

# Management of patients with musculoskeletal disease and burns

Rakesh Bhandary

## Abstract

Musculoskeletal disorders include a wide range of disorders which affect the locomotor system (i.e. muscles, bones, joints and associated connective tissues like tendons and ligaments, which are listed in chapter XIII of the International Classification of Diseases – 10). While the primary pathology affects the locomotor system, a number of these disorders are associated with systemic complications, resulting in increased morbidity and mortality in the perioperative period. Burns are an acute emergency and require anaesthetic or critical care input for assessment of injuries, early surgical intervention or organ support. Major burns are a multisystem disorder and although they account for less than 5% of all new burns cases, their management is complex and requires multidisciplinary approach in a specialized centre. This article highlights the important considerations for perioperative management of these conditions.

**Keywords** Burns; complications; difficult airway; dystrophy; fluid; muscle wasting; myotonia; nutrition; pre-assessment; resuscitation; sepsis; weakness

## Musculoskeletal disorders

This group includes a diverse range of disorders which affect the locomotor system and include disorders as listed in chapter 13 of WHO ICD-10,<sup>1</sup> which may affect:

- joints, such as osteoarthritis, rheumatoid arthritis, psoriatic arthritis, infectious arthritis
- bones, such as osteopenia, osteoporosis, associated fragility fractures, traumatic fractures
- muscles, such as sarcopenia, myositis, mitochondrial myopathy
- spine, such as ankylosing spondylitis, kyphosis, scoliosis, traumatic and pathological spine fracture
- connective tissues, affecting ligaments, tendons and supporting connective tissues such as Ehlers-Danlos disease, Marfan's syndrome.

Strictly speaking, primary disease of muscles such as muscular dystrophy, myotonic dystrophy and myasthenia gravis are neurological disorders and included in Chapter VI of International Classification of Diseases (ICD)-10. However, these have been mentioned in brief due to their association with the locomotor system. Musculoskeletal disease are either congenitally inherited or acquired as summarized in [Table 1](#).

*Rakesh Bhandary MB BS FRCA EDICM FFICM is a Consultant in Anaesthesia and Intensive Care Medicine at University Hospital of North Durham, Durham, UK. Conflict of interest: none declared.*

## Classification of musculoskeletal diseases

	Inherited	Acquired
Joints	Arthrogyposis multiplexa congenital Familial chondrocalcinosis	Infectious arthritis Non-inflammatory arthritis Sero-positive inflammatory arthritis (e.g. rheumatoid arthritis) inflammatory arthritis (rheumatoid arthritis) Sero-negative inflammatory arthritis (psoriatic arthritis) Crystal arthropathy (gout)
Spine	Scoliosis Kyphosis Lordosis Lumbosacral agenesis As component of spina bifida	Ankylosing spondylitis Traumatic fracture Pathological fracture Ankylosing hyperostosis Acquired kyphoscoliosis (as in muscular dystrophy)
Bones	Osteogenesis imperfecta Congenital fibrous dysplasia	Osteomyelitis Osteoporosis Osteomalacia and osteopenia
Muscles	Muscular dystrophy Myotonic dystrophy Mitochondrial myopathy McArdle's disease	Polymyositis Dermatomyositis Myasthenia gravis Rhabdomyolysis Acquired myopathy (steroid, thyrotoxicosis)
Connective tissue	Ehler Danlos Marfan's syndrome	Systemic sclerosis Lupus erythematosus Sjogren's disease Mixed connective tissue disorder

**Table 1**

It is pertinent to understand that musculoskeletal conditions may either involve only a part of the system (e.g. osteoarthritis of hip or knees) or they may be multisystemic in nature (e.g. rheumatoid arthritis, Marfan's syndrome, muscular dystrophies), leading to marked changes in physiology with or without compensation. Such patients may be receiving multiple and complex pharmacological regimens, including potent immunosuppressant drugs, which have an implication in the perioperative period. Due to the above issues, a meticulous preoperative assessment and planning is essential to reduce perioperative complications.

