





# What you will learn

- 1. How have neuromuscular diseases been treated in the past
- 2. 1980s how interdisciplinary approach improves treatment outcome
- 3. 1990s how digitalisation enables networking & quality assurance
- 4. 2000s how 3D-gait analysis & BoNT give a deeper understanding of biomechanics
- 5. 2010s how muscle power training reduces pain & enhances motor learning
- 6. 2020s how genetics & Al enable personalised treatment protocols
- 7. Future what we may expect?
- 8. Diskussion

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Evolvement in the Treatment of Neuromuscular Disorders Early past 1980s 1990s 2000s 2010s 2020s Future

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# Evolvement in the Treatment of Neuromuscular Disorders

Larry past 19003 193		
	Hemiplegia following stroke Tetraplegia following stroke, brain injury TBI Diplegia (CP, encephalitis, HSSP,)	500 A.D.
a socialitier Cortex	Bilateral Spastic Cerebral Palsy BSCP Dystonia, dyskinesia	1862
Basalganglien, Kleiphirg	Akinesia (Parkinson,)	1817
Stammhirn Morocortex	Spinocerebellar ataxia (Friedreich,)	1863
Invester toritonpater Fait	Cerv./thor./lumb./sacr. Paraplegia (MMC,)	1769
Spina Spina motorisches und	Spinal muscular atrophy SMA	1893
senakka Neuron motoriske Kolplante	Amyotrophic lateral sclerosis ALS	
And	Poliomyelitis anterior	1838
Kinget Band	Secondary toxic neuropathy	
Apparat Gelenko-	Hereditary motorsensor. neuropathy HMSN (CMT,)	1887
biomechanik Knorpel-	Progressive muscular dystrophy (DMD,)	1850
Knochen-Struktur	Congenital myopathy, arthrogryposis Myasthenia, myotonia	1841



 Early past
 1980s
 1990s
 2000s
 2010s
 2020s

# Early past of treating Neuromuscular Diseases



632 A.D. Paul of Aegina describes laminectomy for traumatic spinal cord compression

1768 A.D. Smellie describes obstetric plexus palsy





Future

1838 A.D. Heine reports treatment result of surgery for muscular torticollis



Early past 1980s 1990s 2000s

0s 2010s 2020s Future

# Early past of treating Neuromuscular Diseases – 1880s



1813 Ling publishes Swedish therapeutic gymnastics

Johann Dumreicher (1815-1880) develops the 'railway apparatus'





Karl Nicoladoni (1847-1902) performs first tendon transfer in 1881





'Orthopaedics in the service of neurology'

Albert Hoffa (1859-1907 Würzburg & Berlin)

Die Orthopädie im Dien

'We find ourselves here in a field of our activity that was previously little known, but whose development promises the most beautiful fruits!'















#### **Evolvement in the Treatment of Neuromuscular Disorders** 1990s 2000s 2010s Early past 1980s 2020s Future OCCUPATIONAL THERAPY Max 1958 Early past of treating Neuromuscular Diseases - 1950s An Assessment of the Motor Handicap of Children with Cerebral Palsy and of their Response to Treatment By KAREL BOBATH, M.D. (Prague), D.P.M. **Neurodevelopmental Therapies:** and BERTA BOBATH, F.C.S.P Bobath **Castillo-Morales** Chiropractic CIMT **Conductive Education Petö** Cranio-Sacral-Therapy Feldenkrais Karel und Berta Bobath **Hippotherapy** Kabat **1958 Neurodevelopmental** Macmillan Treatment PNF Osteopathy Root SI Jean Ayres Vojta



Early past 1980s 1990s 2000s 2010s 2020s Future

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### Evolvement in the Treatment of Neuromuscular Disorders Early past 1980s 1990s 2000s 2010s 2020s Future

# An interdisciplinary team approach improves treatment outcome

New technologies:

- In-house databases in specialised institutions
- MRI
- Advanced film technology
- International exchange
- Foundation of international scientific societies



EPOS 1982 Paris

Treatment topics:

- Interdisciplinary
   consultation
- Cinematographic
   evaluation of gait
- Reducing spasticity
- Recognising deformities
- Correction of deformities







