

Tomasz Potaczek, MD
Daniel Zarzycki, MD, PhD

Natural history and results of surgical treatment of spine deformities in patients with spinal muscular atrophy type 2 and type 3a



**Department of Orthopedic Surgery and Rehabilitation, Jagiellonian University,
Zakopane, Poland**



Introduction

- ☞ **Spinal muscular atrophy (SMA); diseases that manifests in progressive muscle weakness and flaccid paresis**
- ☞ **Pathogenesis - Degeneration of anterior motor cells of the spinal cord**
- ☞ **Incidence - 1/6-10k births**
- ☞ **Genetics - autosomal recessive, gene SMN1 (*survival of motor neuron*) location, long arm of the 5th chromosome (5q11.2-13.3), incidence of carriers: 1/35-50 births**
- ☞ **Types of SMA^{1,2}:**
 - SMA1 (Werdnig – Hoffman disease, acute)
 - **SMA2** (Sub-acute, non-ambulatory)
 - SMA3 (**SMA3a**, start to walk independently, but loose this ability < 3 y.o age; type SMA3b, Kugelberg-Welander disease)

Introduction

☞ **Scoliosis – dominant orthopaedic problem in non-ambulatory patients**

- „collapsing” scoliosis
- prevents unaided sitting
- decreases pulmonary function



☞ **Benefits of surgery**

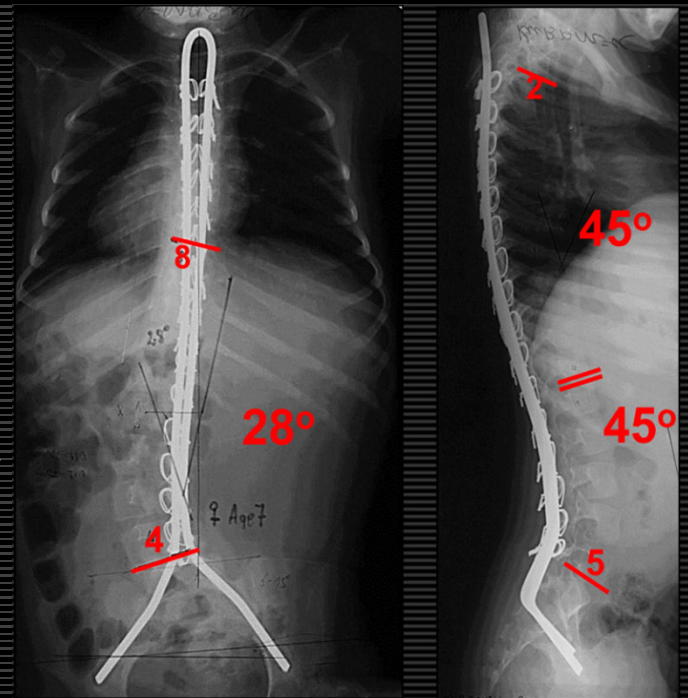
- **Halt progression**
- **Curve correction**
- **Facilitate seating position**
- **Level pelvis**
- **Respiratory status**



Increase quality of life!?!

Aim of paper

- ☞ Evaluate the natural history and results of surgical treatment of spinal deformities in patients with spinal muscular atrophy types SMA2 and SMA3a
 - All patients treated with the same surgical technique
 - Galvestone technique (unit rod or double rod) posterior fusion with Luque wires
- ☞ To establish optimal criteria for surgical treatment depending on
 - age
 - curve magnitude
 - respiratory status



Material

- ➡ **Between 1985 – 2007 173 patients with SMA treated surgically**
 - 139 patients treated surgically due to scoliosis
 - 45 patients with follow-up period ≥ 5 years
 - 17 boys and 28 girls
 - Age at surgery – mean 11.4 years
- ➡ **Mean preoperative follow-up 2 years (1 – 6 years)**
- ➡ **Mean postoperative follow-up was 6.9 years (5 – 15 years)**

