PHYSIOTHERAPY MANAGEMENT IN PATIENTS WITH NEUROMUSCULAR DISEASE AND RESPIRATORY PROBLEMS

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AIMS

• REMINDER OF NMS PROBLEMS: RESPIRATORY
• COUGH ASSESSMENT
• PRE-REFERRAL TO VENTILATION CENTRE WORK UP
• INEFFECTIVE COUGH MANAGEMENT
• OPTIMISING NIV
NEUROMUSCULAR CONDITIONS (NMS)

- RESPIRATORY MUSCLE WEAKNESS
- INABILITY TO COUGH EFFECTIVELY
- IMMOBILE
- KYPHOSCOLIOSIS
- FIXED CHEST WALL
- OSA
- MACROGLOSSIA
EVOLUTION OF RESPIRATORY MUSCLE WEAKNESS

Normal breathing

REM related sleep disordered breathing

Non-REM and REM sleep disordered breathing

Daytime ventilatory failure

Chest Infection

FVC <40% predicted

FVC < 20% predicted

PaCO₂ >6kPa

survival is 9-10 months in DMD

Death

predicted
COUGH ASSESSMENT

• TO MAINTAIN AIRWAY CLEARANCE NEUROMUSCULAR CONDITIONS INDIVIDUALS MUST HAVE AN EFFECTIVE COUGH, THIS REQUIRES A HIGH VOLUME AND FLOW OF AIR BUT THESE PATIENTS OFTEN HAVE AN INEFFECTIVE AND WEAK COUGH.

CAUSES:
• WEAK INSPIRATORY/EXPIRATORY MUSCLES
• SMALL LUNG VOLUMES
• STIFF AND INELASTIC CHEST WALL
• DIFFICULTY COORDINATING THE CLOSURE OF THE GLOTTIS
COUGH MECHANICS

1) INSPIRATORY PHASE: LARGE BREATH/LATER EXPELLED
2) GLOTTIC PHASE: 2 FLAPS OF MUSCLE IN LARYNX CLOSE BLOCKING THE EXIT FOR PREVIOUSLY INSPIRED AIR
3) CONTRACTION OF EXPIRATORY MUSCLES
4) INCREASED INTRA-THORACIC PRESSURE/INCREASED PRESSURE IN SUB-GLOTTIS, TRYING TO BREATHE OUT AGAINST CLOSED GLOTTIS.
5) GLOTTIS OPENS
6) AIR IS EXPELLED
7) SECRETIONS PROPELLED TO CENTRAL AIRWAY
8) EXPECTORATION
FEW POINTS

• NORMAL COUGH EXPELS A VOLUME OF AIR X 4 NORMAL TIDAL VOLUME (THUS NIV ALONE IS NOT THE ANSWER TO IMPROVE COUGH)

• EFFECTIVE ELIMINATION OF SPUTUM ESSENTIAL FOR THE SUCCESS OF CHRONIC AND ACUTE VENTILATORY SUPPORT.

• IN PATIENTS WITH VENTILATORY IMPAIRMENT: 90% OF ACUTE RESPIRATORY FAILURE CAUSED BY SPUTUM RETENTION DURING INFECTIONS.
CONSEQUENCES

- POOR AIRWAY CLEARANCE
- ATELECTASIS/COLLAPSE
- CHOKING/SPUTUM RETENTION/PLUGGING OFF
- LRTI/PNEUMONIA
- RESPIRATORY FAILURE
- RESPIRATORY ARREST
SPUTUM

• PROTECTION DUST/MICRO-ORGANISMS
• CONTINUOUS PRODUCTION/CONVEYER BELT
• GOBLET CELLS: 10-100ML’S/DAY
• GEL/SOL LAYERS: INFLUENCED BY DISEASE/INFECTION: MADE OF PROTEIN & LIPID
• TRANSPORT: CILIA. 200/CELL: CLAWS SLING SHOT MANOEUVRE
• INFECTION, SPUTUM CONTENT, CILIA DYSFUNCTION MEANS COUGH IS ESSENTIAL!
PRE-REFERRAL WISH LIST

