Disease Diagnostics: Focus on MMT and CMAS

Cure JM Conference
Baltimore, MD
Oct 12, 2012

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Assessment of Muscle Strength

- Various methods for assessing strength:
  - Manual muscle testing
  - Quantitative muscle testing
  - Hand held dynamometry
  - Functional tasks

Manual Muscle Testing

Quantitative Muscle Testing

Hand Held Dynamometry

Functional Tasks

Advantages of Each

- Manual muscle testing: ease of administration, can be administered at any location with any level of strength
- Quantitative muscle testing: very objective and reproducible
- Hand held dynamometry: objective, easy to use with children
- Functional tasks: able to test younger children

Disadvantages of each

- Manual muscle testing: subjective, moderate to low inter-rater reliability for individual muscles, esp with mild weakness
- Quantitative muscle testing: very expensive equipment, time consuming, children may not cooperate
- Hand held dynamometry: expensive, poor inter-rater reliability, children have difficulty understanding instructions
- Functional tasks: difficult to quantify changes, not directly measuring strength

Why MMT?

- Widely used in myositis clinical trials as part of primary endpoint
- Commonly and easily used in clinics to follow patient progress, responses to therapy
- Accepted by rheumatologists and neurologists
- Validated tool:
  - Excellent internal reliability
  - Very good to excellent inter- and intra-rater reliability (total scores, not individual muscles)
  - Good construct validity: correlation with other measures of myositis activity
  - Excellent sensitivity to change

Potential Pitfalls of MMT

- Insensitive in detecting mild weakness
- Not for kids < 4 years of age
- Expanded scale commands not completely standardized, defined
- Number of influences on performance
  - Examiner:
    - Allowing patient compensatory strategies
    - Improper positioning
    - Skills of tester: amount of force applied, judgment, feedback
  - Patient:
    - Steroid myopathy, joint contractures, calcinosis, arthritis, myalgias may interfere with strength assessment
    - Comprehension and motivation, effort
    - Motor skill, strength
- Muscle strength does not distinguish disease activity from damage
  - Limited information on clinical meaning of scores

Manual Muscle Testing Procedures

Key to Muscle Grading

	Function of the Muscle	Grade		
No Movement	No contractions felt in the muscle	0	0	Zero
	Tendon becomes prominent or feeble contraction felt in the muscle, but no visible movement of the part	T	1	Trace
Test Movement	**MOVEMENT IN HORIZONTAL PLANE**			
	Moves through partial range of motion	1	2-	Poor-
	Moves through complete range of motion	2	2	Poor
	ANTIGRAVITY POSITION	3	2+	
	Moves through partial range of motion			
Test Position	*Gradual* release from test position	4	3-	Fair-
	Holds test position (no added pressure)	5	3	Fair
	Holds test position against slight pressure	6	3+	Fair+
	Holds test position against slight to moderate pressure	7	4-	Good-
	Holds test position against moderate pressure	8	4	Good
	Holds test position against moderate to strong pressure	9	4+	Good+
	Holds test position against strong pressure	10	5	Normal

Modified from 1993 Florence P. Kendall. Author grants permission to reproduce this chart.

Standardization of the MMT for IMACS: Total MMT24

Muscle Groups	TESTING POSITIONS Anti-Gravity	Gravity Eliminated
Trapezius (shoulder elevators)	Sitting	Supine
Deltoid middle (shoulder abductors)	Sitting	Supine
Biceps brachii (elbow flexors)	Sitting	Sidelying
Wrist extensors	Sitting (pronation)	Sitting (neutral)
Wrist flexors	Sitting (supination)	Sitting (neutral)
Iliopsoas (hip flexors)	Sitting	Sidelying
Quadriceps femoris (knee extensors)	Sitting	Sidelying
Ankle dorsiflexors	Sitting	Sidelying
Neck flexors	Supine	Sidelying
Gluteus medius (hip abductors)	Sidelying	Supine
Neck extensors	Prone	Sidelying
Gluteus maximus (hip extensors)	Prone	Sidelying
Hamstrings (knee flexors)	Prone	Sidelying
Ankle plantarflexors	Prone/Standing	Sidelying

http://www.niehs.nih.gov/research/resources/assets/docs/muscle_grading_and_testing_procedures.pdf

Abbreviated MMT: MMT8

MMT8 is a set of 8 designated muscles tested unilaterally (score range 0-80):

Axial muscles

- Cervical flexion (Neck flexors)