**Evolvement in the Treatment of Neuromuscular Disorders** Future Early past

#### 1980s 1990s 2000s 2010s 2020s

# Painful flat foot deformity -> reconstructive foot surgery

Pain relief & walking ability following medial plantar closed-wedge osteotomy & peroneal transfer



**Evolvement in the Treatment of Neuromuscular Disorders** 1980s Future Early past 1990s 2000s 2010s 2020s

# Painful club foot deformity -> reconstructive foot surgery

Pain relief & walking ability following dorso-lateral closed-wedge osteotomy & posterior tibial transfer



**Evolvement in the Treatment of Neuromuscular Disorders** Future

1980s 1990s 2000s 2010s Early past 2020s

# Painful cavovarus foot deformity -> reconstructive foot surgery

Pain relief & improved walking ability following dorsal closed-wedge osteotomy & Jones procedure



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# **Evolvement in the Treatment of Neuromuscular Disorders**

Early past 1980s 1990s 2000s 2010s 2020s Fut

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#### Evolvement in the Treatment of Neuromuscular Disorders Future Early past 1980s 1990s 2000s 2010s 2020s 3D-gait analysis & BoNT give a deeper understanding of biomechanics New technology: Treatment topics: First internat. databases Pain & Quality of Life ٠ for diseases Are BoNT, ITB, and SDR able Start of multi-center to prevent spasticity and outcome studies progressive deformities? Mid-term outcome by 3D-Do we want to weaken or ٠ **Gait Analysis** strengthen muscles? 35







# Vitious Cycle Of Developing Deformities



**Evolvement in the Treatment of Neuromuscular Disorders** 1990s Early past 1980s 2000s 2010s 2020s Future Pain & Quality Of Life in Children with Disabilities Prevention programms needed for: Risk of • Stroke patients Cerebral palsy 0 **Hip dislocation** 0 Spinal disorders 0 **Kypho-Scoliosis** 0 • Neuropathies (HMSN) in **Muscle diseases** 0 Joint contractures 0 **Collagene disorders** 0 Arthritis 0 Arthrogryposis 0 Syndroms 0 In Europe approx. 30.000 new per year

Evolvement in the Treatment of Neuromuscu Early past 1980s 1990s 2000s 2010s 2020s	lar Disorders
3D-Gait analysis identifies muscle weakness as the ma	in problem
<ul> <li> abnormal muscle activity in orthopaedic patients who had involvement.</li> <li> Muscle weakness was the most important cause of the abnactivity.</li> <li> The triceps surae muscle, represented by the medial gastr significantly more involved.</li> <li> Co-activity of the knee extensors and hamstrings</li> <li> These findings illustrate mechanisms to compensate for n in stance in neurologically intact subjects.</li> </ul>	no neurological normal EMG rocnemius, was nuscle weakness
Brunner R <sup>1</sup> , Romkes J. <b>Abnormal EMG muscle activity during gait in patients withou disorders.</b> Gait Posture. 2008 Apr;27(3):399-407. Epub 2007 Nov 19.	t neurological





	Evolve	ement in t	he Tre	eatme	nt of N	leuror	nusci	ular Disorders	
		Early past	1980s	1990s	2000s	2010s	2020s	Future	
0									
Conse	quences	for treatm	ient						
	abilization of	Light avia							
• 31	Ankle 90°	knee 0° hin	0° spin						
•	Dynamic o	rthotics	v, spir						
•	Lever arm-	restoring su	iraerv						
• St	rengthening	of muscles							
•	Strengther	ning exercise	es						
•	Shortening	g and recons	structive	e surger	у				
•	BoNT-injec	tons in anta	igonists						
•	Dosed leng	gthening of a	antagon	ists					
• Av	oidina musa	le weakenin	a						
•	SDR only i	n selected c	ases						
•	BoNT only	in selected	cases						
•	Lengtheni	ng by multile	evel sur	gery in s	selected	cases			
	-								

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#### Evolvement in the Treatment of Neuromuscular Disorders Early past 1980s 1990s 2000s 2010s 2020s Future

# Muscle power training reduces pain & enhances motor learning

New technology:

