
Revised Upper Limb Module for SMA (RULM FOR SMA)

November 10th

2014

Introduction

The **Revised Upper Limb Module (RULM FOR SMA)** has been devised to assess motor performance in the upper limbs for individuals with Spinal Muscular Atrophy (SMA). Details of the conceptual framework of the scale are detailed in the manual. The Revised ULM has been devised by the ULM Physiotherapy Working Group (Elena Mazzone, Anna Mayhew, Danielle Ramsey, Marion Main, Jackie Montes, Roberto De Sanctis, Lavinia Fanelli, Marlene Vandenhouwe, Allan Glanzman, Julaine Florence, Amy Pasternak, Sally Dunaway). Work is on going to improve the content, validity and reliability of the scale.

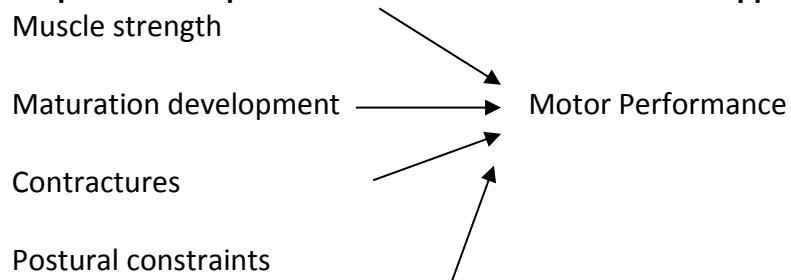
Conceptual Framework of the Revised ULM

The purpose of an upper limb scale for use in Spinal muscular atrophy is to assess change that occurs in motor performance of the upper limb over time. Motor performance in Spinal Muscular Atrophy is defined as a demonstrated ability to perform a skill under certain test conditions. This performance changes with disease progression and /or intervention (including surgery) and is based on the observed response on the day. Motor performance will be impacted by muscle strength, contractures and maturational development (puberty) and the scale aims to incorporate performance of shoulder, elbow, wrist and hand function. Specific domains have yet to be established precisely but domains may relate to functional workspace. I.e. high level shoulder dimension, mid-level elbow dimension, distal wrist and hand dimension. These may however lie on a continuum of ability. Items measuring motor performance relate to meaningful functional activities of everyday life.

Intended population

Ambulant and non-ambulant individuals with Spinal Muscular Atrophy from childhood to adulthood.

Proposed Conceptual framework for Performance of Upper Limb Scale



Equipment Required

• Adjustable table top to fit individual being tested • Tablecloth marked with circles (Appendix 1) • HB pencil • Tokens • 2 Plastic cups (vending cup) one placed inside the other • Selection of metric weights: 200 g – metal cooking weight 500 g - sand weight, cooking metal weight 1 kg – cooking metal weight • 500 g gym sand weight • Ziploc container (8 ounce) • Push light button (Osram Dot) • Plain A4 paper	
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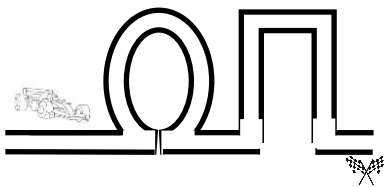
General Instructions

- Ideally the same evaluator should perform evaluations at subsequent assessments
- Items should be performed in the order provided in this manual and the scores should be recorded directly on the Proforma (Revised Upper Limb Module for SMA Source Document-Appendix A)
- Each item will be tested on the dominant / preferred side unless it's a bilateral task or if the scoring options include bimanual option.
- Up to three good acceptable attempts are allowed per task
- The subject should wear loose clothing, T-shirt etc.
- All items are performed with patient sitting in wheelchair or in an appropriate sized chair and with an adjustable height table or surface
- For individuals in a powered chair unable to transfer easily ensure their chair has the seat positioned as parallel as possible to the ground with the backrest in an upright position if tolerated and that the feet are supported
- To ensure a standardized start position adjust the table or surface so that it is placed at armrest level if patient is sitting in wheelchair or is adjusted to the subject's height at umbilicus level if seated on chair
- If it is not possible to assure this starting position the patient should be placed in the sitting position he spends most of the day in with height set individually at a comfortable working position
- Elbows and forearms are supported on armrests or table
- Preferably TLSOs should not be worn during testing. If not possible to test without TLSO then specify if a TLSO is worn and what type
- In all items that include elbow and/or shoulder flexion the arm can slide on armrest onto examining surface. No upper extremity splints or assistive devices are allowed during the test.
- Make a note of significant elbow contractures and mark on the Proforma (Revised Upper Limb Module for SMA Source Document - Appendix A) if the score was limited by these (LBC)
- It is acceptable for the evaluator to demonstrate the task and suggest alternative strategies within scoring options