Proximal muscles

- Shoulder abduction (Deltoids)
- Elbow flexion (Biceps)
- Hip extension (Gluteus maximus)
- Hip abduction (Gluteus medius)
- Knee extension (Quadriceps)

Distal muscles

- Ankle dorsiflexion (Tibialis anterior)
- Wrist extension (Extensor carpii ulnaris and radialis)

Validation studies in adult and juvenile DM/PM have shown that unilateral MMT8 was comparable to bilateral MMT24

Much shorter time needed for testing (< 5-10 minutes); doctors and physical therapists able to perform in clinic

Video Clips of MMT8

Manual Muscle Test Considerations for Assessing Strength in Myositis

http://www.niehs.nih.gov/research/resources/collab/imacs/diseaseactivity/index.cfm

Childhood Myositis Assessment Scale (CMAS)

- Observational performance-based instrument of 14 functional tasks to assess muscle endurance, muscle function, strength
- Weighted more towards proximal and axial muscles, lower > upper extremities (weakest muscles weighted more)
- Scores range from 0-52
- Used in children of all ages, but durational items and situps best obtained in children ≥ 4 years
- Normal values for CMAS available for 9 items, ages 4-9 years
- Trained observers; potentially more objective than other functional assessment tools (CHAQ)
  - Potentially even parents can do at home

Childhood Myositis Assessment Scale (CMAS)

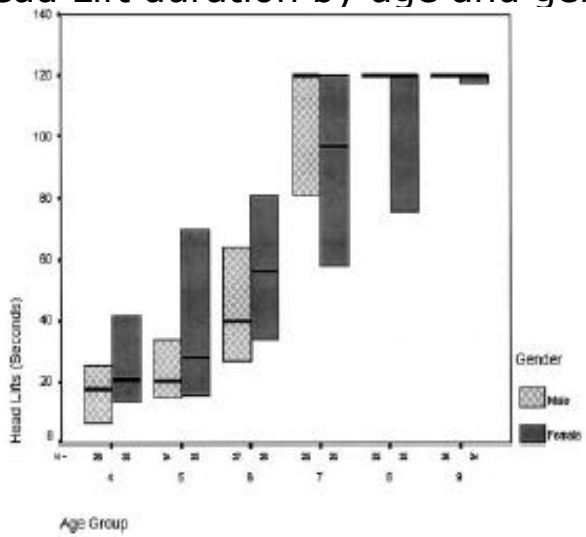
- Widely used in clinics and clinical studies
  - Accepted by pediatric rheumatologists as a core outcome measure
- Well validated
  - Good content validity: developed by Robert Rennebohm, Carol Lindsley and a group of pediatric rheumatologists
  - Very good to excellent inter- and intra-rater reliability
  - Good construct validation: correlation with other myositis activity measures
  - Very good - excellent sensitivity to change

Childhood Myositis Assessment Scale (CMAS)

- Clinical meaning of scores and changes in scores – some data
- Potential pitfalls:
  - 15 minutes to perform (not practical for every clinic visit)
  - Does not discriminate disease activity from damage
    - Joint contractures, calcinosis may result in fixed deficits
  - Patient motivation, cooperation, esp for youngest kids
  - Need some equipment: stop watch, step stool & chair of age-appropriate heights, floor mat

Younger children have difficulty performing head lift and sit-ups

Head Lift duration by age and gender



Sit-ups by age and gender

Table 4. Sit-ups: percentage of each age group that achieved each score

Age group	0	1	2	3	4	5	6
4 years, boys	42	8	19	15	8	0	8
4 years, girls	40	0	16	28	8	4	4
5 years, boys	12	4	20	20	28	4	12
5 years, girls	4	4	16	20	20	24	12
6 years, boys	0	0	8	11	23	27	30
6 years, girls	0	4	4	27	31	23	11
7 years, boys	0	0	0	16	4	20	60
7 years, girls	4	0	4	8	16	20	48
8 years, boys	0	0	0	4	4	24	68
8 years, girls	0	0	8	16	8	28	40
9 years, boys	0	0	8	0	0	19	73
9 years, girls	0	0	0	13	8	8	71

Normal CMAS scores by age and gender

Table 5. Normal CMAS-9 scores*

Age	Girls score (% of maximum)	Boys score (% of maximum)	Maximum
4	27 (73)	26 (70)	37
5	30 (81)	25 (68)	37
6	31 (84)	31 (84)	37
7	32 (86)	32 (86)	37
8	33 (89)	35 (95)	37
9	32 (86)	35 (95)	37

* Ninty percent of healthy children are able to achieve these scores at these ages. CMAS-9 = Childhood Myositis Assessment Scale.

