Mucopolysaccharidoses are a group of autosomal recessive, lysosomal storage disorders characterized by abnormal buildup of glycosaminoglycans. There are different types of MPS. Morquio syndrome, also known as MPS type IV, is an enzyme deficiency disorder of either N-acetylgalactosamine-6-sulfatase (MPS type IV A) or β-galactosidase (MPS type IVB) that results in the inability to break down keratin sulfate and chondroitin-6 sulfate. Morquio syndrome is rare, and its incidence ranges from 1 in 75,000 in Northern Ireland to 1 in 450,000 in Portugal. Individuals with Morquio syndrome may appear normal in their first 2 years of life but subsequently exhibit dwarfism, with short trunk, chest wall deformity and smaller than normal thoracic capacity, cervical spinal stenosis and instability, thoracolumbar kyphosis, hip dysplasia, and genu valgum. Although physical manifestations are severe, patients with Morquio syndrome have normal intelligence.

Odontoid dysplasia is common among affected patients, resulting in atlantoaxial instability and spinal cord compression. Surgical treatments include decompression and prophylactic fusion, during which intraoperative neuromonitoring is important to alert the surgical team to changes in cord function so that they can prevent or mitigate spinal cord injury. This report describes a 16-year-old girl with Morquio syndrome who developed paraplegia due to thoracic spinal cord infarction during foramen magnum and atlantal decompression. This tragic event demonstrates the following: 1) that patients with Morquio syndrome are at risk for ischemic spinal cord injury at levels remote from areas of maximal anatomical compression while under anesthesia in the prone position, possibly due to impaired cardiac output; 2) the significance of absent motor evoked potential responses in the lower limbs with preserved upper-limb responses in an ambulatory patient; 3) the importance of establishing intraoperative neuromonitoring baseline assessments prior to turning patients to the prone position following induction of anesthesia; and 4) the importance of monitoring cardiac output during prone positioning in patients with chest wall deformity.

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Case report

Morquio syndrome, which frequently leads to atlantoaxial instability and subsequent spinal cord compression, Stenosis caused by the buildup of mucopolysaccharides at the occipitocervical junction may also result in spinal cord compression and devastating outcomes, such as myelopathy, spastic quadripareisis, and premature death. Prophylactic posterior cervical fusion with or without posterior surgical decompression is recommended prior to the onset of neurological deficits.

Strengthening of the spinal musculature and development of abdominal compensation is encouraged preoperatively, and an awake positioning test is performed to plan the surgical approach. The prone position is used to expose the foramen magnum and atlantal base, and a cervical spine fusion is performed to prevent subsequent spinal cord compression.

Key Words • Morquio syndrome • spinal cord infarction • paraplegia • chest wall deformity • hypotension • cardiac output • prone position • bolster • spine

Abbreviations used in this paper: MEP = motor evoked potential; MPS = mucopolysaccharidosis; SSEP = somatosensory evoked potential.
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pairment of cardiac output. Intraoperative neuromonitor-
ing, including SSEPs and MEPs, is often performed to
warn the surgical team of changes in sensory and motor
conduction during the operation.5,16

There are few published reports describing the loss or
reduction of evoked potentials during patient positioning
in preparation for operations. In these cases surgeries were
aborted, evoked potentials returned to baseline values, and
to our knowledge no long-term sequelae were reported.16
This report describes a patient with Morquio syndrome
who required an occipitocervical decompression, who
had tolerated an awake test of prone positioning, but who
exhibited transient hypertension when positioned prone
after induction of anesthesia. Blood pressure normalized
promptly, but lower-extremity MEPs were never record-
ed during the procedure. The patient suffered permanent
complete thoracic paraplegia due to cord infarction.

