

PERIOPERATIVE MANAGEMENT OF CHILDREN WITH NEUROMUSCULAR DISORDERS: PROSPECTIVE STUDY

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Keywords: general anesthesia, sedation, neuromuscular diseases, postoperative respiratory complications, non-invasive ventilation, mechanical insufflation-exsufflation

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Running head: Perioperative management in neuromuscular patient

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Running head: Perioperative management in neuromuscular patients

Table 1. Demographic data and pre-existing dependence on technology

Patients (n=167)	
Age (years)	8 ±6
Weight (kg)	26 ±18
Height (cm)	117 ±34
Gender (male)	102 (61%)
BMI (kg/m ²)	17 ±6
Pre-existing dependence on technology	85 (51%)
Mechanical ventilation	57
<i>IMV</i>	8
<i>NIV</i>	43
<i>N-CPAP</i>	6
Cough assistance therapy	70
<i>MI-E</i>	62
<i>MANUAL ASSISTANCE</i>	8
Gastrostomy / Jejunostomy	45
Pace-Maker	1

ASA, American Society of Anesthesiology; IMV, invasive mechanical ventilation; NIV, noninvasive ventilation; n-CPAP, noninvasive continuous positive airway pressure; MI-E, mechanical insufflation–exsufflation

Table 2. Preanesthetic assessment and management

	Patients (n=167)
History of weak cough or prolonged respiratory illnesses	80 (48%)
Pulse-oximetry test (performed/not performed)	155 / 12
SpO ₂ at < 95% in room air	16 (10%)
Peak cough flow measurement (performed/not performed)	19 / 148
Peak cough flow < 270 L/minute	12 (63%)
Spirometry (performed/not performed)	36 / 131
FVC < 50% of that predicted	22 (61%)
FVC < 30% of that predicted	10 (28%)
Carbon dioxide level assessment (performed/not performed)	84 / 83
hypercapnia (PaCO ₂ ≥ 50)	23 (27%)
Sleep respiratory studies (performed/not performed)	77/ 90
altered sleep respiratory study	13 (17%)
Preoperative training in NIV	28
Retraining in NIV	16
Preoperative training in cough assistance	39
Retraining in cough assistance	30
Patients affected by scoliosis	100
Cobb Angle	
10°- 50°	58
50-90°	37
> 90°	5
Gilardeau dysphagia score	
0	58
1	59
2	8
3	6
4	36
Echocardiogram (performed/not performed)	88 / 79

Ejection fraction < 35%	2 (1%)
Preadesthetic location	
Outpatient – Day Surgery Status	16
Inpatient Ward	104
High Dependency Unit /ICU	6/41
Documented DNR order/ treatment limitation	3

FVC, forced vital capacity; MV, mechanical ventilation; NIV, non-invasive ventilation; ICU, intensive care unit; DNR, do not resuscitate

Gilardeau dysphagia score: 0 = able to eat normal diet / no dysphagia; 1 = able to swallow some solid foods; 2 = able to swallow only semi solid foods; 3 = able to swallow liquids only; 4 = unable to swallow anything/total dysphagia

Table 3. Anesthetic technique and airway management

Patients (167 cases)		
PREMEDICATION DRUGS	28 (16.8%)	
Midazolam	26	
Ketamine	2	
REGIONAL ANESTHESIA	14 (8.4%)	
Spinal anesthesia	2	
Peripheral nerve blocks	12	
GENERAL ANESTHESIA	136 (81.4%)	
Halogenated agents	31	
TIVA	105	
SEDATION	43 (25.7%)	
DRUGS (not exclusive)	General anesthesia	Sedation
Propofol	96	28
Sevoflurane	31	-
Thiopentone	7	1
Midazolam	16	23
Dexmedetomidine	11	1
Ketamine	9	13
Fentanyl	85	11
Remifentanil	46	-
Nitrous Oxide	21	-
Rocuronium	46	-
Cis-Atracurium	8	-
Succinylcholine	1	-
Sugammadex	8	-
AIRWAY MANAGEMENT		
Mask only	27	
LMA only	24	
Tracheostomy tube	8	
Standard ETI with a conventional laryngoscope	61	
Bronchoscopic intubation	14	

Videolaryngoscopy intubation	6
Frova catheter use during standard ETI	1

TIVA, total intravenous anesthesia; NIV, non-invasive ventilation; LMA, laryngeal mask airway; ETI, endotracheal intubation.

