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Situs inversus totalis

Spinal muscular atrophy

orphan anesthesia

a project of the German Society
of Anaesthesiology and Intensive Care Medicine

SUPPLEMENT NR. 11 | 2021

OrphanAnesthesia –

ein krankheitsübergreifendes Projekt des Wissenschaftlichen Arbeitskreises Kinderanästhesie der Deutschen Gesellschaft für Anästhesiologie und Intensivmedizin e.V.

Ziel des Projektes ist die Veröffentlichung von Handlungsempfehlungen zur anästhesiologischen Betreuung von Patienten mit seltenen Erkrankungen. Damit will Orphan Anesthesia einen wichtigen Beitrag zur Erhöhung der Patientensicherheit leisten.

Patienten mit seltenen Erkrankungen benötigen für verschiedene diagnostische oder therapeutische Prozeduren eine anästhesiologische Betreuung, die mit einem erhöhten Risiko für anästhesieassoziierte Komplikationen einhergehen. Weil diese Erkrankungen selten auftreten, können Anästhesisten damit keine Erfahrungen gesammelt haben, so dass für die Planung der Narkose die Einholung weiterer Information unerlässlich ist. Durch vorhandene spezifische Informationen kann die Inzidenz von mit der Narkose assoziierten Komplikationen gesenkt werden. Zur Verfügung stehendes Wissen schafft Sicherheit im Prozess der Patientenversorgung.

Die Handlungsempfehlungen von OrphanAnesthesia sind standardisiert und durchlaufen nach ihrer Erstellung einen Peer-Review-Prozess, an dem ein Anästhesist sowie ein weiterer Krankheitsexperte (z.B. Pädiater oder Neurologe) beteiligt sind. Das Projekt ist international ausgerichtet, so dass die Handlungsempfehlungen grundsätzlich in englischer Sprache veröffentlicht werden.

Ab Heft 5/2014 werden im monatlichen Rhythmus je zwei Handlungsempfehlungen als Supplement der A&I unter www.ai-online.info veröffentlicht. Als Bestandteil der A&I sind die Handlungsempfehlungen damit auch zitierfähig. Sonderdrucke können gegen Entgelt bestellt werden.

Bisher in A&I publizierte Handlungsempfehlungen finden Sie unter:

www.ai-online.info/Orphsuppl
www.orphananesthesia.eu

OrphanAnesthesia –

a project of the Scientific Working Group of Paediatric Anaesthesia of the German Society of Anaesthesiology and Intensive Care Medicine

The target of OrphanAnesthesia is the publication of anaesthesia recommendations for patients suffering from rare diseases in order to improve patients' safety. When it comes to the management of patients with rare diseases, there are only sparse evidence-based facts and even far less knowledge in the anaesthetic outcome. OrphanAnesthesia would like to merge this knowledge based on scientific publications and proven experience of specialists making it available for physicians worldwide free of charge.

All OrphanAnesthesia recommendations are standardized and need to pass a peer review process. They are being reviewed by at least one anaesthesiologist and another disease expert (e.g. paediatrician or neurologist) involved in the treatment of this group of patients.

The project OrphanAnesthesia is internationally oriented. Thus all recommendations will be published in English.

Starting with issue 5/2014, we'll publish the OrphanAnesthesia recommenations as a monthly supplement of A&I (Anästhesiologie & Intensivmedizin). Thus they can be accessed and downloaded via www.ai-online.info. As being part of the journal, the recommendations will be quotable. Reprints can be ordered for payment.

Find a survey of the recommendations published until now on:

www.ai-online.info/Orphsuppl
www.orphananesthesia.eu

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orphananesthesia

Anaesthesia recommendations for

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-Hoffman disease	SMA I
Spinal muscle atrophy type II: Dubowitz disease	SMA II
Spinal muscle atrophy type III: Kugelberg-Welander disease	SMA III

Disease summary: 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.

Medicine is in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

Find more information on the disease, its centres of reference and patient organisations on Orphanet: www.orpha.net

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Disease summary

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.

<u>Neurology</u>
- Floppy infants with weak cry
- Atrophic tongue with fasciculation. Deep tendon reflexes are absent or poor.

<u>Respiration</u>
- Needs respiratory support to survive 12–24 months of age.

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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 post-operatively.