## Myasthenia gravis

Myasthenia gravis (MG) is an auto-immune disorder affecting the post-synaptic nicotinic acetylcholine receptors on striated muscles leading to varying degrees of fatigability. The disease is characterized by variable and fluctuating muscle weakness. Depending on the group of muscles affected, myasthenia gravis may be generalized, ocular or bulbar in nature. While acetylcholine receptor antibodies are present in most patients with generalized MG, anti-muscle specific kinase (anti-muSK) and anti-striated muscle (anti-Sm) antibody may be present in patients with ocular and bulbar variant. The mainstay of treatment

includes immunosuppression and acetylcholine esterase inhibitor drugs (e.g. pyridostigmine, neostigmine). Prognosis is generally good except for the elderly age group with thymoma.<sup>2</sup> Predictors of postoperative ventilation include: major body cavity surgery, disease duration >6 years., pyridostigmine requirement >750 mg.day<sup>-1</sup> and co-existing respiratory disease.

**Myotonic dystrophy (Stienert’s disease)**

Myotonic dystrophy is a condition characterized by muscle weakness and myotonia – the inability of muscle to relax following contraction. It is inherited in an autosomal dominant manner with the genetic mutation in the trinucleotide repeat sequence of the DMPK (dystrophia myotonica protein kinase) gene on chromosome 19. It manifests in the third or fourth decade, and is characterized by myotonia and myopathic faces – frontal balding, droopy jaw, and cataracts. Diabetes, atrioventricular conduction abnormality (e.g. third degree AV block), cardiomyopathy, epilepsy, hypogonadotropic hypogonadism and progressive bulbar and respiratory weakness by the fifth decade are the associated features. The treatment is mainly supportive, while sodium channel blockers like procainamide, phenytoin or mexiletine may be used for myotonia.

**Muscular dystrophy**

Muscular dystrophy is a heterogeneous group of inherited disorders characterized by variable pattern of muscle weakness and wasting. The pattern of inheritance maybe X-linked, autosomal dominant or autosomal recessive. Several types are known, with a few summarized in Table 2. Mutation affect genes coding for dystrophin and dystrophin associated glycoproteins (DAG). Both dystrophin and DAGs are important elements for sarcolemmal stability and are present not only in skeletal muscles but also smooth muscles, cardiac myocytes and neurones, hence the multisystem involvement. X-linked and autosomal recessive disorders have an early onset and poor prognosis, while autosomal dominant disorders have a good prognosis and life expectancy. Muscular dystrophies have some common overlapping features:

- variable degree and pattern of muscular weakness and wasting
- cardiomyopathy and AV block
- kyphoscoliosis
- restrictive lung disease secondary to kyphoscoliosis
- epilepsy and learning disability
- diabetes mellitus
- contractures, wheelchair dependence and bulbar weakness in the severe forms.

**Preoperative assessment**

The associated systemic complications of musculoskeletal disorders as highlighted in Table 3 validates the need for a meticulous and sometimes detailed anaesthetic pre-assessment.

A thorough pre-assessment begins will review medical records, which may provide invaluable information about patient’s clinical history, any systemic complications and medication history. Quite a few of these patients have had previous surgeries related to the musculoskeletal disorders or an unrelated pathology, and anaesthetic charts help complement the pre-assessment process. A good patient history, along with chronology of events, complications and medications is vital; however, cardio-respiratory tolerance maybe difficult to assess due to limited patient mobility. It is not unusual for patients to be on immunosuppressant drugs like steroids, disease-modifying anti-rheumatoid drugs (e.g. methotrexate, sulfasalazine, leflunomide, chloroquine) and monoclonal antibody therapy (e.g. infliximab, adalimumab). Long-term steroids can lead to suppression of the hypothalamic–pituitary–adrenal axis and patients require supplementation in the perioperative period as per latest recommendations.<sup>3</sup> Immunosuppressant drugs and monoclonal antibodies increases the risk of postoperative infection and may need discontinuation after discussion with the rheumatology team.

