

Syllabus	NA_IK_18
Topic	Myotonic dystrophy

A 30 year-old man presents for laparoscopic cholecystectomy. He was diagnosed with myotonic dystrophy 10 years ago.

a)

Define myotonia (1 mark)

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b)

Describe the underlying pathophysiology of myotonic dystrophy and Mendelian inheritance (3 marks)

Pathophysiology:

.....

Inheritance:

c)

Fill in the box below to describe the systemic problems associated with myotonic dystrophy relevant to anaesthesia (7 marks)

System	Problems relevant to anaesthesia
Cardiovascular system	1) 2)
Respiratory system	1) 2)
Central nervous system	1)
Gastrointestinal system	1)
Endocrine system	1)

d)

Give four aspects of your induction of anaesthesia specific to myotonic dystrophy (4 marks)

- 1)
- 2)
- 3)
- 4)

e)

Give four intra-operative aims specific to myotonic dystrophy (4 marks)

1)

2)

3)

4)

f)

Name a neuromuscular condition which carries a greater risk of malignant hyperthermia compared to the general population (1 mark)

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	Answer	Mark	Guidance
a)	<ul style="list-style-type: none"> Incomplete muscle relaxation 	1	
b)	<u>Pathophysiology:</u> <ul style="list-style-type: none"> Disorder of sodium and chloride channels Sustained muscle contraction 	1	<i>Extra detail:</i> -Leaving muscles in a hyper excitable state -Repetitive action potentials
	<u>Inheritance:</u> <ul style="list-style-type: none"> Autosomal Dominant 	1	
		1	
c)	<u>Cardiovascular system</u> <ul style="list-style-type: none"> Conduction defects (risk of sudden cardiac death) Cardiomyopathy Congestive cardiac failure Hypotensive and myocardial effects of anaesthetic agents may be exaggerated 	Any 2	
	<u>Respiratory system</u> <ul style="list-style-type: none"> Restrictive lung disease Poor cough Respiratory muscle weakness Central + OSA Risk of aspiration and pneumonia Exaggerated respiratory depressant effect of opioids and IV anaesthetic agents 	Any 2	
	<u>Central nervous system</u> <ul style="list-style-type: none"> Reduced intelligence – issues with capacity and consent Early cataracts – vision Myotonia results from shivering or suxamethonium 	Any 1	
	<u>Gastrointestinal system</u> <ul style="list-style-type: none"> Gastrointestinal reflux 	Any 1	
	<u>Endocrine system</u> <ul style="list-style-type: none"> Increased risk of type 2 diabetes Increased risk of hypothyroidism Adrenal insufficiency 	Any 1	

d)	<ul style="list-style-type: none"> • Use of PPI or prokinetics • Consider awake arterial line • Consider RSI • Judicious use of induction agent • Intubate without muscle relaxant • Avoid suxamthonium • Avoid neuromuscular monitoring 	Any 4	
e)	<ul style="list-style-type: none"> • Avoid hypothermia • Avoid shivering • Monitor BMs • Ensure defibrillator available • Reverse fully with suggamadex • Avoid neostigmine • Avoid nerve stimulator • Judicious use of opioids • Multimodal analgesia 	Any 4	
f)	<ul style="list-style-type: none"> • Central core disease <p>HYPOkalaemic periodic paralysis</p>	1	

References

- 1) Marsh S, Ross N, Pittard A. Neuromuscular disorders and anaesthesia. Part 1: generic anaesthetic management. CEACCP (2011) 11(4)115–118
<https://academic.oup.com/bjaed/article/11/4/115/267032>
- 2) Marsh S, Pittard A. Neuromuscular disorders and anaesthesia. Part 2: specific neuromuscular disorders, CEACCP (2011) 11(4)119–123
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