

Assessment of Hand Contractures in Epidermolysis Bullosa (ACE)

What is the ACE?

The ACE is a hand assessment developed for use with children and young people with Recessive Dystrophic Epidermolysis Bullosa (RDEB). It may also be used with adults with RDEB. It is intended to be used by hand therapists to systematically assess hand contractures typically seen in this condition. The ACE can be used routinely to monitor changes over time. It may also be used to establish a baseline prior to hand surgery and as an outcome measure following surgery.

How do I use it?

The assessment consists of four parts:

Part A: Contracture Assessment

This section considers three component contractures typically seen in RDEB: web spaces, finger flexion and thumb adduction. Each of these component contractures is assessed and then given a severity score. The wrist and forearm motion is also recorded.

Part B: Hand Deformity Grade

The component severity scores from Part A are combined to provide a Hand Deformity Grade ranging from: none, mild, moderate to severe and is used to describe the overall severity of the hand deformity.

Part C: Reported Hand Care Routines

This section records information about the reported routines of the child or young person regarding hand splint use, web space bandaging and glove use.

Part D: Hand Surgery

This section records details of previous hand surgery, records if surgery is being considered and patient/parent opinion of hand appearance and function following surgery.

Assessment of Hand Contractures in Epidermolysis Bullosa (ACE)

Name:
MRN:
NHS no:
DOB:

Date assessed:
Diagnosis:
Age today:

Part A: Contracture Assessment

1: Web space contracture (Pseudosyndactyly)

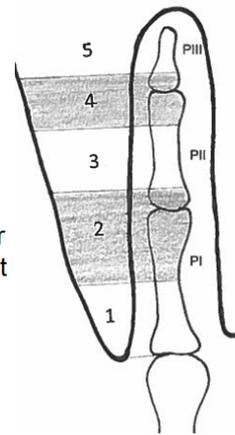
1.1 Assess the 2nd, 3rd and 4th web space for each hand. The thumb and 1st web space are considered in Part 4. Assess the hand from a dorsal view. Use the bones and joints of the adjacent fingers as landmarks to assess the progression of web space contractures using the Contracture Key below.

Contracture Key:

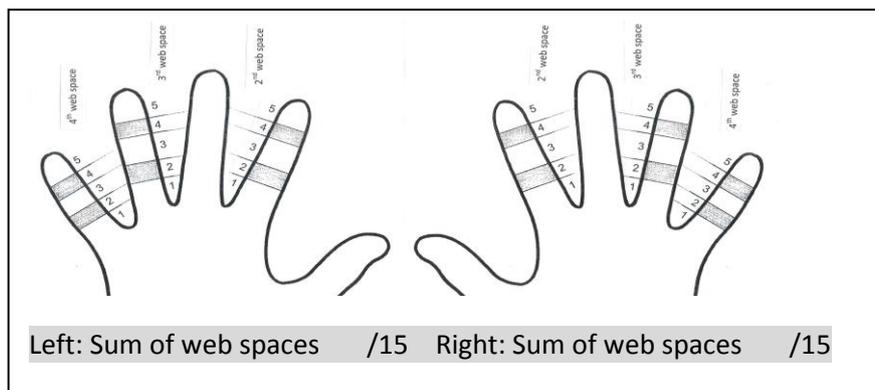
- 1 = up to midway proximal phalanx (PI), does not involve PIP joint
- 2 = beyond midway PI and may involve PIP joint and base of middle phalanx (PII)
- 3 = beyond base of PII, does not involve DIP joint
- 4 = involves DIP joint and may involve base of distal phalanx (PIII)
- 5 = beyond DIP joint and may involve fingertip

To ensure consistency, always measure the 2nd web space against the index finger joints, the 3rd web space against the ring finger joints and the 4th web space against the little finger joints.

Do not measure against the middle finger because it is typically longer with more distal joints that become involved later in web space contracture development.



1.2 Indicate the progression of each web space contracture by shading in the diagram, and then add together the numbers for all three web spaces to give sum for each hand from 0-15. If no contractures, score 0.



