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Rehabilitating individuals with spinocerebellar ataxia: Experiences from impairment-based rehabilitation through multidisciplinary care approach

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Abstract

Spinocerebellar ataxia (SCA) is a rare neurodegenerative disease with progressive course and poor expected outcomes. Therefore, rehabilitation remains the principal form of management especially in advanced disease. Impairment-based rehabilitation through multidisciplinary care approach has proven benefits for functional improvement in individuals with advancing SCA. This concept is based on comprehensive assessments of individualised impairments and functional limitations while exploring contributing environmental and personal factors affecting the person as a whole. From this assessment, individualised rehabilitation goals can be formulated through a multidisciplinary care approach. Neurologists, rehabilitation physicians, physiotherapists, occupational therapists and speech and language pathologists are key individuals involved in the multidisciplinary care for individuals with SCA rehabilitation. Two cases of individuals at different stages of SCA are presented to highlight the rehabilitation approach in providing focused interventions based on individualised impairments through multidisciplinary care. These cases emphasise the importance of understanding the needs of each individuals with SCA so that the rehabilitative therapies prescribed can be tailored to the functional achievements desired.

Keywords: Spinocerebellar ataxia, neurorehabilitation, spinocerebellar ataxia rehabilitation, impairment-based rehabilitation, multidisciplinary care approach

INTRODUCTION

Spinocerebellar ataxia (SCA) is a rare progressive neurodegenerative disease with heterogenous genetic mutations and phenotypes.[1] Up to 40 SCA subtypes have been reported leading to variable neurological presentations including incoordination, postural imbalance, unsteady gait, recurrent falls, oculomotor disturbances and speech difficulties.[2] As curative treatment for SCA has yet to be established and the expected outcomes are poor with restrictions in functional and social activities, rehabilitation remains the mainstay of management especially in an advancing course of the disease. Though the primary aim is supportive, nevertheless, impairment-based rehabilitation through a multidisciplinary care approach has been proven to be beneficial in terms of functional improvement in individuals with advancing SCA.

Concept of impairment-based rehabilitation through multidisciplinary care approach

The lack of evidence and guidelines on rehabilitative treatment in degenerative ataxia confers further difficulties in formulating a standard protocol for rehabilitation in SCA. Hence, an individualised impairment-based rehabilitation approach could be appropriate.

Comprehensive assessment on impairments and functional limitations are based on the International Classification of Functioning, Disability and Health (ICF) framework, a clinical problem-solving tool towards the provision of holistic management.[3] This framework not only evaluates the functional limitations of each individual but also explores contributing environmental and personal factors. Therefore, individualised rehabilitation goals can be formulated through input from a multidisciplinary

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care team, comprising primarily of neurologists, rehabilitation physicians, physiotherapists (PT), occupational therapists (OT) and speech and language pathologists.[4]

The progress of an individualised rehabilitation programsfor SCA individuals is guided through outcome measures monitoring; namely Scale of Assessment and Rating of Ataxia (SARA), Berg Balance Scale (BBS), Timed Up and Go (TUG) and Modified Barthel Index (MBI). The former two measure body structures and function, the latter two measure functional domains. Our primary intention of this review is to highlight impairment-based rehabilitation through a multidisciplinary care approach,with various instruments adopted throughout two different stages of SCA. We emphasise the need to address each patient individually, with the ultimate goal being to promote a certain degree of self-independence while reducing fall risk and improving quality of life.

CASE REPORTS

Patient 1

History and clinical course

A 25-year-old lady presented with a history of progressive worsening of imbalance and recurrent falls since early 2013 associated with an abnormal gait. By late 2015, she had to resign as a cashier; required walking aids for ambulation by late 2016, and subsequently was confined to a wheelchair as her fatigue worsened. Noting that a similar disease progression had affected her mother and two siblings, she finally presented in early 2017 with head titubation and limbs tremors, dysmetria, nystagmus and dysarthria. Computed brain tomography showed global cerebellar and brainstem atrophy, with normal CSF studies, hormonal studies, connective tissue and infective diseases screenings. The diagnosis of SCA was made based on the clinical features, strong family history and exclusion of other causes. She was then admitted for four weeks of intensive rehabilitative therapy with the goal of safe, limited short-distance ambulation within the setting of her room to enable her to perform self-care.

Balance and coordinative training

Focused balance training was done in a standing position while incorporating functional tasks such as holding objects and throwing a ball (Table 1). Sit-to-stand exercises were reinforced on a daily basis in addition to therapeutic flexibility and strengthening exercises of the core trunk muscles (Table 1). To promote the ability for corrective balance from postural sway as a measure of a fall prevention strategy, visual feedback training using centre of pressure motion signals was used. Gait training was delivered with bodyweight supported device on a treadmill. The percentage of supported body weight was gradually reduced and at the third week of intensive training, over-ground gait training was achieved with a walking frame for minimal distance ambulation. As part of environmental adaptation, visual cueing was taught during walking to enhance gait speed and step length. PT delivered this form of training for an hour a day, for 5 times per week.