A. Entry item							
Equipment:	200g weight and plastic cup. Token and a pencil if necessary.						
Starting position:	Sitting, start with hands on lap If ambulant use a chair with a backrest but with no armrests. If they are non-ambulant and it's not practical to transfer them they can remain in their wheelchair with seat as horizontal as possible and backrest upright as possible.						
Instruction	"Lift your hands and arms as high as you can." Trunk side flexion can be up to 20°. If they bend their head to their hands instruct them to keep their head up if they can. For individuals less able you can start by asking them: "Can you bring your hands to your mouth?" For the most able individuals, demonstrate abduction of both arms simultaneously with elbows in extension.						
Scoring details	Score 4: Defined as elbow to shoulder height. Score 2 and 3. They should not do this by bringing their mouth to their hands. If they use significant head and trunk flexion in order to complete the task means they cannot score 2 or 3 for this item.						
A.ENTRY ITEM	0	1	2	3	4	5	6
	No useful function of hands.	Can use hands to hold pencil or pick up a token or drive a powered chair, use phone key pad	Can raise 1 or 2 hands to mouth but cannot raise a cup with a 200g weight in it to mouth	Can raise standardized plastic cup with 200g weight in it to mouth using both hands if necessary.	Can raise both arms simultaneously to shoulder height with or without compensation. Elbow bent or in extension	Can raise both arms simultaneously above head only by flexing the elbow shortening circumference of the movement /using accessory muscles)	Can abduct both arms simultaneously elbows in extension in a full circle until they touch above the head.

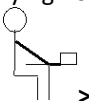
B. BRING HANDS FROM LAP TO TABLE			
Equipment:			
Starting position:	Sitting, start with hands resting on thighs. Block trunk.		
Instruction:	Can you bring hands from lap onto table?		
Tested function	Bring hands from lap onto table.		
Scoring details	0	1	2
	Unable to bring 1 hand onto table	Brings one hand completely to table	Brings two hands completely to table either together or one at a time

C. TRACING A PATH			
Equipment	Standard HB pencil		
Starting position	Sitting, start with hand(s) on table. Table should be set so elbow is at approximately 45° - a functional position. To start, paper can be positioned by the subject but must not be moved thereafter Pencil can be placed in hand and any handhold on the pencil can be used. Write in the usual writing position Make clear to the child that it is not necessary to exactly stay within the lines but just follow the course		
Instruction	Can you complete the path bringing the car to the finish line without stopping or taking pencil off of paper?		
Tested function	Hand function (tripod), holding objects, drawing/writing		
Scoring details			
	0	1	2
	With pencil in hand unable to hold it or make a mark	Able to complete the path but needs to stop or raises pencil from paper	Able to complete the path without stops or raising hand from paper.



D. PICK UP TOKENS			
Equipment:	2 tokens		
Starting position:	Sitting, start with hand(s) on table. Table should be set so elbow is at approximately 45° flexion from full extension (a functional position) If not possible, adapted position is allowed but hands must be on the table. Place tokens on the table in front of the individual.		
Instruction:	Can you pick up these tokens one at a time with one hand and hold them?		
Scoring details	They should not stack the tokens on top of each other before they are picked up. They must hold all tokens in their hand to score 2. Can repeat test up to 3 times if they drop tokens during test		
	0	1	2
	Cannot pick up one token	Can pick up one token	Can pick up and hold 2 tokens

E. PLACE TOKEN INTO CUP ON TABLE OR SHOULDER HEIGHT

Equipment:	Token, cup		
Starting position:	Sitting, start with elbow resting on armrest or lap Token placed in hand Cup placed lying horizontally on table at wrist length with 90° shoulder flexion from the patient. Token placed in hand Cup placed vertically by the examiner at arm length and 90° shoulder flexion from the patient with cup's upper rim at shoulder height under hand.		
Instruction:	Can you place the token in the cup without throwing?		
Tested function	Moving objects in front on table surface and in front at shoulder height		
Scoring detail:	Less than 30° trunk flexion compensation is allowed		
	0	1	2
Place token into cup - On table: horizontal - At shoulder height vertical	Unable to bring token 	Able to bring token to cup lying horizontally 	Able to bring token to cup placed vertically at shoulder height  

