AN INITIAL REPORT ON A GRADING SYSTEM FOR GLOBAL ASSESSMENT OF A CHILD WITH AMYOPLASIA

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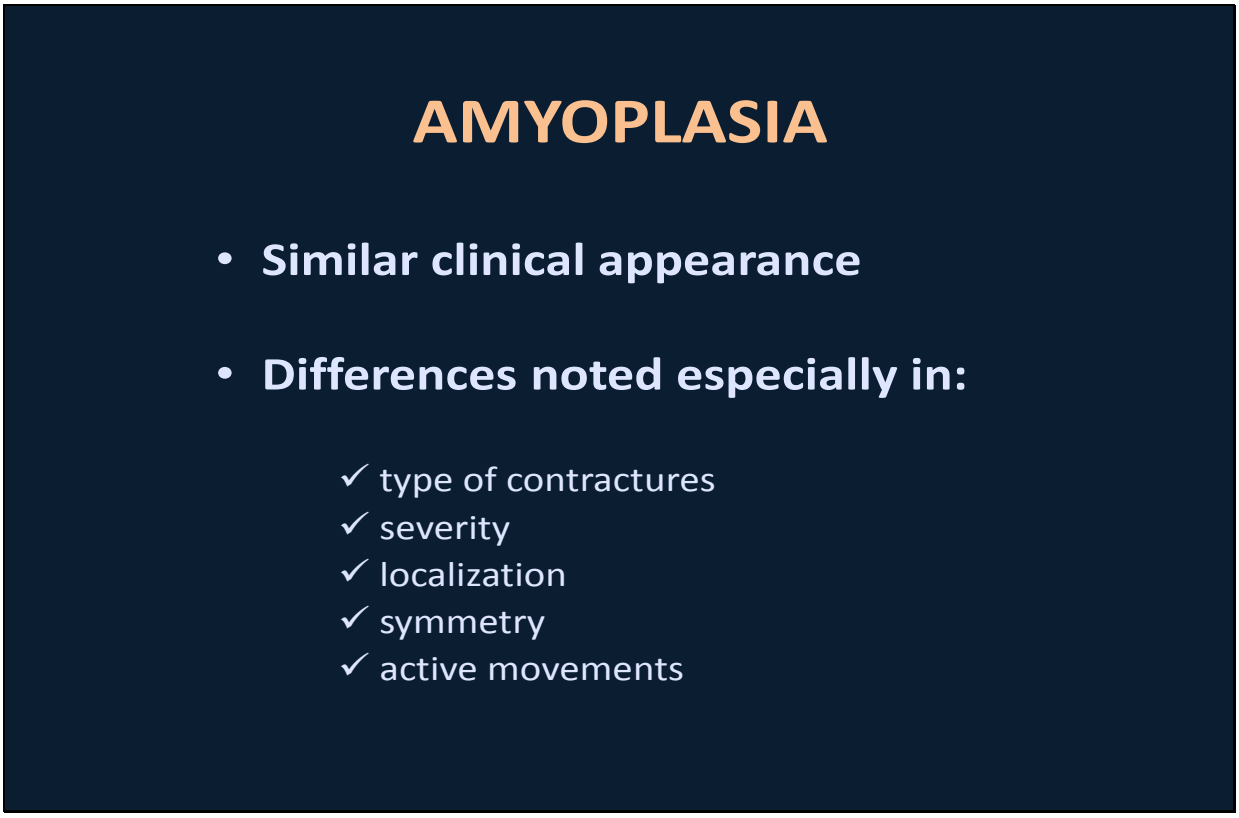
This is a presentation on a proposal of a grading system dedicated to assess the severity of deformities and contractures in children with amyoplasia. It is based on an orthopaedic examination of the upper/lower body and the spine including joint contractures, passive range of joint motion, and the presence or absence of their selected active movements.

Although the scale is designed for amyoplasia, it could possibly be used in other types of arthrogryposis, following certain modifications.

