

## Anaesthesia recommendations for patients suffering from **Spinal muscular atrophy**

**Disease name:** Spinal muscular atrophy

**ICD 10:** G12.0 Infantile spinal muscle atrophy type I  
G12.1 Spinal muscle atrophy childhood form type II  
G12.1 Spinal muscle atrophy juvenile form type III

**Synonyms:** Spinal muscle atrophy type I: Werdnig-Hoffmans disease                      SMA I  
Spinal muscle atrophy type II: Dubowitz disease    SMA II  
Spinal muscle atrophy type III: Kugelberg-Welander disease                              SMA III

Spinal muscle atrophy (SMA) is an autosomal recessive inherited disease; characterised by progressive symmetrical muscle weakness. Clinical severity in SMA ranges from the extremely severe, prenatal onset to the mildest adult onset form. Clinical classification of SMA relies on both age at onset and maximal motor ability achieved by patients.

SMA is usually divided into three types; SMA I, II, III. Sometimes two additional groups are added in the age extremes – SMA 0 and SMA IV. Patients with early onset have a rapid progression.

SMA is the most common genetic cause of infant mortality.

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Medicine in progress



Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

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Find more information on the disease, its centres of reference and patient organisations on Orphanet: [www.orpha.net](http://www.orpha.net)

## **Disease summary**

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### **Common symptoms in SMA:**

#### Neurology

- Progressive muscle weakness: proximal > distal.
- Achieved motor milestones can be lost during progression of disease.

#### Cardiorespiratory

- Respiratory problems and failure is a main problem in patients with SMA I and II
- Scoliosis, nocturnal hypoventilation, aspiration, sleep breathing disorder and weak muscles can contribute to deterioration of both respiratory and cardiac function.

#### Orthopaedic

- Kyphoscoliosis, joint contractures
- Osteopenia

### **Genetics and pathophysiological background:**

#### Genetics:

- SMA is an autosomal recessive disease caused by a deletion in Survival Motor Neuron 1 gene (SMN1 gene). The product of a second gene SMN2 can partly compensate for the loss of SMN1.
- Put simply, the disease is caused by a mutation in SMN1 gene and the severity of the disease is inversely related to the expression of SMN2.

#### Pathophysiology:

- The SMN1 protein is expressed in many cells especially in anterior spinal motor neurons. The loss of SMN protein leads to degeneration of anterior spinal motor neurons and in severe cases degeneration of brainstem nuclei. The protein is involved in RNA processing. The SMN 1 protein is thought to be of importance for integrity of neuromuscular junction.

### **SMA 0 “Prenatal SMA”, Congenital SMA**

- Fatal at birth without immediate artificial ventilation.

### **SMA I Werdnig Hoffman disease “non sitters”**

- Age at onset: birth – 6 months
- Symptoms: Rapidly progressive muscle weakness. Early onset of respiratory failure, hypotonia, weakness. Reduced bulbar function.
- Natural lifespan without respiratory support <2 years

#### Neurology

- Floppy infants with weak cry
- Atrophic tongue with fasciculation's. Deep tendon reflexes are absent or poor.

#### Respiration

- Needs respiratory support to survive 12-24 months of age.

### Heart and circulation

- Cardiac malformations e.g septal defects and/or hypoplastic left heart are reported in children with neonatal SMA. Cardiac arrest and sudden infant death have been reported
- Autonomic dysfunction can develop in advanced disease.

### Nutrition/gastrointestinal

- Bulbar dysfunction
- Regurgitation

### Miscellaneous

- Osteopenia, fractures, thin ribs

There is evidence that anticipatory respiratory management including airway clearance with assisted cough, non invasive or invasive ventilatory support and adequate nutritional care are associated with longer survival in SMA1.

### **SMA II Dubowitz disease “sitters”**

- Age at onset: 6 – 12 months
- Symptoms: Progressive proximal limb weakness in infancy. Legs > arms. Never stand or walk. Ability to sit can be lost due to progression of the disease.
- Natural lifespan without treatment: 70% reach adulthood.