Assistive and adaptive devices for functional improvement

In the compensatory arm, to achieve functional improvement, the patient was trained by OT to decompose complex movements into simple and multiple joint movements. Additional use of weighted cuffs and largely handled utensils helped to improve feeding and grooming whereas quad cane was used to facilitate safe ambulation and prevent falls. Training on wheelchair propulsion was included to enable longer distance mobility as a measure of energy-conserving techniques. The duration of training received was similar to the sessions by the PT.

Maintenance exercises for the preservation of communication ability

The patient continued to have effective communication in addressing her needs. Oral facial muscle training including tongue and jaw range of motion and strengthening exercises were prescribed as part of maintenance exercises to delay the deterioration of her speech symptoms (Table 1). The progression of the patient's rehabilitation training is summarised in Table 2.

Patient 2

History and clinical course

A 38-year-old policeman presented with progressive deterioration in walking function and recurrent falls since early 2012. He required support for ambulation by the end of 2015 and by mid 2016, the patient started to develop an asymmetrical head posture. In early 2017, the

limb tremors worsened the balance and he had to crawl to mobilise. With time, long term crawling led to neuropathic pain at the lower back area that was made worse in a supine position and affected his sleep quality. Alongside dysarthria, his work performance was severely affected. Investigations did not found any metabolic causes or infective pathologies. Magnetic resonance imaging of the brain showed generalised cerebral atrophy. There was a similar history in his paternal grandfather and younger sister. A diagnosis of SCA was made based on the clinical features, family history and exclusion of other causes. For management, the patient desired pain control, ability to stand supported during shower in the toilet, ability for self-feeding and mobile phone usage for communication purpose. He was admitted for four weeks of intensive inpatient rehabilitative therapy aiming for these goals.

Pain management

To manage the low back pain, regular gabapentin and topical analgesic ointment were prescribed with transcutaneous electrical nerve stimulation as a physical modality. With pain being managed effectively as shown in Table 2, the patient was able to sit and sleep upright without considerable difficulties.

Balance and coordinative training

Focused balanced training was initiated with sit-to-stand exercise followed by pivot transfer to promote chair usage so that patient would no longer need to crawl on the floor. Therapeutic exercises were focused at targeted strengthening of the core and proximal limb muscles with concurrent flexibility exercises (Table 1). Mat activities for balance training and whole-body movement exercises were accomplished in quadruped standing and kneeling positions to improve trunk-limb coordination. Gradually, balance training was upgraded to standing position and at the third week, the training was further upgraded to side-to-side stepping balance exercise at the parallel bars. By the end of his training, he had better trunk control in maintaining an upright standing position for showering, had a smoother stand pivot transfer technique and achieved independent sideways ambulation approximately 30 metres distance while holding onto the parallel bar, as demonstrated from the outcome measures shown in Table 2. The PT was responsible for conducting this training for an hour a day during weekdays.

Assistive and adaptive devices for functional improvement

Compensatory strategies were targeted towards dampening upper limbs tremors to improve self-feeding performance and reduce spillage of food. He was taught by the OT on simplification of complex movements in adjunct with weighted cuffs placed on the proximal arm and distal forearm incorporated with dual-task hand function training largely in handling of utensils. As the use of walking frame without assistance was deemed unsafe, a home environmental modification which comprised of wall-mounted hand-railings extending from the living room to the washroom for safer mobility option to promote sideways ambulation was recommended. By adopting these strategies, the patient reported more satisfaction with the performance of ADLs and mobile phone usage as shown in Table 2. Duration spent with the OT was approximately the same as with the PT.

Alternative and augmentative communication tool

The patient produced a considerable amount of incomprehensible words necessitating multiple repetitions and body gestures to improve communication. Improved hand function made it possible for mobile texting as a means of alternative and augmentative communication tool in delivering better communication.

DISCUSSION

SCA, being a neurodegenerative disorder, limits further restorative or curative rehabilitation interventions. A review article on motor training in degenerative spinocerebellar disease demonstrated 3 different approaches for ataxic specific improvements; physiotherapy combined with occupational therapy, coordinative physiotherapy and exergame-based training. Based on these approaches, a new concept of rehabilitative training in degenerative cerebellar ataxia is to emphasise on tailored training regime by applying various training strategies according to individualised needs.[5] Physiotherapy (PT) combined with the occupational therapy approach by Miyai *et al*. provided equivalent training duration and utilised similar outcome measure as described in these two cases.[6] However, their subjects had additional hour over the weekends (either physiotherapy or occupational therapy) and a lower baseline score of SARA (mean score between 11.0 to 12.2). Hence they demonstrated less severe cerebellar impairments that may have

