

APPENDIX A:

STANDARDIZED METHOD FOR CONDUCTING THE CHILDHOOD MYOSITIS ASSESSMENT SCALE (CMAS)

Description of Maneuvers:

The 14 CMAS maneuvers are to be assessed, one after the other, in the order listed on the CMAS Scoring Sheet. (Please see "CMAS Scoring Sheet" and the ERRATUM at the end of this document.)

1. Head elevation (neck flexion): The supine patient is asked to lift his/her head off the examination table for as long as possible (up to 120 seconds). The arms are to be kept still at the patient's sides and are not to be used for support or leverage. The shoulders are to remain stationary. The patient receives credit for raising his/her head off the table if the examiner can freely slide his/her fingers under the patient's occiput. The examiner begins timing from the moment the patient raises his/her head off the table. Timing stops when the patient can no longer keep his/her head off the table and the occiput touches the table, or after 120 seconds have elapsed, whichever comes first. The patient is not required to maintain any particular degree (angle) of neck flexion while the head is off the table.

A score of zero is warranted if the patient is unable to raise his/her head off the exam table so that the examiner can freely slide his/her fingers under the patient's occiput. A patient who is able to lift his/her head off the table for only 1-9 seconds receives a score of 1, and so on (see Scoring Sheet). (If the patient can lift his/her head at least long enough for the examiner to slide his/her fingers under the occiput, the patient will be said to have raised his/her head for at least 1 second.)

2. Leg raise/touch object: The patient is supine with legs extended. In preparation for this maneuver, the examiner passively raises the patient's right foot and rests the heel of the patient's right dorsiflexed foot on top of the extended first toe of the left dorsiflexed foot to measure a distance of 2 "patient foot lengths" above the exam table. Having made this measurement, the examiner replaces the right leg/heel in a position of rest on the table.

The examiner then holds his/her hand or an object (a toy for younger children) 2 patient foot lengths above the patient's resting right heel and asks the child to raise his/her leg so that the right first toe touches the examiner's hand (or an object). The child may use a kicking motion to accomplish this, if necessary. That is, the leg need not be kept straight throughout ascent. However, the pelvis must be kept stationary in a neutral position.

A score of 0 is warranted if the patient is unable to lift the right leg so that it clears the table. A score of 1 is awarded if the patient is able to lift the leg high enough so that it at least slightly clears the table, but not high enough to touch the examiner's hand (or object). A score of 2 is awarded if the patient is able to lift the leg high enough to touch the examiner's hand (or object). Once it is determined whether the patient is able to touch the object, the patient rests the leg for a few seconds before starting maneuver number 3.

3. Straight leg lift/duration: The supine patient is asked to lift his/her straightened right leg off the exam table so that the heel of the right foot is 1 "patient foot length" (the length of the patient's sole, from the posterior heel to the tip of the first toe) above the exam table. The patient is asked to maintain this leg lift for as long as possible (up to 120 seconds). Counting starts as soon as the heel is lifted to the above-mentioned height. During the count, the patient must constantly keep the knee in extension and should be discouraged from raising the heel higher than 1 patient foot length above the table. The pelvis must remain stationary. During the count, the heel might drift downward; when this occurs the count continues, but the patient should be encouraged (repeatedly, if necessary) to again raise the heel to the desired height. Counting stops when the patient can no longer maintain the straightened leg off the table and the heel touches the table, or after 120 seconds have elapsed.

Zero points are awarded if the patient cannot even momentarily lift the straightened leg to the desired height. One point is awarded if the patient can lift the straightened leg to the desired height, but can maintain the leg lift for only 1-9 seconds, and so on (see Scoring Sheet).

4. Supine to prone: The supine patient is asked to roll over to the right into a prone position. Throughout the maneuver, he or she must keep the right arm flexed, and he/she must keep the right wrist inferior to the chin. First, the patient is to roll onto his/her right side, onto the flexed right arm, thereby pinning the flexed right arm against the table. As the patient continues to roll toward a prone position, he/she must pull the flexed right arm out from under his/her torso and free it as he/she finally rolls into a full prone position.