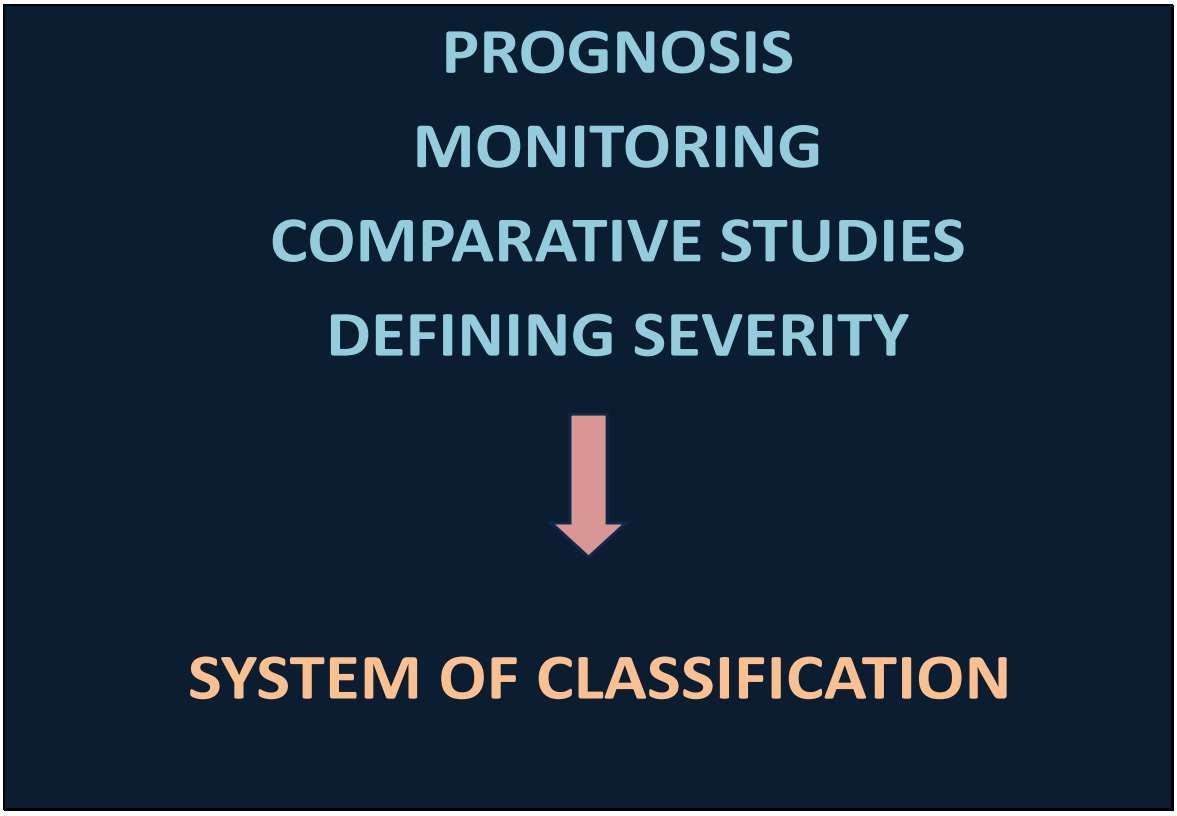


Amyoplasia is characterized by similar clinical appearance i.e. muscle weakness or absence, symmetrical joint contractures: usually adducted internally rotated shoulders, extended elbows, flexed wrists, hips, extended knees, clubfeet, etc.

In clinical practice, however, differences between particular amyoplasia patients are frequently noted, concerning e.g. the type of contractures, whether in flexion or extension, and their severity, understood as the degree to which the contractures cause limitations in passive range of motion. Moreover, not always are all joints involved or the contractures strictly symmetric, and, last but not least, patients vary in their ability for active movement of upper and lower extremities.

These clinical pictures demonstrate how differences in the severity of amyoplasia are often apparent even without a thorough examination.

We believe that those diverse clinical manifestations may require various courses of treatment and result in divergent outcomes and functions obtained by the patients during the follow-up.



Therefore, a system of classification and grading of amyoplasia seems essential to assess the disease prognosis, to monitor the course of treatment, for reliable comparative studies, and to define the severity of the disease.

EXISTING CLASSIFICATION

ARTHROGRYPOSIS MULTIPLEX CONGENITA: FUNCTIONAL CLASSIFICATION AND THE AMC DISC-O-GRAM

U. MENNEN

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The arthrogryposis multiplex congenita (AMC) disc-o-gram is presented as a way to measure the effect of hand therapy, splinting and/or surgery on the AMC patient. It also allows comparative measurement of passive motion (in the baby), active motion (in the young child) and function (in the older child and adult). These measurements are related to a new classification system which, as movement and function improves, indicates the efficacy of management.

