Leiden University Medical Center

duchenne centre netherlands

DMD and orthopedics

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Orthopedic surgeon - epidemiologist

SPIERZIEKTEN NEDERLAND

Duchenne Parent Project

Spieren voor Spieren

Voor kinderen met een spierziekte

disclosure

Nothing to disclose.

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topics

Achilles tendon

Footsurgery in non ambulant patients

Scoliosis

- pelvis
- hips

fractures

Based on the considerations from the lancet article

Birnkrant DJ, Bushby K, et al; DMD Care Considerations Working Group. Diagnosis and management of Duchenne muscular dystrophy, part 2: respiratory, cardiac, bone health, and orthopaedic management. Lancet Neurol. 2018 Apr;17(4):347-361.

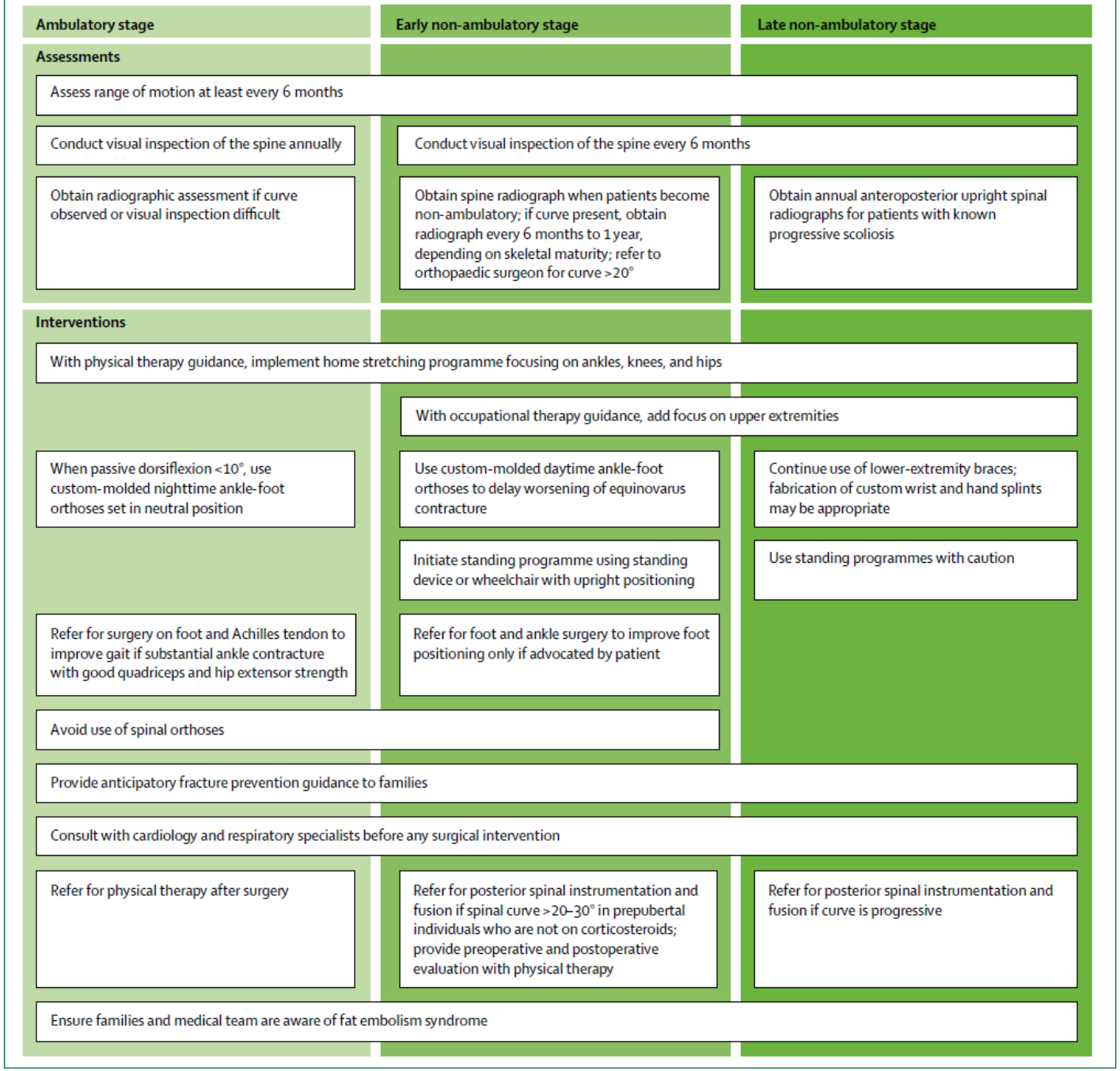


Figure 4: Considerations for orthopaedic and surgical care of patients with Duchenne muscular dystrophy by stage of disease

First the foot

Would you be so kind to stand up?

