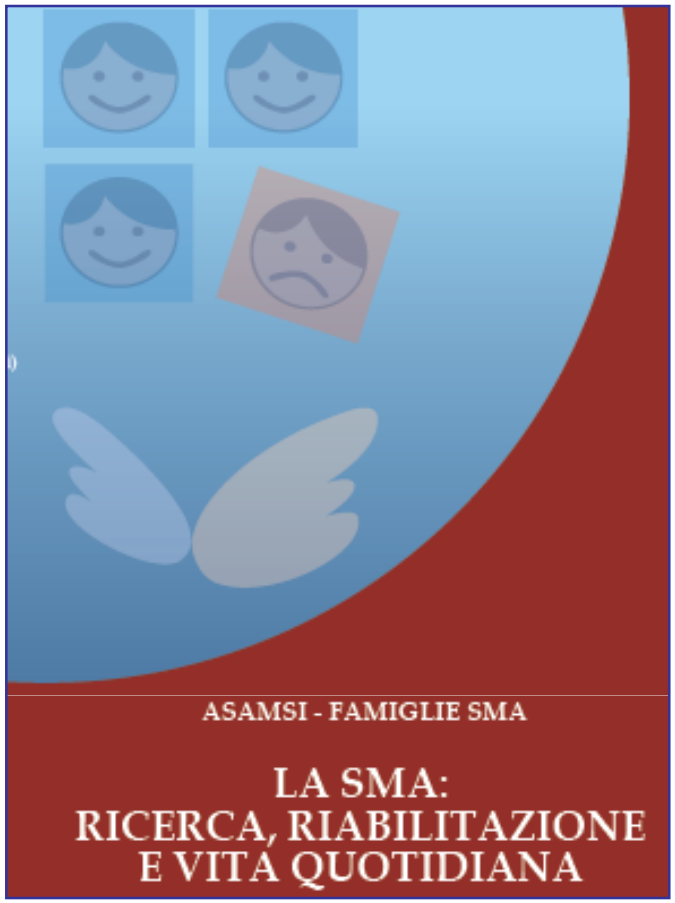
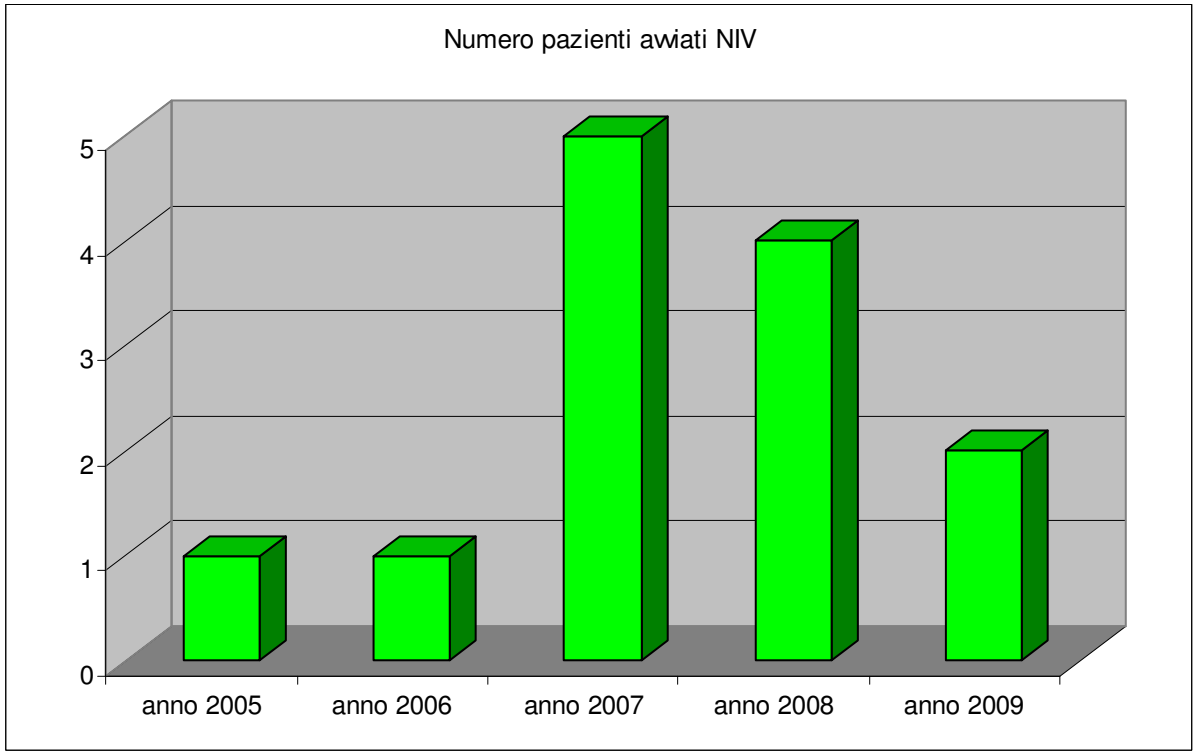