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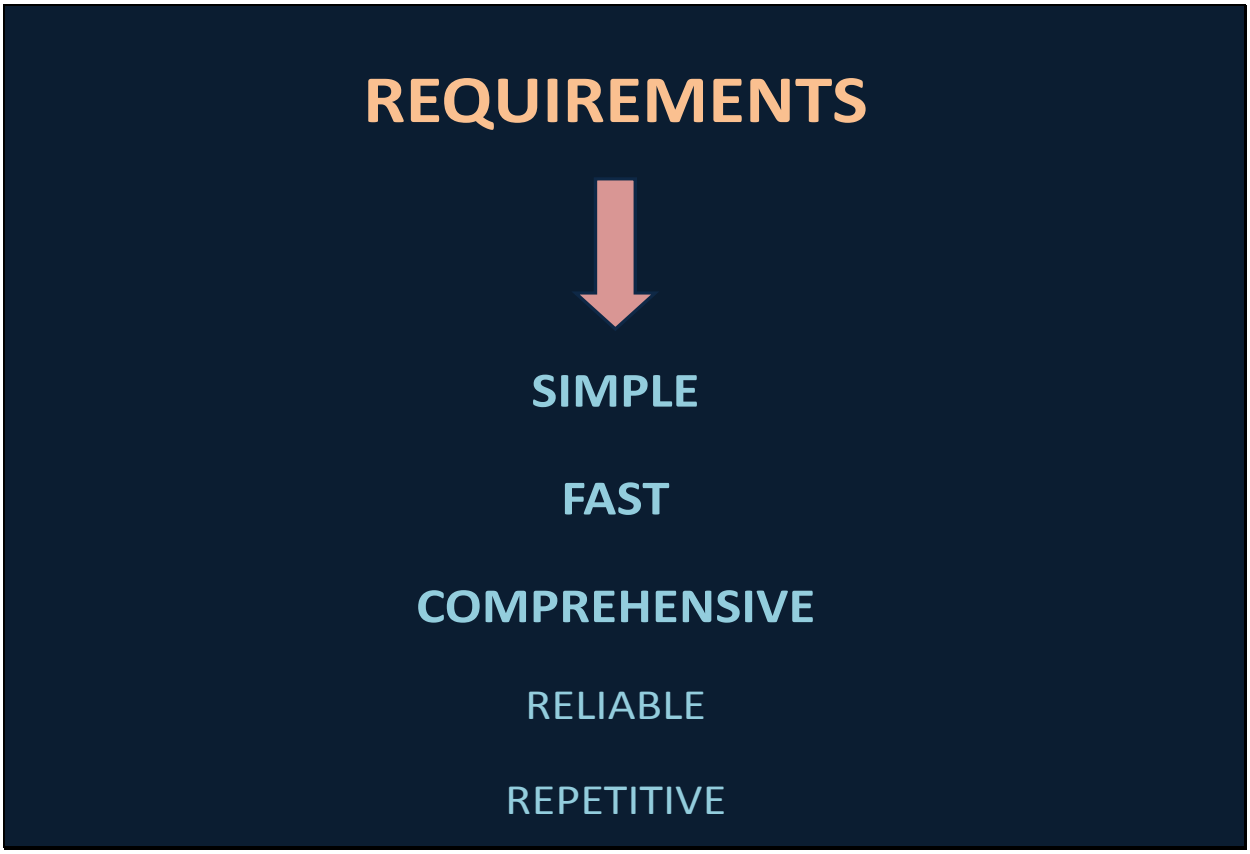
There is already a system designed by U. Menen in 2004, called the disc-o-gram, allowing for comprehensive measurements of ROM and active movements in children with arthrogryposis.

DISC-O-GRAM

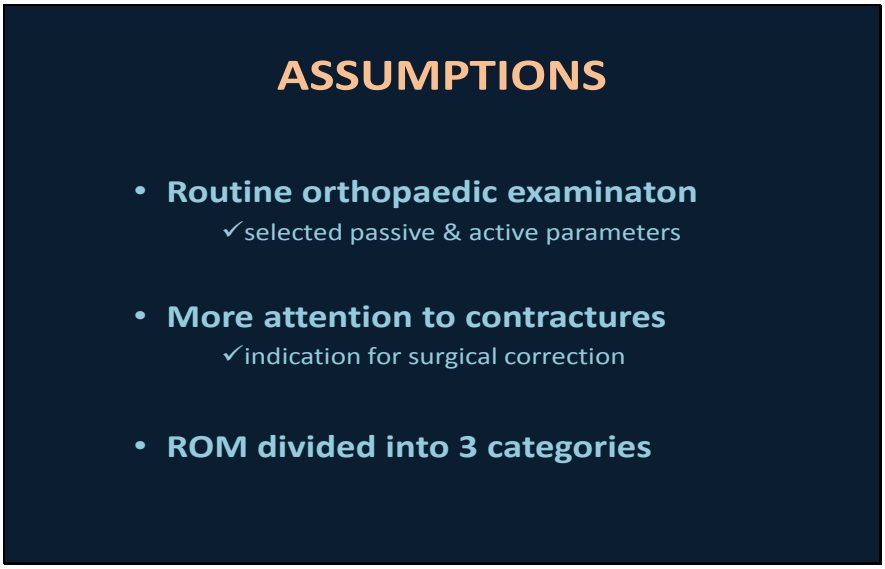
- Accurate, precise
- Complicated
- Difficult for everyday use

However, despite its accuracy, it is also complicated and time-consuming: we tried to use it in our clinical practice but it took nearly 2 hours per patient to complete. Moreover, it is hardly appropriate for infants and young babies.