Lean a little bit forward

And increase your lordosis

Which muscle is tense in your lower leg?

First the foot

Be very careful with Achilles tendon lengthening

we don't do it in the Netherlands with ambulant DMD boys

The greatest risk is to make someone worse!

you need your triceps surae to stand

"no" surgery during ambulatory phase

Seldom surgery during early non-ambulatory phase

Sometimes surgery during late non-ambulatory phase

Feet? In a non-ambulatory patient?

Foot corrections for wheelchair bound patients

Contractures can (and will) be painful

Pressure sores on lateral border of the foot

Inability to wear shoes

Equinovarus deformity

Release of Achilles tendon

Release or transfer for posterior tibial tendon

Often in combination with tenotomy of toe flexors (FHL and FDL)

Scoliosis WITHOUT steroids

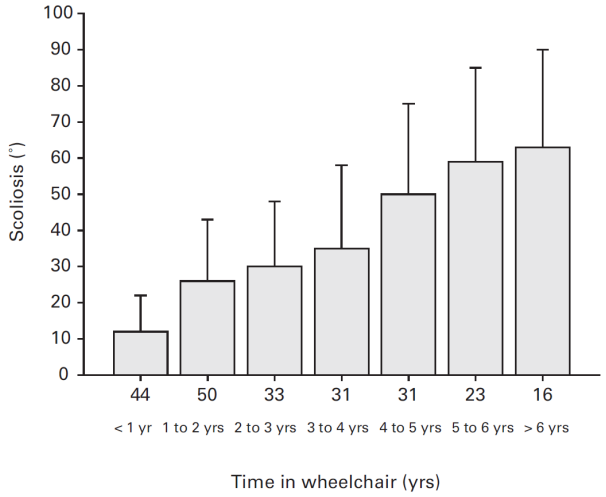
97 % scoliosis > 10 degrees

89 % scoliosis > 20 degrees

75% scoliosis > 30 degrees

n = 88

Time in wheelchair was highly linked with the progression (not the age)



Shapiro F, Zurakowski D, Bui T, Darras BT. Progression of spinal deformity in wheelchair-dependent patients with Duchenne muscular dystrophy who are not treated with steroids: coronal plane (scoliosis) and sagittal plane (kyphosis, lordosis) deformity. Bone Joint J. 2014 Jan;96-B(1):100-5.