Nuovi percorsi della SMA I in Italia: NIV, PEG e ...

Giancarlo Ottonello
U.O. Anestesia e Rianimazione
IRCCS G Gaslini, Genova

Roma, 5-6 Settembre 2009,
V Convegno Nazionale

Bambini affetti da SMA1 avviati alla NIV



N°	M/F	Età diagnosi	Età inizio NIV	Età inizio cough assist	Età attuale	Intubazioni in Elezione per anestesia	Numero intubazioni in urgenza	Numero di estubazioni	gastrostomia con Nissen/PEG
1	f	3	11	11	42	1	2	2	PEG
2	m	2	7	7	35	1	0	1	Nissen
3	m	3	8	8	55	1	4	4	PEG
4	f	4	7	7	46	1	0	1	PEG
5	f	6	12	12	33	1	0	1	Nissen
6	m	5	9	8	27	1	1	1	Nissen
7	m	0	0,5	0,5	dec	0	1	1	Nulla
8	f	5	5	5	26	1	0	1	Nissen
9	f	4	23	23	54	1	0	1	PEG
10	m	4	7	8	32	1	0	1	Nissen
11	f	5	10	10	25	0	0	0	Nulla
12	m	2	12	8	14	0	0	0	Nulla
13	f	3	15	15	96	1	1	1	PEG
media	6 M 7 F	3,5	9,7	9,4	40,4	0,61	0,69	0,92	5 PEG e 5 Nissen

Consensus Statement for Standard of Care in Spinal Muscular Atrophy

Ching H. Wang, Richard S. Finkel, Enrico S. Bertini, Mary Schroth, Anita Simonds, Brenda Wong, Annie Aloysius, Leslie Morrison, Marion Main, Thomas O. Crawford, Anthony Trela and Participants of the International Conference on SMA Standard of Care

J Child Neurol 2007; 22; 1027
DOI: 10.1177/0883073807305788

3. Management of gastroesophageal reflux (GER):

- Short-term use of **acid neutralizers** (e.g. magnesium or calcium carbonate) and/or **inhibitors of acid secretion** (e.g. histamine blockers and proton pump inhibitors (e.g. famotidine, ranitidine, omeprazole) for symptomatic management. However, prolonged use may be associated with a greater risk for gastroenteritis and pneumonia.
- When delayed gastric emptying or diminished motility is present, **prokinetic agents** (e.g.metaclopramide, erythromycin) may be useful.
- Use of **probiotics** such as acidophilus or lactobacillus to help maintain a healthy gastrointestinal flora, particularly following antibiotic treatment or in the setting of prolonged use of acid inhibitors, is an area deserving further study.
- Laparoscopic **anti-reflux Nissen fundoplication** during g-tube placement may be of value in the SMA patient with medically refractory GER, and in whom the benefit is deemed to outweigh the associated surgical and anesthetic risks.