**Examination**

A medical examination is essential with particular emphasis on cardiorespiratory, airway and spinal deformity. Airway and spine

<b>Types of muscular dystrophy (MD)</b>			
<b>Type</b>	<b>Inheritance</b>	<b>Age of onset</b>	<b>Muscle group affected</b>
Duchenne’s MD	X-linked recessive	1–5 years	Pelvic, shoulder girdle, core muscles followed by respiratory and bulbar muscles
Becker’s MD	X-linked recessive	5–25 years	Pelvic, shoulder girdle, core muscles followed by respiratory and bulbar muscles
Emery Dreifuss MD	X-linked recessive (or autosomal dominant)	5–10 years	Humero-scapulo-peroneal muscle group followed by core muscles and respiratory muscles
Facio-scapulo-humeral MD (FSHD)	Autosomal dominant	Any age	Facial, shoulder muscles followed by pelvic & lower limb muscles
Oculopharyngeal MD	Autosomal dominant	Any age	External ocular, face, neck, arm & leg muscles
Distal MD	Autosomal dominant (rarely recessive)	50–60 years	Distal limb muscles
Limb girdle MD (Erb’s MD)	Autosomal recessive	10–30 years	Pelvic and shoulder girdle followed by limb muscles

**Table 2**

### Systemic complications of musculoskeletal diseases

System	Complications	Associated conditions
Cardiovascular	Aortic root dilatation	Marfan's disease, Ehler Danlos
	Aortic valve/mitral valve disease	Marfan's, Ehler Danlos
	Pericardial effusion	Rheumatoid arthritis
	Cardiomyopathy, AV block	Muscular dystrophy, Myotonic dystrophy
Respiratory	Restrictive lung disease	Ankylosing spondylitis, Kyphoscoliosis
	Pleural effusion & nodules	Rheumatoid arthritis
	Pulmonary fibrosis	Rheumatoid arthritis, systemic sclerosis
	Pneumothorax	Marfan's disease
Neurological	Bulbar weakness	Muscular dystrophy, myasthenia gravis
	Epilepsy	Muscular dystrophy, SLE
	Cervical myelopathy	Rheumatoid arthritis
Endocrine	Diabetes mellitus	Myotonic dystrophy, steroid therapy for musculoskeletal disease
	Hypothalamus – pituitary – adrenal suppression	Long term steroid therapy
Others	Difficult airway	Rheumatoid arthritis, ankylosing spondylitis
	Lenticular dislocation	Marfan's, Ehler Danlos
	Renal failure	SLE, methotrexate for arthritis
	Anaemia	Occult bleeding due to NSAID, anaemia of chronic disease
	Bone marrow suppression	DMARDs

**Table 3**

examination may reveal limited mouth opening, fixed flexion deformity of upper thoracic or cervical spine, and limited to non-existent neck extension. Cardiorespiratory examination is essential to look for signs of chronic hypoxia, hypercarbia or right ventricular strain. Neurological examination should be undertaken to document any pre-existing deficit and bulbar dysfunction.

#### Investigations

A full blood count, urea and electrolytes, and coagulation screen are needed in most cases. A full blood count may reveal signs of bone marrow suppression or degree of renal impairment secondary to medications or primary medical condition. Other investigations may be necessary to assess cardiorespiratory function:

- Arterial blood gases may reveal evidence of chronic type 2 respiratory failure secondary to restrictive lung disease.
- Imaging: chest X-ray may reveal thoracic sequelae of musculoskeletal disorders (e.g. pulmonary nodules, fibrosis, or dilated aortic root). If cervical spine (atlanto-axial) instability is suspected based on history or examination, cervical spine x-rays or magnetic resonance imaging (MRI) with interpretation by an expert radiologist maybe requested.
- Cardiorespiratory investigations: lung spirometry may reveal a restrictive pattern of lung disease. Electrography and echocardiogram may show signs of pulmonary hypertension or ventricular strain (left or right). However, these are static tests and in patients with limited mobility, dynamic tests like stress echocardiogram, myocardial perfusion imaging (MPI) or cardio-pulmonary exercise

testing (CPET) need to be considered. The use of perioperative CPET to evaluate the risk of adverse perioperative events has increased over the last decade.<sup>4</sup>