F. REACH TO THE SIDE AND TOUCH TOKEN			
Equipment	Token		
Starting position	Sitting, start with elbows resting. Token placed at 90° shoulder abduction and wrist length from patient. If patient succeeds then place token at 135° shoulder abduction at fingertip length from patient Preferred no arm rests on chair		
Instruction	Can you take the token from my hand?		
Tested function	Take/bring objects at side at shoulder height and above, dressing		
Scoring details	0	1	2
	<i>Target: Shoulder 90° abduction with token at wrist length from patient</i> Can't bring hand to shoulder height 	<i>Target: Shoulder 90° abduction, token at wrist length from patient</i> Brings hand to shoulder height, elbow can be bent or extended	<i>Target: Shoulder 135° abduction, token at fingertip length</i> Brings hand above shoulder height, elbow at least at eye level  OK  NO

G. PUSH A BUTTON LIGHT			
Equipment:	Push button light		
Starting position:	Sitting, start with hand(s) on table. Table should be set so elbow is at approximately 45° - a functional position. The button light is placed on the table between the person's hands on a stiff surface.		
Instruction:	Can you turn the light on by pushing it and hard enough to keep it lit? Try using one hand		
Tested function	Pushing buttons		
Scoring details:	The light should remain on the table. They can only use their hands to activate the light. Squeezing light between fingers or banging on it is not allowed. Elbow cannot be higher than the wrist		
	0	1	2
	Unable to turn the light on with one hand	Able to turn the light on momentarily with fingers and/or thumb of one hand.	Able to turn the light on permanently with fingers and/or thumb of one hand.

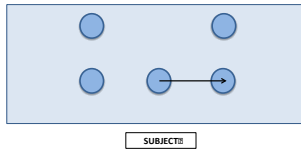
H. TEARING PAPER			
Equipment:	Several sheets of A4 paper		
Starting position:	Sitting, start with hands on table. Table should be set so elbow is at approximately 45° flexion from full extension (a functional position). If not possible, adapted position is allowed but hands must be on the table. Start with piece of paper that has been folded in 4. If unable to tear unfold paper (folded in 2) and ask to tear folded edge but not in the center along the previously folded crease.		
Instruction:	Can you tear this paper starting from the folded edge?		
Scoring details	To score an item they must tear on folded edge and complete the tear across the width of the page in no more than 3 good attempts.		
	0	1	2
	Cannot tear folded piece of paper	Tears the sheet of paper folded in 2, beginning from the folded edge	Tears the sheet of paper folded in 4, beginning from the folded edge

I. OPEN ZIPLOC CONTAINER		
Equipment:	Standard round 8 ounce Ziploc container	
Starting position:	Sitting, start with hands on table. Table should be set so elbow is at approximately 45° - a functional position. Ziploc container placed in midline in front of subject.	
Instruction:	Can you open and take off lid from the container? If not able on first attempt the evaluator can suggest strategy (stabilizing container with one hand against body while prying lid off completely with alternate hand.)	
Tested function	Opening containers	
Scoring details	0	1
	Unable to open.	Open container, on table or against body

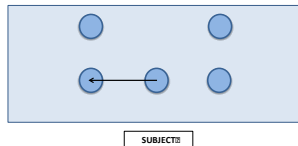
J. RAISE CUP with 200g TO MOUTH			
Equipment:	2 cups one inside each other with 200g weight inside		
Starting position	Sitting, start with hand(s) on table. Table should be set so elbow is at approximately 45° - a functional position. Cup is placed in hand on table in front of subject at midline		
Instruction	Can you raise the cup to your mouth as if you were drinking?		
Tested function	Lifting weight at mouth level. Drinking		
Scoring details	Cup's upper rim should reach mouth level		
	0	1	2
	Unable to get cup to mouth	Cup with 200g to mouth 2 hands	Cup with 200 g to mouth 1 hand