There are several options for gastrostomy tube placement, including insertion via percutaneous methods with endoscopic guidance, or placement via open or laparoscopic surgical techniques[61] together with an antireflux procedure such as Nissen fundoplication. The open surgical technique is associated with a relatively large upper abdominal incision, increased postsurgical pain, and risk for respiratory complications due to diaphragmatic splinting. A laparoscopic surgical technique provides the best possible setting for immediate or early postoperative extubation.[62] Such procedures are typically performed with general anesthesia, although placement using percutaneous methods with endoscopic guidance is performed in some centers with conscious sedation and local anesthesia. Care should be taken to minimize the amount of fasting preoperatively and to resume full nutritional support as quickly as possible following the procedure. Possible pulmonary complications of sedation should be anticipated and may require treatment with noninvasive ventilation (see "Pulmonary Care").

consensus to support this strategy. However, in the spinal muscular atrophy patient with medically refractory gastroesophageal reflux, and in whom the benefit is deemed to outweigh the associated surgical and anesthetic risks, laparoscopic Nissen fundoplication during gastrostomy tube placement is supported as an appropriate intervention.

La Nissen in Videolaparoscopia

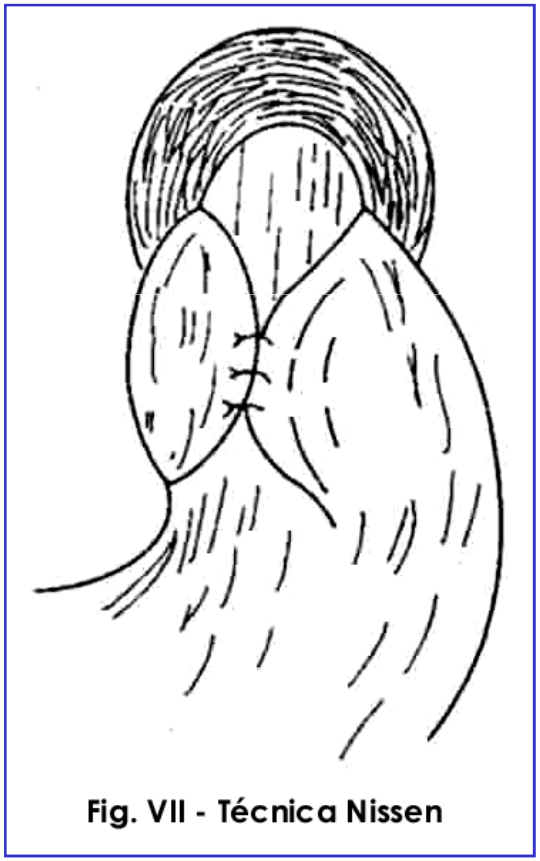


Fig. VII - Técnica Nissen

Gastrointestinal System

- 26/30 sottoposti a valutazione per reflusso gastroesofageo sono stati trattati con fundoplicatio secondo Nissen
- 3/106 pancreatite (gastroparesi (2) acido valproico (1))
- 9: fundoplicatio dopo la gastrostomia

Original Article

Journal of Child Neurology
Volume 22 Number 6
June 2007 727-731

10.1177/0883073807304009
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Laparoscopic Nissen Fundoplication During Gastrostomy Tube Placement and Noninvasive Ventilation May Improve Survival in Type I and Severe Type II Spinal Muscular Atrophy

Nanci Yuan, MD, Ching H. Wang, MD, PhD, Anthony Trela, BS, and Craig T. Albanese, MD

Table 1. Patient Demographics and Diagnosis of Spinal Muscular Atrophy

Case	Gender	Ethnicity	Type of Spinal Muscular Atrophy	Age at Diagnosis, Months
1	Male	Caucasian	I	8
2	Female	Caucasian	I	3
3	Female	Asian	I	5
4	Female	Asian	I	4
5	Male	Asian	II	36
6	Female	Hispanic	I	1.5
7	Male	Caucasian	I	7

Table 2. Patient Demographics: Surgery and Postoperative Results

Case	Age at Surgery, Months	Extubation Time, h	Extubation Type	PICU Stay, Days	Feeds Started, POD	Discharge, POD
1	46	1.5	Room air	1	1	3
2	8	1.0	NPPV	3	1	46
3	6	24	NPPV	2	1	2
4	5	1.0	NPPV	9	1	21
5	69	0.5	NPPV	1	1	5
6	5	1.0	NPPV	7	1	14
7	11	0.5	NPPV	2	2	4

NOTE: PICU, pediatric intensive care unit; POD, post-op day; NPPV, noninvasive positive pressure ventilation.