The profoundly weak patient has difficulty even rolling onto his/her side, and is able to pull the right arm under the torso only slightly or not at all. Such a patient receives 0 points. A slightly stronger patient (score of 1) can roll onto his/her side fairly easily, can pull the right arm much of the way under the torso, but is not able to fully free the arm and, therefore, is unable to fully assume a prone position. Such a patient's right arm remains pinned against the exam table and goes no further. A mildly weak patient (score of 2) easily turns onto his/her side, has some difficulty pulling the right arm under the torso, but does so, and fully achieves the prone position. The patient who performs this maneuver with no difficulty is awarded 3 points.

5. Sit-ups: The patient is asked to perform sit-ups in 6 different ways in the following order:

- 1) The patient is supine on the examination table, keeps hips and knees in full extension, and places his/her palms on his/her upper thighs. The examiner holds the patient's ankles firmly against the exam table ("counterbalancing"). The patient then does a single sit-up, without using the hands or elbows for added support or boosting. During the sit-up, the palms must remain close to (but not clutching) the thighs and the elbows must not touch the exam table. A point is awarded if the child is able to fully complete the sit-up (i.e. assume a full sitting position, with the spine perpendicular to the exam table.)
- 2) Same as number 1, except the patient holds his/her arms folded across the chest (with each hand resting on the opposite shoulder) throughout performance of the sit-up. The elbows may swing away from the chest during the sit-up, but the hands must remain in contact with the shoulders.
- 3) Same as number 1, except the patient holds his/her hands clasped behind the neck/occiput throughout the sit-up.
- 4, 5, and 6) Same as 1, 2, and 3, respectively, except the examiner does not provide counterbalancing.

One point is awarded for each sit-up accomplished.

6. Supine to sit: The patient is asked to go from a supine position on the exam table to a sitting position in which he or she is seated on the exam table, with the legs freely dangling over the side of the table. The hands and arms may be used in any way necessary to achieve this. The profoundly weak patient is unable to get into a sitting position without someone's help and receives 0 points. A slightly stronger individual (score of 1) is barely able to get into such a sitting position, struggles greatly, and takes a long time to do so. A score of 2 is awarded if the patient struggles somewhat and is somewhat slow but does not just "barely make it." A score of 3 is awarded if the patient has no difficulty.

7. Arm raise/straighten: The seated patient is asked to simultaneously raise both arms straight above the head so that the wrists are as high as possible above the head. (The younger patient is asked to "reach to the sky; raise your hands as high as you can." A ball or toy can be held high above the child's head, and the child can be asked to reach or grasp the object with both hands.)

Zero points are awarded if the patient cannot raise his/her wrists up to the level of the acromioclavicular (AC) joint. One point is awarded if the patient can raise the wrists at least up to the level of the AC joint, but not above the top of the head. Two points are awarded if the patient can raise the wrists above the top of the head, but cannot raise the arms straight above the head so that the elbows are in full extension. Three points are awarded if the patient can raise his/her arms straight above the head, so that the elbows

are in full extension and the straightened arms are roughly perpendicular to the exam table. Once it is determined whether the patient can raise his/her arms to the fully straightened position, the patient then rests his/her hands in his/her lap for a few seconds before starting maneuver 8.

8. Arm raise/duration: The seated patient is asked to simultaneously raise both hands from his or her lap to a position in which the wrists are as far as possible above the lap (ideally, as high as possible above the head). The patient is then asked to maintain the wrists in the highest position possible for as long as possible, or until 60 seconds have elapsed.