### Respiration

- Patients with severe SMA II may need respiratory support. Survival into adulthood increases the risk for respiratory problems.
- Respiratory support may be needed during acute disease or postoperatively.

### Heart and circulation

- Heart dysfunction is rare.

### Orthopaedic

- Kyphoscoliosis most invariable develops. Joint contractures

### Nutrition/gastrointestinal

- Difficulty in mouth opening and masticatory weakness can contribute to feeding problems
- Under-nutrition may be seen in severe cases due to eating difficulties
- Obesity due to physical inactivity can be seen in less severe cases.

### Miscellaneous

- Weak abdominal muscles increase the risk for caesarean section or instrumental delivery. Permanent deterioration of muscle function after delivery has been reported.

### **SMA III Kugelberg-Welander disease “walkers”**

- Age at onset: > 18 months
- Symptoms: Onset of proximal limb weakness during childhood. Legs > arms. Heel cord tightness. Scoliosis. Lumbar lordosis. Ability to walk can be lost due to progression of the disease.
- Natural lifespan without treatment: Normal.

### Respiration

- Usually normal
- Respiratory support rarely needed. However during acute illnesses or postoperative respiratory support may be needed.

#### Heart and circulation

- Heart dysfunction is rare. A few reports on conduction abnormalities as well as dilated cardiomyopathy have been reported.

#### Orthopaedic

- Kyfosciosis may develop
- Osteopenia. Increased risk for fractures.

#### Nutrition/gastrointestinal

- Obesity due to physical inactivity is a common problem.

#### Miscellaneous

- Weak abdominal muscles increase the risk for caesarean section or instrumental delivery. Permanent deterioration of muscle function after delivery has been reported.

#### **SMA IV Adult SMA**

- Age at onset: > 5 y, mostly > 30 y
- Symptoms: Onset of proximal leg weakness in adulthood.
- Natural lifespan without treatment: Normal

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### **Typical surgery**

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#### SMA I:

Common procedures: gastrostomy, fundoduplication, tracheotomy, muscle biopsy

#### SMA II:

Common procedures: surgery for scoliosis, talipes equinovarus (club foot), joint contracture release and muscle biopsy

#### SMA III:

Common procedures: surgery for scoliosis and talipes equinovarus, joint contracture release, caesarean section and muscle biopsy

#### SMA IV:

Common procedures: orthopaedic, common surgical procedures in adulthood.

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### **Type of anaesthesia**

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All types of anaesthetic techniques have been used for anaesthesia to SMA patients. None technique is absolutely contraindicated, none is perfect. The same apply for intubation techniques and devices. Every method has been used with success and failure.

The perioperative risks can be considerable and are often related to the respiratory system. Examples of complications are respiratory failure, prolonged intubation, atelectasis, nosocomial infections, upper airway obstruction, and difficult intubation. The risk for perioperative complications are high especially for patients with SMA I and II. Complications are most common in the post-operative period

It is important that the anaesthesia team has knowledge and experience. Perioperative care must be based on pathophysiological and pharmacological knowledge, case reports and common sense. A careful planning of the perioperative period is mandatory.

There are no evidence-based guidelines for the management of anaesthesia to patients with SMA since the disease is rare, clinical variation is enormous and new anaesthetic techniques emerge.

#### Sedation (procedure or conscious) to SMA patients:

In general, sedation is not recommended for SMA patients. The risk with (analgo)-sedation is higher than normal especially in SMA I and II patients, but also SMA III patients can be at risk, due to limited respiratory reserves.

Fibreoptic intubation is a special scenario, where sedation might be considered.

An anaesthetist must always be in charge for sedation of patients with SMA.

- SMA I patients: In spontaneously breathing patient sedation is contraindicated, except in very special situations.
- SMA II patients: In spontaneously breathing patients a decision about sedation must be preceded by a careful evaluation and judgment of respiratory function. Meticulous monitoring and high vigilance is mandatory. These patients are sometimes very difficult to intubate and non-invasive ventilation is an option if assisted ventilation is needed. Dexmedetomidine has been used in an adult and may be an option.
- SMA III patients: If there is no respiratory impairment the SMA III patient may be sedated. However, a careful preoperative evaluation of respiratory function should be done. If any respiratory impairment treat as SMA II

### **Necessary additional diagnostic procedures (preoperative)**

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The preoperative evaluation must be adapted to the clinical picture; which type of SMA is it and how advanced is the disease.