Table 3. Patient Demographics: Long-Term Survival

Case	Survival in Months Since Surgery	Current Survival Age, Months
1	41	87
2	22	30
3	19	25
4	5[a]	Expired
5	4	73
6	4	9
7	1.5	14

a. Expired.

Table 4. Pneumonias in the Months Presurgery and Postsurgery

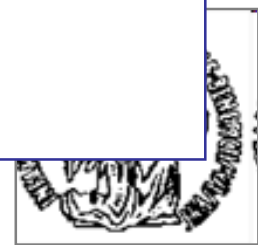
Case	Presurgery 12 Months	Presurgery 3 Months	Presurgery 1 Month	Presurgery Total	Postsurgery 1 Month	Postsurgery 3 Months	Postsurgery 12 Months	Postsurgery Total	Pre-Post[a]
1	2	1	0	3	0	0	0	0	−3
2	NA[b]	1	1	2	0	0	1	1	−2
3[c]	NA[b]	0	1	1	1	1	0	2	+1
4[d]	NA[b]	0	1	1	0	0	Expired	0	−1
5	2	1	2	5	0	0	NA[e]	0	−3
6	NA[b]	1	2	3	0	1	NA[e]	1	−2
7	NA[b]	1	1	2	0	NA[e]	NA[e]	0	−1

a. The difference is calculated during the time available for presurgery and postsurgery comparison.
b. Nonapplicable as patient was less than 12 months of age at time of surgery.
c. This patient has had no pneumonias since 3 months postsurgery.
d. This patient expired from an obstructive apnea at 5 months after surgery.
e. Nonapplicable as patient has not yet reached this postsurgery milestone.

Table 5. Hospitalizations in the Months Presurgery and Postsurgery

Case	Presurgery 12 Months	Presurgery 3 Months	Presurgery 1 Month	Presurgery Total	Postsurgery 1 Month	Postsurgery 3 Months	Postsurgery 12 Months	Postsurgery Total	Pre-Post[a]
1	2	1	0	3	0	0	0	0	−3
2	NA[b]	1	1	2	0	0	1	1	−2
3[c]	NA[b]	0	1	1	1	1	0	2	+1
4[d]	NA[b]	0	2	2	0	0	Expired	0	−2
5	2	1	1	4	0	0	NA[e]	0	−2
6	NA[b]	1	2	3	1	1	NA[e]	2	−1
7	NA[b]	1	1	2	0	NA[e]	NA[e]	0	−1

a. The difference is calculated during the time available for pre- and postsurgery comparison.
b. Nonapplicable as patient was less than 12 months of age at time of surgery.
c. This patient has had no hospitalizations since 3 months postsurgery.
d. This patient expired from an obstructive apnea at 5 months after surgery.
e. Nonapplicable as patient has not yet reached this postsurgery milestone.



Early laparoscopic fundoplication and gastrostomy in infants with spinal muscular atrophy type I

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Received 24 January 2008; revised 5 May 2008; accepted 21 May 2008

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Table 1 Patient demographics

Patient	Sex	Age at diagnosis (mo)	Age at operation (mo)
1	Male	4	11
2	Male	4	7
3	Male	9	14
4	Female	6	12
5	Female	1	5
6	Female	8	14
7	Male	6	9
8	Male	6	8
9	Female	8	10
10	Female	8	12
11	Male	9	13
12	Female	6	8
Mean ± SD	50% male	6.2 ± 2.4	10.1 ± 2.9

Table 2 Perioperative outcomes

Patient	Operative time (min)	Narcotic doses required	Oxygen saturation 4 h (%)	Oxygen saturation 24 h (%)	Time to initiation of feeds (h)	Time to full feeds (h)	Time to first BM (h)	LOS (h)	Complication
1	195	0	100	100	24	45	45	48	–
2	135	0	97	100	21	45	28	70	–
3	165	1	100	99	22	41	42	44	–
4	150	0	99	94	24	44	47	96	–
5	173	0	99	99	24	42	41	120	L lung atalectasis
6	155	0	96	95	22	46	48	60	–
7	161	0	97	98	22	30	54	79	–
8	193	0	95	99	26	42	24	75	–
9	144	0	99	97	21	45	34	74	–
10	274	0	96	100	24	48	28	72	–
11	161	1	100	100	23	45	28	106	–
12	206	0	97	95	20	35	45	74	–
Mean	176 ± 38	<1 dose	98 ± 1.8	98 ± 2.2	23 ± 1.7	42 ± 5.1	39 ± 9.8	77 ± 22	<1

BM indicates bowel movement; LOS, length of stay.

