

## Assessment of Hand Contractures in Epidermolysis Bullosa (ACE-EB)

### What is the ACE-EB?

The ACE-EB is a hand assessment developed for use with children and young people with Recessive Dystrophic Epidermolysis Bullosa – Severe subtype (RDEB-S). It is intended to be used by hand therapists to assess hand contractures typically seen in this condition. It can be used routinely to monitor changes over time and guide clinical treatment. It is recommended to use alongside age appropriate functional and quality of life assessments to understand the impact of hand deformities more holistically. 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 individually the three component contractures typically seen in RDEB-S: web spaces, finger flexion and thumb adduction. Each of these component contractures is assessed and given a *severity score*. Forearm, wrist and thumb range of motion and volar skin bridging of the fingers is also recorded in this part.

#### **Part B: Hand Deformity Grade**

The Hand Deformity Grade (HDG) is used to describe the overall severity of the hand deformity. If used, all three component *severity scores* from Part A are added together to provide a HDG of none, mild, moderate or severe.

#### **Part C: Reported Hand Care Routines**

This section records information about the reported routines of the child or young person regarding hand splints, web space dressings and glove use which are used to help delay hand contracture development.

#### **Part D: Hand Surgery – Patient Reported Outcome Measure**

This section records whether the patient or young person is considering hand surgery. If they have had hand surgery, this part serves as a Patient Reported Outcome Measure of satisfaction with hand appearance and function.

# Assessment of Hand Contractures in Epidermolysis Bullosa (ACE-EB)

Name:	Date assessed:
MRN:	Diagnosis:
NHS no:	Age today:
DOB:	

## Part A: Contracture Assessment

### 1: Web space contracture (Pseudosyndactyly)

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

**Contracture Key:**

- 0 = no web space contracture
- 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 level of each web space contracture by shading in the diagram below. Then add the scores for all three web spaces to give a total for each hand from 0-15. If no contractures, score 0.

Left: Total      / 15

Right: Total      / 15

1.3 Convert total to web severity score for each hand; this score summarises and represents the whole hand. This is used to calculate the Hand Deformity Grade in Part B.

Total	Left web severity score	Right web severity 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 should be taken 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°. Eg. a joint with a 30° flexion contracture (extension deficit) and flexion to 70° is recorded as Ext 30° and Flex 70°. Indicate hyperextension with +.

**Add together the PIP joint extension measurements** of all four fingers to give the total degrees of PIPJ flexion contracture for each hand. This is used to represent finger flexion contracture in the *flexion severity score*. However, all finger measurements are an important part of hand assessment and are recorded here.

Left	MCP		PIP		DIP	
	Ext	Flex	Ext	Flex	Ext	Flex
Index						
Middle						
Ring						
Little						
<b>Total degrees of PIPJ flexion contracture:</b>						

Right	MCP		PIP		DIP	
	Ext	Flex	Ext	Flex	Ext	Flex
Index						
Middle						
Ring						
Little						
<b>Total degrees of PIPJ flexion contracture:</b>						

**2.2 Convert total to flexion severity score;** this score averages and represents all four fingers of the hand. This is used to calculate the Hand Deformity Grade in Part B.

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

**2.3 Observe and record presence of skin bridging.** This refers to palmar skin shortening which forms a bridge of skin across a flexed finger joint. This bridging may progress to involve fusion of the fingertip to the palm.

Left	Indicate level of skin bridging
Index	None / DIPJ / PIPJ / palmar fusion
Middle	None / DIPJ / PIPJ / palmar fusion
Ring	None / DIPJ / PIPJ / palmar fusion
Little	None / DIPJ / PIPJ / palmar fusion

Right	Indicate level of skin bridging
Index	None / DIPJ / PIPJ / palmar fusion
Middle	None / DIPJ / PIPJ / palmar fusion
Ring	None / DIPJ / PIPJ / palmar fusion
Little	None / DIPJ / PIPJ / palmar fusion

## 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°.

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 *thumb severity score*. This score is used to calculate the Hand Deformity Grade in Part B.

Description of thumb adduction contracture	Left Thumb severity score	Right Thumb severity score
<b>No contracture:</b> Pliable 1 <sup>st</sup> web space No CMCJ contracture with full radial and palmar abduction	0	0
<b>Mild contracture:</b> Tightness of 1 <sup>st</sup> web space Reduced radial and/or palmar abduction at end of range	1	1
<b>Moderate contracture:</b> Contracture of 1 <sup>st</sup> web space below the IPJ Contracture of CMCJ restricting radial and palmar abduction Base of thumb may be overlying the palm	2	2
<b>Severe contracture:</b> Contracture of 1 <sup>st</sup> web space involving or beyond IPJ Contracture of CMCJ with no abduction possible Base of thumb is overlying the palm	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°. If compensatory hyperextension is present, indicate with +.

Left Thumb	MCP		IP	
	Ext	Flex	Ext	Flex

Right Thumb	MCP		IP	
	Ext	Flex	Ext	Flex

## Part B: Hand Deformity Grade

**Transfer severity scores** for web spaces, finger flexion and thumb from Part A (in grey) to the table below. Add these to give a total out of 9 which is converted into a Hand Deformity Grade for each hand.

Left	Severity Scores			
Webs				
Fingers				
Thumb				
<b>Hand Deformity Grade:</b>		<b>/ 9</b>		
<b>None</b>	<b>Mild</b>	<b>Moderate</b>	<b>Severe</b>	
0	1-3	4-6	7-9	

Right	Severity Scores			
Web				
Fingers				
Thumb				
<b>Hand Deformity Grade:</b>		<b>/ 9</b>		
<b>None</b>	<b>Mild</b>	<b>Moderate</b>	<b>Severe</b>	
0	1-3	4-6	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 dressings? <i>Describe the dressings:</i>	I do not wear web dressings <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 dressings <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 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"/>

**Additional comments:**

## Part D: Hand Surgery – Patient Reported Outcome Measure

	Left Hand	Right Hand
Are you considering hand surgery? <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"/>

**Additional comments:**

Completed by: \_\_\_\_\_ Designation: \_\_\_\_\_ Date: \_\_\_\_\_