SMA I: preoperative respiratory evaluation and consultation is absolutely mandatory. The assessment of respiratory function should include physical examination, measurement of respiratory function, if possible, and evaluation of cough effectiveness. Cardiac malformations are more prevalent among patients with severe SMA I and a preoperative cardiac consultation, with ECG and echocardiography, is strongly recommended.

SMA II: preoperative respiratory consultation is strongly recommended. If the patient has severe scoliosis, respiratory insufficiency or suffers from obstructive sleep apnoea it must be remembered that, this can contribute to impairment of cardiac function. Preoperative cardiac investigations should be performed liberally and on wide clinical indications.

SMA III: Routine preoperative evaluation and laboratory investigations performed liberally and on clinical indications.

#### **SMA II and III – planning spinal epidural anaesthesia**

The anatomy of the spine should be investigated prior to any spinal or epidural anaesthesia. Apart from clinical investigation it can be necessary to consult an orthopaedic surgeon, and perform MR and/or CT scan. In the end, these investigations can give good help but provide

no guarantee for successful blockade. For the experienced anaesthetist ultrasound guidance can be helpful.

## **Ethics**

There can be ethical issues to take in consideration in the treatment of spinal muscle atrophy patients. It is important to remember that children and adolescents with SMA II and III report a good quality of life regardless of their functional status.

SMA I patients: Care of patients with SMA I often leads to ethical questions. When is treatment only causing prolonged suffering? With advanced treatment life can be prolonged to a state where the disease has progressed to a locked-in state with intact emotions and intellect. Clinicians and family can have different views on the child's quality of life.

SMA II and III patients: Even if the course is very different there could be ethical issues as well. What are the anaesthesia risks? Has *pro primo non nocere* (first do no harm) precedence over all other arguments?

Acute situations are not the time for discussions about escalation or limitation of treatment or level of care. Parents – and if possible patients – should be involved in discussions and decisions. Decisions must always be documented and well known.

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## **Particular preparation for airway management**

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Endotracheal intubation with direct laryngoscopy is the most commonly used technique for endotracheal intubation of SMA patients. However, the risk for difficult intubation is increased among SMA patients. Failed intubations causing death has been described.

There are several causes for difficult intubations e.g.

- Limited mobility of the cervical spine caused by joint contractures and/or previous spinal surgery.
- Limited mouth opening caused by ankylosis of the mandibular joint. In SMA patients reduced mouth opening is increasing with age.

### **Anaesthesia**

- It is of utmost importance to evaluate the probable intubation conditions preoperatively.
- The team responsible for SMA patients should have necessary skills, knowledge and experience. Anaesthesia to infants with SMA I is a challenge.
- Be prepared for difficult intubation even if a problematic intubation is not anticipated.
- Have all types of “difficult airway devices” easy available.
- A physician competent to perform a tracheotomy/coniotomy should be in the theatre if a very difficult intubation is anticipated
- Direct laryngoscopy may become more difficult with progression of the disease.

The choice of airway must be based on clinical situation and the knowledge that almost all SMA I and some SMA II patients need postoperative respiratory support. All types of airway devices from classical mask anaesthesia to tracheotomy have been used.

Non-invasive ventilation is an excellent bridge from intubation to spontaneous ventilation.

### **Airway devices:**

- *Endotracheal tube* inserted by direct laryngoscopy is a common scenario.