Consequently, we consider this method difficult and inconvenient in everyday use when treating children with amyoplasia.



To be widely accepted as a comfortable tool in everyday practice, such system of grading should be fast and simple, possible to carry out during a routine examination of the child, appropriate for any age, and allowing for a comprehensive assessment of the whole body and basic function of the musculoskeletal system.

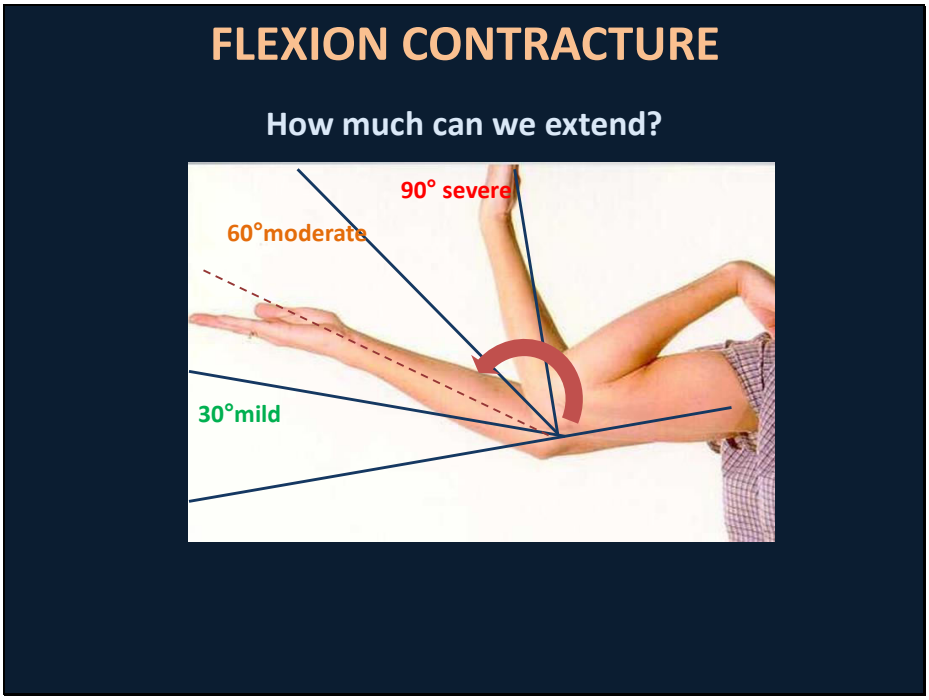


To achieve the previously mentioned goals, the system of examination:

1. **cannot be based on overly precise measurements of the range of motion, i.e. to 1 degree accuracy, as this is time-consuming and non-repeatable;**
2. **should rather be based on the categorization of the range of motion and contractures;**
3. **and should focus on selected – prioritised – active movements;**

Otherwise, the classification will be non-repeatable, time-consuming, and too difficult to be used in practice. We decided to focus more on passive contractures, as they constitute most frequent indications for surgical treatment in amyoplasia. Contractures are also easily measured regardless of the age of the patient, whereas for an active movement examination we need a cooperative patient.

To avoid time-consuming and non-repeatable measurements of joints arch of motion, we sub-divided them into three categories of contractures: mild, moderate, and severe.



To explain the method of 'mild-moderate-severe' examination, let us use the example of a flexion contracture of the elbow, where we ask to what degree we can passively extend the forearm.