- First national register's longterm outcome studies
- Long-term outcome by 3D-Gait Analysis



Treatment topics:

- BoNT & ITB cannot prevent development of deformities
- Underestimated pain
- Muscle power training
- Minimal-invasive surgery

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Improving Orthotics

Evolvement in the Treatment of Neuromuscular Disorders Early past 1980s 1990s 2000s 2010s 2020s Future
Muscle power training reduces pain & enhances motor learning
Causes of pain
Non-use     Lack of active & passive muscle training causes "neurogenic neuroinflammation"
Over-use syndrome     Lack of control & weakness cause compensatory muscle activity & hyperactivity
Deformities     Malalignments of extremities and spine cause pressure pain
Spasticity, dystonia     Involuntary movements cause pain

# Muscle power training reduces pain & enhances motor learning



Evolvement in the Treatment of Neuromuscular Disorders Early past 1980s 1990s 2000s 2010s 2020s Future
The use of minimally invasive techniques in multi-level surgery for children with cerebral palsy: preliminary results
<sup>1</sup> Oxford Gait Laboratory, Nuffield Orthopaedic Centre, Headington, Oxford OX3 7LD, UK. nicky.thompson@noc.nhs.uk J Bone Joint Surg Br 2010 Oct;92(10):1442-8. doi: 10.1302/0301-620X.92B10.24307.
This study compares the initial outcomes of minimally invasive techniques for single-event multi-level surgery with conventional single-event multi-level surgery. The minimally invasive techniques included derotation osteotomies using closed corticotomy and fixation with titanium elastic nails and percutaneous lengthening of muscles where possible. A prospective cohort study of two matched groups was undertaken. Ten children with diplegic cerebral palsy with a mean age of ten years six months (7.11 to 13.9) had multi-level minimally invasive surgery and were matched for ambulatory level and compared with ten children with a mean age of 11 years four months (7.9 to 14.4) who had conventional single-event multi-level surgery. Gait kinematics, the Gillette Gait Index, isometric muscle strength and gross motor function were assessed before and 12 months after operation.
The minimally invasive group had significantly reduced operation time and blood loss with a significantly improved time to mobilisation. There were no complications intra-operatively or during hospitalisation in either group. There was significant improvement in gait kinematics and the Gillette Gait Index in both groups with no difference between them. There was a trend to improved muscle strength in the multi-level group. There was no significant difference in gross motor function between the groups. We consider that minimally invasive single-event multi-level surgery can be achieved safely and effectively with significant advantages over conventional techniques in children with diplegic cerebral palsy.

Clinical 1 studies 2 2010s 3 4 5 7 8
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	Evolvement in the Treatment of Neuromuscular Disorders Early past 1980s 1990s 2000s 2010s 2020s Future
Mussle	
wuscie	bower training
	Training of muscular power
	<ul> <li>signif. findings in peripheral and central pareses</li> </ul>
	•Dodd et al. 2002; Darrah et al. 1997
Repetitiv	ve Training
	Repetitive training
	signif. improvements of motor functions
	• Bütefisch et al. 1995; Sterr et al. 2002

# Whole Body Vibration Therapy

Whole Body Vibration (WBV) in CP patients "... may be a useful tool for improving muscle strength and balance in children with diplegic cerebral palsy."

Effect of whole-body vibration on muscle strength and balance in diplegic cerebral palsy: a randomized controlled trial. EI-Shamy SM. Am J Phys Med Rehabil. 2014 Feb;93(2):114-21.





Evolvement in	hthe Tr	eatme	nt of I	Neuro	musci	ular Dis	orders
Early past	1980s	1990s	2000s	2010s	2020s	Future	

# **Locomotion Therapy**

Starting postoperatively:

- •1-2 days following
  - $\circ$  minimal-invasive SEML surgery
- •3-5 days following
  - $\circ\,$  Tendon transfers with joint stabilization
  - Ostotomies/locking plates < 50 kg BW</p>
- 4 weeks following
  - $\circ$  Osteotomies/locking plates > 50 kg KG
  - $\circ\,$  Foot reconstruction surgery with joint stabilization
- 6-8 weeks following
  - $\circ~$  hip reconstruction surgery incl. open reduction, bilat. pelvic osteotomies