Zero points are awarded if the patient cannot even momentarily raise his/her hands so that the wrists are above the level of the top of the head. One point is awarded if the wrists can be raised above the top of the head, but can be maintained in this position for only 1-9 seconds. Two points are awarded if the wrists can be maintained above the top of the head for only 10-29 seconds, and so on (see Scoring Sheet). Counting starts as soon as the wrists are lifted above the top of the head, and counting stops as soon as the wrists fall below the level of the top of the head (or until 60 seconds elapse). Throughout the counting, the child should be encouraged to keep the forearms perpendicular to the exam table, but counting continues as long as the wrists remain above the level of the top of the head, even if the forearms drift out of the perpendicular plane. Throughout the counting, the patient is encouraged to keep the wrists as high as possible, but it is to be understood that the patient may not be able to raise the arms so that the elbows achieve full extension. Throughout counting, the cervical spine should be kept in a neutral position.

9. Floor sit: The patient is asked to stand alone in the middle of the examination room, away from any potentially supporting chairs or tables. The patient is then asked if he/she thinks he/she can safely descend into a sitting position on the floor without using a chair for support. If the patient is hesitant to try, he/she should not be encouraged to try. Such a patient should then be offered a chair to use for support during descent.

The profoundly weak patient (score of 0) would not be able to go from a standing position to a sitting position on the floor without simply falling into a sit (which is not allowed or encouraged). Such a patient is afraid to try and refuses to try, even if allowed to hold onto a chair during descent. The slightly stronger patient (score of 1) is willing to try if allowed to use the chair for support and, with the availability of the chair, is able to descend into a sitting position. He/she is not able to safely descend without the use of the chair. A score of 2 is awarded if the patient can safely descend unassisted (without use of a chair for support), but has at least mild difficulty doing so. Such a patient descends abnormally slowly, does not have full control over the muscles or full balance as he/she descends, and/or excessively relies on supporting him/herself by placing his/her hands on his/her thighs, knees, or floor as he/she descends. The patient who requires no compensatory maneuvering and has no difficulty descending receives a score of 3.

10. All-fours maneuver: The patient begins this maneuver while in a prone position on the floor. The patient is then asked to rise up to an all-fours position, so that he or she is bearing all weight on the hands and knees. Once assuming this position, the patient is asked to keep the back straight and to raise the head up so that he/she can look straight ahead (with the plane of the face nearly perpendicular to the floor). Then, the patient is asked to creep (crawl) forward so that all 4 weight-bearing points are moved to new positions.

A score of 0 is warranted if the patient is unable to go from a prone position to an all-fours position. That is, the patient is too weak to rise up into a weight-bearing position on his/her hands and knees. The patient is unable to even momentarily assume an all-fours position. A score of 1 is warranted if the patient is able to assume an all-fours position, but only barely and/or weakly so. The patient achieves the all-fours position, but is unable to raise the head to look straight ahead. A score of 2 is warranted if the patient can achieve and maintain an all-fours positions, with back straight and head raised; but the patient cannot creep (crawl) forward. A score of 3 is awarded if the patient can solidly maintain an all-fours position, raise head, look straight ahead, and creep (crawl) forward. A score of 4 is awarded if the patient,

while in an all-fours position, is able to maintain balance while raising the head and extending and lifting one leg above the level of the body.

11. Floor rise: The patient is seated on the floor, away from any chair or table. The patient is then asked to get into a kneeling position. He or she may place his/her hands on the floor for support as he/she assumes this kneeling position. The patient is then asked to remain kneeling on the right knee, but to raise the left knee so that the left foot is planted in front of him/her, with the left knee and left hip each in 90 degrees of flexion. The patient is then asked to rise from this kneeling position to a standing position without using a chair for support and without placing his/her hand(s) on the knee(s), thigh(s), or the floor, if possible. A chair is provided if the child is unable to rise without the use of a chair for support.

A score of 0 is warranted if the child is unable to rise to a stand even if allowed to place his/her hands/forearms on a chair for support. A score of 1 is awarded if a patient is able to rise, but only if using a chair for support. A score of 2 is awarded if the patient is able to rise to a stand without use of a chair, but needs to place one or both hands on the knees/thighs or floor during ascent. A score of 3 is awarded if the child can ascend without placing his/her hand(s) on the knee(s), thigh(s), or floor, but has at least some difficulty during ascent (struggles and/or ascends slowly). A score of 4 is awarded if the patient has no difficulty during ascent.

