

Anaesthesia recommendations for patients suffering from

Dermatomyositis

Disease name: Dermatomyositis

ICD 10: M33.90

Synonyms: Adult dermatomyositis, polymyositis, idiopathic inflammatory myopathy, juvenile dermatomyositis (onset < 18 yrs)

Disease summary: Dermatomyositis is a chronic degenerative disease of the muscle fibres and connective tissue caused by CD4+ and T cell mediated microvasculopathy, perifascicular atrophy and muscle microinfarcts. It is a part of the idiopathic inflammatory myositis trio which includes polymyositis, dermatomyositis and inclusion body myositis. The main pathology is considered to be the perimysial ischemia leading to atrophy and degeneration. The ischemia is a result of classical complement pathway triggered by C1-q attaching to the injured endothelium leading to membrane attack complex formation and deposition in the endothelium of the capillaries. The capillary occlusion ensued by this deposition and the subsequent reperfusion is the main cause for tissue damage. The trigger for inflammation is considered to be a combined interplay of genetic, immunologic predisposition and viral infection.

When the onset is less than 18 years of age, it is termed juvenile dermatomyositis.

Incidence is more frequent in females than males with peak onset between 30-60 yrs. 30% adults are left with mild to severe disability.

Medicine in progress



Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong



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

The disease mainly involves skin and muscles but other organs are variably involved. Proximal muscle weakness is very prominent. Skin involvement involves rash. The cutaneous manifestations are a result of the vasculopathy or photosensitivity; manifestations include various eruptions, such as heliotrope rash and Gottron's papules etc. Heart failure and rhythm disturbances, cardiac valvular abnormalities, anaemia, polyneuropathy, subacute cerebellar degeneration and aspiration/ interstitial pneumonia can be present. Gastroesophageal reflux disease and gastrointestinal bleeding / perforation is also reported.

In paediatric patients calcinosis and lipodystrophy is common. Chronic calcinosis can lead to muscle encasement, restrictions and joint contractures. Children may have remissions but most have residual skin lesions and muscle atrophy.

Adult dermatomyositis has a very strong association with carcinomas (45%) of lung, breast, or Non-Hodgkin's lymphoma. Gastrointestinal reflux disease (GERD) is one of the other systemic involvements. Overlap syndromes include inflammatory myopathy and connective tissue disorders like scleroderma, systemic lupus erythematosus (SLE) and Sjogrens syndromes.

Treatment is mainly corticosteroids and if resistant, immunosuppressants like azathioprine, methotrexate and cyclosporine. Immunoglobulin infusion, immunoabsorption and plasmapheresis have also shown to be effective.

Typical surgery

Apart from other surgical problems not associated with dermatomyositis, patients are likely to undergo surgery for gastroesophageal reflux disease, hiatus hernia, muscle contractures and joint problems in overlap syndromes. These patients are prone to gastric ulceration and high risk for small intestine and oesophageal perforation due to thrombosis of the small vessels of the bowel hence can present for emergent exploratory laparotomy. Other possible surgeries include carcinomatous mass excisions and chemoport placements.

Type of anaesthesia

Both regional and general anaesthesia use has been documented depending on the surgery involved.

Necessary additional diagnostic procedures (preoperative)

Detailed clinical examination is important to look for the current involvement and baseline status of the patient. Contracture affecting airway, respiratory and cardiac involvement and existing muscle weakness are important assessment and documentation areas.

Tests include routine blood tests like complete blood count to rule out anaemia, active infection and thrombocytopenia. Creatinine kinase and aldolase should be tested for muscle breakdown, aspartate aminotransferase (AST/SGOT), alanine aminotransferase (ALT/SGPT) and lactate dehydrogenase to assess liver function and document a baseline. Additionally renal function tests should be done.

Cardiac workup with electrocardiogram and echocardiogram to rule out rhythm disturbances and valvular or structural abnormalities is recommended. Cardiac enzymes can be done to rule out myocarditis.

Chest xray or high resolution CT scan (HRCT) to rule out pneumonia and interstitial lung disease can be done. Pulmonary function tests, spirometry, carbon monoxide diffusion and arterial blood gas is obtained if the respiratory compromise expected.

Any finding suggestive of malignancy may be further worked up by appropriate tests like CT scan and MRI.