#### Intraoperative management

- *Intraoperative drugs:* induction agents and muscle relaxants should be used with caution. Depolarizing muscle relaxants (suxamethonium) should be avoided in myotonic dystrophy, as this may precipitate sustained contraction, and in certain muscle diseases like mitochondrial myopathy, along with inhalational anaesthetic agents, may precipitate malignant hyperthermia. If non-depolarizing muscle relaxants (NDMR) are used, a smaller dose (nearly one-tenth) of short or intermediate acting agent is advisable. Total intravenous anaesthesia (TIVA) is a safer alternative to inhalational anaesthesia.
- *Airway and ventilation:* utmost caution must be exercised in airway management, due to the possibility of difficult airway or unstable cervical spine (as in rheumatoid arthritis). Patients may need awake fibre-optic intubation, or asleep intubation using difficult airway equipment and with help immediately available. Extension of the neck should be avoided, as this may lead to severe neurological deterioration. Ventilation may be clinically challenging due to marked restrictive lung disease. Pressure control ventilation with high positive end-expiratory pressure (PEEP) akin to ARDSnet protocol maybe beneficial.
- *Positioning and skin care:* attention to positioning and skin care is an important aspect of intraoperative care. Positioning may be challenging due to immobile or restriction in joint movement, kyphoscoliosis or contractures. Gel

pads and a pressure-relieving mattress are essential to reduce the risk of damage at pressure points. Skin may be fragile and prone to easy breakdown in patients on long term steroids. Hence, it is important to forewarn patients about inadvertent injury despite diligence in care.

- **Monitoring:** basic monitoring as per AAGBI recommendations is the minimum standard of care for all anaesthetized patients. As the complexity of clinical condition or complexity of surgical procedure increases, advanced invasive monitoring may be required. Cardiac output monitoring also guides intraoperative fluid and inotrope or vasopressor administration.
- **Regional anaesthesia:** where possible, regional anaesthesia may be a safer alternative to general anaesthesia with airway management. Upper limb surgeries can be done under regional blocks (e.g. shoulder arthroscopy under interscalene block) or surgery below T10 dermatome under intrathecal or epidural anaesthesia. In certain conditions however, central neuraxial block may not be possible, as in ankylosing spondylitis, or provide asymmetrical block, as in kyphoscoliosis.

### Postoperative care

Postoperative care in these patients also require careful planning. Management at emergence from anaesthesia requires some precautions as induction to reduce the risk or avoid damage to the cervical spine. Extra measures like anti-sialagogues may be required in patients with bulbar involvement to reduce risk of aspiration.

Analgesic management can be challenging at times as some patients may be on long term opioids. Besides WHO ladder of analgesia, options include patient-controlled analgesia (PCA) with opiates, or intravenous infusion of ketamine or lignocaine. Where possible, epidural analgesia or regional analgesia with nerve catheters (e.g. sciatic nerve/femoral nerve/interscalene catheter) can be used to avoid systemic opiates.

Depending on the complexity of surgery and medical comorbidities, patients may need admission to level 1 care or a critical care unit.

### The patient with burns

Major burns (i.e. burns involving >20% of total body surface area or (TBSA)) are multisystemic disorder requiring a multidisciplinary approach. Mortality from burns has reduced significantly in the last four decades due to advances in surgical and critical care management of burns. The incidence of burns in England and Wales is approximately 130,000 per year with major burns accounting for 4.7/100,000 population (approximately 3000/year). The overall mortality for burns stands at 1.5% with patients with major burns, with major burns accounting for >10% of total mortality.<sup>5</sup>

### History and assessment

Burns may occur as an isolated injury, may be associated with inhalational injury or noxious gas poisoning, or as part of polytrauma. Hence, a history of circumstances surrounding the injury can give valuable information about the nature, probability and extent of other injuries. Attempts should be made to obtain full

medical history of the patient on admission to the emergency department.

### Primary survey

The initial assessment and management of severely burnt patient is similar to any trauma patient. Sequential assessment is essential to avoid missing any serious associated injury.

- **Airway with cervical spine control:** all burn patients should receive high-flow oxygen through a non-rebreathing face-mask on presentation. If the airway is compromised, it should be secured with tracheal intubation. Low GCS (<8), stridor, hypoxemia or hypercapnia, stridor, deep facial or full-thickness neck burns are indications for securing a definitive airway early on. An uncut endotracheal tube of 8.0 mm or above is the default choice to allow bronchoscopy at a later stage and prevent the tube from submerging into the mouth in case of facial oedema. Succinylcholine is safe within the first 24 hours of burns injury but should be avoided for a year at least due to the increased risk of hyperkalaemia.