- Positioning Plan
- Stretching Program including Mobilisation of Thorax
- Quick Access to GP/Rescue Antibiotics/LOW Threshold for Hospital Admission (THIS IS MES RECORD?/ON-CALL PHYSIO ORGANISED)
- Regular Monitoring of Sputum Growth
- Mucolytic Consideration
- Good Mouth Care Plan
- Respiratory Physiotherapy Plan: Including Assessment and Facilitation of Cough
- Oral Suction Device
- Peak Cough Flow Measure Weekly Record
- Method for Managing Poor Cough Mechanics: MAC and/OR LVR and MiE
POSITIONING

• REGULAR TURNING PROGRAMME (FREQUENT POSITIONING CHANGES)
• BALANCE BETWEEN PREVENTING/POSITIONING CONTRACTURES AND OPTIMISING RESPIRATORY FUNCTION/VENTILATION/PERFUSION
• MAINTAINING THORAX: STRETCHING/MOBILISATION OF SPINE AND RIBS
• STANDING PROGRAM
• CONSIDERATION: EATING/DRINKING/AT RISK OF POOLING SALIVA
GOOD ORAL HYGIENE/MOUTH CARE

• GOOD ORAL CARE BY A PROFESSIONAL CAN REDUCE THE RISK OF ASPIRATION PNEUMONIA CAUSED BY MICRO-ASPIRATION

• CULTURE TESTS OF BACTERIA OBTAINED FROM PATIENTS WHO CONTRACTED ASPIRATION PNEUMONIA REVEALED THAT ORAL RESIDENT BACTERIA WERE MOST COMMON.

• CLEAN TONGUE/SOFT PALATE (REMOVE ANAEROBIC BACTERIA) (ELECTRIC TOOTHBRUSH AND WATER FLOSSERS CAN ASSIST)

• ENSURE ORAL MOISTURE RETENTION IF PEG/RIG FED

• ENSURE DENTAL CARIES FILLED (MORE COMPLICATED IF 24 HOUR VENTILATED BUT VERY ACHIEVABLE)
EXPIRATORY MUSCLE WEAKNESS

• NEUROMUSCULAR-INDUCED EXPIRATORY-MUSCLE WEAKNESS CAN HAVE A GREATER EFFECT ON COUGH EFFICIENCY.

• PATIENTS WITH MILD-TO-MODERATE EXPIRATORY-MUSCLE WEAKNESS CAN EXPERIENCE LIMITATION IN EXPIRATORY PRESSURE.

• THE INABILITY TO GENERATE ADEQUATE EXPIRATORY PRESSURE (REGARDLESS OF HOW MUCH GAS VOLUME IS INHALED) RESULTS IN LOWER DYNAMIC AIRWAY PRESSURE AND LOWER EXPIRATORY FLOW VELOCITY.

• PATIENTS WITH DUCHENNE MUSCULAR DYSTROPHY MAY HAVE SUFFICIENT VENTILATION AND YET BE AT RISK FOR PULMONARY CONGESTION AND ACUTE RESPIRATORY FAILURE ASSOCIATED WITH WEAK COUGH.
WHICH PATIENTS ARE AT RISK OF CHEST INFECTIONS?

POTENTIALLY ALL OF THEM

- POOR VOICE QUALITY: ARTICULATION/VOLUME
- FREQUENT INFECTIONS
- OBVIOUS WEAK COUGH
- PATIENTS C/O CHOKING ON SPUTUM
- REDUCED PEAK COUGH FLOW MEASURE
- REDUCED LUNG FUNCTION
PEAK COUGH FLOW

- Maximum flow recorded immediately following opening of the glottis
- Rapidexpulsive phase of cough
- Effective airway clearance needs PCF in excess of 360L/min
- Vital capacity above 1.5L essential component for cough generation
- Expiratory muscle strength to generate high thoraco-abdominal pressures

(Crude outcome measure but is useful feedback for clinician and patient)
CRITICAL LEVELS OF PEAK COUGH FLOW

• >360L/MIN OK
• <270L/MIN INTRODUCE STRATEGIES FOR ASSISTED AIRWAY CLEARANCE
• <160L/MIN ADDITIONAL ASSISTED AIRWAY CLEARANCE STRATEGIES MUST BE USED (ACPRC)

