

Emergency Care In Duchenne Muscular Dystrophy

Clinical guidance for acute presentations and emergency management



Example of our downloadable Emergency Card template

For families and HCPs

In an emergency situation take with you:
- emergency card
- medicines
- ventilation machine and Cough Assist
- instantly inform your Neuromuscular Team

Remember Stress Dose if:
- unable to take steroids in orally (vomiting, diarrhea)
- any stress situation (fracture, severe inflammation, surgery)
- offer your doctor PJ Nicholoff protocol (also after admission)

Watch out for oxygenotherapy – emphasize the need to use an ambubag.

In case of a fall/fracture: watch out for signs of Fat Embolism Syndrome:
- difficulty breathing
- pain in your chest or head
- a change in your behavior, like confusion, dizziness or not acting like yourself

This is an emergency, contact your doctor.

For health care professionals

Duchenne Muscular Dystrophy

Elevated risk for:
- Fat Embolism Syndrome (also with small fractures)
- hypercapnia (avoid oxygen alone, provide ventilation support f.e. ambubag)
- ventilation (BiPap) may be needed
- adrenal crisis (when using corticosteroids, stress dose is required)
- rhabdomyolysis (avoid suxamethonium, avoid inhaled anaesthetics and neuro-muscular blocking drugs)
- ketoacidose (fasting is a serious risk)
- aspiration (avoid laying patient flat if breathing is compromised)
- changing cardiac medication is a risk (hypotension is common)

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1. Adrenal Insufficiency and Stress-Dose Steroid Management

Most people with DMD receiving long-term corticosteroids are at risk of **HPA-axis suppression** and may be unable to mount an adequate cortisol response during acute illness, trauma or surgery. Patients tapering or recently discontinuing steroids should also be considered at risk until HPA-axis recovery is confirmed. **Steroids must not be abruptly stopped or omitted**, particularly during illness. Adrenal insufficiency may present with nausea/vomiting, abdominal pain, anorexia, marked fatigue or weakness, headache, confusion or unexpected motor deterioration; severe cases may progress to hypotension, hypoglycaemia, altered consciousness and cardiovascular collapse. Maintain a low threshold for stress-dose steroids in an unwell steroid-treated patient.

Minor illness (e.g. mild afebrile cold with the patient otherwise well): continue the usual steroid regimen. Moderate illness (fever, significant respiratory infection, marked fatigue, or mild diarrhoea/vomiting with oral medication retained): give oral stress-dose steroids – hydrocortisone 30–50 mg/m²/day divided every 6–8 hours (maximum 15 mg every 6–8 hours), or prednisone/prednisolone 8–12 mg/m²/day divided every 12 hours, until recovery and for a further 24 hours. Severe illness or suspected adrenal crisis (persistent vomiting preventing oral medication, severe diarrhoea with impaired absorption, reduced consciousness, major trauma, severe physiological instability or inability to take oral steroids): give immediate IM/IV hydrocortisone 100 mg/m² (maximum 100 mg); if BSA is unavailable, 25 mg (<1 year), 50 mg (1–5 years), 100 mg (≥6 years). Ongoing severe illness may require IV hydrocortisone 50–100 mg/m²/day every 4–6 hours or by continuous infusion, with concurrent assessment and treatment of blood pressure, glucose, hydration and the underlying illness.

Hydrocortisone, prednisone and prednisolone can be used for stress coverage. The optimal stress dose for deflazacort is uncertain, and vamorolone should not be relied upon for stress coverage; patients receiving either should receive additional hydrocortisone when stress dosing is required. Every steroid-treated person with DMD should have an individual emergency steroid plan and clearly documented adrenal-insufficiency risk. In patients receiving vamorolone, particularly those also

taking cardiac medications affecting the mineralocorticoid axis, diarrhoea/vomiting may warrant electrolyte monitoring, including sodium, because of the risk of hyponatraemia.

> ***Any person with DMD discontinuing corticosteroids should be considered at risk of adrenal insufficiency during acute illness, with prompt stress-dose steroid treatment.***

2. Respiratory Failure: Emergency Management

Respiratory failure is among the most critical emergencies in Duchenne muscular dystrophy (DMD) and remains a leading cause of hospitalization and death. Progressive weakening of the respiratory musculature impairs both effective ventilation and airway clearance, predisposing patients to infection, atelectasis, and acute respiratory decompensation. A respiratory tract infection and/or aspiration pneumonia can precipitate acute respiratory failure.

