Chapter

International Classification of Functioning, Health and Disability (ICF) Conceptual Approach towards Spinocerebellar Ataxia

Kevin Triangto, Steven Setiono and Herdiman Bernard Purba

Abstract

Spinocerebellar Ataxia (SCA) is an autosomal dominant disease with progressive decline towards functional capacity. Although studies had shown that there are various SCA types, physical medicine and rehabilitation approach would focus mostly on functional aspects in each individuals. Analysis through International Classification of Functioning, Disability, and Health would assist clinicians to identify activity and participation aspects of SCA, mostly revolves around mobility function. Good correlation of mobility with quality of life was also reported, and thus it is only natural that this becomes the main focus of rehabilitative intervention. Approximately one hour physical exercise session focusing on postural control and balance was proven to be effective in improving disease related measurement tool, functional capacity, and quality of life. These benefits could be improved through newer therapies such as exercise games and virtual reality, virtually creates a rapidly changing environment, thus providing training through anticipatory actions. It is speculated that neuroplasticity through self-recognition of errors are the main physiology of recovery in SCA. Finally, it could be seen that rehabilitation intervention remains to be a cornerstone in current ataxia therapy, with goals of achieving exercise gains while alleviating the natural functional decline of the disease.

Keywords: Spinocerebellar Ataxia, physical medicine, rehabilitation, balance exercise, neuroplasticity

1. Introduction

Spinocerebellar Ataxia (SCA) is well known to be an autosomal dominant progressive disease that significantly affect quality of life [1]. Despite the many types of SCA based on genetic code variations, which are reflected in varying severity of symptoms, studies have shown that balance problem remained to be the mainstay reason in quality of life reductions [2]. Due to the appearance of all these symptoms, it is still a challenge in prioritizing problems to be managed in order to provide the best impact. One common language that is generally used as a functioning concept in physical medicine and rehabilitation field, is known as the International Classification of Functioning, Disability, and Health (ICF) concept [3]. Several investigations had shown that utilizing this concept in rehabilitation

would substantially enhance success due to better prioritization of problems. Although rehabilitation had been demonstrated to be a cornerstone in ataxia therapy, reports about ICF in SCA rehabilitation is still very much lacking [4].

While rehabilitation could alleviate several symptoms of SCA, many concerned if these therapies could match the speed of natural progression in this disease [4, 5]. As a general overview of rehabilitation interventions, it could be majorly divided into technique based rehabilitation exercises, and the utilization of modalities [2, 6, 7]. Exercises are then more specifically classified to each professionals in the rehabilitation team, namely physiotherapist, occupational therapist, and speech therapist. Each of the rehabilitation team play an essential part towards the holistic care of SCA subjects, and hence has to be well apprehended. On the other hand, modern therapy modalities have emerged as adjuncts to conventional therapy [8, 9]. These newer modalities are targeted to improve neuroplasticity in SCA subjects as the residual brain potential towards better functioning [10]. Therefore, this chapter is dedicated to review the comprehensive management of SCA from the physical medicine and rehabilitation point of view.

2. International classification of functioning, disability and health (ICF) concept on spinocerebellar ataxia

Over the years there seem to be growing evidence on the language of physical medicine and rehabilitation, especially in defining disability and its impact to both the individual and the society they live. The most recent terminology utilizes the International Classification of Functioning, Disability and Health (ICF) concept [3]. The ICF concept is a supplement of the 10th revision of International Statistical Classification of Diseases and Related Health Problems by World Health Organization (ICD X), that comprehensively describes an individual's health condition while still accounting their performance in community [3]. This concept could then ease physiatrists in creating both treatment goals and therapy focus which are tailor made for the individual.

Utilizing ICF in the daily practice requires the use of several core sets which are specified for the disease, but this however has been a challenge as not all diseases have their specific core sets published. As SCA have no specific core set yet, it is then recommended for physiatrist to adapt to an existing core set which has similar properties, and chronic stroke core set would seem most fitting to an ongoing central neurological disorder [11]. Aside from utilizing the core set, it is important to focus on some components of the ICF concept that could become the focus of the treatment plan, emphasizing on setting achievable goals by using several measuring tools.

In a glance it could be seen that the ICF concept starts with describing the body structure and body function after stating the diagnosis [3]. Afterwards, activity and participation should be listed as to describe the individuals' challenges in performing activity of daily living or even in the community level [3]. The next subsection of ICF involves description of environmental factors that would affect the individual, be it from physical environment or the community they are involved in [3]. Last but not least, personal factors should be addressed as well, knowing that adherence and motivation would affect the success of a rehabilitation program.