Inhalational injury is defined as the aspiration of heated fluids or noxious products of incomplete combustion. Its probability increases if the incident occurred in an enclosed space with delayed escape or rescue. It takes three distinct forms:

1. Supraglottic inhalational injury – occurs above the vocal cords and is associated with hoarseness of voice, inspiratory stridor and swollen uvula
  2. Infraglottic inhalational injury – occurs below the vocal cord and associated with sloughing of epithelial lining, inflammation, atelectasis and obstruction.
  3. Chemical inhalational injury – carbon monoxide (CO) poisoning should be suspected in any unconscious patient. CO binds avidly to haemoglobin, shifting the oxygen dissociation curve to the left and also inhibits cytochrome oxidase system. This leads to tissue hypoxia and metabolic acidosis. Diagnosis and extent of CO poisoning can be confirmed through co-oximetry on ABGs. Treatment is oxygen, which reduces the half-life of carboxyhaemoglobin from 240 to 60 min. Cyanide poisoning should be suspected in burns patient with persistent lactic acidosis despite adequate fluid resuscitation. Intravenous hydroxycobalamin should be administered if acute cyanide poisoning is suspected.
- **Breathing:** tracheal position, chest movement and breathing should be assessed clinically as burns can compromise respiration. Full-thickness or deep dermal circumferential chest wall burn can lead to mechanical restriction of respiration and requires early escharotomy. Blast injuries may cause penetrating injuries causing pneumothorax; or it may cause pulmonary contusions and acute lung injury.
  - **Circulation:** major burns is associated with significant hypovolemia but at a later stage. Hypovolemia at presentation should prompt immediate assessment or investigation for bleeding. Two large bore intravenous cannulae should be inserted. Usual sites of cannulation may not be available but inguino-femoral regions tend to be relatively spared and can be used for large bore venous access of

central line insertion. If intravenous access proves difficult, intraosseous access or venous cut down should be considered. Warmed Hartmann's solution should be infused intravenously to clinical effect till percentage of burns and fluid requirement is estimated

- **Disability:** a rapid assessment of conscious level should be carried out by calculating the GCS score and noted along with size and reactivity of pupils to light
- **Exposure and estimation:** as with any trauma, patients with burns need to be exposed and examined, with a log-roll if indicated, to assess for presence of other injuries and to estimate the burn area. Burn area can be assessed using a standard Lund-Browder chart or Wallace's Rule of 9. However, caution should be exercised as patients of burns are prone to hypothermia
- **Fluids:** intravenous fluid resuscitation is required in adults with >15% TBSA burns or 10% TBSA burns with smoke inhalation; lack of fluid resuscitation will lead to hypovolaemia shock, organ dysfunction and ultimately death. The Parkland formula for fluid resuscitation is the most widely used formula for fluid resuscitation internationally. This formula estimates fluid requirement in the first 24 hours from time of burn (not presentation) at 4 ml/kg body weight per percentage of burn. The choice of fluid is Hartmann's solution and half is given in first 8 hours, followed by the other half in 16 hours. This formula is used in conjunction with regular review of physiological parameters and resuscitation end points, specifically urine output, the aim being 0.5 ml/kg per hour of urine output.

#### Anaesthesia for surgical management of burns

Early tangential excision and autologous split-thickness skin grafting remains the standard of care in burns centre worldwide. Some patients may be listed for escharotomy, which involves excision of circumferential burns to body parts.

In addition to standard pre-assessment, specific features of the history and physical examination deserve special focus in patient with burns as highlighted in Table 4. The intraoperative management provides its own set of challenges.