Scoliosis WITH steroids

Most data on DAILY steroids

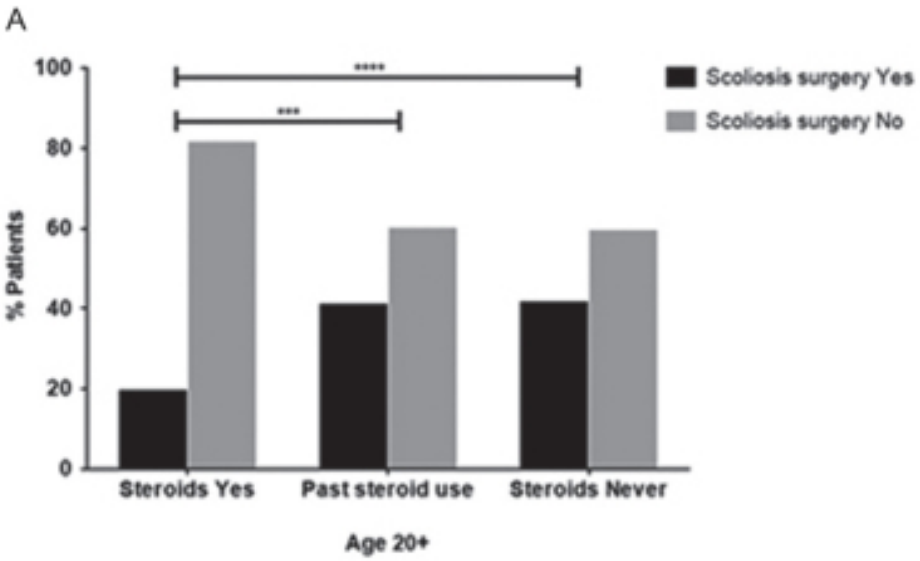
Decline from about 90% to 20%

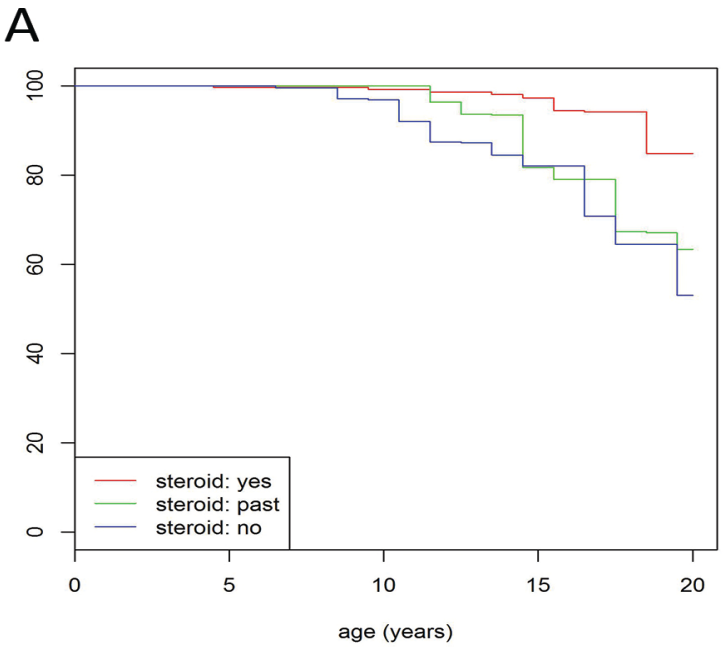
And curves are less severe

Sanzarello I, Merlini L, Traina F, Rosa MA, Faldini C. Corticosteroid Treatment Impact on Spinal Deformity in Duchenne Muscular Dystrophy. Int Sch Res Notices. 2014 Oct 29;2014:965235

Lebel DE, Corston JA, McAdam LC, Biggar WD, Alman BA. Glucocorticoid treatment for the prevention of scoliosis in children with Duchenne muscular dystrophy: long-term follow-up. J Bone Joint Surg Am. 2013 Jun 19;95(12):1057-61.

Scoliosis WITH steroids





N = 836

A total of 9% (*n* = 488) of the registry patients have had scoliosis surgery.

Koeks Z et al; Clinical Outcomes in Duchenne Muscular Dystrophy: A Study of 5345 Patients from the TREAT-NMD DMD Global Database. J Neuromuscul Dis. 2017;4(4):293-306.

Window of opportunity

Curves will progress (and progress **fast**)

Bracing is not useful

When you postpone referral for surgery:

- surgery becomes more difficult
- lungs and heart
- BMI increase

Refer to spine surgeon **before**:

- curve is over 40 degrees
- BMI is over 40
- ejection fraction is < 40%
- FVC is < 40%

Perioperative complications after scoliosis surgery

Higher incidence of complications as compared to scoliosis surgery for other neurological diseases.