NB. 90% OF EPISODES OF RESPIRATORY FAILURE DEVELOP BECAUSE OF INABILITY TO CLEAR THE AIRWAYS IN NMD

THE GREATER THE PCF THE LESS RISK OF RESPIRATORY COMPLICATIONS (KANG AND BACH 2000)
EFFECT OF A COLD ON RESPIRATORY MUSCLE STRENGTH

• POPONICK ET AL AJCCM, 1997
• PHYSIOLOGICAL CHANGE IN RESPIRATORY MUSCLES DURING INFECTION CAUSES RESPIRATORY MUSCLE WEAKNESS, LASTS FOR 21 DAYS!
• IF PCF DOES NOT EXCEED 270-300L/MIN WHEN WELL THEN PATIENTS ARE AT RISK OF HAVING A PCF<160L/MIN WHEN THEY ARE UNWELL. BACH ET AL., 1997 CHEST
TREATMENT

IS THERE ANY POINT IN TRADITIONAL RESPIRATORY PHYSIOTHERAPY?

ACBT, MANUAL TECHNIQUES, POSTURAL DRAINAGE, AD, THE VEST, FLUTTER & PEP.

THESE TECHNIQUES MAY BE OF SOME BENEFIT EARLY STAGES PRIOR TO LOSS OF VC

BUT
I LOVE A GOOD ACTIVE CYCLE BREATHING BUT YOU NEED TO FACILITATE COUGH!

• UNASSISTED BREATH STACKING AND MANUALLY ASSISTED COUGH (MAC)
• BREATH STACKING: BAG OR VENTILATOR MI:E (MECHANICAL INSUFFLATION-EXSUFFLATION)
• BREATH STACK AND MAC
• BREATH STACK AND MI:E
BREATH STACKING

• THE PATIENT STACKS AIR BY CONSECUTIVELY DELIVERING VOLUMES VIA A VOLUME VENTILATOR OR AMBU BAG (GLOSSOPHARYNGEAL CAN AIR STACK WITHOUT VENT/BAG)

• WITHOUT EXHALING THE PATIENT BUILDS ON EACH BREATH UNTIL LUNGS ARE MAXIMALLY EXPANDED

• MAIN EVIDENCE IN NMS
BREATH STACKING

- UNASSISTED BREATH STACKING
- LUNG VOLUME RECRUITMENT (LVR) BAG
- VENTILATOR
BREATH STACKING

• INCREASE COUGH PEAK FLOW (BACH 2003)
• MAINTAIN MAX INSUFFLATION CAPACITY
• MAINTAINS CHEST WALL MOVEMENT/DECREASES CONTRACTURES (KANG, 2000)
• MOBILISE SECRETIONS
• IMPROVES TV/VC/TLC (CONCALVES, 2006)
LVR BAG

• A LVR bag is used to deliver volumes of air that the glottis holds to volumes greater than VC until the maximum insufflation capacity (MIC) is reached. The intrathoracic pressures are thereby elevated.

• The breaths are stacked one after the other as the LVR contains a one-way valve preventing the individual breathing out through the bag.
PATIENT INDICATIONS

• LOW VITAL CAPACITY (VC)
• POOR PEAK COUGH FLOW (PCF)
• INEFFECTIVE COUGH AND INABILITY TO CLEAR SECRETIONS
• PATIENT COMPETENT AND ABLE TO CONSENT (MAY BE WORTH TRIALLING IF PATIENT DOES NOT HAVE CAPACITY AS SOME PATIENTS STILL BENEFIT)
• INCREASE LUNG VOLUME
• IMPROVE COUGH EFFECTIVENESS
• DECREASE ATELECTASIS
• INCREASE MECHANICAL COMPLIANCE
• OPTIMISE THORACIC RANGE OF MOTION
• INCREASE SPEAKING VOLUME
• MAY IMPROVE TOILETING AND BOWEL CONTROL

Improve quality of life
MANUAL ASSISTED COUGH?
BTS/ACPRC GUIDELINES

• MANUAL ASSISTED COUGH (MAC) SHOULD BE USED TO INCREASE PCF IN PATIENTS WITH NMS DISEASE.

