

Improvement of walking ability of three inhibitors by rehabilitation

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Introduction

Haemophilia patients with inhibitors have often severe bleeding complications including multiple joint bleeds. In addition, it is common for such patients to develop arthropathy leading to reduce their quality of life. Recently, bypassing agents show effectiveness to prevent bleeds resulting in improvement of qualities of life in some haemophilia patients with inhibitors.

We report three haemophilia patients with high inhibitor titers, who improved their abilities to move from wheelchair, walk with a cane and orthotics by means of rehabilitation with administration of the prophylactic use of bypassing agents.

RESULTS	Case1: (Before/After)	Case2: (Before/After)	Case3:(Before/After)
ROM(passive)	Lt knee ext: (-45/-20)	Not remarkable change	Lt knee ext:(-50/-25)
MMT	Lt knee ext: (0/3)	All joints 5	Lt knee ext:(4/4)
ADL	Wheelchair/Walk	Confined to a wheelchair/Walk with insole	Wheelchair/Walk with bilateral axilla crutch
Hospital days	240 days	140 days	185 days
Bleeding episode	Left knee @ 5/ Right Iliopsoas muscle @ 1	Right elbow @1	Left knee @1 /Both elbows @1
Bleeding control	Single bolus infusion of rFVIIa 6.0mg per vial and rest,ice,compression for bleedings of joints or muscles.		
Physiotherapy after bleeding	Affected Joint: passive exercise is started from one to seven days later according to swelling and pain at the joint Others: Resistive exercise is started next day		
Follow up	Follow up:7 months Go to college with walking	Follow up:8 months Walk for 1 hour or 5km	Follow up:8 months Walk with bilateral axilla crutch for 30 minutes

Conclusions

For patients with inhibitors,bleeding control seems to be not easy for patients with inhibitors,who are prone to have developed haemophilia arthropathy.

On the contrary ,bypassing agents have contributed significantly to control various types of bleedings,recently prophylactic use with bypassing agents is also utilized for physiotherapy to improve the QOLs.

Cases presented improved the joint function and walking ability in the outdoor significantly by combining the prophylactic use of bypassing agents and physiotherapy.

We suggest that physiotherapy with the support of prophylactic use of bypassing agents should be promising options for such patients with impaired joint function.

1. H.A.Cooper,et al:Rationale for the of high dose rVIIa in a high-titre inhibitor patient with haemophilia B during major orthopaedics.Haemophilia 2001,7,517-522
2. G.Young,et al:Prophylactic recombinant factor VII in haemophilia patients with inhibitors.Haemophilia 2005,11,203-207


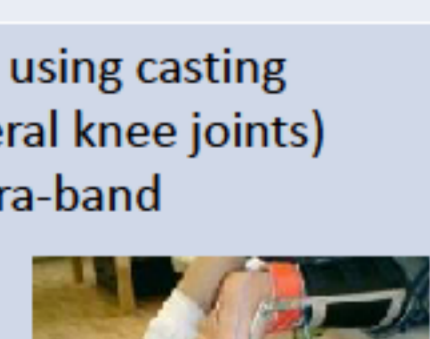
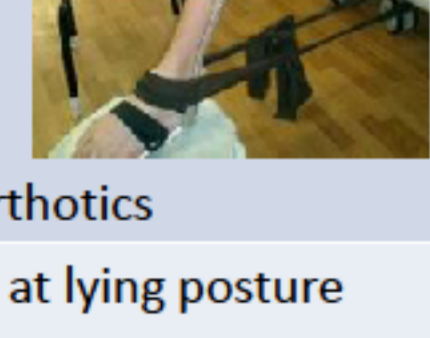
Case 1: Exercise Points:Reinforcement of Lt-Quadriceps

Haemophilia A , 19 years old, 45kg, 17.9BU/ml(HR)
He developed an inhibitor at the age of 3. Following numerous bleedings, his left knee had been a target joint and he could not walk at the age of 18 . No surgical treatments have been performed.

		Hip		Knee		Ankle		Elbow	
		R	L	R	L	R	L	R	L
XP	Arnold scale	II	II	II	IV	II	IV	II	IV
ROM (°)	flexion	125	130	160	160	50	40	150	140
	extension	15	-15	-10	-45	15	15	-5	-25
MMT	flexion	4	2	4	3	4	2	4	3
	extension	4	2	4	0	4	2	4	3

Findings	Findings
SMD*	Rt=LT
Target Joint	Lt-Knee

Problem list	Finding
Short range objective	① Reinforcement of Lt-Quadriceps ② Increase Lt-knee extension
Long range objective	① Walking without support
Prophylaxis	Bolus infusion of rFVIIa: 4.8mg(106mcg/kg)@ 3-5 times/week
Physical treatment	① Low frequency treatment: Lt-Quadriceps ② ROM exercise: Lt-knee extension with Hot pack ③ Walking exercise with orthotics

MMT of Lt-Quad	Clinical course with physiotherapy	prophylaxis	Bleeding
0 Week	# Reinforce of quadriceps muscles power with low frequency treatment # ROM exercise of Lt-knee extension after hot pack	4.8mg@ 5 times Per week	
3rd Week	# SLR exercise with low frequency treatment & orthotics  # Walking exercise with orthotics & caster walker	4.8mg@ 5 times Per week	Lt-Knee (3)
5th Week	# Knee extension exercise using casting board (started with bilateral knee joints) # Knee exercise using Thera-band  # Walking exercise with orthotics	4.8mg@ 5 times Per week	Lt-Knee (3)
6th Month	# Knee extension exercise at lying posture  # Walking exercise with orthotics	4.8mg@ 3 times Per week	Rt-Iliopsoas (1)
8th Month	# SLR exercise without orthotics # Knee extension without orthotics # Standing and Walking without orthotics	4.8mg@ 3 times Per week	