Case Report

History and Examination. This 16-year old girl with
Morquio syndrome presented with MR imaging changes
secondary to spinal cord compression at C-1. She had un-
dergone a posterior C-1 laminectomy for craniocervical
stenosis and atlantoaxial hypermobility at the age of 8
years and had remained neurologically normal. Routine
follow-up MR imaging studies obtained at the age of 16
years demonstrated recurrent spinal stenosis at the level
of the atlas (the anteroposterior diameter of the spinal
canal measured 8 mm), with abnormal hyperintensity in
the cord extending from the foramen magnum to C2/3 on
T2-weighted sequences (Figs. 1A and 2). These findings
were interpreted to reflect evolving cervical myelomalacia.
Cervical spinal CT scans revealed the bone morphol-
yogy characteristic of Morquio syndrome throughout the
cervical spine and atlantoaxial instability secondary to
a hypoplastic odontoid. Generalized platyspondyly was
also seen with associated widening of the atlantoaxial
distance, which measured 9 mm on flexion and 7 mm
with extension.

Clinical examination at that time showed that her up-
per cervical sensory function was intact, and there was
no evidence of lower bulbar cranial nerve dysfunction.
Her vibration sense in her lower limbs was minimally de-
creased, but it was normal in her upper limbs. Deep ten-
don reflexes were brisk at her elbows and knees. However,
ankle jerks could not be elicited and she had no plantar
responses. Muscle power was limited, at 3.5/5 in upper
and lower limbs, because of the joint changes due to her
underlying disease. She was independently ambulatory,
although she used a motorized wheel chair for mobiliza-
tion outside of her home.

The SSEPs revealed normal latency from her upper
and lower limbs. However, the amplitude of her right-sid-
ed response was less than that on her left, and both were
less than normal. In view of her progressive spinal cord
compression, operative decompression and fusion was
recommended.

Operation. Prior to the operation, the patient’s toler-
ance for the prone position was tested with her awake, by
hanging her lie prone, on transversely positioned supports,
with her head flexed on her chest for several minutes. No
neurological symptoms were provoked by cervical flexion,
prone positioning, or both. It was believed that she would
tolerate the positioning necessary for the operation (Fig. 2).

After successful induction of anesthesia, the patient
was placed in a pin fixation headrest and was turned to the
prone position on transverse upper chest and pelvic bolsters,
on the Jackson table. Cranial cervical alignment was main-
tained during the roll from the supine to prone position.
Following placement in the prone position, blood pressure
initially increased, and then normalized in response to an-
algesic administration (Fig. 3). Baseline SSEPs and MEPs
were measured after positioning and prior to the incision.
Initial MEPs demonstrated good responses in the upper
limbs bilaterally, but no response in the lower limbs. There

Fig. 1. Sagittal T2-weighted MRI studies obtained in a 16-year-old
girl with Morquio syndrome. A: In May 2010, abnormal hyperintensity
on T2-weighted sequences (arrow) was seen at the posterior lip of the
foramen magnum, extending to the level of C2/3. The spinal canal has
an anteroposterior diameter of 8 mm. B: In September 2010, imme-
diately postoperatively, a new area of hyperintensity was seen between
C-7 and T3/4 (arrowhead) on T2-weighted sequences, with hyperinten-
sity at the craniocervical junction (arrow) still present. C: In Decem-
ber 2010, a marked narrowing of the cord from C-7 to T2 was observed,
along with a small focus of hyperintensity at T2/3 on T2-weighted se-
quencies (arrowhead). Hyperintensity on T2-weighted sequences was
persistent at the craniocervical junction (arrow).
was no technical or anesthetic explanation for the absence of MEPs to the lower limbs, and it was assumed that this was due to her underlying disease. The SSEPs showed poor responses in the lower limbs and asymmetrical responses in the upper limbs, with a greater left-sided suppression.

During the decompression, the SSEPs and MEPs (upper limbs normal, lower limbs absent) remained stable and unchanged from the baseline. While the left side of the atlantal arch was being drilled off under magnification, using a matchstick bur, the left upper-limb MEPs were lost, without apparent change in anesthetic or vital signs. The absence of the left MEP response persisted throughout the operation. During the removal of the right side of the arch, a brief transient loss of MEPs to the right arm followed by a quick rebound to baseline was noted. This was associated with a drop in blood pressure. The fusion was deferred, given the change in evoked potentials. By raising the mean arterial pressure to 85 mm Hg, the left MEPs reappeared. The patient awoke from anesthesia with a complete sensory-motor paraplegia to the T-4 level, and intact upper-limb sensory and motor function.

