orphanaesthesia

Anaesthesia recommendations for

Arthrogryposis multiplex congenita

Disease name: Arthrogryposis multiplex congenita (AMC)

ICD 10: Q74.32

Synonyms: Arthrogryposis, amyoplasia congenita, congenital arthromyodysplasia, Guérin-Stern syndrome, myodystrophia fetalis deformans

Disease summary: AMC is the clinical description of a congenital syndrome with an estimated incidence of 1:3,000-10,000 [25,30,38]. Already within the uterus, the movement frequency of the foetus is reduced due to multiple – mostly symmetrical – joint contractures. Primarily, the great joints of extremities, the spine [12] but also the otolaryngologic region [10,11] are affected - with varying severity. Furthermore, a reduced muscle mass and a merely developed subcutaneous tissue are notable. During growth, numerous deformities are generated. Other organ systems can be involved (CNS, cardiovascular system, lungs, gastrointestinal tract, urogenital tract, abdominal wall). In most cases, patients develop normal intelligence.

Details of the cause of this nonprogressive disease are not clear. The reduced frequency of foetal movements seems to be significant – for different reasons [16]. Among others, external factors (e.g. oligohydramnion, uterine septi), primarily metabolic changes in muscle cells ("myopathic form") or a disturbance of the anterior cornual cells of the spinal cord ("neuropathic form") are discussed as possible triggers. Thus it is not surprising that numerous syndromes can be associated with AMC (e.g. Freeman-Sheldon syndrome [1,2], Brown syndrome [3], Bruck syndrome [4], ARC syndrome [5,46]). Even maternal diseases (Myasthenia gravis [7,44]) or drug consumption during pregnancy [6,31] as well as maternal/fetal infections as caused by de zika virus [52] can lead to AMC of the unborn.

The clinical classification can be performed according to three degrees of severity ("Munich classification") [37]:

Type 1: Primary affection of the extremities, possibly neck and trunk muscles; e.g.:

- Primary affection of hands and feet; part of this is the contractural arachnodactylia (so called distal arthrogryposes; autosomal dominant)

- Affection of all extremities – including shoulder and hip joint (60-80%) with symmetrical internal rotation of the shoulders, fixed extended elbows, flexion and extension contractures of the knee joints, as well as lower legs of cylindrical shape, talipes equinovarus (approx. 85%) (so called amyoplasia; in most cases sporadical).

Type 2: Primarily midline malformations; affections of extremities (vide type 1) as well as malformations of different organs (e.g. diaphragmatic hernia, pronounced scoliosis), pterygium. A differentiation of further subgroups in distal arthrogryposis is carried out.

Type 3: Further dysmorphic disorders and malformations; disorders of the CNS.

These are multifaceted syndromes of which AMC is just one aspect; the syndrome severity is originated by the additional malformation.

Frequently, patients with AMC have to undergo recurrent surgical interventions. In this context, several anaesthesiologic particularities have to be observed.

Medicine is in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

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Typical surgery

In the first place, patients with AMC have to undergo interventions in the field of (paediatric) orthopaedics [39]. Existing deformities are corrected (e.g. club feet [25], hip luxations and hip contractures, patellar luxations and Achilles tendon shortenings [25]).

Also upper extremity interventions are common [23,28].

After the first year of life, up to 65% of the patients develop scoliosis, which possibly turn out to be in need of correction [12].

Frequently, muscle biopsies are already performed in infants [22].

Possibly, further interventions are necessary depending on accompanying malformations (e.g. inguinal hernia, syndactyly, hypospadia, omphalocele [10], cryptorchism, renal calculi [17], gastrointestinal reflux [20], plastic deformities), underlying diseases or causative syndromes.

In many cases, the interventions last for several hours.

Type of anaesthesia

Main problems arising from anaesthesia in patients with AMC are the potentially difficult respiratory tract, poor venous conditions (which improve beyond infancy) as well as intraoperative hyperthermia [17-19,25,27,30]. Frequently, patients with AMC have to undergo recurrent surgical interventions already in infancy.

The preoperative establishment of regional anaesthesia seems to be advantageous – as the sole procedure or in combination with general anaesthesia. In small infants, regional anaesthesia is performed while the patients are asleep.

Normally, peripheral regional techniques work well – as described in the literature regarding upper and lower extremities [22,23,30]. Ultrasonography increases the success rate of peripheral nerve blocks in children with arthrogryposis because the response of nerve stimulation is often difficult to elicit due to the contractures [47]. However, replacement of normal muscle tissue with fat and fibrous tissue may aggravate visibility of nerves when using ultrasound.