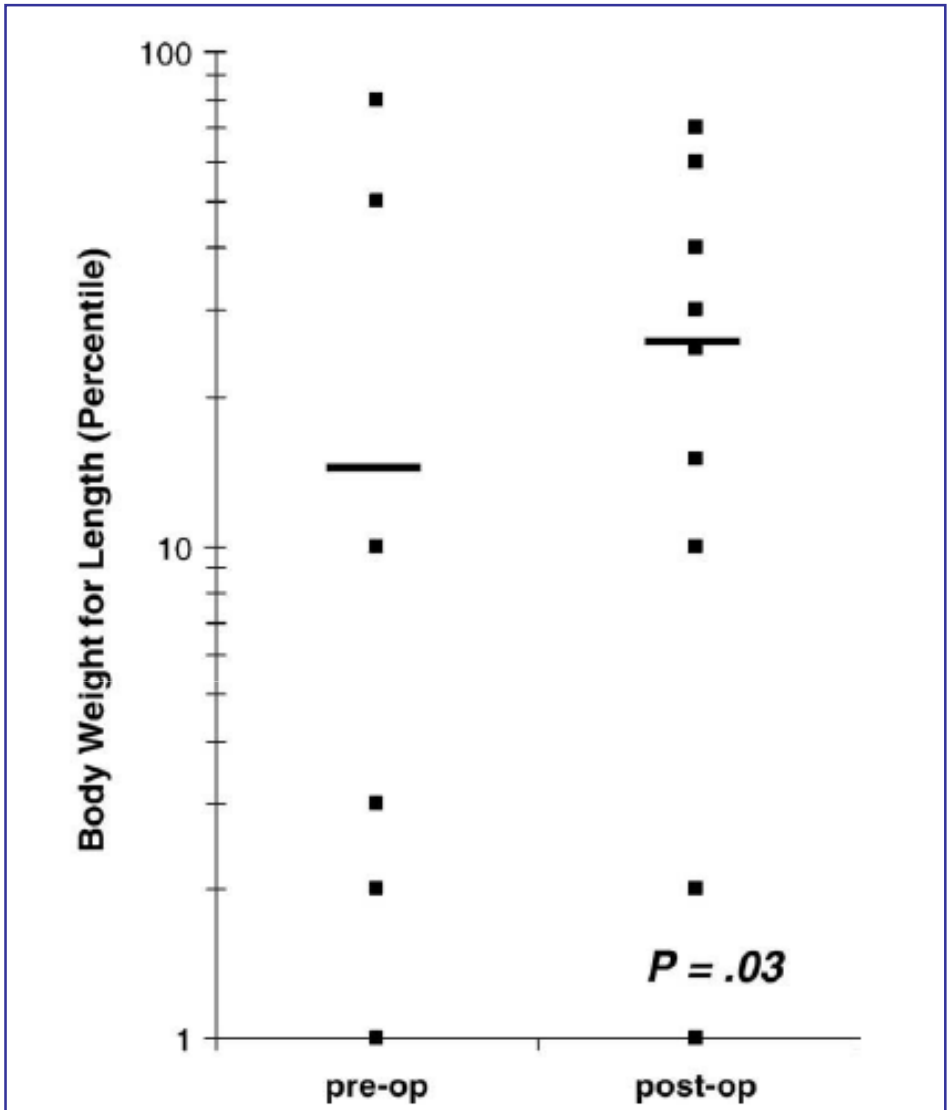


Fig. 1 Body weight-for-length percentiles for children with SMA type I after surgery. Mean weight-for-length percentile is shown for the cohort both pre- and postoperatively. Preoperative growth parameters shown are those recorded for each child at the time of operation. Postoperative growth parameters were assessed at each subsequent visit but are reported here at 12 months postoperatively or the most recent follow-up visit if 12-month data were not yet available.