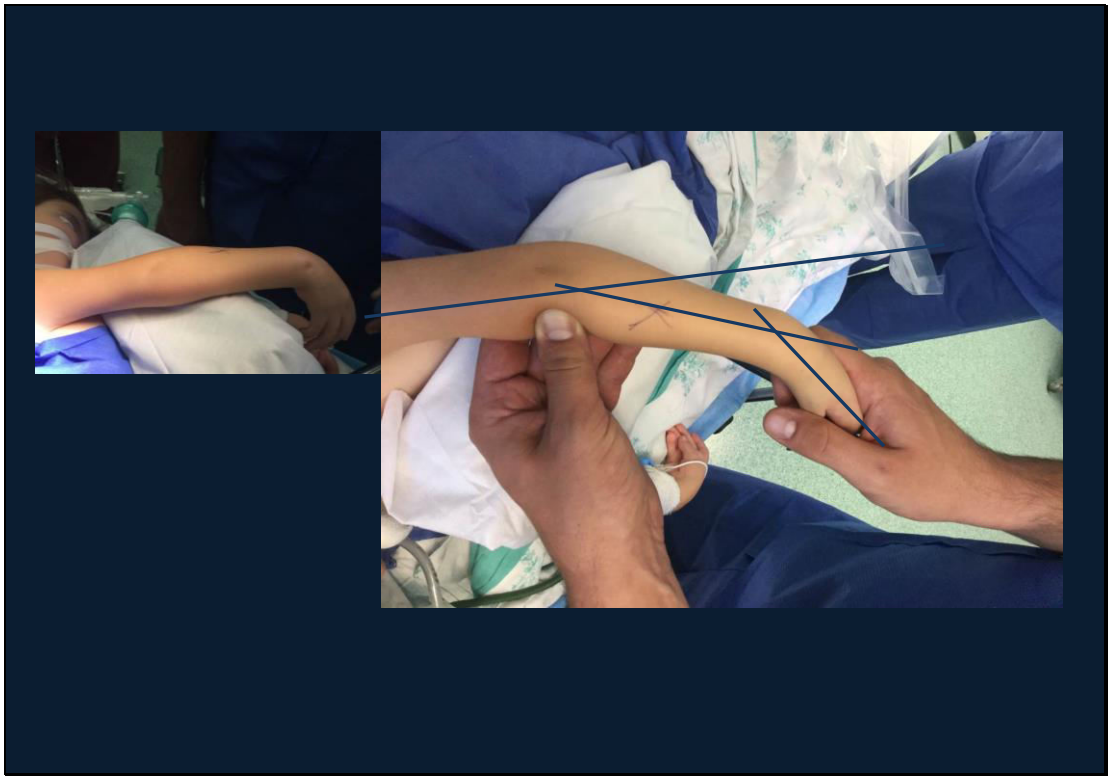
If full extension is considered 0°, the flexion contracture of 0-30° would be a mild flexion contracture.

If we can move the elbow from maximal flexion to between 30-60°, it would constitute a moderate contracture.

If we can extend the elbow to only between 60-90° or less, it would be a severe flexion contracture.

The range of motion presented in the picture allows for extension from full flexion to about 45°, so it is a moderate contracture.

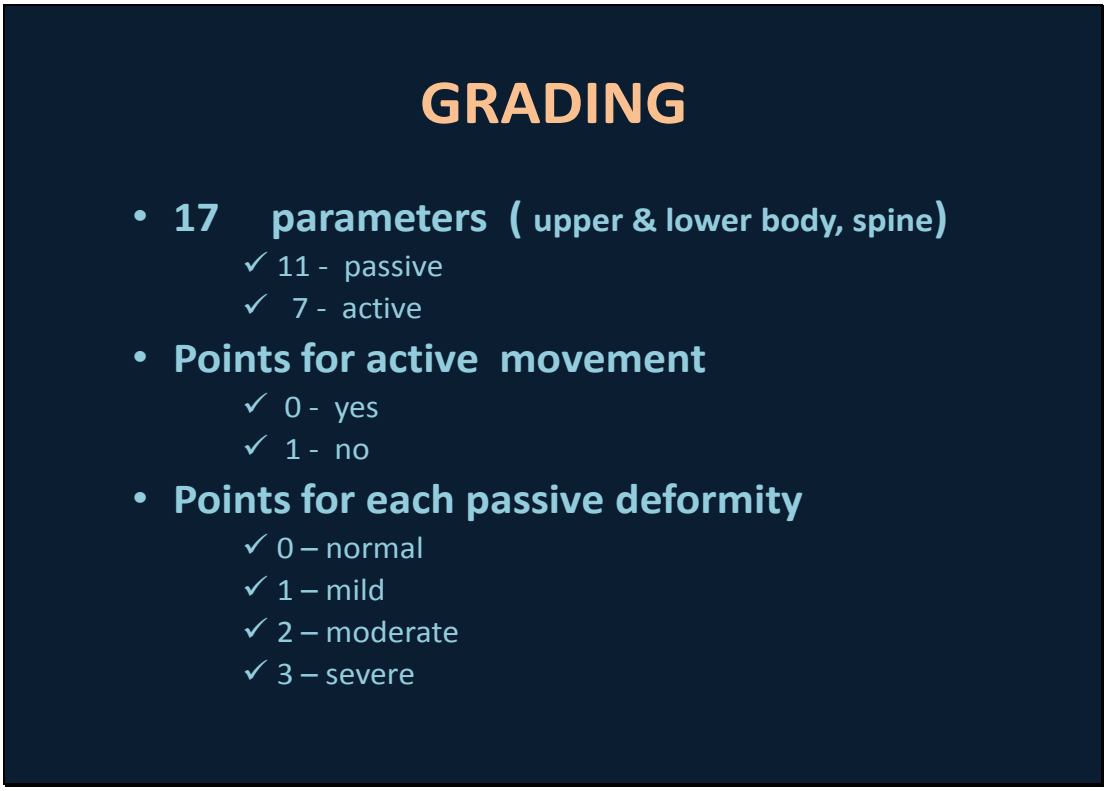
Measurements '0-30-60-90°' are easy to perform during a clinical examination, and can even be done without a handheld goniometer. They are also more reliable and repeatable than precise measurements with 1 degree accuracy, as is needed in the disc-o-gram.