Early warning signs include frequent chest infections and inability to cough effectively. Late signs include: tachypnea, day-time fatigue, breathlessness that interrupts speech, morning headaches, disturbed sleep secondary to nocturnal hypoventilation, weight loss. In chronic respiratory insufficiency, daytime oxygen saturations may be normal, however, carbon dioxide and or serum bicarbonate levels may be raised, especially in the mornings. In acute on chronic respiratory failure oxygen saturations will be reduced.

Emergency management centers on restoring adequate ventilation. Non-invasive ventilation (e.g., BiPAP) is the first-line intervention, with invasive mechanical ventilation reserved for severe or refractory cases. Mechanical insufflation-exsufflation (cough-assist) devices support secretion clearance. Oxygen therapy must be administered with caution and under close monitoring: supplemental oxygen without ventilatory support may exacerbate hypoventilation and precipitate carbon dioxide retention, worsening respiratory failure which can present as loss of consciousness and/or respiratory arrest. Prompt, protocol-driven intervention is essential to limit complications and improve outcomes. If non-invasive ventilation

or carbon-dioxide monitoring is not available, ambu bag ventilation may be used to provide the pressure and volume needed for effective ventilation. If there is evidence of an acute infection appropriate broad spectrum antibiotics should be prescribed and adrenal replacement therapy with hydrocortisone for those patients who are steroid treated.

> *Respiratory complications remain a leading cause of morbidity and mortality in DMD.*

3. Cardiac Complications and Emergency Cardiac Care

Cardiac involvement is a major cause of morbidity and mortality in DMD. Progressive myocardial degeneration leads to dilated cardiomyopathy and an increased risk of arrhythmia and heart failure. Usually cardiomyopathy is asymptomatic, however, once severe cardiac failure may develop. Presenting features of cardiac failure may include dyspnea (and lower tolerance for horizontal position), and frequent manifestations of low cardiac output, which can be easily underdiagnosed. These include fatigue, loss of weight, abdominal pain and poor appetite. Palpitations or tachycardia may reveal arrhythmic complications such as atrial fibrillation or, less frequently, ventricular tachycardia, which can signal deteriorating cardiac function.

In the emergency setting, rapid assessment with electrocardiography, echocardiography, and laboratory investigation is essential. Blood troponin I should preferably be used, as troponin T is frequently elevated and is not specific to myocardial injury. Treatment may include aldosterone antagonists, loop diuretics, ACE inhibitors, beta-blockers, SGLT-2 inhibitors and antiarrhythmic agents to stabilize cardiac function. Early, proactive cardiac management has been shown to improve long-term outcomes and reduce the risk of sudden cardiac events. Hypotension and sinus tachycardia is not unusual in patients with DMD and cardiac medication shouldn't be discontinued unless they are symptomatic. However, treatment doses should be titrated against the blood pressure and renal function monitored regularly.

> *Cardiac disease is now a leading cause of death in DMD, accounting for a substantial proportion of mortality.*

4. Rhabdomyolysis: Signs and Emergency Treatment

Rhabdomyolysis is a serious condition characterized by the rapid breakdown of skeletal muscle and the release of intracellular contents into the circulation. In DMD, it may be precipitated by trauma, infection, or certain medications, including some anesthetic agents. Typical features include muscle pain, weakness, and dark (tea-colored) urine. If left untreated, rhabdomyolysis can progress to acute kidney injury and life-threatening electrolyte disturbance.

Emergency treatment centers on aggressive intravenous fluid resuscitation to preserve renal function, together with close monitoring of electrolytes and urine output. Early recognition and intervention are critical to prevent severe complications and support recovery. Due to low muscle mass, not creatinine, but cystatin C is used as a marker of renal function in Duchenne patients. In an acute situation the serum urea and blood electrolytes (sodium, potassium, calcium and phosphate) should be monitored.