1.3 Select web space score for each hand. This is used to calculate the Hand Deformity Grade in Part B.

Sum of web spaces	Left web space score	Right web space score
0	0	0
1-5 (Mild)	1	1
6-10 (Moderate)	2	2
11-15 (Severe)	3	3

2: Finger Contracture

2.1 Measure the passive range of motion (PROM) of finger joints using a goniometer and record in table below. Measurements need to be made in a position which eliminates soft tissue restrictions and allows true joint ROM. Normal PROM for the MCP joints is 0-90°; PIP joints 0-100° and DIP joints 0-90°. A joint with a 30° flexion contracture and flexion to 70° would be recorded as Ext -30° and Flex 70°. Indicate extension beyond 0° with +.

Add together the PIP joint extension measurements (disregarding minus signs) of all four fingers to give the combined degrees of flexion contracture for each hand. Only PIPJ flexion contractures are used in the Hand Deformity Grade in Part B because they typically develop before MCP joint contractures, have greater functional impact than DIP joint flexion contractures and are more accessible for accurate measurement. Other PROM measurements are an important part of hand assessment and may be recorded here.

Left	MCP		PIP		DIP	
	Ext	Flex	Ext	Flex	Ext	Flex
Index						
Middle						
Ring						
Little						
Combined degrees of PIPJ flexion contracture:						

Right	MCP		PIP		DIP	
	Ext	Flex	Ext	Flex	Ext	Flex
Index						
Middle						
Ring						
Little						
Combined degrees of PIPJ flexion contracture:						

2.2 Select PIPJ score for each hand. This is used to calculate the Hand Deformity Grade in Part B.

Combined degrees of PIPJ flexion contracture	Left PIPJ score	Right PIPJ score
0°	0	0
1-133° (Mild)	1	1
134-266° (Moderate)	2	2
267-400°+ (Severe)	3	3

2.3 Comment on active ROM (consider presence of palmar skin tightness, bridging across finger joints and/or bony re-absorption). These observations are not used to calculate the Hand Deformity Grade in Part B.

3: Wrist and Forearm Contracture

Measure the PROM of wrists and forearms and record in table below. Normal PROM of wrist flexion is 0-80°, wrist extension 0-70°, radial deviation (RD) 0-20°, ulnar deviation (UD) 0-30°, pronation 0-90° and supination 0-90°. These measurements are not used to calculate the Hand Deformity Grade in Part B.

Left	Ext	Flex	RD	UD	Pro	Sup

Right	Ext	Flex	RD	UD	Pro	Sup

4: Thumb Contracture

4.1 Assess adduction contracture of thumbs for each hand using description below and select score. This score is used to calculate the Hand Deformity Grade in Part B.

Description of thumb adduction contracture	Left Thumb score	Right Thumb score
No contracture: Normal 1 st web space with full radial and palmar abduction. Cylindrical grasp is possible, where the thumb and fingers make full contact with an object.	0	0
Mild contracture: Tightness of 1 st web space with reduced radial and/or palmar abduction at end of range. Cylindrical grasp is slightly reduced. Mild hyperextension of MCPJ and/or IPJ may be present.	1	1
Moderate contracture: Contracture of 1 st web space up to or involving IPJ with restricted radial and/or palmar abduction. CMCJ may be contracted with base of thumb overlying the palm. Cylindrical grasp is significantly reduced. Hyperextension of the MCPJ and/or IPJ is present.	2	2
Severe contracture: Contracture of 1 st web space beyond IPJ with no abduction or cylindrical grasp possible. Adapted pinch may be possible.	3	3

4.2 Measure the PROM of thumb joints and record in table below. Normal PROM of the thumb MCP joint is 0-50°, and IP joint is 0-80°. These measurements are not used to calculate the Hand Deformity Grade in Part B.

Left Thumb	MCP		IP	
	Ext	Flex	Ext	Flex

Right Thumb	MCP		IP	
	Ext	Flex	Ext	Flex

Part B: Hand Deformity Grade

Transfer scores for web spaces, finger PIPJs and thumb from Part A (in grey) to the table below. These add up to give a Hand Deformity Score from 0-9.