- Securing the airway maybe challenging due to reasons already stated or it may be difficult to maintain and fix in place due to facial burns or application of dressings or ointment.
- ECG chest leads may have to be stapled to skin and pulse oximeter probe may have to be applied to unusual body parts like pinna, nasal columella or tongue. Invasive monitoring can help with cardiac output measurement to guide fluid and vasopressor therapy.
- Major burns patient may have added pulmonary insult due to inhalation injury or sepsis related acute adult respiratory distress syndrome (ARDS). A protective lung ventilation strategy should be initiated to minimize ventilator associated lung injury.
- Insidious blood loss is a problem with tangential excision and split thickness grafts. 50–100 ml of blood loss for every percentage of burn should be expected and prepared for, and cross-matched blood should be available.
- The hypermetabolic state resets the body's core temperature to 38°C, making burns patients susceptible to

### Major preoperative concerns for patient with burns

Concerns	Implication
• Age and co-morbidities	Increased risk of mortality
• Facial burns/inhalational injury	Airway patency and management
• Pulmonary injury/lung injury	Difficulties with ventilation
• Adequacy of resuscitation	Exacerbation of shock with use of vasodilatory drugs
• Haematological issues	Anaemia and deranged coagulation
• Gastric stasis	Risk of aspiration
• Repeated surgeries	Repeated starvation despite hypermetabolic state
• Associated injuries	Difficulty with positioning
• Presence of infection	Exaggerated SIRS response during surgery
• Altered drug response	Titration of drug doses (e.g. muscle relaxants)
• Others	Difficult venous access, mental status

Table 4

hypothermia. Warm air blowers may be difficult to apply, so using an underbody heated mattress, warm and humidified theatre environment and warmed IV fluids can help reduce risk of hypothermia.

- Positioning could be challenging due to pattern of burns or they may need frequent change of position to make donor or excision sites accessible.
- Drugs require titration; suxamethonium should be avoided after the first 24 hours as mentioned earlier. The hypermetabolic state means enhanced drug elimination, hence patients need incremental doses of other muscle relaxant drugs
- Full-thickness burns are relatively painless as the nerve endings are also damaged. For others, unless contra-indicated, morphine is the mainstay of analgesia. Morphine requirement is increased in burns and peaks between week 10 and week 20 post-burns. Ketamine can be added as an adjuvant for analgesia, while benzodiazepines, clonidine or dexmedetomidine may be added for anxiolysis.

#### Intensive care unit management

Four interventions in critical care have improved outcomes for major burns in ICU.

- Fluid resuscitation, as mentioned earlier, is important to avoid hypovolemia related renal and multi-organ failure. The concept of 'fluid creep' and subsequent fluid overload due to inaccurate estimation can be avoided by using invasive cardiac output monitoring like transpulmonary thermodilution, lithium dilution or pulse contour analysis methods.
- Ventilation and recognition of its complication has been another important aspect of improving outcomes in the ICU. Protective lung ventilation strategy with high PEEP, low inspiratory pressures, titrating tidal volume to 6 ml/kg

of ideal body weight, permissive hypercapnia and realistic SpO<sub>2</sub> of 88%–92% has shown to improve outcomes.

- Nutrition is an extremely vital component of patient care. Burns cause a hypermetabolic state resulting in protein loss, reduction in lean body mass and hyperglycaemia. This leads to impaired wound healing, susceptibility to wound infection, longer inpatient admissions, organ failure and death. Early nasogastric or nasojejunal feeding with supplemental vitamins, amino acids and insulin therapy where required, has shown to decrease wound healing times.<sup>6</sup>
- Sepsis control, and early recognition, is a key component of critical care for the burns patient. There is a separate burn specific sepsis criteria as mentioned below. Burns specific sepsis include the following criteria, and presence of more than three criteria should trigger concern for sepsis:<sup>7</sup>
  - temperature >39°C or <36°C
  - progressive tachycardia >110/min
  - progressive tachypnoea (>25 breathes/min in spontaneously breathing patient or minute volume >12 l/min in ventilated patient)
  - thrombocytopenia <100,000/μl (not applied until 3 days after initial resuscitation)
  - hyperglycaemia (untreated blood glucose >12.5 mmol/l or >25% increase in insulin requirement)
  - feed intolerance >24 hours

Burns sepsis mandates aggressive treatment with antimicrobials and antifungals if needed. While white cell count and C-reactive protein are the current commonly used sepsis markers, pro-calcitonin (PCT) has been shown to be a useful marker in the adult population. ◆

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