Heart and circulation

- Heart dysfunction is rare.

Orthopaedic

- Kyphoscoliosis most invariably 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.

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Respiration
- Usually normal
- Respiratory support rarely needed. However, during acute illness or post-operatively, respiratory support may be needed.

Heart and circulation
- Heart dysfunction is rare. There are a few reports on conduction abnormalities as well as dilated cardiomyopathy.

Orthopaedic
- Kyphoscoliosis 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 years, mostly > 30 years
- Symptoms: Onset of proximal leg weakness in adulthood
- Natural lifespan without treatment: Normal.

Typical surgery

SMA I:

Common procedures: gastrostomy, fundoplication, 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.

Type of anaesthesia

All types of anaesthetic techniques have been used to administer anaesthesia to SMA patients. None of these techniques are absolutely contraindicated, none are perfect. The same applies to intubation techniques and devices. Every method has been used with success and failure.

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The peri-operative 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 risks for peri-operative 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. Peri-operative care must be based on pathophysiological and pharmacological knowledge, case reports and common sense. A careful planning of the peri-operative period is mandatory.

There are no evidence-based guidelines for the management of anaesthesia in 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 SMAI and II patients, but also SMA III patients can be at risk, due to limited respiratory reserves.

Fibre-optic intubation is a special scenario, in which sedation might be considered.

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

- SMA I patients: In a spontaneously breathing patient, sedation is contraindicated, except in very special situations.
- SMA II patients: In spontaneously breathing patients, a decision for 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 will be 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 pre-operative evaluation of respiratory function should be done. If there is any respiratory impairment, the patient should be treated like SMA II patients.

Necessary additional pre-operative testing (beside standard care)

The pre-operative evaluation must be adapted to the clinical picture: which type of SMA is it and how advanced is the disease.

SMA I: Pre-operative 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 pre-operative cardiac consultation with ECG and echocardiography is strongly recommended.

SMA II: Pre-operative 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 an impairment of cardiac function. Pre-operative cardiac investigations should be performed liberally and on wide clinical indications.

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SMA III: Routine pre-operative 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 might be necessary to consult an orthopaedic surgeon, and perform MR and/or CT scan. In the end, these investigations can give good help but are no guarantee for successful blockade. For the experienced anaesthetist, ultrasound guidance might be helpful.

Ethics

There can be ethical issues to take into 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 the discussions and decisions. Decisions must always be documented and well known.

Particular preparation for airway management

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 intubation 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 increases with age.

Anaesthesia

- It is of utmost importance to evaluate the probable intubation conditions pre-operatively.
- 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" easily 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.

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The choice of airway must be based on clinical situation and the knowledge that almost all SMA I and some SMA II patients need post-operative 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 intubation by way of direct laryngoscopy is a common scenario.
- Airway guidewire can be helpful.
- Video laryngoscopic intubation can be an excellent alternative, more and more used.
- Classical mask anaesthesia has been used for short superficial procedures.
- LMAs 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 case of tracheal granulomas or tracheal stenosis.
- LMA can be a rescue device in situations of failed intubation.
- Flexible bronchoscope. Fibre-optic intubation has been used in cases with difficult airway. Fibre-optic intubation might be impossible.
- Retrograde intubation has been described.
- Tracheotomy or cricothyrotomy (coniotomy) is the last alternative.

SMA I: Many patients will already have respiratory support, e.g. NIV or via tracheostomy. All will need post-operative respiratory support.

SMA II: A few patients will already have respiratory support. Some will need post-operative respiratory support.

SMA III:

- Older patients may need respiratory support post-operatively.
- Tracheotomy
- After a tracheotomy, a SMA I or II patient will lose the ability of 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.

Particular preparation for transfusion or administration of blood products

There are no particular recommendations or disease-related hazards.

Particular preparation for anticoagulation

Coagulopathy is not a symptom of SMA.

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

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Particular precautions for positioning, transportation and mobilisation

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: Post-operative mobilisation is of outmost importance. Intense physiotherapy with airway clearance techniques can prevent airway secretion retention and treat hypoventilation. Assisted coughing techniques can be very valuable.

Intrahospital and transport on land (ambulance): Proper monitoring of the clinical status. If an 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. An anaesthetist should be responsible and accompany the patient.