- *Airway guidewire* can be helpful.
- *Videolaryngoscopic intubation* can be an excellent alternative, more and more used.
- *Classical mask anaesthesia* has been used for short superficial procedures.
- *LMA* have been used and can be an alternative in shorter superficial procedures. Pressure supported ventilation with Proseal® LMA may be appropriate for patients with SMA II and III undergoing superficial surgery.
- *LMA* is an alternative in the case of tracheal granulomas or tracheal stenosis.
- *LMA* can be a rescue device in situation of failed intubation.
- *Flexible bronchoscope*. Fiberoptic intubation has been used in cases with difficult airway. Fiberoptic intubation can be impossible.
- *Retrograde intubation* have been described.
- *Tracheotomy or cricothyrotomy (coniotomy)* is the last alternative.

**SMA I:** Many patients will already have respiratory support e.g. NIV or via a tracheostomy. All will need postoperative respiratory support.

**SMA II:** A few patients will already have respiratory support. Some will need postoperative respiratory support.

**SMA III:**

- Older patients may need respiratory support postoperatively
- *Tracheotomy*
- After a tracheotomy a SMA I or II patient will lose the ability to any spontaneous respiration and also the ability to learn to speak. Patients who undergo tracheotomy often eventually die from complications related to the tracheal cannula.

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### **Particular preparation for transfusion or administration of blood products**

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There are no particular recommendations or disease related hazards.

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### **Particular preparation for anticoagulation**

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Coagulopathy is not a symptom of SMA.

However, some SMA patients have risk factors for thromboembolic postoperative complications such as immobilisation, overweight and/or pregnancy. This must be taken in consideration in the perioperative plan.

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### **Particular precautions for positioning, transport or mobilisation**

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**Positioning:** Scoliosis, joint contractures and osteopenia are very common among patients with SMA. Positioning of the patient during surgery is important, in order to prevent decubitus. Positioning should be adapted to the clinical picture. Vulnerable areas should be padded and joints and contractures should not be overstretched.

**Mobilisation:** Postoperative mobilisation is of outmost important. Intense physiotherapy with airway clearance technique can prevent airway secretion retention and treat hypoventilation. Assisted coughing techniques can be very valuable.

Intrahospital and transport on land (ambulance): Proper monitoring for the clinical status. If endotracheal airway tube or other airway device is *in situ* it should be very well secured. A plan and devices for handling accidental extubation should be in place. Anaesthetist should be responsible and come with the patient.

Air transport: Air travel can be dangerous for SMA patients with limited respiratory reserves. The oxygen level in the aircraft cabin is the same as approximately on 3000 m height. The lower oxygen concentration increases the respiratory work and can cause overt respiratory insufficiency. This risk is probably most dangerous for SMA infants.

Travel in general: Nowadays it is not uncommon for children with SMA to travel to other countries with their parents. Accidents and illnesses may occur, and the child may need anaesthesia and/or intensive care while abroad. There may be language barriers, and the standard of care may be suboptimal for the child. It is advisable that parents carry information regarding what kind of anaesthetic and intensive care problems should be anticipated for their child.

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### **Probable interaction between anaesthetic agents and patient's long-term medication**

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There is no specific drug for treatment of SMA. Many types of drugs and other treatments are and have been, tested for the treatment of SMA. There is no evidence based treatment or cure. There are many trials going on and it is possible that effective treatment can be found.

The possible interactions between any "anti-SMA drug" and anaesthetic drugs must always be controlled prior to anaesthesia. However for many experimental "anti-SMA drugs" data about interactions is not available. Ongoing trials can be found on [www.clinicaltrials.gov](http://www.clinicaltrials.gov).

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### **Anaesthesiologic procedure**

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An experienced team, prepared for complications, should perform anaesthesia for all patients, especially patients with severe/moderate SMA. A careful planning of the perioperative period is mandatory. Postoperative respiratory care is of paramount importance for a good outcome in patients with respiratory symptoms.

The anaesthetist in charge must personally meet the patient and evaluate the airway and what problems and obstacles any kyphoscoliosis and joint contractures can cause.

Anaesthetic risks vary variably between different types of SMA. Early onset of symptoms and end-stage disease indicates a high risk. The most common risks are related to respiratory disease and bulbar dysfunction.

It must be kept in mind that children with severe SMA have normal intellectual and emotional capacities. They also report a good satisfaction with life.