**Postoperative Course.** Immediate postoperative MR images revealed a new area of hyperintensity on T2-weighted sequences, with diffusion restriction in the cord between C-7 and T3/4, which was suggestive of ischemic insult from T-1 to T-4. There was no change in the cervicomedullary cord signal compared with preoperative findings (Fig. 1B). At 14 days postsurgery, the patient was breathing comfortably using her diaphragm and had normal results on a motor and sensory examination at and above T-4. She had impaired sensation below T-4 and no perceptible sensation below T-10, with flaccid total paraplegia and neurogenic bladder and bowel.

The MR imaging studies obtained at 3 months postoperatively showed near complete resolution of the hyperintensity of the cord between C-7 and T3/4 on T2-weighted sequences. However, a marked narrowing of the cord from C-7 to T-2 was observed, along with a small focus of hyperintensity at T2/3 that was suggestive of myelomalacia. The hyperintensity at the cranio cervical junction and focal kyphosis at C7/T1 were unchanged (Fig. 1C). Follow-up studies obtained at 12 months showed persistence of the complete sensory-motor paraplegia, with evolving spasticity.

**Discussion**

For patients with Morquio syndrome, odontoid dysplasia is common and can lead to atlantoaxial instability and spinal cord compression. Impingement of the spinal cord may result in devastating neurological deficits. Decompression and fusion have been suggested to

![Fig. 2. Lateral images showing the chest wall deformity and the potential for cardiac compression, with the patient in the prone position on a transverse chest bolster. A: Sagittal T2-weighted TrueFISP (fast imaging with steady state precession) study demonstrating the cardiac volume and orientation. B: Lateral chest radiograph showing rib and sternal deformity.](image)

![Fig. 3. Vital signs record from induction to positioning prone. The initial increase in blood pressure occurred when the patient was positioned prone and was thought to be due to a lightening in the depth of anesthesia. The blood pressure then normalized with adjustment of the anesthetic.](image)
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prevent or mitigate the effects of cord compression. In our patient, preoperative MR and CT images suggested severe peripheral myelopathy at the craniovertebral junction, and atlantoaxial instability secondary to her hypoplastic odontoid. Therefore, cervical decompression and fusion from the occiput to C-4 was recommended.

Spinal cord injuries during surgical procedures are reported to occur in approximately 1 in 1000 cases. The causes of such injury can include the following: 1) local trauma at the site of instability during prone positioning, traction, or curvature correction; or 2) ischemia, most commonly due to radicular artery injury during the procedure. Impaired cardiac output resulting in a cord infarction due to hypoperfusion in vascular border zones is less commonly recognized. The effects of prone positioning on cardiac output have been investigated by several authors. Tabara et al., in a study of healthy adult males, demonstrated that the prone position can lead to unregulated postural hypotension. In the setting of lumbar spine surgery, hypovolemia may aggravate blood pressure instability by decreasing cardiac preload through preoperative volume depletion due to fasting, or due to leg positioning that would allow blood pooling during the operation. The anesthesia administered may also have an important role.

The role of bolsters has also been studied in spine surgery. Park measured intraabdominal pressures and blood loss in adults positioned on narrow and wide, longitudinally oriented bolsters on a Wilson frame. He found that bolsters that were too narrow resulted in abdominal compression and increased bleeding. In the study by Dharmavaram et al., longitudinal bolsters were found to have the least effect on cardiac function. In the 34-month-old toddler reported by Alexianu et al., the cardiac compromise seen with transverse bolsters was not replicated with longitudinal bolsters.

Alexianu et al. and Bafus et al. have reported individual patients with pectus excavatum and scoliosis who exhibited systemic hypotension when placed in the prone position for scoliosis correction. In both cases, cardiac output was compromised in the prone position as a result of impaired cardiac filling. Transesophageal echocardiography was used to demonstrate these phenomena. Recognizing this possibility, it behooves the anesthesia team to maintain arterial pressure in the presurgical range and not to allow it to fall in an attempt to reduce operative blood loss.

