

DISEASE ACTIVITY SCORE (DAS) INSTRUCTIONS

OVERVIEW: THE DAS IS A VALIDATED INSTRUMENT WITH A TOTAL SCORE 20. THIS INSTRUCTION SHEET INCLUDES DATA THAT MAY BE USED IN THE NEXT VERSION.

A) NECK FLEXOR STRENGTH

1. The examiner's hand is placed on the child's forehead
2. The child is asked to lift her head from the examining table
3. The examiner's (placing the effort in the appropriate age range) evaluates the strength of the effort. The number of cm (back of head to mat) is recorded.

The MRC muscle strength score is also recorded.

B) SIT UP (DIFFICULTY CLEARING SCAPULA)

The child is asked to perform a sit up alone, without any counterbalance. Lack of age appropriate performance is counted as "weak" for the scoring. *Any deviation from age-appropriate performance is scored as 1.* There are 5 levels of evaluation in performing a sit up without counterbalance (the level is checked in a box):

1. The child is unable to clear the scapula off the exam table when the child tries to do a sit.
2. The child clears 1/3 or less of the scapula. That is, 1/3 or less of the scapula is visible to the examiner when the child tries to do a sit up without counterbalance. The child has enough abdominal strength to lift head and neck from the table (arms extended), but not to elevate the upper thorax more than top 1/3 of the scapula (usually about 10-20 cm from top of table). This type of sit up is often followed by rolling to one side, leaning on an elbow to get up.
3. The child is able to sit up with arms extended.
4. The child can perform a sit up with arms crossed on torso.
5. The child can sit up with hands behind the head.

C) UPPER PROXIMAL STRENGTH

1. The child sits on the examining table with the arms extended to the side at shoulder height and the examiner places the hands between the shoulder and the elbow and attempts to lower arms.
2. The child holds the arm in front of body with elbows flexed, and the examiner attempts to extend the arm. The MRC muscle strength score is also recorded.

D) LOWER PROXIMAL STRENGTH

1. The child lies on back on the table and raises leg so that the foot is at least 18° from mattress.
2. The examiner places hand midway between the hip and the knee and attempts to push the leg down on the mattress. The MRC muscle strength score is also recorded.

E) GOWER'S SIGN

The child sits on the floor (making sure that the examining gown is not in the way) and is asked to hold her hands at ear level and get up. Any unsteady maneuver or touching the body or the room's equipment (floor included) is noted and considered an "assisted" maneuver.

F) ABNORMAL GAIT

Any deviation from normal gait is recorded as 1.

G) DIFFICULTY SWALLOWING:

The child is asked if he had difficulty swallowing a cracker, and the answer is recorded.

H) NASAL SPEECH

The child is asked to say the alphabet up to the letter "E", and to say the letter "E" three times. The quality of speech is noted assigning 1 to any deviation from normal.

I) ATROPHIC CHANGES

Skin atrophy or thinning, in the absence of erythema.

J) ERYTHEMA

Overall degree and severity of inflammation and induration.

1. Atrophic changes only: dermal or epidermal cutaneous atrophy, with or without telangiectasia, in the absence of erythema.
2. Mild erythema: pale pink, no induration,
3. Moderate erythema: brighter pink to red, with or without induration.
4. Severe erythema: more intense pink to red with induration.

K) EYELID ERYTHEMA

Red/purple discoloration of the eyelid.

L) EYELID BLOOD VESSEL DILATION

Small vessels apparent at the margin of the eyelid.

M) EYELID BLOOD VESSEL TELANGECTASIA

Greater degree of eyelid vessel dilation, involving the middle portion of the eyelid.

N) NAILFOLD ERYTHEMA (CUTICLES)

Periungual erythema.

O) NAIL BED TELANGIECTASIA

Dilation of the small vessels adjacent to the edge of the nail, which includes end row capillaries and other vessels.

P) PALATE VESSEL DILATION

Dilated blood vessels evident on the hard palate.

Q) GOTTRON'S PAPULES

1. Mild Gottron's papules: a few areas (~2.0 cm or less) of localized cutaneous papules, usually skin color.
2. Moderate Gottron's papules: several locations of cutaneous involvement, medium sized papules, light to medium pink in color.
3. Severe Gottron's papules: Bright red, extensive involvement and larger papules, most often over multiple areas, including the MCP, PIPs, elbows, knees.

To calculate the **TOTAL DAS SCORE** for a JDM patient it is necessary to add points for two components:

- Muscle weakness
- Skin involvement

Muscle Weakness

1. Functional status
2. 0-3 points possible
3. Weakness
4. 0 or 1 point FOR EACH of the 8 categories; 8 total

RANGE OF POSSIBLE POINTS FOR MUSCLE WEAKNESS: 0 TO 11

Skin involvement

3. Type 0-4 points
4. Distribution 0-3 points
5. Vasculitis 0 or 1 point, IF ANY are "present"
6. Gottron's papules 0 or 1 point, IF ANY is "present"

RANGE OF POSSIBLE POINTS FOR SKIN INVOLVEMENT: 0 TO 9

To obtain the **TOTAL DAS SCORE**: add the two components muscle weakness (range 0 to 11) and skin involvement (range 0 to 9).

RANGE OF POSSIBLE POINTS FOR THE TOTAL DAS SCORE: 0 TO 20