Rennebohm et al., 2004 Arthritis Care and Res

1. **HEAD LIFT:**
0 = Unable 3 = 30-59
1 = 1-9 sec 4 = 60-119 sec
2 = 10-29 5 = ≥ 2 min # of sec ____

2. **LEG RAISE/TOUCH OBJECT:**
0 = Unable to lift leg off table.
1 = Able to clear table, but cannot touch object (examiner's hand).
2 = Able to lift leg high enough to touch object (examiner's hand).

3. **STRAIGHT LEG LIFT/DURATION:**
0 = Unable 3 = 30-59 sec
1 = 1-9 sec 4 = 60-119 sec
2 = 10-29 sec 5 = ≥ 2 min #of sec ____

4. **SUPINE TO PRONE:**
0 = Unable. Has difficulty even turning onto side; able to pull right arm under torso only slightly or not at all.
1 = Turns onto side fairly easily, but cannot fully free right arm and is unable to fully assume a prone position.
2 = Easily turns onto side; has some difficulty freeing arm, but fully frees arm and fully assumes a prone position.
3 = Easily turns over, fully frees right arm with no difficulty.

5. **SITS-UPS:**
Hands on thighs, with counterbalance ____
Hands across chest, with counterbalance ____
Hands behind head, with counterbalance ____
Hands on thighs, without counterbalance ____
Hands across chest, without counterbalance ____
Hands behind head, without counterbalance ____
Total Sit-up Score (0-6) ____

6. **SUPINE TO SIT:**
0 = Unable by self.
1 = Much difficulty. Very slow, struggles greatly, barely makes it. Almost unable.
2 = Some difficulty. Able, but is somewhat slow, struggles some.
3 = No difficulty.

7. **ARM RAISE/STRAIGHTEN:**
0 = Cannot raise wrists up to the level of the A-C joint.
1 = Can raise wrists at least up to the level of the A-C joint, but not above top of head.
2 = Can raise wrists above top of head, but cannot raise arms straight above head so that elbows are in full extension.
3 = Can raise arms straight above head so that elbows are in full extension.

8. **ARM RAISE/DURATION:** Can maintain wrists above top of head for:
0 = Unable 3 = 30-59 sec
1 = 1-9 sec 4 = ≥ 60 sec
2 = 10-29 sec #of sec _____

The **maximum possible total score** for the 14 maneuvers is **52** (52 "points of muscle strength/function").

9. **FLOOR SIT:** Going from a standing position to a sitting position on the floor:
0 = Unable. Afraid to even try, even if allowed to use a chair for support. Child fears that he/she will collapse, fall into a sit, or harm self.
1 = Much difficulty. Able, but needs to hold onto a chair for support during descent. Unable, or unwilling to try if not allowed to use a chair for support.
2 = Some difficulty. Can go from stand to sit without using a chair for support, but has at least some difficulty during descent. May need Gower's. Descends somewhat slowly and/or apprehensively; may not have full control or balance as maneuvers into a sit.
3 = No difficulty. Requires no compensatory maneuvering.

10. **ALL FOURS MANEUVER:**
0 = Unable to go from a prone to an all-fours position.
1 = Barely able to assume and maintain an all-fours position. Unable to raise head to look straight ahead.
2 = Can maintain all-fours position with back straight and head raised (so as to look straight ahead). But, cannot creep (crawl) forward.
3 = Can maintain all-fours, look straight ahead and creep (crawl) forward.
4 = Maintains balance while lifting and extending one leg.

11. **FLOOR RISE:** Going from a kneeling position on the floor to a standing position:
0 = Unable, even if allowed to use a chair for support.
1 = Much difficulty. Able, but needs to use a chair for support. (Unable if not allowed to use a chair.)
2 = Moderate difficulty. Able to get up without using a chair for support, but needs to place one or both hands on thighs/knees or floor. (Unable without using hands.)
3 = Mild difficulty. Does not need to place hands on knees, thighs or floor, but has at least some difficulty during ascent.
4 = No difficulty.