For upper extremity surgery, axillary, supraclavicular and infraclavicular blocks are recommended. For lower extremity surgery, paravertebral, lumbar, epidural, iliofascial, femoral and sciatic nerve blocks provide pain relief. The use of ropivacaine (0,2%, 0,375%, 0,5% and 0,75%), bupivacaine (0,5%) and lidocaine (2%) is described. Alternatively a subcutaneous tunnelled catheter can be an option [54].

Occasionally, catheter placement can be complicated due to extremity contractures [23]. For better postoperative mobilization and sufficient pain therapy, it is recommendable to choose a catheter-based procedure.

Moreover, intraoperative infiltrations of wound edges have proven to be of value.

Due to existing spinal changes and ensuing unpredictable spread of local anaesthetics [51], regional anaesthesia methods close to the spinal cord are partly described to be impossible

(spinal anaesthesia [22]) or insufficient (unilateral epidural block [34]). In some case reports, a single shot spinal anaesthesia [42] or a continuous spinal anaesthesia appeared to be practicable [32,33].

As far as caudal anaesthesia is concerned, in infants good experiences have been made with the single shot method, which is applied most frequently [25,30]. However, the placement of a caudal catheter is possible as well [24]. The length of stay of a caudal catheter which is positioned close to the anal region should be limited in order to avoid the formation of abscesses; moreover, it should be checked at close intervals [25].

Analgo-sedation: Basically, there are no contraindications. In patients with AMC, a slightly increased responsiveness towards respiratory depressant drugs is assumed. With a patient history of gastroesophageal reflux, an increased risk for the aspiration of gastric content has to be taken into account [17,30].

Necessary additional pre-operative testing (beside standard care)

Basically, no particular preoperative diagnostics are required in patients with AMC. Due to the multifaceted characteristics and causes of this disease, an exact evaluation regarding severity of the disease, head shape, contractures as well as spinal and thoracic deformities is indispensable. Anamnestic data regarding previous operations and intubations provide valuable information regarding intubation risks which have to be anticipated.

Blood sampling: In the case of smaller interventions and inconspicuous anamnestic data, blood sampling can be avoided before surgery or rather postponed to the period after induction of anaesthesia – due to the well-recognized difficulty in venous access.

ECG/UCG: There is no specific congenital heart disease in connection with AMC. In literary sources, reports on anomalies like a persistent Ductus arteriosus, coarctations of the aorta and aortic stenoses are mostly evaluated as sporadic coincidences of this syndrome [21,30].

Thoracic radiography, lung function, blood gas analysis: These examinations should be carried out depending on the existence and the severity of a scoliosis, pulmonary hypoplasia, thoracic deformity or myopathy as well as the size and type of the intended intervention. Under these circumstances, a restrictive ventilation disorder might exist which should be clarified [12,17].

Specific questions regarding recurrent aspirations, pneumonia and reflux is part of every anamnesis.

In case of agitated children, blood gas analysis can be performed using capillary blood gas samples.

Screening for obstructive sleep apnoea in patients with a history of snoring is recommended. Arthrogryposis may lead to OSA and sleep hypoventilation [50].

Imaging of the cervical spine: In AMC patients with clinical symptoms related to cervical spine instability, it is recommended to carry out a neuroradiological assessment of the craniocervical junction. A case of atlantoaxial subluxation because of AMC has been described [13]. Scoliosis comprising the cervical spine or a Klippel-Feil syndrome can lead to these symptoms [30].

Further investigations: The organ system involved most frequently is – secondary to the musculoskeletal system – the urogenital tract (10-42%). In this respect, hypospadia and renal anomalies (duplex kidneys or the lack of kidneys, functional impairments of the kidney similar to Fanconi's syndrome) [15] are relevant in terms of anaesthesiology. Patients with the extremely rare ARC syndrome (arthrogryposis, renal dysfunction, cholestasis) [5] are particularly affected by kidney function impairments, hepatic disorders, hypothyroidism and congenital heart disease.

Particular preparation for airway management

In up to 25% of AMC patients, a difficult or rather impossible direct laryngoscopy and/or intubation can be anticipated [17,30]. Typical causal findings are for example: narrow oral aperture, mandibular hypoplasia (micrognathia), high arched palate, reduced tongue mobility, appearance similar to that of Pierre Robin syndrome, short neck, torticollis and omega-formed epiglottis. Cleft palates and big haemangiomas on the face have been described as well [8-11,26,30].

In most cases of difficult intubation, good conditions for ventilation through a laryngeal mask airway have been reported. However, there are a case reports which describe the failure of ventilation through a laryngeal mask airway because of massive anatomical changes [9,42,49].