There are no case reports on anaesthesia to patients with SMA 0 however very large risks can be anticipated.

## Respiration

Complications from respiratory system is the dominating perioperative problem especially among SMA I and SMA II patients. A difficult airway is more common than in the normal population.

With a reliable airway in place ventilation is rarely a problem during surgery, however postoperative respiratory complications are a major cause of morbidity and mortality.

## Heart and circulation

Perioperative cardiovascular complications are rare. However it must be remembered that cardiac malformations are sometimes seen among patients with severe neonatal SMA.

## Gastrointestinal tract

The risk for gastro-oesophageal reflux and pulmonary aspiration must be evaluated and proper prophylactic measures undertaken.

## Nutrition/Metabolism

Hypoglycemia is seen in patients with low muscle and fat mass. These patients do not tolerate prolonged fasting well. Vigilance for hypoglycaemia is important. Blood glucose should be measured during the perioperative period and during acute illnesses in patients with severe SMA.

Hyperglycemia may be a problem among immobilised patients with overweight.

## Anaesthesia drugs

### Neuromuscular blocking agents:

- Intubation without neuromuscular blockers can be a good alternative, whenever possible.
- Succinylcholine: In the literature it is strongly recommended that suxamethonium/succinylcholine should be withheld from any patient with neuromuscular disease due to the risk of hyperkalaemia. SMA is a neuromuscular disease. However it should be noted that there are no reports in the literature of hyperkalaemia after the use of suxamethonium/succinylcholine.
- Non-depolarizing neuromuscular blockers: Sensitivity to non-depolarizing blockers appears to vary and can be prolonged. The dose must be titrated and the effect monitored carefully, both clinically and with a neuromuscular monitor.
- Neuromuscular monitoring can be unreliable in SMA patients. Muscle strength must be evaluated clinically before extubation. Do not rely only on train-of-four (TOF) measurements. Postoperative muscle weakness can be seen in spite of neuromuscular reversal and four equal twitches on TOF measurement.
- Even if not studied specifically in SMA patients, reversal of relaxation with sugammadex (Bridion®) seems to be a very good alternative that gives a rapid and reliable reversal of rocuronium and vecuronium relaxation. Sugammadex can also be a "rescue drug" in problematic situations for example failed intubation after rapid sequence induction. Sugammadex may not be available everywhere.

### Inhalational agents

- Inhalational agents appear to be a good choice in many situations. There are no reports of any MH or MH like reactions, rhabdomyolysis or neurological complication.

### Opioids

- Short-acting opioids are well suited for intraoperative use. Careful titration and monitoring (SaO<sub>2</sub>, EtCO<sub>2</sub> or TcCo<sub>2</sub>) are mandatory monitoring, especially in the postoperative period in SMA I and II patients, since respiratory depression could be disastrous. Opioid free general anaesthesia in combination with caudal anaesthesia have been used for gastrostomy and fundoplication in SMA I patients.

### Regional anaesthesia

- Regional anaesthesia could be a very good alternative, however not without obstacles.
- Wound infiltration anaesthesia is recommended whenever possible.
- Many different types of regional anaesthesia have been used successfully.
- Abnormal anatomy of the spine and rods for correction of scoliosis can make it impossible to administer spinal or epidural anaesthesia. Caudal anaesthesia has been used successfully to infants. Regional blocks can be technically difficult and the distribution of local anaesthetics could be altered. Failed effect of epidural blockade has been reported.

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### **Particular or additional monitoring**

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The monitoring must be adapted to the clinical situation for each patient and type of surgery.

In some special cases, such as kyphoscoliosis surgery, intraoperative neurophysiological investigations may be needed. Anaesthesia technique must be adjusted to the requirements of the neurophysiological investigations.

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### **Possible complications**

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Possible complications are many varying from decubitus to death.

