New Zealand Paediatric Neurology Clinical Network

Duchenne Muscular Dystrophy and Perioperative Management

Clinician Resource

Key points

- DMD results in global muscle dysfunction and can affect respiratory and cardiac function.
- Appropriate pre-anaesthesia assessment can identify those at highest risk and aid in planning care during and after the procedure. Assessment is guided by clinical review augmented by physiological and blood tests.
- Many patients will be on daily steroids, which need to be continued in the perioperative period.
- Depolarising neuromuscular blocking agents are contraindicated in DMD
- Inhalational anaesthetics should be avoided where possible.
- Early post-operative mobilisation is important to avoid deconditioning.

Background

Duchenne muscular dystrophy (DMD) is part of a group of disorders in which there is a complete or partial deficiency of the muscle protein dystrophin and affects 1 in 5000 boys [1]. Children with this condition present with delayed motor milestones, and a constellation of other symptoms including toe walking and muscle pseudohypertrophy. Muscle weakness in DMD is invariably progressive. Respiratory and cardiac complications of DMD generally first manifest in the second decade of life.[2]

Individuals with DMD are at risk of many perioperative complications, both due to the primary muscle dysfunction (rhabdomyolysis and hyperkalaemia) and secondary cardiac/respiratory compromise. Because of this, the perioperative period requires careful management to ensure the best care of those with DMD.

Pre-anaesthesia Assessment

Pre-anaesthesia assessment is key to the successful management of all patients with DMD. A thorough assessment of the stage and impact of the disease should be performed by their lead clinician with respect to their cardiovascular and respiratory health. Consideration of assessment by a multidisciplinary team possibly including a neurologist, respiratory physician, cardiologist, physiotherapist and dietician should be made.[3] Consideration of the nature of the planned anaesthesia (duration, type, body position etc) and procedure (type of surgery) are equally important.

Baseline blood testing including a full blood count (to screen for anaemia), renal function and electrolytes (to

Box 1 Preoperative Management of DMD		
Check Diagnosis/screen for disorders		
Examination	Respiratory exam	
	Cardiac exam	
	Neuro exam	
	Scoliosis	
Investigation to consider	1st line	2nd line
	FBC	Echocardiogram
	Renal function	Blood gases
	Electrolytes	CXR
	Vitamin D	Polysomnography
	Lung function tests	Overnight oximetry
	ECG	
Nutritional optimisation		
Consider medications	Long term steroids = if unable to eat IV therapy	
	Opioid sensitivity	

ensure normal preoperative GFR and potassium homeostasis) can be useful tools in assessing disease activity. Elevated transaminases (AST and ALT) will be in line with the creatine kinase, as these enzymes are muscle- and liver-derived; this elevation does not indicate liver dysfunction if the GGT is normal. Assessment of the adequacy of Vitamin D intake is important for wound healing and muscle strength.

Nutritional status should be optimised prior to major surgical interventions, to ensure optimal muscle strength, wound healing and recovery time.

Cardiac function should be assessed with a routine ECG and cardiac examination, and there should be a low threshold for performing an echocardiogram to assess cardiac function [4]. Resting tachycardia should be an absolute indication for formal assessment of pre-operative cardiac function by echocardiogram (or cardiac MRI if an appropriate window for echocardiogram cannot be achieved).

Respiratory assessment ought to consider the individual's degree of weakness, history of respiratory difficulties (eg pneumonias), prior anaesthetic history, symptoms of sleep disordered breathing (snoring, morning headaches, etc), current use of respiratory support (non-invasive ventilation), strength of cough and need for regular airway clearance manoeuvres (including cough assist) and the presence of additional risk factors (scoliosis, pulmonary hypertension). This will guide the need for further assessment such as pulmonary function tests (FVC, peak cough flow), chest x-rays, blood gas, oximetry and other sleep physiology tests. More detailed assessment is recommended prior to major thoracic procedures (eg scoliosis surgery) – Starship Clinical Guideline. Forced vital capacity (FVC) provides an indication of respiratory muscle strength and has some predictive value for anaesthetic complications. A FVC of less than 50% suggests some increased risk and less than 30%, high risk. If not already receiving ventilatory support (eg NIV), more detailed physiological assessment and/or training in NIV (non-invasive ventilation) may be appropriate pre-anaesthetic to facilitate care afterwards, particularly for major surgeries [4, 5]. As some guide, FVC is typically still normal at age 8-10 years but has fallen by 14-18 years (although, individuals vary) [7]. Similarly, in those >12 years of age, a peak cough flow <270l/min may identify those whom would benefit from cough-assist techniques and undergo pre-anaesthetic training [6, 8]. If in doubt, consultation with a respiratory or sleep specialist is recommended.

Many children with DMD will be on ongoing (daily) steroid medication. Corticosteroids are the only medical therapy shown to slow the decline of muscle strength and progression of scoliosis and respiratory dysfunction in DMD. Continuation of steroids around surgery is essential to prevent an adrenal crisis and if children are not able to continue their oral steroids, IV administration should be planned for. Further advice is available from the Starship Steroid Management Guideline, or an endocrinologist.

Anaesthetic and intra-operative considerations

Children with DMD are at risk of a number of complications of anaesthesia. In those viewed at risk, anaesthesia ought only be performed in a centre with appropriate expertise and post operative (ICU) care including access to skilled physiotherapists.

While these children are not at risk of the malignant hyperthermia syndrome seen in neuromuscular disorders associated with ryanodine receptor gene mutations, they are at risk of anaesthesia-induced rhabdomyolysis (AIR). This potentially life-threatening complication of anaesthesia has been well described with use of depolarising agents such as suxamethonium in boys with DMD [9, 10].

Depolarising agents can cause skeletal muscle breakdown, leading to a release of myoglobin, creatine kinase and potassium. The use of such agents is contraindicated in children with DMD [6].

Rhabdomyolysis has been described with use of inhalational/halogenated anaesthetics with hyperkalaemic cardiac arrest also occurring when succinylcholine was used. [7]. Use of intravenous anaesthetic agents should be considered where possible [8]. Difficult IV access is a feature with some of these patients and many centres allow brief use of inhalational agents to secure IV access before switching to an intravenous anaesthetic technique, when this is required.

Careful management of physiological state to prevent hypotension or major fluid shifts are important in cases where a cardiomyopathy is present [9].

Post-operative management

Respiratory considerations continue to be paramount in the post-anaesthetic period, and the use of specific extubation criteria and practice is recommended. Proposed extubation criteria /practice includes [4]:

- Presence of minimal airway secretions
- Access /use of specific airway clearance techniques (eg cough-assist)
- Oxygen saturations >94% without supplemental oxygen for more than 12 hours
- Direct transfer from invasive ventilation to NIV in those using NIV pre-anaesthetic or viewed as high risk (FVC <30%)

Effective analgesia and early mobilisation is key to the management of patients with DMD as deconditioning can occur quickly. Patients can permanently transition from an ambulant stage to a non-ambulant stage of the illness if early mobilisation is not prioritised [10].
Opioid analgesia should be carefully titrated to effect as patients with DMD are generally opioid sensitive [13].

References

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