In this example, an examiner assesses contractures: elbow in extension and wrist in palmar flexion:

The elbow can passively be flexed to less than 30°, which is a severe extension elbow contracture, and is the cause for surgical release that is being performed on the child.

When it comes to the wrist, the examiner tries to put the hand passively into the neutral position, and is able to bring it to more than 30 ° from neutral, which, again, is a severe wrist flexion contracture.

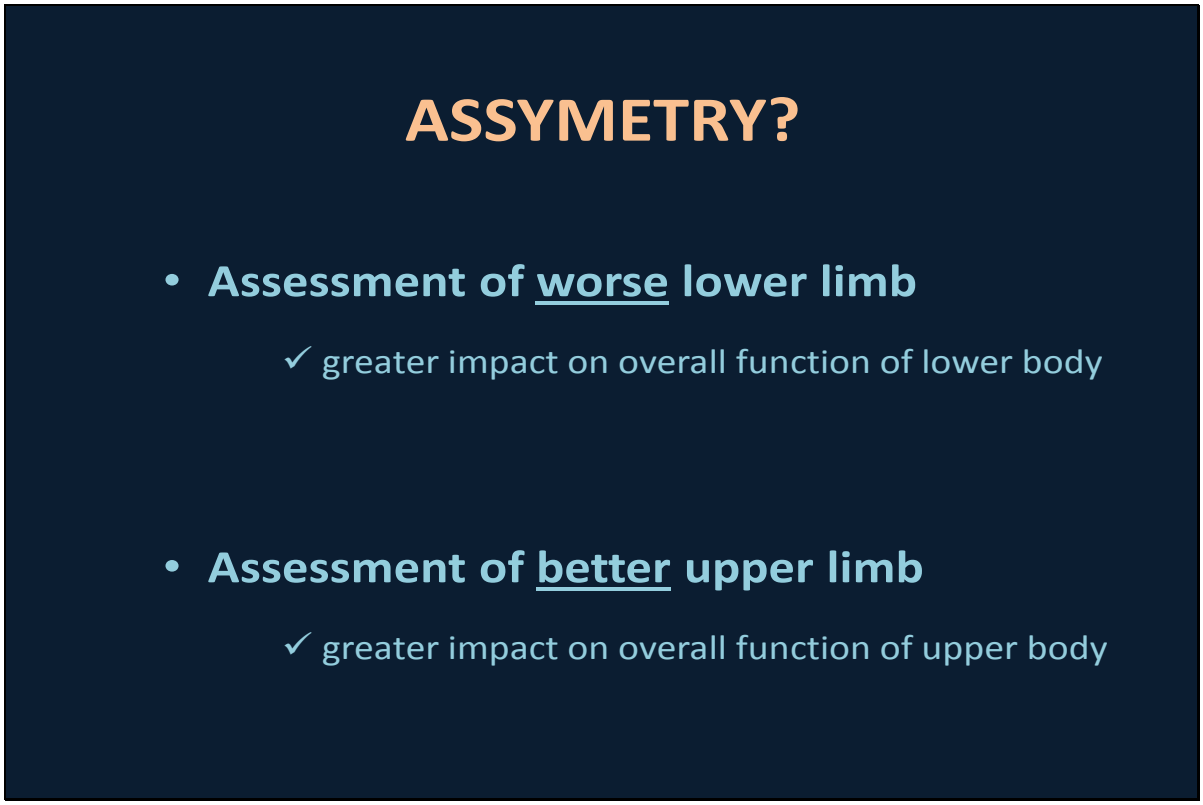


For the whole body classification, we assessed 11 passive parameters (contractures), and 7 active movements.

For active movements, we noted only the presence or absence of active movements, without grading the muscle power or range of active motion.

Although those parameters remain important, to assess them accurately would last very long and require a cooperative child. Therefore, we decided to introduce a 0 to 1 scale for active movement, marking the mere presence or absence of the selected movements.