> ***Rhabdomyolysis is a serious condition involving rapid muscle breakdown that demands prompt recognition.***

5. Ketoacidosis

Normoglycaemic ketoacidosis can develop rapidly in patients with DMD especially when the patient is dehydrated or unwell, leading to profound acid-base disturbances. Symptoms may include acute shortness of breath and fatigue.
If recognised, this complication is easy to treat with an intravenous glucose-infusion. Extra caution is needed in perioperative care and while treated with Dapagliflozin (group).

6. Hypertransaminasemia

Transaminases are highly concentrated in muscle cells. Consequently, muscular dystrophies such as DMD, lead to hypertransaminasemia. Elevation in ALT and AST is most striking during the early stages of disease.

7. Elevated Creatine kinase (CK) levels in Duchenne Muscular Dystrophy

In patients with DMD, the absence of dystrophin leads to progressive muscle fiber degeneration and increased muscle membrane permeability. As a result, creatine kinase (CK) leaks from damaged muscle cells into the bloodstream, causing markedly elevated serum CK levels.

8. Trauma, Fractures, and Fat Embolism Syndrome

Patients with DMD are at heightened risk of fracture owing to muscle weakness and reduced bone density, frequently compounded by long-term corticosteroid therapy. Even minor falls may cause significant injury and can precipitate complications such as fat embolism syndrome (FES). FES occurs when fat globules enter the circulation and occlude blood vessels, potentially producing respiratory distress, neurological impairment, and skin changes such as a petechial rash.

Emergency management of trauma in DMD includes careful immobilization of fractures, adequate analgesia, and close monitoring for complications. Early recognition of FES is critical, as it requires immediate supportive care to prevent life-threatening deterioration, especially support with non-invasive ventilation.

9. Dental Procedures and Emergency Precautions

Dental procedures in patients with DMD require particular care because of the risk of respiratory compromise, cardiac complications, aspiration, and adverse reactions to anesthesia. Even routine dental treatment may precipitate an emergency if appropriate precautions are not observed. Careful planning, coordination with the patient's medical team, and awareness of DMD-specific risks are therefore essential to ensure patient safety. Patients with DMD may have been taking steroids for many years and are thus at risk of adrenal insufficiency. Patients undergoing significant dental procedures, such as dental extractions, may require additional oral hydrocortisone for 48 hours to cover the procedure.

10. Anesthesia Risks and Emergency Precautions

Anesthesia poses unique and serious risks in DMD, making meticulous planning essential in any surgical or emergency setting.DMD patients should be admitted to high dependency or intensive care following any general anaesthetic. Many DMD patients will have been taking steroids for many years, all surgical procedures should be covered with hydrocortisone to prevent an adrenal crisis.

Certain anesthetic agents, particularly depolarizing muscle relaxants such as succinylcholine (suxamethonium), can trigger severe reactions including hyper-kalemia, rhabdomyolysis, and cardiac arrest, **and must be strictly avoided**. Total intravenous anesthesia (TIVA) is the recommended alternative, accompanied by continuous monitoring of cardiac and respiratory function. Emergency preparedness requires established protocols for the rapid management of complications. Awareness of these risks among all healthcare providers is critical to patient safety. Being informed about the latest results from cardiac and respiratory tests is of the essence.

> ***Anesthesia presents significant and potentially life-threatening risks in DMD patients.***

11. Emergency Department Protocols for Duchenne Patients

Emergency departments should adopt specialized protocols when treating patients with DMD to ensure safe and effective care. Initial assessment should prioritize airway, breathing, and circulation, with particular attention to respiratory and cardiac status. Excessive oxygen administration without adequate ventilatory support should be avoided, as it may worsen carbon dioxide retention; continuous monitoring of oxygen and carbon dioxide levels is essential (see section on respiratory failure).

Patients and caregivers should provide a detailed medical summary, including diagnosis, current medications, and baseline function, to guide clinical decision-making. A multidisciplinary approach involving neurology, cardiology, and pulmonology is often necessary for optimal care.

> *Emergency physicians must adopt specialized, DMD-specific protocols.*

12. Pain Management

Pain should be assessed and treated promptly in people with Duchenne muscular dystrophy (DMD), particularly following trauma, fracture or surgical intervention. Analgesia should be individualised and, where possible, a multimodal approach using non-opioid and opioid-sparing strategies should be employed. Paracetamol/acetaminophen may be used as first-line analgesia, with NSAIDs considered for short-term use where not contraindicated; gastroprotection should be considered, particularly in patients receiving corticosteroids. Opioids should be used cautiously and at the lowest effective dose because respiratory muscle weakness and reduced respiratory reserve increase the risk of opioid-induced respiratory depression; constipation is also common in DMD and may be exacerbated by opioids. Patients receiving opioids require appropriate respiratory monitoring. For severe acute pain, particularly associated with fractures or surgery, regional anaesthesia/nerve blocks may be considered in conjunction with specialist anaesthetic input.