Total 110 patients, 26 DMD

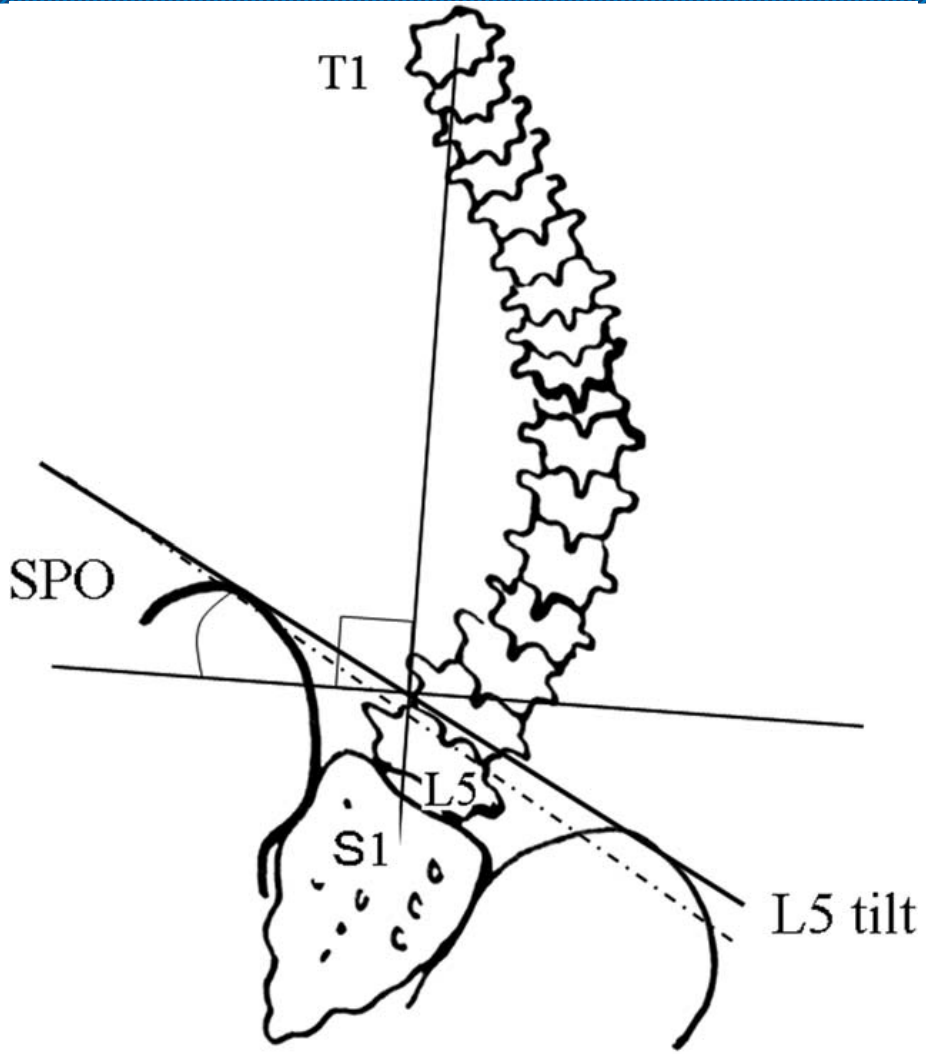
Wound infection 19% versus 5%

Hepatotoxicity only in DMD (N=4) and associated with blood loss

As group comparable blood loss

Duckworth AD, Mitchell MJ, Tsirikos AI. Incidence and risk factors for post-operative complications after scoliosis surgery in patients with Duchenne muscular dystrophy : a comparison with other neuromuscular conditions. Bone Joint J. 2014 Jul;96-B(7):943-9.

Pelvic tilt



Fusion to L5 or Sacrum?

A lot of debate

With newer instrumentation it is easier to perform

- duration of surgery
- blood loss
- infections

In the Dutch scoliosis guideline “with severe pelvic obliquity”

In the Lancet review **> 15 degrees**: fusion to sacrum

Research question!

Duckworth AD, Mitchell MJ, Tsirikos AI. Incidence and risk factors for post-operative complications after scoliosis surgery in patients with Duchenne muscular dystrophy : a comparison with other neuromuscular conditions. Bone Joint J. 2014 Jul;96-B(7):943-9.

Only to L5?

TABLE 2. Radiographic Measurements in the Study Group				
	Postop	Postop	2-yr Postop	The Ultimate
Coronal Cobb angle	70° (51°–88°)	15° (5°–25°)	16° (6°–28°)	17° (6°–27°)
Sagittal thoracic curve (T3–T12)	7° (−8° to 22°)	20° (12°–33°)	21° (12°–32°)	21° (13°–34°)
Thoracolumbar junction (T12–L2)	18° (2–25°)	3° (−5° to 8°)	3° (−6° to 10°)	2° (−5° to 8°)
Sagittal lumbar curve (L1–S1)	20° (−18° to 58°)	42° (12°–58°)	43° (13°–55°)	43° (12°–56°)

TABLE 3. Radiographic Measurements in the Study Group							
L5 Tilt (°)				Pelvic Obliquity (°)			
Preop	Immediate Postop	2-yr Postop	The Ultimate	Preop	Immediate Postop	2-yr Postop	The Ultimate
9 (2–14)	2 (0–4)	2 (0–5)	2 (0–5)	15 (9–25)	5 (3–8)	6 (3–8)	6 (3–8)

*Segmental Pedicle Screw Instrumentation and **Fusion Only to L5** in the Surgical Treatment of Flaccid Neuromuscular Scoliosis*
M. Takaso et al
Spine 2018;43:331–338

Outcome after scoliosis surgery

While most studies of Duchenne muscular dystrophy scoliosis **focus on technical and radiographic indices**, functional status is a more important factor to consider in the management of Duchenne muscular dystrophy. The objectives of the current study were to compare the pulmonary function, radiographic outcome, and **functional recovery**, with use of validated questionnaires, **in surgically and non-surgically** treated patients with Duchenne muscular dystrophy who have scoliosis.

Conclusions: Surgery in patients who had Duchenne muscular dystrophy with scoliosis improved function and decreased the rate of deterioration of forced vital capacity compared with patients treated conservatively. However, the muscle power and forced vital capacity decreased in both groups.