2.1 Body structure and body function

The focus of body structure in spinocerebellar ataxia is obviously the cerebellum, in the spinocerebellum (center) portion. It is known that the spinocerebellum

gathers a large volume of sensory information from the peripheral organs, as well as relaying information from the motor cortex [12, 13]. Etiologically this is caused by autosomal dominant mutation on the SCA gene, and this would disrupt the connection between multiple layers of cerebellum [1]. Prior studies had shown that severity of symptoms would correlate to the cerebellar areas involvement, and differs between various types of SCA [2, 7, 14]. Ultimately these changes result in functional disturbances as well as learning difficulties [15, 16].

One of the main body function disorders in SCA that should be addressed is balance and postural control [9, 17]. Consistently studies had shown that balance is disturbed both during static and dynamic, hence implies difficulty in performing effective gait, and maintaining standing position [9, 18, 19]. Since cerebellum also becomes a relay center for agonist and antagonistic complimentary contractions, it is natural to see that spinocerebellum lesion would affect effective voluntary muscle contractions, which could also present as central hypotonia [4, 6]. As discussed earlier, learning difficulties in SCA would span its impact from impaired conditional skill learning, up until reduced ability in adapting changes from environment [15, 16]. All these errors in cerebellar signaling would also result in poor coordination, as there are several mismatches in relaying sensory information to produce an effective motor response, as well as poor intra-limb coordination in spinocerebellum damage [20, 21]. Limited study are available in exploration of autonomic function disorder, and it was reported that overactive bladder is most commonly seen in SCA 2 [22]. Therefore, despite the local extent of damage in body structure, the functional impact is notoriously destructive.

2.2 Activity and participation

Having the body function described, it could be inferred that there would be a wide range of activity and participation disorders in SCA patients. One of the most reported problem in activity would be gait efficacy, as the lesion will interfere limb advancing patterns, as well as poor alternative terrain adaptability [8, 19]. Despite the inferior quality of life in motor control, the patients could still communicate as there are supposedly no barrier in this [23]. Even so, several studies revealed that cognitive impairments are found in SCA with varying severity [15, 16]. A number of studies have also reported that difficulty in verbal memory, learning, and fluency are commonly seen [16, 24]. These cognitive disorders would sum into a restriction in several community participation, but motor abilities still remained to inflict the most significant effect to quality of life in SCA [25]. Reports have mentioned the involvement of visuospatial and executive functioning abilities being reduced in SCA3, as it may correlate with reduced cerebellar perfusion [16]. Learning abilities in particular, were consistently shown to be retained in prodromal SCA2, as neural plasticity may still be prominent with Brain Derived Neurotrophic Factor playing its role, and other parts of the brain compensating for the functional deficit [15]. These evidences of preserved learning abilities in progressive disorders, must be recognized in the highest priority, knowing it would be the key to efficient rehabilitation for SCA cases [26–28].

2.3 Environmental and personal factors

The main environmental factor issues that was addressed for SCA is the difficulty to walk on varying level terrains [13, 29, 30]. It should be remembered that stable walking in varying level terrains require several functions ranging from cognition, vision, limb control, and balance. Severe fatigue was also seen in 69% of SCA patients, and thus different SCA types would result in different lesion focus

and function disorder [31]. Moreover, since learning abilities are also compromised, a combination of these symptoms would ultimately result in terrain adaptation barriers [1, 4]. These should be identified in each patients, and correlated to their living environment in order to formulate an effective intervention.

Depression were consistently reported in several studies on SCA, and this could correlate significantly with quality of life [22, 30]. As mentioned previously that mobility is the main concern, depression levels were also seen lower in those subjects with better mobility [6, 25]. Even when other causes of depression may revolve around memory and learning ability disorders, most SCA subjects would have learning difficulty in limb control, which becomes a vicious cycle and a hazard for them to perform well in mobile activities of daily living [5]. Therefore, early detection of depression is important, especially in the personal factors subsection of ICF.

Although physical medicine and rehabilitation approach to SCA might differ between studies, it's observable that the main focus is always towards body function, activity and participation. This focus is generally uncommon to be seen in the published studies, since most of these studies would focus on exploring various types of SCA, such as SCA1, SCA2, SCA3, SCA6 and SCA7 that are commonly found in the community [1, 7, 14, 27]. Shortly put, rehabilitative approach to SCA would place its greatest weigh on identifying disorders of body function through physical examination, rather than determining the SCA type through genetic testing [6]. In any case, each individual must receive tailor made interventions even when they are in the same SCA type.