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

Traveling 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 also need anaesthesia and/or intensive care while abroad. There may be language barriers and the standard of care abroad might be suboptimal for the child. It is advisable that parents procure information about any anaesthetic and intensive care problems to be anticipated for their children.

Interactions of chronic disease and anaesthesia medications

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

Potential interactions between any “anti-SMA drug” and anaesthetic drugs must always be controlled prior to anaesthesia. However, for many experimental “anti-SMA drugs” data relating to interactions are not available. Ongoing trials can be found at www.clinicaltrials.gov.

Anaesthetic procedure

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. Post-operative 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 might be likely to cause.

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Anaesthetic risks vary among the different types of SMA. An early onset of symptoms and end-stage diseases indicate that there is 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 about anaesthesia administered to patients with SMA 0, but risks can be anticipated to be very high.

Respiration

Complications arising from the respiratory system is the dominating peri-operative 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, but post-operative respiratory complications are a major cause of morbidity and mortality.

Heart and circulation

Peri-operative cardiovascular complications are rare, but 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 taken.

Nutrition/metabolism

Hypoglycaemia 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 peri-operative period and during acute illnesses in patients with severe SMA.

Hyperglycaemia may be a problem among immobilised patients with overweight.

Anaesthetic 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-depolarising neuromuscular blockers: The sensitivity to non-depolarising 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 clinically evaluated before extubation. Do not rely only on train-of-four (TOF)

measurements. Post-operative muscle weakness can be seen in spite of neuro-muscular reversal and four equal twitches during TOF stimulation.
- Even if not studied specifically in SMA patients, reversal of relaxation with sugammadex seems to be a very good alternative and 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 (SaO2, EtCO2 or TcCo2) are mandatory, especially in the post-operative period in SMA I and II patients, since respiratory depression could be disastrous Opioid-free general anaesthesia in combination with caudal anaesthesia has been used for gastrostomy and fundoplication in SMA I patients.

Regional anaesthesia
- Regional anaesthesia could be a very good alternative, but it is 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 in infants. Regional blocks can be technically difficult and the distribution of local anaesthetics could be altered. A failed effect of an epidural blockade has been reported.

Particular or additional monitoring

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. The anaesthesia technique must be adjusted to the requirements of the neurophysiological investigations.

Possible complications

Possible complications are many, varying from decubitus to death.

Examples are:
- Post-operative respiratory failure
- Post-operative respiratory infection
- Airway obstruction
- Failed intubation
- Tracheal granuloma caused by several intubations
- Decubitus
- Hypoglycaemia.

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Post-operative care

Post-operative care is of outmost importance. Patients in need of nocturnal respiratory support will need it in the post-operative period and in acute illness. The risk of postoperative respiratory complications is related to the pre-operative respiratory situation.

To treat post-operative 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 will be 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 aid” should, if possible, be brought to the hospital and used in the post-operative period since the availability of these devices in the hospital might be limited.

SMA 0, I and II

- All patients with SMA I and many with SMA II will need post-operative care in an ICU setting.
- SMA I patients need almost always post-operative respiratory support. They need hospitalisations longer than normal even after small procedures. A shorter hospital stay has been described also after standardised procedures performed by a single experienced surgeon.
- Some SMA II patients need post-operative respiratory support.

SMA III

- There might be a need for post-operative respiratory support in SMA III patients with an advanced stage of the disease and/or who are of old age.

Oxygen

- An 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 EtCO2 or TcCO2 measurements can help to optimise oxygen delivery and ventilation.

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

Post-operative pain management must be individualised 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 muscles and underdeveloped lungs.
- Paracetamol/acetaminophen and ibuprofen are useful.
- Regional anaesthesia has been used.
- Post-operative pain may exacerbate respiratory compromise.

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Disease-related acute problems and effect on anaesthesia and recovery

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 diagnosis SMA is almost always known in advance. There are no reports on problems with differential diagnoses pre-operatively.

Ambulatory anaesthesia

SMA I: It is rarely feasible.

SMA II: Ambulatory anaesthesia can be an option after short procedures in patients without respiratory problems. Post-operative 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 post-operative unit/ICU for post-operative 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

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. A good planning for delivery and for an emergency caesarean section is therefore important.

Spinal or epidural anaesthesia is preferable, as always, and often feasible, but 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 have been described.

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

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