Table 1: Rehabilitation strategies through multidisciplinary care approach

Rehabilitation strategies	Type of exercises and training	
	Case 1	**Case 2**
Therapeutic exercises	Flexibility exercises Spine – extensors and lateral rotators Shoulder – flexors and extensors	Flexibility exercises Spine – extensors and lateral rotators Shoulder – flexors and extensors
	Targetedstrengthening exercises Core – spine extensors and lateral rotators, abdominal flexors Upper limb – elbow flexors and extensors Lower limb – hip flexors and extensor, knee extensors	Targeted strengthening exercises Core – spine extensors and lateral rotators, abdominal flexors Upper limb – elbow flexors and extensors Lower limb – hip flexors and extensor, knee extensors
	Aerobic exercises Static cycling	Aerobic exercises Static cycling
Balance and coordination training	Static balance Standing on one leg	Static balance Quadruped standing Standing on one leg
	Dynamic balance Standing with one foot performing stepping to the front, side and back Visual feedback training with pressure motion signals	Dynamic balance Quadruped standing Kneeling
	Whole-body movement Sit-to-stand exercise	Whole-body movement Quadruped standing Kneeling Sit-to-stand exercise Side-stepping
Mobility training	Bodyweight supported treadmill training Over-the-ground body weight supported gait training	Transfer training Stand pivot transfer training – from bed to chair, chair to bed Transfer training from floor to chair, chair to the floor
Functional training	Joint movement decomposition to simple one joint movement	Joint movement decomposition to simple one joint movement
	Adaptive device – weighted cuff, large handle utensils	Adaptive device – weighted cuff, large handle utensils
	Dual task training Handling objects while standing – holding and throwing ball	Dual-task training Handling objects at seated level – feeding, mobile phone usage
Communication training	Oral facial muscles exercises Tongue – range of motion and strengthening exercises Jaw – strengthening exercises	Oral facial muscles exercises Tongue – range of motion and strengthening exercises Jaw – strengthening exercises AAC tool – using a mobile phone

Table 2: Summary of outcome measures

Outcome measure	Case 1		Case 2	
	1st week of rehabilitation	4th week of rehabilitation	1st week of rehabilitation	4th week of rehabilitation
TUG	1 min 36 secs	1 min 36 secs	NA	NA
NRS	NA	NA	9	1
BBS	29/56	29/56	4/56	9/56
SARA	16/40	16/40	24.5/40	22/40
MBI	61/100	66/100	77/100	81/100

TUG = Timed Up and Go; NRS = Numerical Rating Scale; BBS = Berg Balance Scale; SARA = Scale for Assessment and Rating of Ataxia; MBI = Modified Barthel Index; NA = Not applicable

led to higher score changes at the end of the intervention period.

Coordinative physiotherapy is a strategy that emphasises activating and demanding the control mechanism for balance control and multi-joint coordination.[8] While using similar exercise principles for conditioning, i.e. range of motion, strengthening, balance and mobility training, this particular approach studied by Ilg *et al*. highlighted the importance of selecting compensatory feedback, either in the form of somatosensory, visual or vestibular inputs to prevent falls.[8] In addition to utilising SARA outcome measure as a monitoring tool, they used the goal attainment score to determine the relevance of this intervention on daily life. The latter assessment would undoubtedly demonstrate improvement in these two described cases, since both achieved the functional goals despite pre and post-intervention SARA scores that were relatively unchanged or minimally changed.

These two approaches were concurrently applied on these two cases based on a standardised rehabilitative intervention framework that focused on five domains; therapeutic exercises, balance and coordination training, mobility training, functional training and communication training (Table 1). The prescribed interventions were tallied with the individual's impairments, through multidisciplinary care in accordance to the principles of the ICF model for establishing individualised goals.

In addition to the rehabilitative interventions, evidence on the benefits of neuromodulation therapy for degenerative cerebellar ataxia has yet to be studied. A systemic review on cerebellar neuromodulation therapy (including transcranial magnetic stimulation, transcranial direct current stimulation and deep brain stimulation) among several movement disorders have demonstrated improvements in the ataxic symptoms, walking distance, number of tandem steps and standing capacities for individuals with degenerative spinocerebellar ataxia.[9] However, such therapies are infrequently accessible and highly selective, implying the need to select the right type of individuals for such an expensive treatment mode.

Better still, the exponential growth in numbers of devices for promoting self-independency has been intensively explored among individuals with neurological disorders, including spinal cord injury and stroke. However, such applications have not been widely researched in SCA, possibly due to its progressive nature requiring frequent change of devices throughout the disease course.

In conclusion, formulating rehabilitation goals for individuals with SCA needs to be individualised. Impairment-based rehabilitation through multidisciplinary care approach is beneficial for functional improvement. Adopting mixed rehabilitative strategies with the use of appropriate monitoring instruments at different stages of SCA will lead to achievement of the desired goals.

DISCLOSURE

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