Suk KS, Lee BH, Lee HM, Moon SH, Choi YC, Shin DE, Ha JW, Song KM, Kim HS. Functional outcomes in Duchenne muscular dystrophy scoliosis: comparison of the differences between surgical and nonsurgical treatment. J Bone Joint Surg Am. 2014 Mar 5;96(5):409-15.

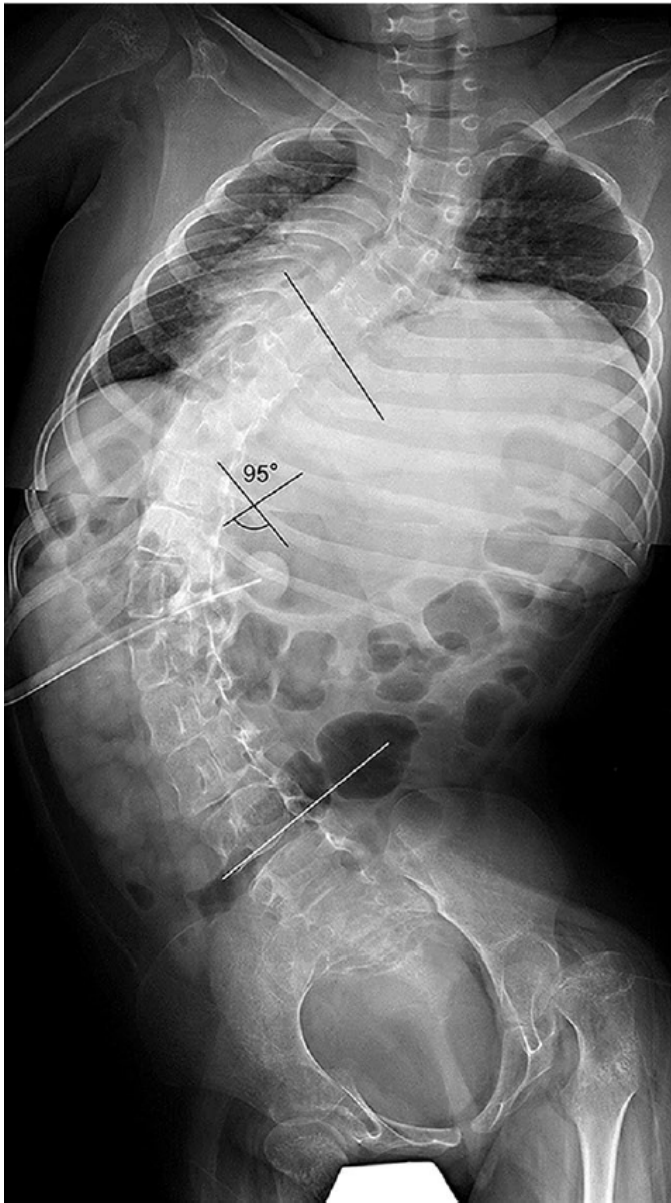
Trunk-pelvis-hip

Recently more literature on this important topic

Don’t only focus on the spine

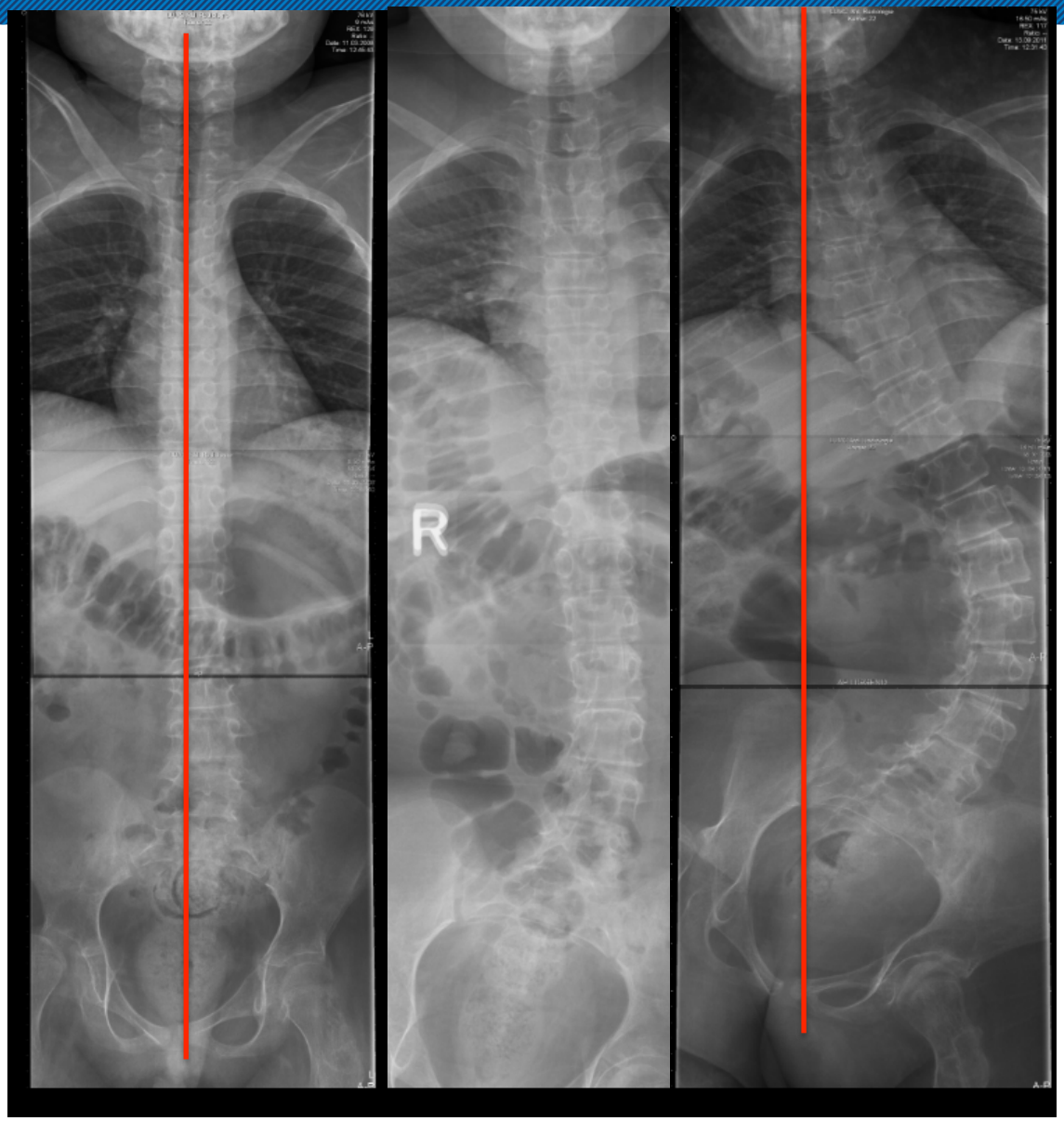
A triad of deformities

- thoracolumbar scoliosis
- pelvic obliquity
- femoral head (hip) subluxation/ dislocation



Research question!