Examples are:

- Postoperative respiratory failure
- Postoperative respiratory infection
- Airway obstruction
- Failed intubation
- Tracheal granuloma caused by several intubations.
- Decubitus
- Hypoglycemia

## Postoperative care

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Postoperative care is of utmost importance. Patients in need of nocturnal respiratory support will need it in the postoperative period and in acute illnesses. The risk for postoperative respiratory complications is related to the preoperative respiratory situation.

To treat postoperative hypoventilation and secretion retention non-invasive ventilation (NIV) and aggressive airway clearance techniques should be used. NIV is an extremely useful bridge from intubation to spontaneous ventilation. It is important to inform and educate the child and parents if NIV and airway clearance techniques are going to be used.

Any personal device for non-invasive ventilation and a “coughing aides” should, if possible, be brought to the hospital and used in the postoperative period since the availability in the hospital for these devices may be limited.

### SMA 0, I and II

- All patients with SMA I and many with SMA II will need postoperative care in an ICU setting.
- SMA I patients' needs almost always postoperative respiratory support. They need longer than normal hospital stay even after small procedures. However shorter hospital stay has been described after standardized procedures performed by a single experienced surgeon
- Some SMA II patients need postoperative respiratory support.

### SMA III

- There can be a need for postoperative respiratory support in SMA III patients with advanced disease and/or in old age.

### Oxygen

- Excessive use of oxygen is not recommended. Too much oxygen can mask hypoventilation due to muscle weakness.
- Only acceptable saturation (94 – 95%) should be sought. Blood gas analysis and EtCO<sub>2</sub> or TcCO<sub>2</sub> measurements can help to optimise oxygen delivery and ventilation.

There are reports on severe postoperative respiratory problems in spite of well-planned postoperative care with respiratory support in an ICU setting.

Postoperative pain management must be individualized and multimodal.

- Wound infiltration with local anaesthetics is recommended whenever possible.
- Carefully titrated opioids in combination with careful monitoring can be used. It must be remembered that opioid induced respiratory depression is more dangerous in patients with weak muscle and underdeveloped lungs.
- Paracetamol/acetaminophen and ibuprofen are useful.
- Regional anaesthesia has been used.
- Postoperative pain may exacerbate respiratory compromise.

## **Information about emergency-like situations / Differential diagnostics**

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*caused by the illness to give a tool to distinguish between a side effect of the anaesthetic procedure and a manifestation of the disease*

Since the clinical spectrum is extremely wide the anaesthesia procedures in emergency situations must be adapted to the clinical situation.

Emergency-like situations are mainly related to the respiratory system.

The diagnose SMA is almost always known in advance. There are no reports on problems with differential diagnoses preoperatively.

## **Ambulatory anaesthesia**

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SMA I: It is rarely feasible.

SMA II: Ambulatory anaesthesia can be an option after short procedures in patients without respiratory problems. Postoperative care/observation should usually be longer than for healthy children. Any ambulatory anaesthesia should be performed in a hospital setting with access to devices for the difficult airway and a postoperative unit/ICU for postoperative care.

Day care surgery may be appropriate in selected cases after minimally invasive surgery.

SMA III: Decisions about ambulatory anaesthesia should be based on and adapted to the clinical picture.

## **Obstetrical anaesthesia**

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Pregnancy is not uncommon in SMA II and SMA III patients. Permanent deterioration of the neurological symptoms after pregnancy is reported in about 30% of the patients.

Weak muscle power and pelvic disproportions are not uncommon and therefore caesarean section is more prevalent than in a normal population. Anaesthesia can be a difficult challenge. It is important with a good planning for delivery and for an emergency caesarean section.

*Spinal or epidural anaesthesia* is preferable, as always, and often feasible, however it can be impossible due to scoliosis and rods close to the spinal canal.

*General anaesthesia* is another option. The risk for airway problems is increased. Both uncomplicated intubations as well as emergency tracheotomy due to intubation failure are described.

*Local infiltration anaesthesia* for caesarean section is almost never used in our time and not mentioned in modern textbooks, however in extreme situations and with a skilled and gentle obstetrician and a cooperative patient it could be an alternative.

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*Please note that this guideline has not been reviewed by an anaesthesiologist but by two disease experts instead.*

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