According to all literature sources which have been consulted, mask respiration was possible without any problems. Particularly whenever anaesthetic intubation is indispensable, it is recommendable to have the (paediatric) respiratory aids in store which are generally used in the clinic or medical practice – beyond the laryngeal mask airway [54].

It is reported that successful intubations have been carried out via laryngeal mask airway, Cook airway exchange catheter, by performing flexible fiberoptic intubation as well as by using a McCoy laryngoscope with integrated Fogarty catheter [8-10]. The need for cricothyroidotomy in an emergency situation may occur [49].

Particular preparation for transfusion or administration of blood products

In AMC patients, usually no increased bleeding tendency is observed compared to patients without AMC. Again, a detailed bleeding anamnesis in every individual case determines the preoperative planning.

However, in the case of ARC syndrome (arthrogryposis, renal dysfunction, cholestasis) a functional thrombocyte disorder is described. One case of unexpectedly strong haemorrhage after percutaneous liver biopsy and one case with a life-threatening spontaneous nasal bleeding were described. [14,46].

Caution is advised in the case of patients who take valproic acid. The following are side effects relevant to haemorrhage: thrombocytopenia as well as a prolonged bleeding time because of a reduced fibrinogen concentration, factor VIII and inhibition of the secondary phase of platelet aggregation. If there is sufficient time before major surgical interventions (neurosurgery, hip operations), the active antiepileptic agent should possibly be changed to another substance class under expert medical supervision. It has been proven successful to start operations with expected abundant blood loss (e.g. pelvic osteotomy, scoliosis correction in terms of long-segment fusion) with cell-saver collection unit (reservoir).

Distal extremities surgery is frequently performed under local circulatory arrest (Tourniquet).

Particular preparation for anticoagulation

There is no evidence of a generally increased risk of thrombosis or the requirement for regular intake of anticoagulants by patients with AMC. However, as well in these cases, the anamnesis should include questions regarding e.g. a family tendency to develop thrombosis (thrombophilia).

Postoperatively – in particular after lower extremity interventions and during long periods of immobilisation – in general an adequate thrombosis prophylaxis has to be considered from the beginning of puberty on.

Particular precautions for positioning, transportation and mobilisation

Frequently, children with AMC are very slim as a consequence of reduced subcutaneous tissue, reduced muscle mass and due to nutrition problems which possibly exist in case of the neurogenic form of AMC (reflux, dysphagia, aspirations [11,17,30]).

Together with the pre-existing contractures, positioning patients for surgery becomes more difficult.

In order to avoid decubitus, acribic placement of cushions under all parts of the body touching the base is obligatory.

Already in the initial phase of anaesthesia, especially in small children, sufficient thermal management has to be remembered.

Due to the reduced flexibility of the joints and the resulting lack of physical exercise, already foetuses and newborn babies can develop osteopenia. The risk of fracturing the long hollow bones during birth is increased in affected children [35]. Osteoporosis already exists in infancy [23]. This underlines the importance of careful positioning, even though there is no evidence for the clinical relevance of osteoporosis in this respect.

Interactions of chronic disease and anaesthesia medications

There is no typical long-term medication in the case of AMC. As mentioned before, a detailed anamnesis should be carried out regarding concomitant diseases.

Anaesthetic procedure

It makes sense to give anxiolytic oral premedication by administering a benzodiazepine (e.g. Midazolam) in a careful dosage (0,4-0,5mg/kg) [25]; however, this is not prescribed by all authors [23].

Due to the usually poor vein quality and the mostly paediatric patient population, in many cases inhalational induction of anaesthesia is necessary. In this respect, unproblematic use

of Sevoflurane, Halothane [28,30], but as well the use of a N2O/O2 mixture [25] is described. In some cases it is not possible to obtain peripheral venous access and the primary placement of a central venous catheter is required [25,30] but since the introduction of ultrasound the ultrasound-guided IV placement may be helpful [53].

For intravenous induction of anaesthesia, Propofol and Thiopental have proved of value [20,23,25,28,30]. There are positive reports on ketamine/ midazolam as well [17,23,43].

Due to low muscle mass as well as due to the neurogenic and myopathic changes, AMC patients may show sensitive reactions to inhalational and intravenous anaesthetics, non-depolarizing muscle relaxants and to opiates [17,23,30,34]. Preference is given to derivates of these groups of drugs with short-term efficacy. In this way, the risk of postoperative complications can be reduced [18,19,30]. There are no absolute contraindications as far as certain narcotics are concerned.

Anaesthesia can be maintained by administering intravenous or inhalation anaesthetics (as well in combination with N2O).