Patel J, Shapiro F. Simultaneous progression patterns of scoliosis, pelvic obliquity, and hip subluxation/dislocation in non-ambulatory neuromuscular patients: an approach to deformity documentation. J Child Orthop. 2015 Oct;9(5):345-56.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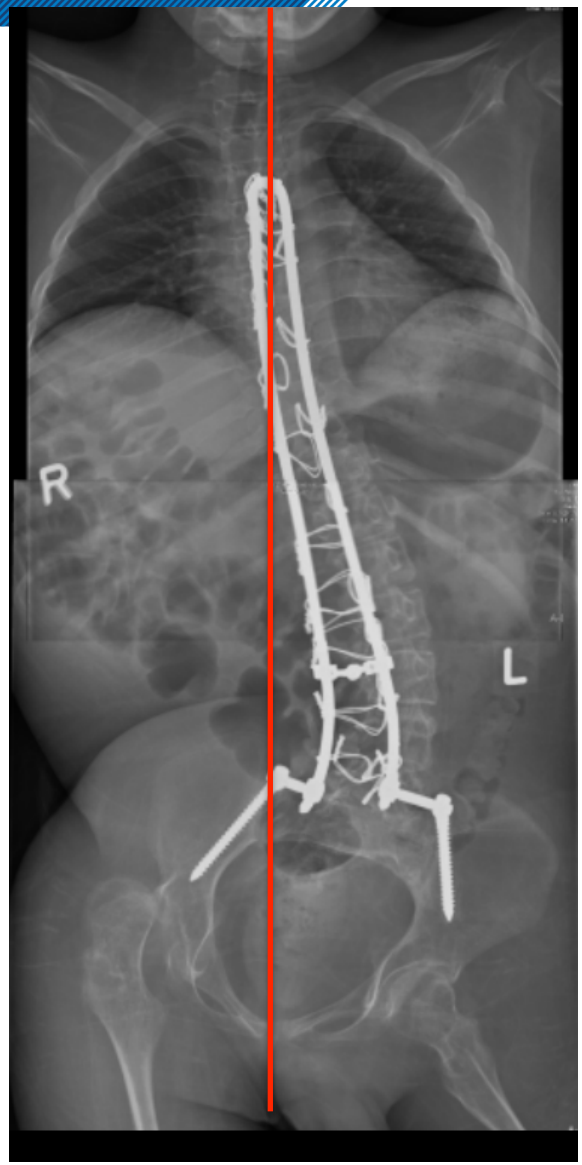
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Decline in upper extremity function after spine fusion

We all know some patients, but hardly any publication

Until recently…. From the Nijmegen group

n = 213, web based questionnaire

Scoliosis is negative associated with UE function.