Particular preparation for airway management

Skeletal deformity with restricted temporomandibular joint and cervical spine movement are seen in overlap syndromes hence detailed pre op assessment is invaluable. Videolaryngoscope, fiberoptic bronchoscope, LMA, bougie should be ready on the airway cart in anticipation of difficult airway.

Risk of aspiration is high due to poor coordination of swallowing, pooling of secretions in vallecula, decreased intestinal motility, decreased gastric emptying and oesophageal reflux due to diaphragmatic weakness. Therefore, airway protection strategies are to be used. Rapid sequence induction can reduce the aspiration risk to some extent. Rapid sequence induction is also recommended in case of oesophageal perforation to prevent increase in pneumomediastinum and to prevent the spread of oesophageal contents further into the mediastinum. A double lumen tube may be required if thoracic lavage is being considered.

Particular preparation for transfusion or administration of blood products

None.

Particular preparation for anticoagulation

Patients with dermatomyositis/polymyositis have an increased incidence of venous thromboembolism and the risk increases with old age, immunoglobulin therapy, steroids and other co-morbidities. Patients can be on long term antithrombotic prophylaxis. It is recommended to have patients on perioperative antithrombotic prophylaxis. The risk of continuation of the anticoagulants in the perioperative period should be carefully weighed against the risk of bleeding and in discussion with the surgeons.

Particular precautions for positioning, transport or mobilisation

Patients may have joint abnormalities and deformities making appropriate positioning very challenging. A reverse Trendelenburg position during induction of general anaesthesia may reduce chances of aspiration. Skin fragility and osteoporosis is common from long term steroid treatment hence adequate padding for bony prominences is mandatory. Adequate insulation is important if Raynaud's phenomenon is present.

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

Most patients are on long term medications like steroids or immunosuppressants. Cyclophosphamide and doxorubicin can cause cardiomyopathy, myositis, arrhythmias and heart failure hence can make patients very sensitive to induction agents. Careful titration of medications is of utmost importance. Cyclophosphamide can also cause hepatocellular failure hence, for medications metabolised by liver, doses need to be adjusted. Cyclophosphamide inhibits pseudo cholinesterase hence can prolong the duration of action for succinylcholine. Concurrent use of NSAIDS with methotrexate can reduce its excretion leading to potentially fatal toxicity therefore careful use of NSAIDS is endorsed.

Anaesthesiologic procedure

Frequent hospitalizations and intravenous medications can make IV access difficult. Muscle contractures and joint abnormalities can lead to difficult positioning.

Primary concerns are difficult airway, use of muscle relaxants and risk of aspiration.

Airway manipulations may be problematic due to the muscle contractures. Pre induction airway evaluation and possible airway challenges should be recognised and anaesthesia plan made accordingly. Awake intubation, fiberoptic bronchoscope or videolaryngoscope use can help in securing the airway.

Conflicting evidence exists regarding use of muscle relaxants. Patients may have abnormal response to skeletal muscle relaxants (SMR). If surgery does not demand muscle relaxation their use is best avoided.

The use of succinylcholine is mostly not recommended for the risk of hyperkalaemia. The risk of malignant hyperthermia is very unproven. Depolarising skeletal muscle relaxants are preferably avoided in lieu of quicker acting nondepolarising muscle relaxants. Enhanced effect and delayed onset of nondepolarising muscle relaxants has been documented. Prolonged duration of vecuronium and pancuronium has been reported. Delayed onset and prolonged duration of rocuronium is described in literature. Reports of increasing sensitivity to atracurium exist. Titration of muscle relaxants based on train of four monitoring can prevent overdosing and delayed recovery. Steroid induced myopathy can also increase the sensitivity to SMR further therefore titration is the best approach. No documented deviation from normal for reversal of muscle relaxants is recommended. Conflicting reports on onset of action for sugammadex exist but efficacy seems to be the same as for normal patients.

Airway protection from aspiration from the reflux disease is the other challenge. Rapid sequence induction and modified cricoid pressure can prevent aspiration to some degree. Extubation is recommended after adequate tidal volumes and vital capacity are reached and when the patient is awake, preferably in lateral position to avoid aspiration. Reports of use of LMA classic/proseal for surgeries on these patients exist however considering the high risk of aspiration, it should be preferably avoided.