12. **CHAIR RISE:**
0 = Unable to rise up from chair, even if allowed to place hands on sides of chair seat.
1 = Much difficulty. Able, but needs to place hands on sides of seat. Unable if not allowed to place hands on sides of seat.
2 = Moderate difficulty. Able, but needs to place hands on knees/thighs. Does not need to place hands on sides of seat.
3 = Mild difficulty. Does not need to place hands on seat, knees or thighs but has at least some difficulty during ascent.
4 = No difficulty.

13. **STOOL STEP:**
0 = Unable.
1 = Much difficulty. Able, but needs to place one hand on exam table (or examiner's hand).
2 = Some difficulty. Able, does not need to use exam table for support, but needs to use hand on knee/thigh.
3 = Able. Does not need to use exam table or hand on knee/thigh.

14. **PICK-UP:**
0 = Unable to bend over and pick up pencil off floor.
1 = Much difficulty. Able, but relies heavily on support gained by placing hands on knees/thighs.
2 = Some difficulty. Has some difficulty (but not "much-difficulty"). Needs to at least minimally and briefly place hand(s) on knees/thighs for support. Is somewhat slow.
3 = No difficulty. No compensatory maneuver necessary.

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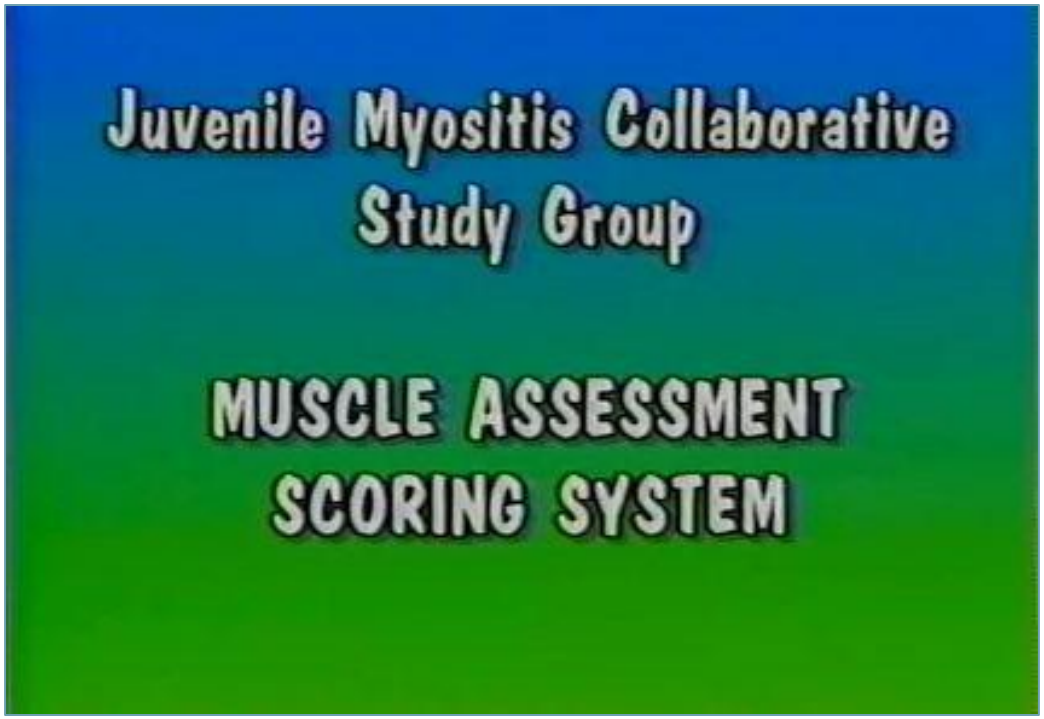
CMAS VIDEO CLIPS

Demonstration of the CMAS and its scoring



http://www.niehs.nih.gov/research/resources/collab/imacs/diseaseactivity/index.cfm

Acknowledgments

- EAG, NIEHS and Rehab Medicine, CC, NIH:
  - Frederick Miller, Michael Harris-Love, Joe Shrader, Michaele Smith, Galen Joe, Jeanne Hicks
- Collaborators:
  - Robert Rennebohm, Carol Lindsley, Daniel Lovell, Adam Huber, JDM Disease Activity Collaborative Group
- IMACS Group and PRINTO
- Our patients and their families
- Our funding/sponsors:

NIEHS National Institute of Environmental Health Sciences — *National Institute of Environmental Health Sciences*

National Institute of Arthritis and Musculoskeletal and Skin Diseases

The NIH Clinical Center — *NIH Clinical Center*

THANKS!

QUESTIONS?