But in the article no distinction is made between operated and not operated patients.

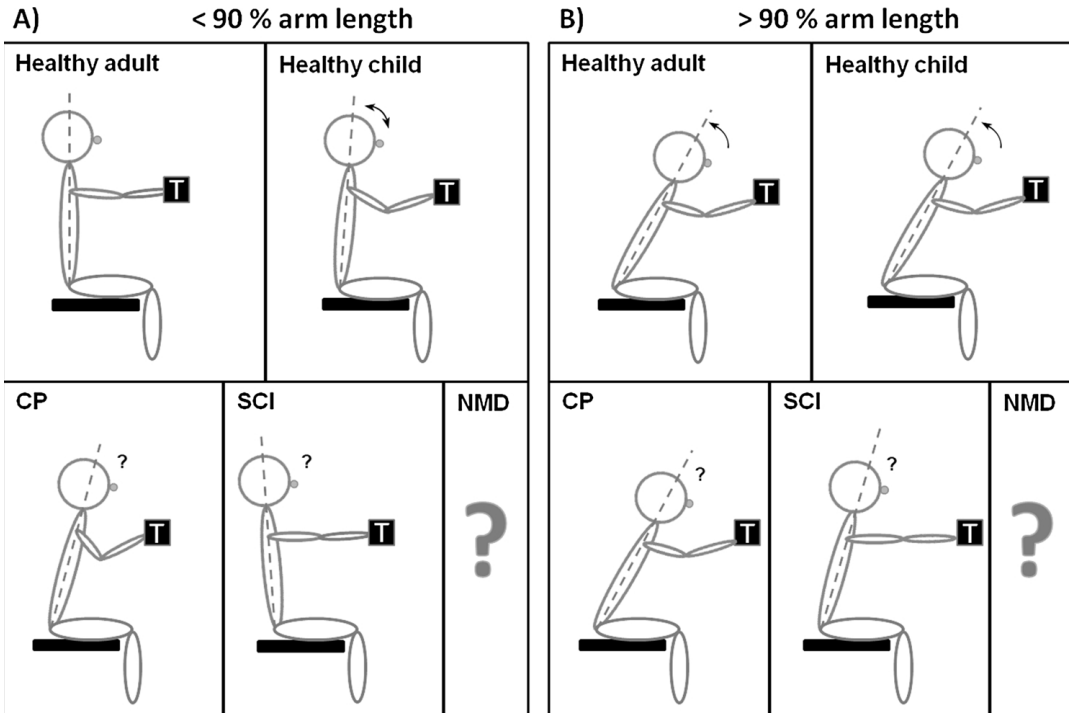
Research question!

Variables associated with upper extremity function in patients with Duchenne muscular dystrophy
Mariska M. H. P. Janssen, Jan C. M. Hendriks, Alexander C. H. Geurts, Imelda J. M. de Groot
J Neurol. 2016; 263(9): 1810–1818.