Table 4. Postanesthetic care and outcomes

	Patients (167 cases)
Disposition following anesthesia	
Home discharge (phone follow-up)	7
Hospital ward	64
Intensive care unit	100
Use of MV and MI-E after surgery	
NIV	60
MI-E	81
NIV and MI-E	49
Invasive ventilation	53
Postoperative analgesia (not exclusive)	
Epidural analgesia	6
Acetaminophen or NSAIDs	143
Opioids	64
Postoperative complications	27 (16.2%)
Surgical complications*	3
Cardio-vascular complications	1
Pulmonary complications	14
<i>Atelectasis</i>	3
<i>Secretion retention</i>	4
<i>Respiratory failure</i>	9
<i>Prolonged intubation</i>	7
<i>Re-intubation</i>	5
Hypothermia	5
Gastrointestinal dysmotility	4
Deaths in the first week after surgery	0 (0%)
Deaths during hospitalization	3 (1.2%)
ICU LOS after surgery (days), mean (SD)	8 (\pm14)
Hospital LOS after surgery (days), mean (SD)	12 (\pm17)

MV, mechanical ventilation; MI-E, mechanical insufflation–exsufflation; NIV, non-invasive ventilation; NSAIDs, Nonsteroidal anti-inflammatory drugs; ICU, intensive care unit; LOS, length of stay. * Surgical complications were dehiscence of surgical suture in two cases and surgical site infection in one case.

Table 5. Predictors of hospital length of stay

VARIABLES	IRR	95% CI	P-VALUE
PREOPERATIVE			
✓ Gender (F vs M)	0.89	0.65-1.22	p = 0.463
✓ Age	0.98	0.95-1.01	p = 0.127
✓ Cough assistance	1.10	0.78-1.59	p = 0.596
✓ Long-term MV	1.65	1.19-2.31	p < 0.001
✓ Cobb angle > 50°	1.90	1.26-2.91	p < 0.001
✓ Dysphagia	1.91	1.29-2.85	p < 0.001
INTRAOPERATIVE			
✓ Major spine surgery	3.03	1.88-4.92	P < 0.001
✓ Urgent surgery	3.20	1.93-5.59	P < 0.001
✓ General anesthesia	1.94	1.93-5.59	P < 0.001
POSTOPERATIVE			
✓ Postoperative cough assistance	1.33	0.98-1.8	P = 0.049
✓ Postoperative admission			
• day surgery unit	1.40	0.42-4.9	P = 0.589
• ward	4.82	1.84-14.1	P = 0.002
• ICU	9.64	3.2-29.9	P<0.001
✓ Duration of IMV			
• IMV < 24 h	0.75	0.51-1.1	P = 0.138
• IMV ≥ 24 and < 168 h	1.95	1.27-3.0	P = 0.002
• IMV ≥ 168 h	3.84	1.89-8.7	P < 0.001
✓ Post-operative complications	1.27	0.28-2.0	P = 0.284
✓ Use of opioids	1.26	0.91-1.8	P = 0.140

Legend: F, female; M, male; ICU, intensive care unit; IMV, invasive mechanical ventilation; IRR, incidence rate ratio; CI, confidence interval; DF, degree of freedom. For all the dichotomous variables except for gender in which the reference category is female, the reference variable is the absence of the variable characteristic. For the variable of the post-operative admission the reference category is day-hospital.



Fig 1. Mechanical insufflation-exsufflation device. It allows for non-invasive airway clearance. This device removes secretions by applying a positive pressure to airway (insufflation), then rapidly shifting to negative pressure (exsufflation). The rapid shift in pressure produces a high expiratory flow of 360- 660 L/min simulating a natural cough.