orphanaesthesia

Anaesthesia recommendations for

Emery-Dreifuss muscular dystrophy

Disease name: Emery-Dreifuss muscular dystrophy (EDMD)

ICD 10: G71.0

Synonyms:

Disease summary: Benign Scapuloperoneal Muscular Dystrophy, Hauptmann-Thannhauser Muscular Dystrophy, EDMD 1 (X-linked affecting EMD gene), EDMD 2/3 (autosomal dominant/recessive affecting LMNA gene)

Other laminopathies may be phenotypically similar.

Joint contractures (Achilles tendons, elbows and spine). Posterior neck and lower back involvement may result in a rigid spine.

Humeroperoneal muscle weakness by adolescence, later involving the proximal limb girdles.

Cardiac conduction abnormalities (PR prolongation & complete heart block), atrial and ventricular arrhythmias and dilated cardiomyopathy usually beginning in the 2nd – 3rd decade of life.

Medicine is in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

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

Emergency information

	AIRWAY / ANAESTHETIC TECHNIQUE	prepare for a difficult airway (restricted neck movement, joint contractures) – assessment of aspiration risk necessary (reduced gut motility) – no definitive (dis)advantage for GA (TIVA / balanced) or RA – RA techniques might be challenging (joint contractures)
B	**BLOOD PRODUCTS (COAGULATION)**	prepare for intra-operative increased bleeding risk (mechanism not fully understood)
C	**CIRCULATION**	cardiac conduction abnormalities, atrial / ventricular arrhythmias and DCM may occur – judicious use of fluids and a means of external pacing (should it be necessary) will reduce risks of cardiac failure – availability of DC cardioversion recommended – consider IBP
D	**DRUGS**	no risk for MH – restrictive use of opioids / muscle relaxants – it may be prudent to avoid suxamethonium and inhalational anasthetics during first decade of life (induction of anaesthesia-induced rhabdomyolysis, which may mimic MH type picture – however, evidence lacking)
E	**EQUIPMENT**	patient positioning / mobilisation with caution (contractures) – neuromuscular monitoring recommended – anticipate respiratory complications in PACU / IMC / ICU

Typical surgery

Tendon release; Corrective spinal surgery; Caesarean section; Permanent pacemaker / defibrillator insertion.

Type of anaesthesia

All forms of anaesthesia are possible. Intubation and central neuro-axial blocks may be difficult due to joint contractures.

Although evidence is lacking it may be prudent to avoid suxamethonium and inhalational anaesthetics during the first decade of life to avoid anaesthesia-induced rhabdomyolysis.

Necessary additional pre-operative testing (beside standard care)

Electrocardiogram (ECG).

Echocardiography and 24 hour ambulatory ECG telemetry are recommended.

Cardiac electrophysiological testing should be considered in patients with conduction defects.

Particular preparation for airway management

There is the potential for reduced gut motility, aspiration risk should be assessed and managed. The airway plan should include options for dealing with a difficult airway secondary to restricted neck movement.

Particular preparation for transfusion or administration of blood products

May be at increased risk of intra-operative bleeding; mechanism not fully understood.

Consider anti-fibrinolytics and early treatment of acquired coagulopathy.

Particular preparation for anticoagulation

Antiplatelet or anticoagulation prophylaxis for atrial fibrillation and atrial flutter is recommended barring contraindication.

Particular precautions for positioning, transportation and mobilisation

Take care when positioning contractures.

Interactions of chronic disease and anaesthesia medications

Not reported.

Anaesthetic procedure

An opioid sparing technique and careful titration of muscle relaxants will limit the impact on post-operative respiratory function.

Judicious use of fluids and a means of external pacing should it be necessary (pads or temporary pacing wire) will reduce the risks of cardiac failure. Electrolyte monitoring and DC cardioversion should be available.

Potentiation of neuromuscular blockade by hypothermia should be avoided.

Particular or additional monitoring

Invasive arterial pressure monitoring is prudent. Central venous pressure monitoring may be of benefit.

Neuromuscular blockade should be monitored routinely.

Possible complications

Decompensation of existing cardiac abnormalities:

- Conduction defects, including complete heart block and atrial standstill
- Cardiac failure
- Ventricular and supraventricular arrhythmias.

Prolonged neuromuscular blockade.

Post-operative care

Patients are at risk of respiratory compromise. High dependency care should be considered particularly following intra-abdominal or thoracic surgery.

Disease-related acute problems and effect on anaesthesia and recovery

Anaesthesia induced rhabdomyolysis may present as a Malignant Hyperthermia type picture. Hyperkalaemia and life-threatening arrhythmias are possible.

Ambulatory anaesthesia

Joint contractures and muscle weakness may affect gait and stability.

Obstetrical anaesthesia

Require early investigation to inform decision making.

Elective Caesarean Section may be necessary, and the feasibility and impact of central neuro-axial blocks need to be assessed.

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