Compensation mechanisms

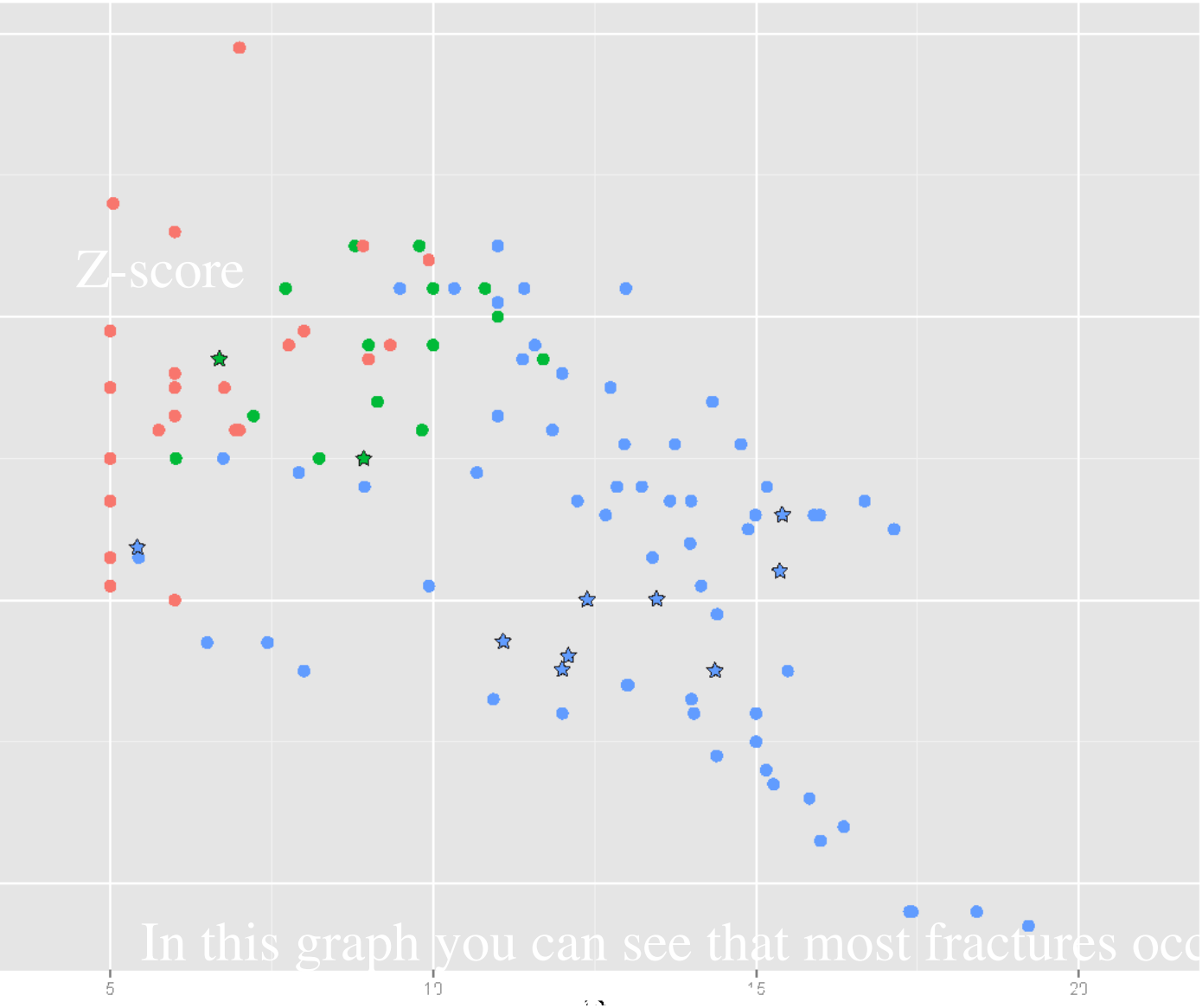
No rotation or flexion from the spine after fusion

Importance of good hip function



Peeters LHC, de Groot IJM, Geurts ACH. Trunk involvement in performing upper extremity activities while seated in neurological patients with a flaccid trunk - A review. Gait Posture. 2018 May;62:46-55.

Fractures (unpublished LUMC data)



No fracture
fracture

Mobile
Partial wheelchair bound
Full wheelchair bound

In this graph you can see that most fractures occ

Vast majority of the lower extremity fractures are in non-ambulant boys
In mobile boys more upper extremity fracture

Treat as a fracture in non-DMD boys

only exception when a long immobilisation can be prevented

Risks and benefits for the fracture and the operation

Vertebral fracture assesment

As good as lateral X-ray

Lower radiation

However: at a different department and not available in all hospitals

Crabtree NJ, Chapman S, Högler W, Hodgson K, Chapman D, Bebbington N, Shaw NJ. Vertebral fractures assessment in children: Evaluation of DXA imaging versus conventional spine radiography. Bone. 2017 Apr;97:168-174.

Peri operative team

In the guidelines:

- cardiologist
- pulmonologist
- physiotherapist

We would suggest to add

- anesthesiologist
- intensive care physician
- physiatrist
- occupational therapist

summary

All considerations are applicable for the Dutch situation except:

We don't do Achilles tendon lengthening in ambulant boys	research
Variation between spine surgeons to fuse until L5 or sacrum	accept
Variation in vertebral fracture assesment	implement
Lower leg fractures mostly nonoperative	accept
new guideline trauma in children: refer to specialist centre	
Not in all hospitals possibility to make sitting spine X-rays	implement