Results – natural history of the deformity

	SMA2	SMA3a
No of patients	25	20
Age of SMA diagnosis	1.8 y.o (1 – 3)	2.8 y.o (1 – 8)
Wheelchairbound	1.3 y.o (1 – 3)	5.2 y.o (1 – 10)
Scoliosis onset	4.8 y.o (3 – 8)	7.0 y.o (5 – 10)
<i>Annual curve progression pre-op</i>	<i>11.2°/year</i> (0° - 50°)	<i>14.9°/year</i> (0° - 34°)
<i>Annual AVT progression pre-op</i>	<i>9mm/year</i> (-15-42mm)	<i>15.4mm/year</i> (0-73mm)

Results – perioperative data

	SMA2	SMA3a
Age of surgery	10.5 y.o (5 – 21)	12.6 y.o (8 – 21)
Curve magnitude (°)	101.7° (38-158°)	111.3° (43-150°)
FVC (%)	40.6% (21 – 83)	54.1% (19 – 86)
Time of surgery (hours)	3.5 hours (2.5 – 5.5)	3.8 hours (2 – 7)
Intraoperative blood loss (ml)	620 ml (150 – 2200)	712 ml (150 – 1350)

Results – according to *age* at surgery

	Group A (below ≤ 11 y.o.)	Group B (above > 11 y.o.)
Cobb angle pre-op($^{\circ}$)	94,2$^{\circ}$ (38 – 139)	122,1$^{\circ}$ (53 – 158)
Final correction (%) of Cobb angle	53,7% (30,1 – 84,1)	42,6% (18,3 – 71,7)
Pelvic tilt pre-op ($^{\circ}$)	21,7$^{\circ}$ (0 – 85)	31,9$^{\circ}$ (0 – 60)
Final correction (%) of pelvic tilt	57,8% (-12,6 – 100)	47,7% (8,0 – 100)
AVT pre-op(mm)	82,0 mm (18 – 165)	100,9 mm (45 – 223)
Final correction (%) of AVT	42,8% (6,7 – 88,9)	30,9% (10,5 – 80,0)

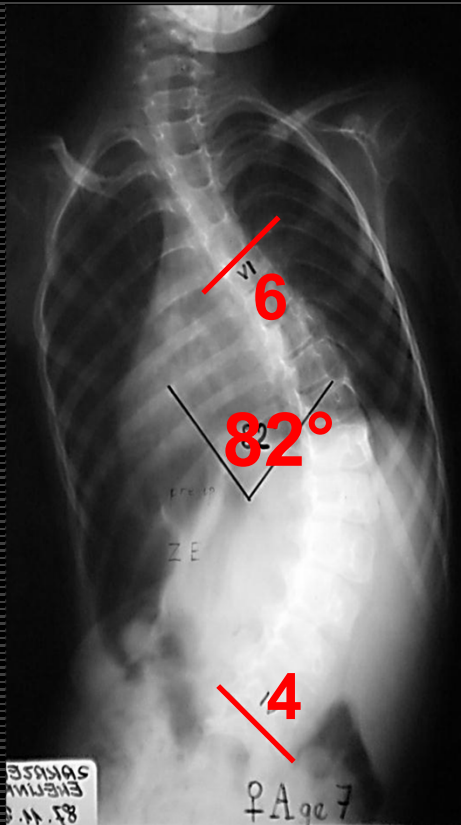
Results – according to *pre-operative curve magintude*

	Group I (40°-80°)	Group II (81°-120°)	Group III (121°-160°)
No of patients	10	16	19
Age at surgery (years)	9.6 (7 – 13)	10.6 (5 – 21)	13.1 (7 – 21)
Final curve correction (%)	51.4% (24.3 – 66.6)	44.7% (18 – 82.9)	34.4% (11.8 – 54)
Loss of correction during f-up (%)	6.6% (-1.9 – 18.6)	5.9% (-2.4 – 17.1)	6.5% (-3.7 – 18.5)
No of pt's with major curve progression (>10°)	3 (33%)	3 (19%)	5 (26%)
Total no of reoperations	1(10%)	4 (25%)	9 (47%)

