

DUCHENNE PARENT PROJECT EMERGENCY CARD

I'm affected by Duchenne Muscular Dystrophy (DMD), that includes the following COMPLICATIONS:

RESPIRATORY COMPLICATIONS (they usually appear after loss of ambulation):

- ✓ **restrictive ventilatory defect**
- ✓ **chronic hypercapnia** caused by hypoventilation first just at night and then also during the day
- ✓ **cough weakness**
- ✓ **obstructive sleep apnea** (it may appear before loss of ambulation, mainly in patients suffering from obesity or adeno-tonsillar hypertrophy)
- ✓ **acute respiratory failure** (the most frequent causes are: 1) upper respiratory tract infection which may lead to secretion retention and mucus plugging, 2) atelectasis, 3) pneumonia, 4) pulmonary embolism, 5) drugs that further reduce muscle strength such as benzodiazepines, 6) pneumothorax, 7) fat embolism in the presence of long-bone or vertebral fractures, 8) cardiogenic pulmonary edema in the presence of left ventricular dysfunction)

HEART DISEASE (they usually appear after 10 years of age): they can cause **heart failure** or **hypotension** through one of the following mechanisms:

- ✓ **dilated cardiomyopathy**
- ✓ **arrhythmias**
- ✓ **atrio-ventricular block**

COMPLICATIONS RELATED TO CHRONIC CORTICOSTEROID THERAPY:

- ✓ severe hypotension and/or loss of consciousness due to **acute adrenal insufficiency** as the patient may have little to no ability to produce cortisol in response to stress (risk factors for adrenal crisis include physical stress such as infection, trauma, or surgery),
- ✓ **gastrointestinal bleeding** due to peptic ulcer disease

SCOLIOSIS (more likely after loss of ambulation) increases risk of chronic respiratory failure

SWALLOWING PROBLEMS (more likely in adulthood) may cause:

- ✓ **inhalation pneumonia**
- ✓ **coughing or choking when eating or drinking**

OSTEOPOROSIS (more likely after loss of walking and in patients treated with corticosteroid) increases the risk of **vertebral and femur fractures**

MANAGEMENT IN EMERGENCY DEPARTMENT: ALERTS FOR PHYSICIAN

In case of ACUTE RESPIRATORY FAILURE (at high risk after loss of ambulation):

- ✓ Consider hypoventilation and secretions retention in the presence of hypercapnia or hypoxemia (i.e., SpO₂ <95% in room air)
- ✓ Treat with non-invasive ventilation (NIV) and frequent application of a cough assistance device (or manual assisted coughing if device is unavailable); use the patient's home equipment when available; if the patient is unconscious or has a severe swallowing impairment, consider elective tracheal intubation
- ✓ O₂ must be never used without associating it with NIV; if supplemental oxygen is required titrate oxygen therapy to achieve SpO₂ 94-98% and monitor PCO₂ and pH performing serial arterial blood gas analysis.
- ✓ Low threshold for use of antibiotics is recommended for chest infections
- ✓ In case of NIV failure intubation and invasive mechanical ventilation are indicated in the presence of an acute reversible event unless there is an advance directive stating otherwise
- ✓ Difficult intubation should be always considered; in this case intubation guidelines for difficult airway management should be taken into account
- ✓ Perform a chest x-ray as soon as possible; if no infectious cause of respiratory crisis is evident, consider non-infectious causes (atelectasis, pneumothorax, pulmonary embolism, fat embolism, cardiogenic pulmonary edema); if chest x-ray is not conclusive, perform a computerized tomography pulmonary angiography to rule out pulmonary embolism and anterior pneumothorax)
- ✓ After recovery from the acute illness these patients should be promptly extubated to NIV combined with assisted coughing
- ✓ Indications for a tracheotomy can be evaluated, but it should not be considered in the acute phase, rather only in the case of multiple failures of weaning protocol which must include the use of NIV and cough assistance device

In case of ACUTE CARDIAC COMPLICATIONS:

- ✓ Obtain an electrocardiogram and a portable chest radiograph; remember that in these patients Q wave may be caused by chronic fibrosis related to dystrophic myocardial involvement
- ✓ Obtain an echocardiogram and early consultation with a cardiologist

- ✓ Measure blood levels of B-type natriuretic peptide
- ✓ Treat congestive heart failure and arrhythmias
- ✓ NIV may be indicated in case of pulmonary edema
- ✓ Patients with higher degree of AV blocks may need a cardiac pacemaker
- ✓ Ventricular assist device (VAD) may be considered in patients with advanced heart failure

In case of FRACTURES:

- ✓ If the patient is ambulant before fracture, internal fixation is preferable to casting as it helps to preserve muscle and speeds a return to walking
- ✓ In case of long-bone or vertebral fractures consider fat embolism syndrome if patient has dyspnoea or altered mental status