K. MOVING 200gr WEIGHT ON TABLE HORIZONTALLY

Equipment	- Tablecloth with drawn 2 circles 20cm apart (see Appendix 1) placed with center circle aligned with patient's midline - Weights 200g		
Starting position	-Sitting, start with elbow resting, hand on tablecloth next to weight placed on midline circle The weight is moved from the midline circle to outer circle on tested side in one motion.		
Instruction	Can you lift this weight from the center circle to the outside circle with one hand?		
Tested function	Hand function (pinch), forearm function, moving objects on horizontal plane		
Scoring details	For score 2 a lift is defined as hand and forearm clear of all surfaces for duration of movement and the weight must be completely inside the outer circle at end of task. Score 0 if uses two hands on weight		
	0	1	2
Between horizontal circles (INNER TO OUTER CIRCLE)	Unable	Slide 200g	Lift 200g weight

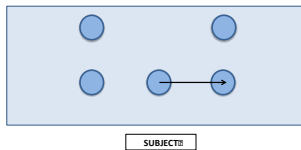


Right-handed subject example

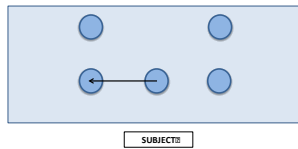


Left-handed subject example

L. MOVING 500gr WEIGHT ON TABLE HORIZONTALLY			
Equipment	- Tablecloth with drawn 2 circles 20cm apart (see Appendix 1) placed with center circle aligned with patient's midline - Weights 500g		
Starting position	-Sitting, start with elbow resting, hand on tablecloth next to weight placed on midline circle The weight is moved from the midline circle to outer circle on tested side in one motion		
Instruction	Can you lift this weight from the center circle to the outside circle with one hand?		
Tested function	Hand function (pinch), forearm function, moving objects on horizontal plane		
Scoring details	For score 2 a lift is defined as hand and forearm clear of all surfaces for duration of movement and the weight must be completely inside the outer circle at end of task. Score 0 if uses two hands on weight.		
	0	1	2
Between horizontal circles (INNER TO OUTER CIRCLE)	Unable	Slide 500g	Lift 500g weight

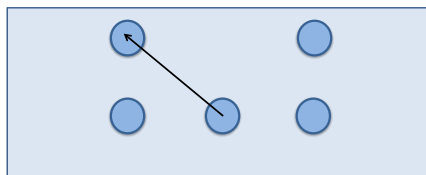


Right-handed subject example



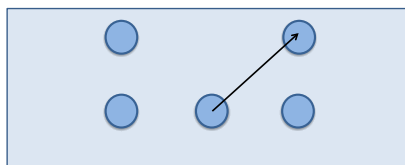
Left-handed subject example

M. MOVING WEIGHT ON TABLE DIAGONALLY			
Equipment	- Tablecloth (see Appendix 1) placed with center circle aligned with patient's midline - Cooking weight 200g		
Starting position	-Sitting, start with elbow resting, hand on tablecloth next to weight - The weight is moved across midline from the center circle to the upper outer circle on opposite side.		
Instruction	Can you lift this weight from this circle to this circle with one hand?		
Tested function	Hand function (pinch), forearm function, moving objects on horizontal plane		
Scoring details	For score 2-a lift is defined as hand forearm and elbow clear of all surfaces for duration of movement. For a score of 2 subject can be in full elbow extension without moving out of base of support and keeping hand along the line, although not reaching the opposite diagonal circle		
	0	1	2
	Unable	Slide 200g	Lift 200



SUBJECT

Right-handed subject example



SUBJECT

Left-handed subject example

N. BRING 500G WEIGHT FROM LAP TO TABLE			
Equipment:	500gr sand weight fastened in a ring shape		
Starting position:	Sitting, start with hands resting on thighs and weight (500 gr) on patient's lap. Examiner's hand placed in midline at subjects shoulder level and wrist length. Block excessive trunk flexion		
Instruction:	Can you bring hands/weight from thighs onto table with 2 hands?		
Tested function	Bring objects from lap onto table or shoulder height with 2 hands		
Scoring details	0	1	2
	Unable to bring weight to table using two hand	Brings weight onto table using two hands	Brings weight at eye level using two hands 