> *Pain medicine should be chosen with particular attention to respiratory status and gastroprotection.*

13. Fluid Management

Fluid management should be individualised according to body weight, clinical volume status and cardiac function. In patients with shock, an initial bolus of 10 mL/kg of isotonic crystalloid may be given, followed by prompt reassessment of perfusion and signs of fluid overload. In patients with known or suspected cardiac dysfunction, smaller boluses (e.g. 5 mL/kg) should be considered, with early involvement of cardiology/critical care. Routine maintenance fluids should also be prescribed according to weight and reduced when cardiac or renal dysfunction is present.

> ***Fluid management should always take the patient's cardiac function into account, with lower volumes and closer monitoring in those with known or suspected cardiac dysfunction.***

14. Acute gastrointestinal issues

Acute gastric dilatation, gastroparesis and severe constipation should be considered in any person with DMD presenting with acute abdominal pain, distension, vomiting or reduced bowel movements. Gastrointestinal smooth-muscle dysfunction can cause severe gastric or intestinal dilatation and pseudo-obstruction, which may mimic mechanical bowel obstruction. Early abdominal imaging and gastroenterology/surgical consultation are recommended when symptoms are severe or atypical. Where mechanical obstruction, volvulus or perforation is not demonstrated, conservative management including bowel decompression, treatment of constipation, appropriate fluids and prokinetic therapy should be considered before proceeding to surgery.

> ***In DMD, severe constipation and gastrointestinal dysmotility should be recognised as potential causes of acute abdominal symptoms to avoid misdiagnosis and unnecessary surgical intervention.***

15. More Information, Relevant resources

Diagnosis and management of Duchenne muscular dystrophy, part 1: diagnosis, and neuromuscular, rehabilitation, endocrine, and gastrointestinal and nutritional management
Read report

Diagnosis and management of Duchenne muscular dystrophy, part 2: respiratory, cardiac, bone health, and orthopaedic management
Read report

Diagnosis and management of Duchenne muscular dystrophy, part 3: primary care, emergency management, psychosocial care, and transitions of care across the lifespan
Read report

PJ Nicholoff Steroid Protocol (stress doses)
Read report

Adult North Star Network (ANSN): Consensus Guideline For The Standard Of Care Of Adults With Duchenne Muscular Dystrophy
Read report

Primary Care and Emergency Department Management of the Patient With Duchenne Muscular Dystrophy https://publications.aap.org/pediatrics/article-abstract/142/Supplement_2/S90/11546/Primary-Care-and-Emergency-Department-Management?redirectedFrom=fulltext

Fat embolism syndrome in Duchenne muscular dystrophy, an underdiagnosed complication
https://pmc.ncbi.nlm.nih.gov/articles/PMC12958405/

Fatal Fat Embolism Syndrome Without Recognized Fracture in Patients With Duchenne Muscular Dystrophy: Two Case Reports
https://www.pedneur.com/article/S0887-8994(26)00155-4/fulltext

Ketoacidosis in Duchenne muscular dystrophy: A report on 4 cases
https://www.sciencedirect.com/science/article/abs/pii/S0960896617315080





Awareness of these risks among all healthcare providers is critical to patient safety



Contributors

Design & Layout
Hans Bulthuis, Hans. Concept & ontwerp, Netherlands

Photo Credits
Duchenne Parent Project Netherlands

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About the World Duchenne Organization
The World Duchenne Organization is an umbrella organization of national patient organizations for DMD and Becker MD (BMD) around the world. This organization aims to inform parents and people with DMD/BMD based on latest research, promote good standards of care, and actively participate in efforts develop safe and effective treatments for DMD/BMD and find a cure.

www.worldduchenne.org

Landjuweel 34
3905 PG Veenendaal
The Netherlands

+31 20 62 750 62
info@worldduchenne.org

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