12. Chair rise: The patient is seated in an arm-less chair that is of “appropriate size” for the patient’s age. (When a person sits in a chair of “appropriate size “ and sits with the lower legs planted perpendicular to the floor, the distal thigh will be slightly higher than the proximal thigh). The feet are to be placed any distance apart, but the toes must be kept pointing forward (no out-toeing). The patient is allowed to rock forward, if necessary, during ascent.

A score of 0 is warranted if the child is unable to rise to a stand, even if allowed to place the hands on the sides of the chair seat. A score of 1 is awarded if the child is able to rise to a stand, but needs to place the hand(s) on the side(s) of the seat in order to do so. A score of 2 is awarded if the child is able to stand up, but needs to place one or both hands on the knee(s) or thigh(s) to do so. A score of 3 is awarded if the child does not need to use his/her hands at all as he/she ascends to a stand, but has at least some difficulty during ascent (is slow, struggles somewhat, and/or needs to rock). A score of 4 is awarded if the child has no difficulty going from a sit to a stand.

13. Stool step: An “age appropriate “ stool is placed next to the exam table, and the patient is asked to step up onto the stool. An “age appropriate” stool has a height equal to approximately 1/3 of the distance from the plantar aspect of a child’s heel to that child’s mid-patella. For an older child this would be a stool that is 6-8-inches (15-20 cm) high. The patient is encouraged to try to step onto the stool without placing a hand on the exam table (or on the examiner’s hand/forearm for a shorter child) for support, and without placing a hand (or hands) on her stepping knee/thigh for support.

A score of 0 is awarded if the child is unable (or appropriately unwilling to try) to step onto the stool even when allowed to place one hand on the exam table (or on the examiner’s hand/forearm) for support. (Such a patient would be afraid of losing his/her balance if not allowed to place a hand on the table for support).

A score of 1 is awarded if the child is able to step onto the stool, but needs to place one hand on the exam table (or on the examiner’s hand/forearm) in order to do so. A score of 2 is awarded if the patient is able to step onto the stool without placing a hand on the exam table (or examiner’s arm) but needs to place his/her hand(s) on the knee/thigh in order to do so. A score of 3 is awarded if the patient is able to step onto the stool without placing his/her hand(s) on either the exam table or his/her stepping thigh/knee.

14. Pick-up: The patient stands in the middle of the exam room and is asked to bend over to pick up a pen or pencil off the floor and to return to an erect standing position.

A score of 0 is warranted if the patient is unable to pick up the pencil and return to an erect standing position. A score of 1 is awarded if the patient is able to pick up the pencil and return to a standing position, but relies heavily on support gained by placing his/her hand(s) on the knees/thighs and is barely able to perform the maneuver. A score of 2 is awarded if the patient has some difficulty, but not extreme difficulty, i.e., if the patient needs to at least briefly place his/her hand(s) on the knees/thighs for support and/or is at least somewhat slow in performing the maneuver. A score of 3 is awarded if the patient has no difficulty, requires no compensatory maneuvering and performs the maneuver quickly.

Maximum Possible Score for the 14 Maneuvers is **52 points** (52 “points of muscle strength/function”).

ERRATUM: There is an important discrepancy between the CMAS Scoring Sheet found on this web-site and the scoring sheet published in the original CMAS article (*Lovell DJ, Lindsley CB, Rennebohm RM, et al. Development of validated disease activity and damage indices for the juvenile idiopathic inflammatory myopathies: The childhood myositis assessment scale (CMAS). Arthritis Rheum 1999; 42: 2213-2219*). The discrepancy is due to mis-prints in that article regarding the scoring of maneuvers 4, 8, and 12 (see Tables 2 and 3 in the 1999 article). The authors of the 1999 article acknowledge the mis-prints in that article and fully agree that the “CMAS Scoring Sheet” and the “Description of Maneuvers” found on this web-site are correct.