O. BRING BOTH ARMS ABOVE HEAD - <i>Shoulder abduction</i>			
Starting position:	Arms along side or beyond armrests		
Finish position	Extended arms above head		
Instruction	"Raise your arms above your head out to the side – try and keep straight elbows"		
Scoring details	Score from Entry item (Brooke score)		
	0	1	2
	Unable	Can raise both arms simultaneously above head only by flexing the elbow (using compensation) (Brooke 5)	Can abduct both arms simultaneously elbows in extension in a full circle until they touch above the head (Brooke 6)

P. BRING 500g ABOVE SHOULDER HEIGHT WITH EXTENDED ARM- <i>Shoulder abduction</i>			
Equipment:	500g weight		
Starting position:	-Sitting, start with hands on lap. Elbows unsupported. -Place weight in their hand if able to perform task without weights. Ask them to use a full palmar grasp to hold the weight. Preferred no arm rests on chair		
Finish position	Arm extended out to side above head (elbow to eye level)		
Instruction	Hand on lap – “give me the weight” Always take weight from them once they have reached maximum height (to avoid eccentric muscle work).		
Tested function	Bringing objects at shoulder height and above		
Scoring details	Less than 30° trunk flexion compensation is allowed -The proper movement without compensation is: simultaneous combination of shoulder external rotation with shoulder abduction and elbow extension. Movement has to be controlled and the patient has to maintain the finishing position - Compensation: Absence or asynchrony of one of the previously mentioned movement components, additional or uncontrolled movement Other arm must remain in the individuals lap or on armrest		
	0	1	2
	Unable to lift 500g weight even with compensation	Able to lift 500g weight with compensation	Able to lift 500g weight without compensation

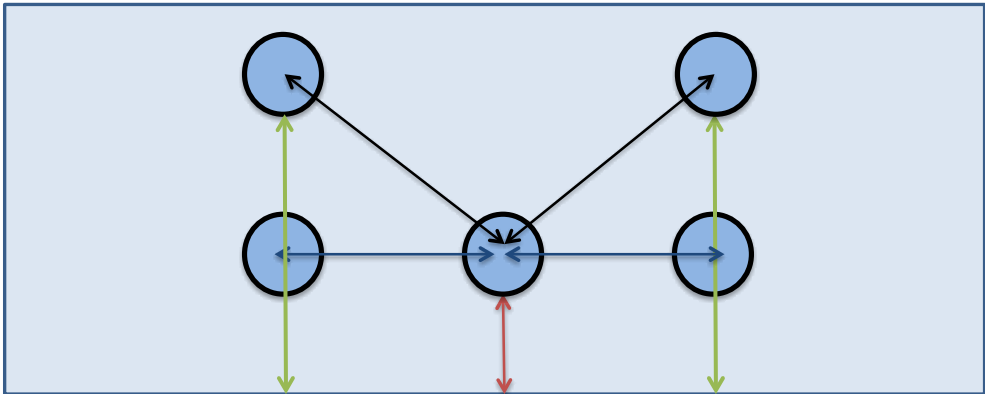
Q. BRING 1kg ABOVE SHOULDER HEIGHT WITH EXTENDED ARM- <i>Shoulder abduction</i>			
Equipment:	1kg weight		
Starting position:	-Sitting, start with hands on lap. Elbows unsupported. -Place weight in their hand if able to perform task without weights. Ask them to use a full palmar grasp to hold the weight. Preferred no arm rests on chair		
Finish position	Arm extended out to side above head (elbow to eye level)		
Instruction	Hand on lap – “give me the weight” Always take weight from them once they have reached maximum height (to avoid eccentric muscle work).		
Tested function	Bringing objects at shoulder height and above		
Scoring details	The proper movement without compensation is: simultaneous combination of shoulder external rotation with shoulder abduction and elbow extension. Movement has to be controlled and the patient has to maintain the finishing position -Compensation: Absence or asynchrony of one of the previously mentioned movement components, additional or uncontrolled movement Other arm must remain in the individuals lap or on armrest		
	0	1	2
	Unable to lift 1kg weight even with compensation	Able to lift 1kg weight with compensation	Able to lift 1kg weight without compensation