Left	Score
Webs Spaces	
Fingers PIPJ	
Thumb	
Left Hand Deformity score:	/ 9

Right	Score
Web Spaces	
Fingers PIPJ	
Thumb	
Right Hand Deformity score:	/ 9

Finally, the score is converted to a Hand Deformity Grade to describe the severity of the whole hand deformity.

Left Hand Deformity Grade			
None (0)	Mild (1-3)	Moderate (4-6)	Severe (7-9)

Right Hand Deformity Grade			
None (0)	Mild (1-3)	Moderate (4-6)	Severe (7-9)

Part C: Reported Hand Care Routines

	Left Hand	Right Hand
In general, how frequently do you wear hand splints? <i>Describe the splint:</i>	I do not wear splints <input type="checkbox"/> Often: 4 to 7 nights/week <input type="checkbox"/> Sometimes: 1 to 3 nights/week <input type="checkbox"/> Variable (describe) <input type="checkbox"/>	I do not wear splints <input type="checkbox"/> Often: 4 to 7 nights/week <input type="checkbox"/> Sometimes: 1 to 3 nights/week <input type="checkbox"/> Variable (describe) <input type="checkbox"/>
In general, how frequently do you wear web space bandaging? <i>Describe the bandaging:</i>	I do not wear web bandaging <input type="checkbox"/> Every day and night <input type="checkbox"/> Every day or night <input type="checkbox"/> Variable (describe) <input type="checkbox"/>	I do not wear web bandaging <input type="checkbox"/> Every day and night <input type="checkbox"/> Every day or night <input type="checkbox"/> Variable (describe) <input type="checkbox"/>
In general, how frequently do you wear EB gloves? <i>Describe the gloves:</i>	I do not wear gloves <input type="checkbox"/> Every day and night <input type="checkbox"/> Every day or night <input type="checkbox"/> Variable (describe) <input type="checkbox"/>	I do not wear gloves <input type="checkbox"/> Every day and night <input type="checkbox"/> Every day or night <input type="checkbox"/> Variable (describe) <input type="checkbox"/>

Please use this space to make any additional comments about hand care routines:

Part D: Hand Surgery

	Left Hand	Right Hand
If you have not had hand surgery, are you considering it? <i>Comments:</i>	Yes <input type="checkbox"/> No <input type="checkbox"/> Don't know <input type="checkbox"/>	Yes <input type="checkbox"/> No <input type="checkbox"/> Don't know <input type="checkbox"/>

Previous hand surgery date and details:		
Following your surgery how do you feel about how your hand looks? Patient/parent (please circle)	Very satisfied <input type="checkbox"/> Satisfied <input type="checkbox"/> Neither satisfied nor dissatisfied <input type="checkbox"/> Dissatisfied <input type="checkbox"/> Very dissatisfied <input type="checkbox"/>	Very satisfied <input type="checkbox"/> Satisfied <input type="checkbox"/> Neither satisfied nor dissatisfied <input type="checkbox"/> Dissatisfied <input type="checkbox"/> Very dissatisfied <input type="checkbox"/>
Following your surgery how do you feel about how your hand works? Patient/parent (please circle)	Very satisfied <input type="checkbox"/> Satisfied <input type="checkbox"/> Neither satisfied nor dissatisfied <input type="checkbox"/> Dissatisfied <input type="checkbox"/> Very dissatisfied <input type="checkbox"/>	Very satisfied <input type="checkbox"/> Satisfied <input type="checkbox"/> Neither satisfied nor dissatisfied <input type="checkbox"/> Dissatisfied <input type="checkbox"/> Very dissatisfied <input type="checkbox"/>
Are you considering further hand surgery? Patient/parent (please circle)	Yes <input type="checkbox"/> No <input type="checkbox"/> Don't know <input type="checkbox"/>	Yes <input type="checkbox"/> No <input type="checkbox"/> Don't know <input type="checkbox"/>

Please use this space to make any additional comments:

Completed by: _____ Designation: _____ Date: _____