
- Tofani M, Blasetti G, Lucibello L, et al. Seated postural control measure: Italian translation and validation in children with cerebral palsy. *Prosthet Orthot Int* 2021; 45: 378–383.
- Vialle R, Thévenin-Lemoine C and Mary P. Neuromuscular scoliosis. *Orthop Traumatol Surg Res* 2013; 99(1 suppl): S124–39.
- Kotwicki T, Durmala J and Czubak J. Bracing for neuromuscular scoliosis: orthosis construction to improve the patient's function. *Disabil Rehabil Assist Technol* 2008; 3: 161–169.
- Roberts SB and Tsirikos AI. Factors influencing the evaluation and management of neuromuscular scoliosis: a review of the literature. *J Back Musculoskelet Rehabil* 2016; 9: 613–623.
- Blomkvist A, Olsson K, and Eek MN. The effect of spinal bracing on sitting function in children with neuromuscular scoliosis. *Prosthet Orthot Int* 2018; 42: 592–598.
- Holmes KJ, Michael SM, Thorpe SL, et al. Management of scoliosis with special seating for the non-ambulant spastic cerebral palsy population—a biomechanical study. *Clin Biomech* 2003; 18: 480–487.