R. BRING HAND ABOVE SHOULDER HEIGHT WITH EXTENDED ARM- <i>Shoulder flexion</i>			
Starting position:	-Sitting, start with hands on lap. Elbows unsupported. Preferred no arm rests on chair		
Finish position	Arm extended out to front above head (elbow to eye level)		
Instruction	"Reach out and touch my hand"		
Tested function	Bringing objects at shoulder height and above		
Scoring details	Less than 30° trunk flexion compensation is allowed - The proper movement without compensation is : simultaneous combination of shoulder flexion and elbow extension. Movement has to be controlled and the patient has to maintain the finishing position - Compensation: Absence or asynchrony of one of the previously mentioned movement components, additional or uncontrolled movement Other arm must remain in the individuals lap or on armrest		
	0	1	2
	Unable	Able with compensation	Able without compensation

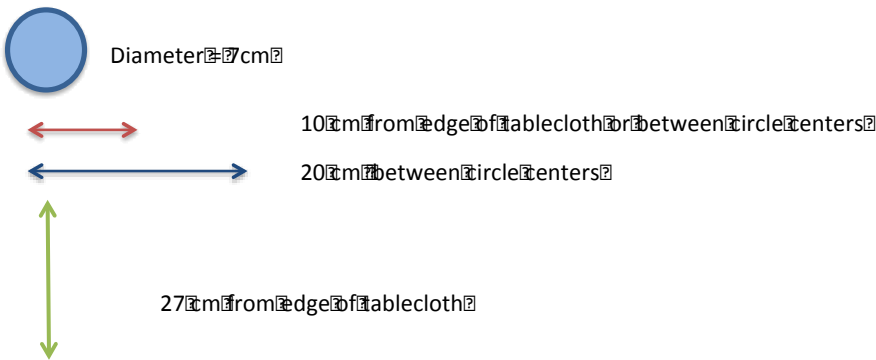
S. BRING 500g ABOVE SHOULDER HEIGHT WITH EXTENDED ARM- <i>Shoulder flexion</i>			
Equipment:	500g weight		
Starting position:	-Sitting, start with hands on lap. Elbows unsupported. -Place weight in their hand if able to perform task without weights. Ask them to use a full palmar grasp to hold the weight. Preferred no arm rests on chair		
Finish position	Arm extended out to front above head (elbow to eye level)		
Instruction	Hand on lap – "give me the weight" Always take weight from them once they have reached maximum height (to avoid eccentric muscle work).		
Tested function	Bringing objects at shoulder height and above		
Scoring details	Less than 30° trunk flexion compensation is allowed The proper movement without compensation is : simultaneous combination of shoulder flexion and elbow extension. Movement has to be controlled and the patient has to maintain the finishing position Compensation: Absence or asynchrony of one of the previously mentioned movement components, additional or uncontrolled movement Other arm must remain in the individuals lap or on armrest		
	0	1	2
	Unable to lift 500g weight even with compensation	Able to lift 500g weight with compensation	Able to lift 500g weight without compensation

T. BRING 1kg weight ABOVE SHOULDER HEIGHT WITH EXTENDED ARM- <i>Shoulder flexion</i>			
Equipment:	1kg weight		
Starting position:	-Sitting, start with hands on lap. Elbows unsupported. -Place weight in their hand if able to perform task without weights. Ask them to use a full palmar grasp to hold the weight. Preferred no arm rests on chair		
Finish position	Arm extended out to front above head (elbow to eye level)		
Instruction	Hand on lap – “give me the weight” Always take weight from them once they have reached maximum height (to avoid eccentric muscle work).		
Tested function	Bringing objects at shoulder height and above		
Scoring details	Less than 30° trunk flexion compensation is allowed The proper movement without compensation is : simultaneous combination of shoulder flexion and elbow extension. Movement has to be controlled and the patient has to maintain the finishing position Compensation: Absence or asynchrony of one of the previously mentioned movement components, additional or uncontrolled movement Other arm must remain in the individuals lap or on armrest		
	0	1	2
	Unable to lift 1kg weight even with compensation	Able to lift 1kg weight with compensation	Able to lift 1kg weight without compensation

Appendix 1 Tablecloth dimensions and layout



Black circles and lines are drawn on tablecloth



Version	Effective	Key Updates
1.0	10 Nov 2014	Original Version

Revised Upper Limb Module for SMA Lead Clinical Evaluator and Trainer:

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