• COMBINE MAC WITH A MAXIMUM INSUFFLATION STRATEGY: LVR/MI: E

• ABDOMINAL THRUSTS SHOULD BE UNDERTAKEN IN FRONT OF THE PATIENT (WHERE POSSIBLE) TO ASSIST COMMUNICATION

• IF COUGHING IS INADEQUATE WITH JUST MI: E COMBINE WITH MAC
CONTRAINDICATIONS TO MAC

• HAEMOPTYSIS (STOP IF EXPERIENCING CHEST PAIN)
• PNEUMOTHORAX
• POST SURGERY TO FACE, NOSE, MOUTH, LUNGS, STOMACH, OESOPHAGUS
• BULLOUS DISEASE (CAUTIOUS IN COPD WITH KNOWN EMPHYSEMATOUS LUNGS)
• RAISED INTER-CRANIAL PRESSURE
• IMPAIRED CONSCIOUSNESS/INABILITY TO COMMUNICATE
• INFLATED TRACHEOSTOMY CUFF (UN CUFFED/CUFF DEFLATED ONLY)
• CAUTIOUS NAUSEA/VOMITING OR FATIGUED
• CAUTIOUS IF HAVING ENTERAL FEEDING (TURN OFF FEED)
MECHANICAL INSUFFLATOR: EXSUFFLATOR (MI:E)
SYMPTOMS TO MONITOR FOR WORSENING RESPIRATORY FUNCTION

- More frequent or pronounced sob on exertion
- Occasional mild sob while at rest
- Sob while lying on your back or waking with sob
- Difficulty sleeping or sleeping more often
- Waking with a headache that goes away after being awake for awhile or periodic/constant headache
- Dizziness during or after exertion
- Ringing in ears
- Constant fatigue or new muscle twitching
- Occasional confusion
- Difficulty concentrating
OPTIMISING VENTILATION IN PATIENTS WITH NMS DISEASE

PARALLEL REVIEW OF:

• SYMPTOMS AND SUBJECTIVE FEEDBACK

• PULSE OXIMETRY, TRANSCUTANEOUS CO₂, BLOOD GASSES

• VENTILATOR SD CARD DOWNLOAD: IDENTIFY CAUSES OF HYPOVENTILATION

• COMPLETE CARE OF THE PATIENT WITH NMS DISEASE REQUIRES MEASURES TO ASSIST IN LUNG VOLUME RECRUITMENT AND SECRETION CLEARANCE.
OPTIMISING VENTILATION IN PATIENTS WITH NMS DISEASE

IMPROVEMENTS IN ALVEOLAR VENTILATION CAN BE IMPROVED BY:

• 10ML/KG RATIO BUT INCREASED PS NOT WELL TOLERATED IN RESTRICTIVE DISORDERS
• MINIMISING EPAP TO ALLOW MAXIMUM PS IF NO EVIDENCE OF OBSTRUCTIVE SLEEP APNOEA
• CHANGING TO PRESSURE CONTROL MODE BY CONTROLLING INSPIRATION TIME: DIFFERENT VENTILATORS ACCOMPLISH THIS IN DIFFERENT WAYS, BUT OVERALL THE CHANGE PREVENTS PREMATURE CYCLING (COMMON IN RESTRICTIVE DISORDERS)
• SHORTENING THE RISE TIME IF TOLERATED BY PATIENTS CAN INCREASE THE TIME AT MAXIMUM INSPIRATORY PRESSURE WITHIN A SET INSPIRATORY TIME, WHICH CAN INCREASE TIDAL VOLUME
• REDUCING THE BACK UP/SET RESPIRATORY RATE MAY BE NEEDED TO MAINTAIN A LONGER INSPIRATORY TIME AND INSPIRATORY : EXPIRATORY OF 1:1
• % OF PATIENT-_TRIGGERED BREATHS CAN BE FOLLOWED AS AN INDIRECT MEASURE OF NOCTURNAL MUSCLE REST
KEY LEARNING POINT

- In patients with ventilatory failure NIV is very effective.
- However, in the majority of cases, secretions are excessive and NIV alone is likely to fail.
- If we can manage sputum effectively it can improve both quality of life and life expectancy.
- The earlier ineffective cough is assessed and managed the more successful the patient outcome.
QUESTIONS
REFERENCES

NICE 2010 MND GUIDELINES/ NICE 2016 MND GUIDELINES


SNIFF NASAL INSPIRATORY PRESSURE: SIMPLE OR TOO SIMPLE?

J-W. FITTING ERJ MAY 1, 2006 VOL. 27 NO. 5 881-883


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