	Evolvement in the Treatment of Neuromuscular Disorders Early past 1980s 1990s 2000s 2010s 2020s Future
Clinical studies 2010s	<ol> <li>Frequent movement in-between fascias reduces development of contractures.</li> <li>Early surgical programms may reduce percentage of hip dislocations down to 0%.</li> <li>Rediscovered minimal-invasive surgical techniques facilitate postoperative mobility and painfree exercises.</li> <li>New implants permit full weigth-bearing postoperatively.</li> <li>Perioperative adjustment of dynamic orthoses helps to reduce immobilisation.</li> <li>Verticalisation reduces spasticity and improves brain metabolism.</li> <li>Locomotion and whole-body-vibration facilitate neuronal integration.</li> <li>Robotic-assisted locomotion therapy enables early postoperative training with guided legs and joints.</li> <li>Power exercises and repetitive training show positive effects on daily life activities, reduction of pain, and quality of life.</li> <li>Education programs for patients and parents improve knowledge, confidence, compliance, and outcome of treatment.</li> </ol>



![](_page_28_Picture_1.jpeg)

![](_page_28_Picture_2.jpeg)

Early past 1980s 1990s 2000s 2010s 2020s

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#### **Evolvement in the Treatment of Neuromuscular Disorders** Early past Future

1980s 1990s 2000s 2010s 2020s

# Genetics & AI enable personalised treatment protocols

New technology:

- First international databases for rare diseases
- Genetic diagnostics
- Gene therapy
- Tools that use artificial intelligence

![](_page_29_Picture_21.jpeg)

Treatment topics:

- Early gene therapy
- Very early physiotherapy
- Detecting pain
- Preventing non-use, hyperactivity, and pain
- Using Al-tools for personalised diagnostics and treatment protocols

![](_page_29_Picture_29.jpeg)

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Future

![](_page_30_Figure_1.jpeg)

SMA	Nusinersen	-> production of SMN protein	
SMA SMA	Onasemnogene Risdiplam	-> sustained expression of SMN protein -> production of SMN protein	
DMD	Deflazocort	-> antiinflammatory	
	Ataluren	-> translation of full-length dystrophin	
Fabry	Agalsidase beta	-> decreases accumulation of Gb3	

![](_page_31_Picture_1.jpeg)

**Disease-modifying treatments** 

When considering early treatment, aspects such as response variability, cost, and safety must be taken into account.

Stronger evidence is needed to support early intervention in several conditions, but the low incidence of rare diseases poses challenges to obtaining reliable data.

Systematic reviews and meta-analyses can help summarize evidence, but they have limitations and biases.

Finally, NBS is a powerful tool that has the potential to revolutionize the course of diseases where early intervention is crucial.

Laurane Mackels, Laurent Servais. The Importance of Early Treatment of Inherited Neuromuscular Conditions. 2024 Mar 5;11(2):253–274. doi: 10.3233/JND-230189

![](_page_32_Picture_1.jpeg)

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Evolvement in the Treatment of Neuromuscular Disorders Early past 1980s 1990s 2000s 2010s 2020s Future

# What we may expect?

- 1. More precise & earlier diagnosis
- 2. More causal therapies -> other pediatric orthopedic disease patterns
- 3. More datas about very early training of muscular function
- 4. Prevention of non-use, compensation, hyperactivity, deformities, and pain
- 5. Improved social partizipation
- 6. Registers and access to institutions
- 7. Profound change in research e.g. automated meta-analyses, ...
- 8. Profound change in education e.g. utilisation and control of AI

![](_page_33_Picture_23.jpeg)

![](_page_34_Figure_1.jpeg)

Evolvement in the Treatment of Neuromuscular Disorders Early past 1980s 1990s 2000s 2010s 2020s Future
History of prevention of secondary damage in cerebral palsy

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![](_page_35_Picture_2.jpeg)

![](_page_36_Figure_1.jpeg)

![](_page_36_Picture_2.jpeg)

![](_page_37_Picture_1.jpeg)

![](_page_37_Picture_2.jpeg)