Hemodynamic instability may be more likely among patients suffering from AMC. Hypotension with the need for vasopressors occurs also without underlying heart disease. There is no known direct link to cardiomyopathy [53]. Vasoactive medication should be available.

The described perioperative hyperthermia (differential diagnosis: malignant hyperthermia) in case of AMC is discussed under the topic “typical differential diagnoses”.

As a precaution, the use of Succinylcholine should be renounced. Although in many cases this drug has been used without causing any problems [27,34] - Succinylcholine can lead to high potassium increases in case of an underlying myopathic component of AMC [28,30].

Whenever possible, it is very reasonable to combine narcosis with regional anaesthesia for AMC patients. This leads to a reduced demand for anaesthetics, stable circulatory conditions, to the maintenance of spontaneous breathing (if necessary), prevention of stress and sufficient perioperative pain therapy. The latter is advantageous especially for the immediate postoperative phase and essential for the success of the operation [18,19,23,51].

Acetaminophen adjusted to weight can be used for postoperative pain therapy [54].

There are no described particularities regarding the application of local anaesthetics. The combination of local anaesthetics with adjuvants like clonidine in the case of peripheral regional anaesthesia [25] or of sufentanil, clonidine or rather adrenaline in the case of epidural (caudal) anaesthesia have been applied successfully [19,25].

Particular or additional monitoring

The type of monitoring should be chosen according to the type and extent of surgery, also considering potentially involved organ systems.

As a part of standard monitoring, the surveillance of temperature and CO2 is of particular importance, even in case of small interventions [25,28]. The reason is that AMC is frequently accompanied by an unspecific intraoperative hypermetabolic reaction (hyperthermia) [27,28,30]. Under these circumstances, the patient possibly develops acidosis and

hyperkalaemia. Repetitive intraoperative measurements of pH values and K+ concentrations might provide valuable information.

The application of relaxometry after administration of muscle relaxants is advisable (difficult to calculate duration of neuromuscular blockade bearing the risk of postoperative residual curarisation (PORC)) [17,23,30,34]).

Investigations regarding the measurement of anaesthetic depth (e.g. BIS) in AMC patients are not available.

Regarding the frequently extended duration of operations (and regarding the possibility of indwelling caudal / epidural catheters), a urinary catheter has been proven to be useful.

In the case of reflux or aspiration anamnesis, the placement of an intraoperative nasogastric tube can be taken into consideration.

Possible complications

Frequent problems in patients with AMC, which are described in literature, have been mentioned in the previous chapters or are described under “typical differential diagnoses”.

Post-operative care

Postoperatively, AMC patients seem to have a predisposition for respiratory problems. Frequent occurrence of post-extubation stridor after difficult intubation is reported [30]. An increased sensitivity to neuromuscular blockers and opioids was described [51].

In case of neurogenic components of the AMC and potentially additional residual effects of anaesthesia, patients sometimes develop a reduced control of the upper respiratory tract and swallowing disorders during the recovery phase. Especially patients with known restrictive pulmonary disorder (e.g. severe scoliosis, pulmonary hypoplasia, myopathia, pre-existing recurrent aspirations) show a postoperative tendency towards hypoventilation and atelectasis formation causing prolonged oxygen requirement [17-19,30].

Nevertheless, the risk of fatal respiratory complication (e.g. pneumonia) also may be increased in distal arthrogryposis type of AMC when orthopaedic alterations lead to difficulty to excrete sputum sufficiently [48].

Consequently, it is recommendable to monitor the patient’s respiratory function at close intervals for an extended period of time (e.g. pulsoxymetry). This applies particularly whenever opiates are used postoperatively (in paediatric patients). If there are no major problems, supervision can be performed at a normal ward.

Remarkably uncomplicated (pulmonary) course in the anaesthetic recovery room are reported, if a regional procedure has been implemented in addition to general anaesthesia and patients have received sufficient pain therapy after the operation [18,19,23-25].

Pain therapy: Please compare to chapter „Regional anaesthesia“.

The enormous importance of sufficient pain therapy has already been pointed out.

If it is not possible to apply regional anaesthesia, starting at the age of approx. 6 years the use of weight-adapted PCA (patient-controlled analgesia) can be taken into consideration in AMC patients[18].

Postoperatively, after ruling out known contraindications, routine pain medication can be applied according to the recommendations of the WHO (e.g. Paracetamol) [23].

In terms of differential diagnosis it has to be considered that in arthrogryposis, patients can develop pain with a neuropathic component. The description of successful pain therapy with Gabapentin in a newborn infant with AMC after therapy with non-opioids had failed is available [31].