Table 3 Pre- and postoperative hospitalizations

Patient	Preoperative hospitalizations (12 mo)	Precipitating factor	Postoperative hospitalizations (12 mo)	Precipitating factor
1	0		0	
2	1	RD	2	RD
3	0		0	
4	0		0	
5	2	Dehydration, RD	0	
6	0		0	
7	1	Pneumonia[a, b]	1	RSV bronchiolitis
8	0		1	RD
9	1	Influenza	0	
10	2	RD	2	RD
11	4	RD[a]	0	
12	0		0	
Total	11		6	

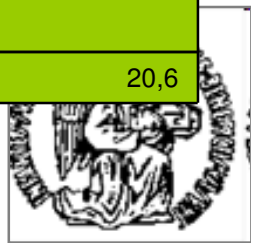
RD indicates respiratory distress; RSV, respiratory syncytial virus.

[a] Mechanical ventilation required at one or more hospitalizations.

[b] Confirmed by chest x-ray.

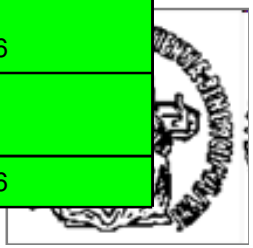
Età all'intervento PEG o Nissen-PEG

sesso	Diagnosi	DATA NASCITA	Gastrostomia Con Nissen/PEG	Età attuale	Età all'intervento PEG	Età all'intervento Nissen
f	SMA1	14/03/2006	PEG	42	11	
m	SMA1	14/10/2006	Nissen	35		19
m	SMA1	06/02/2005	PEG	55	13	
f	SMA1	08/11/2005	PEG	46	25	
f	SMA1	10/01/2007	Nissen	33		26
m	SMA1	15/06/2007	Nissen	27		14
m	SMA1	2008	Nulla	dec		
f	SMA1	03/07/2007	Nissen	26		14
f	SMA1	08/03/2005	PEG	54	28	
m	SMA1	25/01/2007	Nissen	32		30
f	SMA1	14/08/2007	Nulla	25		
m	SMA1	04/07/2008	Nulla	14		
f	SMA1	24/08/2001	PEG	96	17	
					18,8	20,6



Decorso perioperatorio
Nissen PEG

				Età attuale	Età all'intervento		Durata Intervento		gg degenza		alimentazione
m	SMA1	06/02/2005	PEG	55	13		70		10		24/72
f	SMA1	08/11/2005	PEG	46	25		145		6		24/60
f	SMA1	08/03/2005	PEG	54	28		95		4		24/60
f	SMA1	14/03/2006	PEG	42	11		55		20		24/72
f	SMA1	24/08/2001	PEG	96	17						
			-	58,6	18,8		91,25		10		24/66
			-								
f	SMA1	10/01/2007	Nissen	33	26		160		9		24/96
m	SMA1	15/06/2007	Nissen	27	14		145		8		24/96
f	SMA1	03/07/2007	Nissen	26	14		175		6		24/96
m	SMA1	25/01/2007	Nissen	32	30		170		5		24/96
m	SMA1	14/10/2006	Nissen	35	19						
				30,6	20,6		162,5		5,6		24/96



Condizioni cliniche post-operatorie PEG vs Nissen PEG

Evento	PEG	Peg + Nissen
Episodi di urgenze	0	5/5
Problemi risolti con Cough Machine	¼	2/5
Necessità ventilazione 24h/die	0	1/5
Modificazione parametri ventilatore	0	2/5
Modificazione parametri cough machine	0	5/5
Episodi di emergenze	0	2/5
Necessità manovre di rianimazione	0	1/5
Interventi 118	¼	1/5
Ricoveri ospedalieri	2/4	0

Conclusioni

- La ventilazione meccanica non invasiva nei bambini SMA1: una scelta con i genitori
- L'alimentazione: necessità di una procedura chirurgica
- Timing e modalità per efficacia e tollerabilità: la Nissen – PEG in videolaparoscopia.