Each contracture was graded according to the method described earlier, with 0 points for no contracture (normal joint, normal range of passive movements), and 1-3 points for mild, moderate and severe contractures, respectively.



In cases with asymmetry between left and right extremities, which occasionally occurs as a natural course of the disease, or may result from surgical interventions, we decided to assess the WORSE lower limb, as it has a greater influence on the overall lower body function and mobility, and the BETTER upper limb, as, again, more able upper extremity has a greater impact on the overall upper body function.

UPPER LIMB		POINTS
SHOULDER PASSIVE EXTENSION	< 60°	3
	60 - 90°	2
	> 90°	1
SHOULDER ACTIVE EXTENSION?	yes	0
	no	1
ELBOW FLEXION CONTRACTURE (VALUE)	> 60°	3
	30-60°	2
	< 30°	1
ELBOW EXTENSION CONTRACTURE (PASSIVE FLEXION)	< 30°	3
	30-60°	2
	> 60°	1
ACTIVE ELBOW FLEXION?	yes	0
	no	1
WRIST PASSIVE CORRECTION	over 30° from neutral position	3
	between 30° and neutral position	2
	overcorrection	1
FINGERS PASSIVE FLEXION	not possible, fixed contractures	3
	possible but unable to touch palm	2
	able to touch palm	1
ACTIVE FINGERS FLEXION?	yes	0
	no	1
ACTIVE THUMB FLEXION?	yes	0
	no	1

To make the examination easy, we have created a table for each patient's examination, for upper body, lower body and the spine. In the left column selected parameters are presented; the middle one contains the definition of mild/moderate/severe deformities, and on the right, there are points for each deformity, which are added when the examination is completed.

This is the chart for upper limbs.

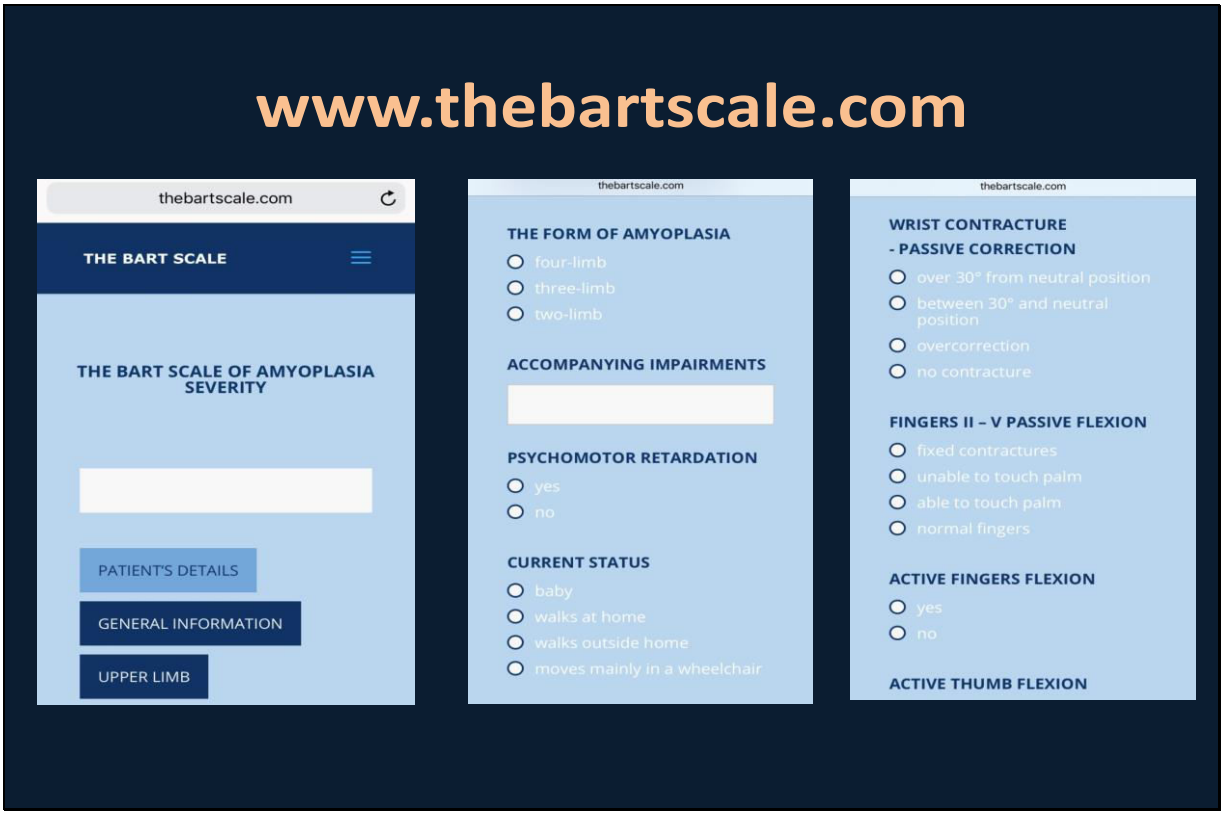
LOWER LIMB & SPINE		POINTS
HIP CONTRATURES	contracture / position over 45°, hip dislocation, passive flexion < 45°	3
	30-45°	2
	< 30°	1
HIP ACTIVE FLEXION?	yes	0
	no	1
KNEE FLEXION CONTRACTURE (VALUE)	> 60°	3
	30-60°	2
	< 30°	1
KNEE EXTENSION CONTRACTURE (PASSIVE CORRECTION)	recurvatum, knee dislocation	3
	passive flexion 0 - 60°	2
	passive flexion > 60°	1
ACTIVE KNEE EXTENSION?	yes	0
	no	1
FOOT	complex deformity (i.e. VT, clubfoot) unsatisactory result	3
	single deformity, satisfactory result	2
	plantigrade, bracable, good result	1
ACTIVE FOOT DORSIFLEXION?	yes	0
	no	1
FIXED SPINE DEFORMITY (SCOLIOSIS, LUMBAR LORDOSIS)		3
NO SPINE DEFORMITY		0