If the patient takes chronic corticosteroid therapy PREVENT ADRENAL INSUFFICIENCY in case of physical stress such as infection, trauma, or surgery

- ✓ Supplemental steroid doses may be necessary. Appropriate stress doses are based on degree of medical or surgery stress
- ✓ For major stress administer 100 mg hydrocortisone by slow intravenous injection or intramuscular followed by 50 mg every 8 hours for 24 hours; in less critical situations consult the PJ Nicholoff Steroid protocol or obtain early consultation with an endocrinologist

ANAESTHETIC PRECAUTIONS in patients undergoing surgery

- ✓ Obtain a pre-operative evaluation including lung function tests, sleep study and cough assessment. If respiratory muscle weakness is present (i.e. forced vital capacity less than 50% of predicted value or peak cough less than 270 l/min), familiarization with ventilatory support is warranted prior to procedure in case it is required post-operatively.
- ✓ Patients should also undergo careful assessment of heart function as well as optimization of cardiac therapies in the pre-operative period.
- ✓ Rule out swallowing and feeding difficulties.
- ✓ Determine if stress steroid dosing is necessary by consulting the PJ Nicholoff Steroid protocol (e.g., for minor procedures or surgery under local anesthesia take usual morning steroid dose and no extra supplementation is necessary; on the other hand, for major surgical stress give 100mg of i.v. hydrocortisone before induction of anesthesia + 50 mg every 8 hours for 24 hours).
- ✓ The preoperative evaluation should also include the assessment for a difficult airway management.
- ✓ The use of regional or local anesthesia offers a significant advantage in term of avoidance of anesthetic drugs and reduction of postoperative respiratory complications, mainly in patients with reduced respiratory function. As a consequence, it should be adopted whenever possible.
- ✓ Inhaled anaesthetics and succinylcholine should be avoided to prevent rhabdomyolysis. Thus, Total Intra-Venous Anesthesia (TIVA) must be used if general anesthesia is required. Ultrashort acting anesthetics (e.g., propofol associated with remifentanyl) should be preferred.
- ✓ These patients may experience increased sensitivity to sedatives and neuromuscular blockade. As a consequence, the depth of anesthesia and the neuromuscular function should be monitored in order to titrate the appropriate dose of those drugs. In addition, the effect of muscle relaxant should be completely reversed at the end of surgery (e.g., rocuronium must be used and must be reversed by sugammadex).
- ✓ Post-operative high dose morphine infusions should be avoided.
- ✓ Post-operative admission to an Intensive Care Unit should be considered in any patient who receive high dose morphine infusions or who is at risk for respiratory or cardiac complications.
- ✓ Patients with decreased respiratory muscle strength require close monitoring and aggressive respiratory management including early extubation to NIV with aggressive use of cough assistance device. O2 must be never used without associating it with NIV

In case of **CHOKING** use a cough assistance device (or manual assisted coughing if device is unavailable); if it is ineffective consider emergent tracheal intubation. In case of severe swallowing impairment elective gastrostomy may be considered.

In case of **LOSS OF CONSCIOUSNESS** rule out: i) carbon dioxide narcosis (in the presence of severe respiratory impairment); ii) heart failure (in presence of heart disease); iii) fat embolism (in the presence of long-bone or vertebral fractures); iv) embolic ischemic stroke (in the presence of severe heart failure); v) adrenal insufficiency (in the presence of chronic corticosteroid therapy).

REFERENCES

- ✓ Hull J, Aniapravan R, Chan E, et al. British Thoracic Society guideline for respiratory management of children with neuromuscular weakness. *Thorax* 2012;67(Suppl. 1):i1–40
- ✓ Birnkrant DJ, Katharine Bushby H, Carla M Bann CM, et al for the DMD Care Considerations Working Group. Diagnosis and management of Duchenne muscular dystrophy, part 3: primary care, emergency management, psychosocial care, and transitions of care across the lifespan. *Lancet Neurol.* 2018 May;17(5):445-455.
- ✓ Racca F, Del Sorbo L, Mongini T, et al. Respiratory management of acute respiratory failure in neuromuscular diseases *Minerva Anestesiol* 2010; 76: 51-62
- ✓ Racca F, Mongini T, Wolfler A, et al. Recommendations for anesthesia and perioperative management of patients with neuromuscular disorders. *Minerva Anestesiol* 2013;79(4):419–33
- ✓ Kinnett K, Noritz G. The PJ Nicholoff Steroid Protocol for Duchenne and Becker Muscular Dystrophy and Adrenal Suppression. *PLoS Curr.* 2017; doi:10.1371/currents.md.d18deef7dac96ed135e0dc8739917b6e