Induction agents are to be used cautiously given the high incidence of cardiomyopathy and cardiac failure. Cardistable drugs like etomidate can be titrated to effect. It is imperative to be prepared for the various rhythm abnormalities and conduction abnormalities. Narcotics are to be used cautiously to prevent respiratory depression.

Interstitial lung disease and other pulmonary diseases increase perioperative mortality and morbidity to a great extent. Pre op assessment with evaluation of this risk is advisable. Supine position and anaesthesia could worsen oxygenation and ventilation. After general anaesthesia extubation can become difficult and may warrant ventilatory support.

Regional anaesthesia is preferred whenever possible as it can prevent aspiration complications and the necessity to monitor muscle relaxation. However, deformities and muscle contractures can make positioning difficult for administration of spinal or epidural. Low dose spinal anaesthesia prevents the respiratory compromise associated with high level block. Epidural supplementation can prolong the duration of anaesthesia and help titrate the dose without resulting in a high level block. Peripheral nerve block should be used whenever possible.

Not many cases are reported for anaesthesia in paediatric patients with dermatomyositis. As in adults, depolarizing muscle relaxants are preferably avoided. Careful titration of nondepolarising SMR is necessary. Airway can be difficult due to calcinosis associated joint contractures making positioning or range of motion restricted.

Steroids are the commonest drugs used to treat DM and can cause hyperglycaemia, skin fragility and osteoporosis. Blood sugar management pre and intra operative is important. Stress dose is valid if patient is on high dose steroid regimen.

Strict asepsis is important as most patients are on chronic immunosuppressants.

Particular or additional monitoring

Apart from standard monitoring like pulse oximetry, EKG, blood pressure, end tidal CO₂, gas monitors and temperature, depending on the case and the organ involvement, other supplementary monitoring should be included.

Cardiomyopathy, heart failure or pulmonary hypertension should include arterial blood pressure, central venous pressure, pulmonary artery and transesophageal echocardiogram as deemed necessary. Train of four monitor is very crucial for muscle relaxant dosing and redosing as DM is a waxing and waning disease with periodic flares and some residual weakness. It is essential to monitor neuromuscular blockade at baseline and after a muscle relaxant. Further doses should be based on the blockage level.

The adequate number of negative inspiratory force and vital capacity before extubation may help to ensure optimum muscle strength to prevent complications of hypoventilation and aspiration.

Possible complications

Aspiration pneumonia, residual muscle weakness, ventilatory inadequacy/depression is likely in the postoperative period.

Postoperative care

It is of paramount importance to watch for adequate ventilation and prevent aspiration. Maintenance of euthermia is vital for patients with Raynauds phenomenon. Narcotics for pain control are avoided to prevent respiratory depression or short acting narcotics can be used with caution. Other supplementary approaches for pain control should be used like epidural analgesia or transverse abdominis block to prevent atelectasis in abdominal surgeries.

Long surgeries with significant fluid shifts, cardiac or pulmonary involvement or residual weakness may benefit from ventilation and ICU monitoring.

Information about emergency-like situations / Differential diagnostics

Persistent muscle weakness post procedure could be residual muscle relaxant effect or the muscle weakness from the disease itself or myositis. This can be prevented by avoiding muscle relaxants or titrating the dose to effect. Adequate reversal and ventilation is the appropriate treatment.

Respiratory compromise can also be due to opioid overdose and can be reversed with naloxone if deemed essential.

It is safer to ventilate the patients till adequate muscle strength is regained and the respiratory drive is normalised, than to extubate and expose them to hypoxia and aspiration risk.

Ambulatory anaesthesia

Even if patients have no residual weakness or have recovered sufficiently, the stress of surgery and the anaesthesia drugs can cause some degree of muscle weakness making patients prone to respiratory depression or aspiration. It is prudent to have the patient observed to prevent any complications.

Short cases under local anaesthesia without any sedatives can be observed before discharging home. Judicious approach to the extent of surgery, the magnitude of the disease and the requirement of narcotics for pain control should guide the postoperative course with a very low threshold for overnight admission for observation.

Obstetrical anaesthesia

Pregnancy can exacerbate the disease manifestation. Regional anaesthesia has been found safe for caesarean section in pregnant women.

Pregnancy adds to the risk of aspiration and adequate prophylaxis and precautions should be taken. Rapid sequence induction is advised if general anaesthesia is planned.

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