Intraoperative neuromonitoring, using SSEPs and MEPs, is commonly performed to inform the surgical team of changes in cord function due to the procedure or anesthesia, so that steps can be taken to reduce the risk of spinal cord injury. Nuwer and colleagues reported that SSEPs identified 90% of clinically relevant neurological events. The SSEP response requires a period for signal averaging. This may be as long as several minutes. Because only the ascending pathways are being monitored, SSEP monitoring alone yields false-positive and false-negative results, and thus its usefulness in forecasting postoperative motor function is limited.

Motor evoked potentials do not require averaging of large sample sizes and can provide timely alerts to the team. Persistent changes in MEPs were reported to be stronger factors than changes in SSEPs in predicting potential postoperative neurological deficits. Schwartz and colleagues demonstrated in their large multicenter study that the sensitivity and specificity of SSEPs in predicting motor loss were 43% and 100%, respectively, whereas those of MEPs were both reported to be 100%. These authors highlighted the benefit in combining SSEPs and MEPs for intraoperative neuromonitoring. Similarly, other evidence also supported this multimodal approach for intraoperative neuromonitoring to prevent spinal cord injury.

Monitoring of SSEPs and MEPs was performed during the procedure in our case. The baseline MEPs showed no response in the lower extremities but good responses bilaterally in the upper limbs. Because the MEPs were not measured prior to turning the patient to a prone position, the effect of this manipulation or of the orientation of bolsters on the MEPs could not be assessed.

Our patient became hypertensive when positioned prone. Although there had been no blood pressure response to skull fixation, it was presumed that the increase in systolic and diastolic pressures was due to a lightening in the depth of anesthesia. We propose that this sympathetic response, in conjunction with impaired cardiac filling due to positioning and subclinically compromised cord circulation at the level of the kyphosis, conspired to create the ischemic cord injury.

Patients with skeletal dysplasia are susceptible to neurological injury during manipulation of the head and neck and during the turn to a prone position for posterior spinal surgery. When this has occurred, the injury is usually located at the level of maximal compression or instability. Because baseline evoked responses in the supine position following anesthesia induction and prior to positioning were not obtained, we cannot define when during the procedure the spinal cord injury occurred. When responses were obtained, the critical significance of the absence of lower-limb MEPs in the face of preserved upper-limb responses was not appreciated. We had assumed that if injury were to occur, it would have been at the level of the craniovertebral compression, and that the lack of lower-limb MEP response was due to the patient’s Morquio syndrome.

Patients with skeletal dysplasia and chest wall deformity appear to be at risk for cord injury at sites remote from maximal compression. The causes of such injury may include regional spinal cord hypoperfusion secondary to spinal deformity; impaired cardiac filling due to cardiac or great vessel compression in a patient with a small thoracic cavity and deformed chest wall, in particular when the patient is positioned on transverse bolsters; or to induced arterial hypotension in an attempt to limit blood loss. Awake preoperative testing of the patient’s tolerance for the prone position may not reveal the patient’s susceptibility to the effects of this position under anesthesia. We recommend that 1) cardiac output be assessed prior to and after placement in the prone position; and 2) MEPs and SSEPs be performed after anesthesia induction but prior to placement in the prone position, after final positioning, and during the procedure.

Cardiovascular instability in the prone position
should prompt a return of the patient to the supine position and reevaluation of the arrangement of the bolsters, on the assumption that compromise to cardiac filling and great vessel circulation may have occurred and is related to positioning. Unexpected MEP results in an ambulatory patient, as in this patient, should alert the surgical team to effects on the spinal cord at level(s) other than those assumed to be at greatest risk and should prompt action based on the monitoring results.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this report.

Author contributions to the study and manuscript preparation include the following. Conception and design: Cochrane, Tong. Acquisition of data: Tong, Chen. Analysis and interpretation of data: all authors. Drafting the article: Cochrane, Tong. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Cochrane. Study supervision: Cochrane.

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