Disease-related acute problems and effect on anaesthesia and recovery

Presumably the phenomenon most frequently discussed relating to AMC patients exposed to general anaesthesia is the question whether there is a predisposition of MH (malignant hyperthermia) or not.

It is known that in some cases AMC comes along with an increase in intraoperative body temperature (described up to 38.8 OC). In such cases, the patient possibly develops an increase in end-tidal CO2 and consecutive acidosis. In the reported cases, patients did not develop cyanosis; postoperative tests to detect the presence of urine myoglobin were negative. The surgical interventions could be concluded; the patients recovered quickly after surgery. No specific therapy was necessary. Active cooling (from 38 OC on) was sufficient. If muscle biopsies were carried out after this event, they were all unremarkable [20,27-30]. Besides others, one aspect ruling out MH as the causative disease is the fact that the phenomenon of intraoperative hyperthermia occurs during trigger-free anaesthesia as well [28,30]. A retrospective survey examined 396 narcosis in 67 patients with AMC and – despite exposition to known trigger substances (Halothane/Succinylcholine) – there was no case of MH [27]. Presumably, the probability to develop a hyperthermia depends on the primary genesis of AMC (neurogenic/myogenic) [28]. In a series of cases this was a 33% [30]. One retrospective study did not find evidence of increased odds of intraoperative hyperthermia or hypermetabolic responses but it could not rule out the possibility that a meaningful association exists [53].

Cases described in the literature which report the suspicion of MH are probably often a depiction of this hypermetabolic reaction, which occurs without any definitely identifiable reason or defect. In this context, hyperthermia in the case of arthrogryposis is thus described without other signs of MH [26,29].

However, there is a description (1984) of two MH cases in AMC patients. Both cases have been adequately confirmed in terms of diagnostics [40]. Literature of 2009 take a clear stand in this respect: In a total of two reported cases, a slight association of arthrogryposis and MH is assumed. This assumption is reinforced by a further case report of MH in literature [53]. Succinylcholine should be avoided [42].

In summary, the intraoperative hypermetabolic reaction observed frequently in AMC patients shows many differences compared to MH. For example there are no definite trigger factors, and it can be controlled by merely symptomatic measures. An adequate monitoring (see above) should be available [27-29].

Of course, the occurrence of MH in case of arthrogryposis – as in the case of every other anaesthesiology patient who has not been tested negative for MH – cannot be excluded and should be taken into consideration when corresponding symptoms are observed.

Ambulatory anaesthesia

Frequently, surgical patients with AMC are children. Due to the possible perioperative problems, they are considered as risk patients [19].

There are no recommendations regarding an outpatient procedure in the case of AMC. In many cases, an outpatient management is probably not reasonable due to the intensive postoperative orthopaedic therapy.

Outpatient anaesthesia in (pediatric) AMC patients should be an exception and only be carried out after having thoroughly weighed benefits and risks. (An imaginable constellation would be e.g.: minimal intervention, AMC without considerable concomitant malformations, good compliance of patient and/or the parents and a sufficiently extended aftercare).

Obstetrical anaesthesia

Most descriptions of anaesthesia in the case of AMC refer to paediatric patients. Case reports of narcosis in the case of arthrogryposis in adults can hardly be found – apart from gynaecological patients undergoing a Caesarean section.

Frequently, this type of delivery is primarily planned because of pelvic and spinal anomalies [32-34].

In most cases, the female patients have undergone anaesthesia several times in their childhood and therefore their medical history can easily be taken. [34]. A pre-existing problematic respiratory tract can deteriorate due to additional, pregnancy typical changes.

If suitable, neuraxial blockade should be preferred over general anaesthesia with rapid sequence induction.

Uneventful deliveries under continuous spinal anaesthesia are described [32,33]. However, back puncture can be difficult or not even practicable [22,34]. Irregular diffusion of the local anaesthetic solution in the case of peridural anaesthesia is possible [34].

In one case, a small tube (6,0 ID) and the change of the spatula was necessary after rapid sequence induction. The administered Succinylcholine had a clearly prolonged clinical action time [34].

If not the mother, but the unborn child is affected by AMC, repetitive deliveries turn out to be difficult because of the increased number of breech presentations. The child is at risk of suffering fractures [35]. Primary caesarean section can be necessary due to foetal head hyperextension [44]. Also ex utero intrapartum treatment (EXIT) procedures for the purpose of airway intervention including tracheostomy of the foetus are described [45]. A retrospective study concludes that new-born babies with arthrogryposis who require artificial respiration at the time of birth, have a bad prognosis [36].

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