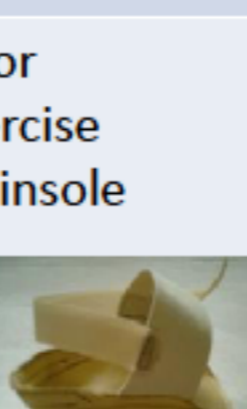
Case 2: Exercise Points: Reinforcement of lower extremity muscles

Haemophilia A , 19 years old, 42Kg, 84.6BU/ml(HR)
He developed an inhibitor at the age of 6. As a result of a great number of bleedings in lower extremity, he become ambulatory and eventually he was confined to a wheelchair.

		Hip		Knee		Ankle		Elbow	
		R	L	R	L	R	L	R	L
XP	Arnold scale	V	III	III	II	V	II	V	I
ROM (°)	flexion	60	70	90	140	60	50	115	145
	extension	-20	20	0	0	-10	0	-50	-10
MMT	flexion	2	3	2	4	3	4	3	4
	extension	2	3	2	4	3	4	3	4

Findings	Findings
SMD*	2cm Rt<Lt
Target Joint	Lt-ankle, Rt-elbow

Problem list	Finding
Short range objective	① Reinforcement of muscles:body,upper extremities nad lower extremities ② move using wheelchair
Long range objective	① Walking with T-cane
Prophylaxis	Bolus infusion :aPCC 4000U@ 3 times/week(~9 weeks) aPCC 3000U@ 3times/week(9~16 weeks) rFVIIa: 4.8mg(114mcg/kg)@ 3 times/week(16weeks~)
Physical treatment	① Muscles exercise:body,upper extremities and lower extremities ② Wheelchair transfer training and mobility training wheelchair ③ Standing balance exercise and walk between parallel bars

MMT	Clinical course with physiotherapy	prophylaxis	Bleeding
0 Week	# Muscles exercise by active movement # Wheelchair transfer training and mobility training wheelchair	aPCC 4000U @ 3 times Per week	
2rd Week	# Muscles exercise to strengthen the body, upper extremity and lower extremity using weight Stand up and sit down motion exercise from chair # Standing balance exercise using DIJOC BOARD 	aPCC4000U@ 3 times Per week	
7th Week	# Walking exercise using caster walker # Walking exercise with Lt-upper extremity in parallel bars 	aPCC 3000U @ 3 times Per week (9th week~)	
10th Week	# Sitting and standing-up on the floor # Stairs up-and-down operation exercise # Walking exercise with T-cane and insole 	aPCC 3000U @ 3 times Per week	Pain:Rt-hip and Rt-ankle No bleeding
16th Week	# To utilize public transportation # Continuance of muscles exercise to strengthen with weight	4.8mg@ 3 times Per week	Rt-elbow(1)


Case 3: Exercise Points: Increase of ROM of Lt-knee extension

Haemophilia B , 18 years old, 40Kg, 1.9BU/ml (HR)
He developed an inhibitor at the age of 2. He have had many bleedings at upper extremities and lower extremities. Especially his right knee have been a target joint and showed extension contracture. As a result, he could not walk from the age of 10 ; he usually used wheelchair.

		Hip		Knee		Ankle		Elbow	
		R	L	R	L	R	L	R	L
XP	Arnold scale	V	III	V	III	III	IV	V	V
ROM (°)	flexion	60	110	140	130	35	25	120	115
	extension	-40	0	-80	-50	15	5	-40	-55
MMT	flexion	3	3	3	3	3	3	3	3
	extension	1	3	2	4	3	3	3	3

Findings	Findings
TMD**	6cm Rt<Lt
Target Joint	Lt-Knee, Both elbows

Problem list	Finding
Short range objective	① Increase ROM Lt-knee extension ② Increase of Body and upper extremity and lower extremity muscles power
Long range objective	① Standing by Lt-lower extremity
Prophylaxis	Bolus infusion of rFVIIa: 4.8mg(120mcg/kg)@ 3 times/week
Physical treatment	① Muscles exercise:Body,upper extremities and lower extremities ② ROM exercise: Lt-knee extension and Lt-hip extension with Hot pack ③ Standing exercise between parallel bars

ROM of Lt-knee	Clinical course with physiotherapy	prophylaxis	Bleeding
0 Week	# Muscles exercise of body,upper extremities and lower extremities # ROM exercise of Lt-knee extension and Lt-hip extension after hot pack	4.8mg@ 3 times Per week	
3rd Week	# Muscles exercise with weight on upper extremities and lower extremities # ROM exercise of Lt-knee extension and Lt-hip extension after hot pack 	4.8mg@ 3 times Per week	Lt-Knee (1) Decrease ROM of Lt-knee ext
6th Week	# Reinforcement of muscles with weight and numbers # Knee extension exercise using weight # Standing balance exercise of Lt-lower extremity	4.8mg@ 3 times Per week	
12th Week	# Walking exercise between parallel bars # Changed walking exercise with bilateral axilla crutch after bleeding episode	4.8mg@ 3 times Per week	Lt-Elbow(1) Pain and stopped walking
19th Week	# Walking exercise with bilateral axilla crutch to outdoor # Continuance of ROM exercise Lt-knee extension and muscles exercise	4.8mg@ 3 times Per week	Rt-Elbow(1) Pain and stopped walking

* SMD:spinomalleolus distance
**TMD:trochantomalleolus distance