Here is the chart for lower limbs and the spine.

AMYOPLASIA SEVERITY

	MILD	MODERATE	SEVERE	EXTREME
UPPER LIMBS	0-6	7-10	11-13	14-19
LOWER LIMBS & SPINE	0-4	5-8	9-12	13-15
GENERAL ASSESSMENT	0-10	11-17	18-24	25-34

After the examination is completed, the points are added and the severity of the upper limbs, lower limbs and the spine can be calculated. Both upper/lower body and the spine calculations constitute the general assessment and allow for amyoplasia grading into four categories/types of severity: mild, moderate, severe and extreme.



To make the severity assessment and grading even more convenient, we have created an online version of the scale, which is available for free on www.thebartscale.com.

Using the online version, the examiner is guided by a questionnaire, clicking on the selected results of assessment, and the system performs the calculations automatically once the whole examination has been completed. The result can be emailed to the examiner.

METHODS

- **Interobserver study**
- **24 patients graded by 3 investigators**
  - ✓ 17 : four-limb involvement
  - ✓ 6 : upper-limb involvement
  - ✓ 1 : lower-limb involvement
- **Av. age: 64,2 mo. / 5.3 y**
  (range: 2 - 246 mo. / 22y)

To prove reproducibility of the Bart Scale system, we performed an inter-observer study, where 24 patients with amyoplasia, aged (on average) 5.3 years at the time of the examination, were assessed independently, but simultaneously, by three investigators. The team of examiners consisted of orthopaedic surgeons and physical therapists experienced in the treatment of children with amyoplasia.

GRADING ASSESSMENT

- **Kendall's W : concordance**
  - ✓ values 0 to 1; closer to 1 indicate strong agreement
- **Friedman's test : differences**
- **P-value <= 0.05.**

We measured the reproducibility with statistical tools, including Kendall's W coefficient to assess the concordance between the investigators, and Friedman's test to assess differences. P value over 0.05 was considered statistically significant.

RESULTS

ASSESSMENT	NUMBER	DIFFERENCE (%)			
		0	<=1	<=2	<=3
UPPER LIMBS	**24**	**41,7**	**75,0**	**100,0**	**100**
LOWER LIMBS	19	15,8	**73,7**	**100,0**	100
GENERAL	18	16,7	**50,0**	**88,9**	100

ASSESSMENT	KENDALL 'S W	P-VALUE
UPPER LIMBS	0,966	< 0,001 *
LOWER LIMBS	0,963	< 0,001 *
GENERAL	0,986	< 0,001 *

The differences between the raters were frequent, but in most cases they were equal to or less than 2 points, which turned out to be statistically insignificant.

On the other hand, the concordance between raters proved to be significant – Kendall's W coefficient was very close to 1 in all categories, meaning that the concordance was high, making the whole grading reliable.

CONCLUSION

- Fast & easy after short training
- SIFNIFICANT high concordance
- Investigator dependence – INSIGNIFICANT
- Reliable & accurate system

We conclude, therefore, that the proposed system of grading is fast and easy to perform during everyday practice, especially when using the examination chart or the Bart Scale online version, as it takes about 10 minutes to complete after minimal training.

It is characterised by high concordance between the raters. The differences, although they did occur, were insignificant, meaning that the system is investigator-independent.