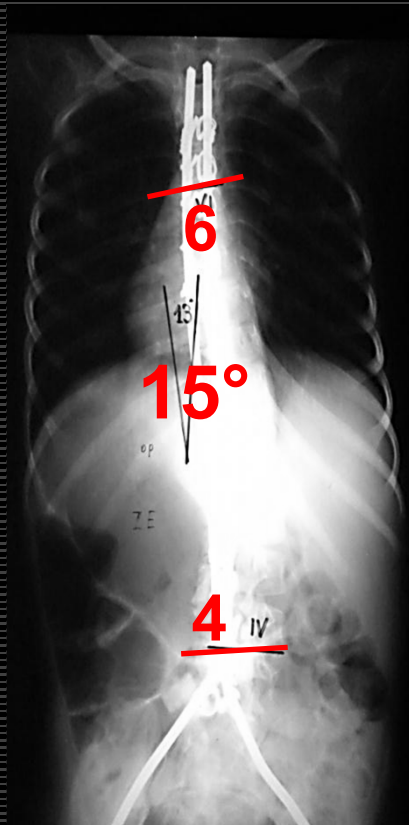
Results – acc to *pulmonary status*

	Group 1 (FVC < 30%)	Group 2 (FVC 31-60%)	Group 3 (FVC > 61%)
No of patients	9	26	10
No of days of mechanical ventilation	1.3 (1 – 3)	1.3 (1 – 2)	1
No of patients requireing tracheostomy	1 (11.1%)	2 (7.6%)	0
No of reoperations	0.1 (0 – 1)	0.5 (0 – 3)	0

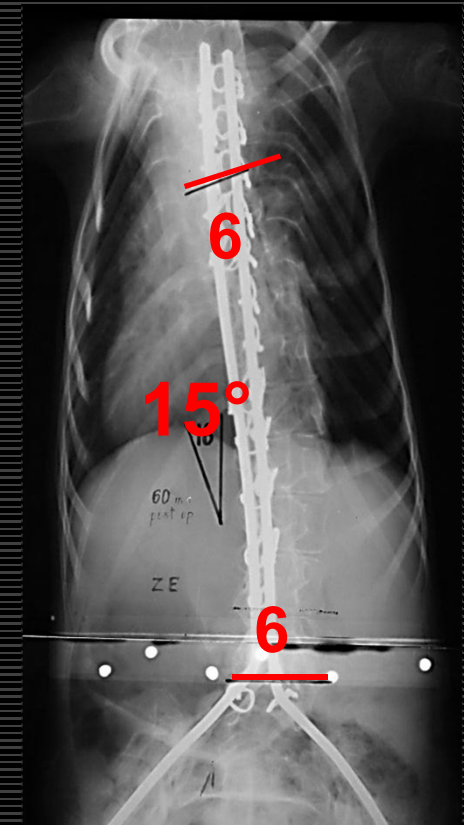
**Patient female, Z.A., SMA type 2,
age at surgery – 7 y.o
follow-up 5 years**



a) pre-op a-p, age 7



b) post-op a-p, lateral, age 7



c) post-op a-p, age 12

Complications

Early ex:

- CSF leakage – 1 patients
- Temporary neurological deficits – 1 patient
- Wound healing problems – 2 patients
- Rod malposition – 2 patients

Late ex:

- Pelvic rod loosening – 3 patients
- Too short range of fusion – 2 patients
- Late infection – implant removal – 1 patient
- Rod transposition – 1 patient

Complications

	SMA2	SMA3a
No of patients requiring revision surgery	7 patients 28%	1 patient 5%
Total number of revision surgeries	13 Reop rate = 52%	1 Reop rate = 5%
Early (<3 months)	4 patients 16%	2 patients 10%
Late (>3 months)	3 patients 12%	1 patient 5%
Minor (soft tissue procedures)	1 patient 4%	1 patient 5%
Major (hardware removal/exchange)	6 patients 24%	0

Conclusions

- ➡ Onset of deformity occurs 2 years after becoming wheelchairbound in SMA3a and 4 y.o age in SMA2
- ➡ Rapid curve progression SMA3a>SMA2
- ➡ No age limits for surgical treatment
- ➡ Surgical treatment allows to achieve good and stable correction
- ➡ Best results in lower curves 40°-80°
- ➡ No major respiratory cxs even in patients with initial low FVC

Referneces

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