3. Rehabilitation strategies for spinocerebellar ataxia

In response to the stated functional problems, various rehabilitation strategies have been implemented and studied over the years [2, 6, 7]. Surely mobility and balance interventions have been one of the main focus in SCA studies, however through time, studies have widened their range of focus into endurance, cognition, and speech [30]. Rehabilitation strategies may be a combination of several mode of interventions, such as exercise, physical modality, and sensory stimulation [9, 14, 27].

Comprehensive examination is required prior to these interventions, as each patients should receive tailormade intervention, and thus not all of these strategies are administered to all patients. Through utilization of ICF conceptual analysis, then clinicians should focus on body function problems identified in SCA patients, as shown on **Table 1** that depicts the common functional problems seen in SCA subtypes. This subsection will discuss these strategies in detail to give an overview of what is being studied in the published studies.

3.1 Physiotherapy interventions

Majority of the published studies have mentioned how physiotherapy plays a big role in mobility interventions of SCA patients, be it through conventional therapy or through exergames and virtual reality [9]. Since physiotherapy intervention is primarily focused on achieving better gait control, it would naturally revolve on improving balance, strength, endurance, and posture simultaneously [18, 27]. This finding has resulted into a more focused exercise sessions in the recent studies, aiming mainly on trunk balance, as these would be positively reflected in significant improvement of Scale for the Assessment and Rating of Ataxia (SARA) score [19, 38].

Although there are no published guidelines on SCA mobility exercise, several published studies from Cuban Centre for the Research and Rehabilitation

No	Function	SCA Type
1	Sensory and Motor Cortex [13]	6
2	Ataxia & Cognition [15]	2
3	Falls, Balance Impairment, & Functional Mobility [29]	1, 2, 3, 6
4	Non-motor & Extracerebellar [22]	2
5	Cognitive [24]	6
6	Action perception cerebellar recruitment [26]	6
7	Dystonia [32]	1, 2, 3, 6
8	Cognitive & Socio-cognitive [16]	1, 2, 3, 6, 7
9	Clinical & Genetic of Brain MRI Changes [33]	1, 2
10	Motor & Cognitive – brain volume [34]	7
11	Autonomic Function [35]	2
12	Non ataxic manifestations [36]	2
13	Dysphagia [37]	3, 6

Table 1.
Spinocerebellar functional problems in common SCA types.

of Hereditary Ataxia (CIRAH) had effectively shown the benefits of intensive neurorehabilitation as they have conducted [18, 27]. The whole therapy lasted for four hours per day, five days per week, lasting for 12 weeks in total, hence to the authors' knowledge, this is the longest exercise duration seen per day. Daily tasks include both physiotherapy and occupational therapy interventions, with several breaks in between to restore both energy and training focus. It could be resumed that majority of the exercises given in CIRAH's daily tasks include static balance improvement, and positional changes, all to improve trunk control and complement daily living tasks being trained by occupational therapists. Another important component that should be noted is the coordination exercises, which trains intralimb coordination [18, 27]. These sets of exercises had proven to be very effective in improving cerebellar symptoms, as reflected in constant improvement of SARA scores of both SCA subjects in early prodromal stage and SCA2 diagnosed 11 years mean post onset [18, 27].

Other studies had utilized shorter sessions as compared to the CIRAH neurorehabilitation schedule, but all these had shown significant improvement of SARA scores. One such study reported that partial Body Weight Supported Treadmill Training could improve balance significantly, and general positive trend in improving mobility, endurance, and quality of life [19]. It could be possible that these studies alike are more focused on providing intervention in trunk control, which is parallel to the fact that trunk ataxia has better prognosis as compared to limb incoordination in SCA. Another concern lies that there are controversies in the outcome measurement of SARA scores, as they are very sensitive in detecting cerebellar symptoms, but not for extracerebellar symptoms. Despite those controversies, SARA scores could still be utilized as it correlates closely to functional abilities, and thus would pertain to be an effective evaluation tool in SCA studies.

Additionally, consistent evidence revealed that trunk ataxia could have better rehabilitation prognosis as compared to limb ataxia [8, 39]. The main problem persists that the rate of degeneration at every year must be matched with beneficial gains from exercise, and thus effective regimens would be the primary choice as a rehabilitation goal. The natural progression of SARA score in SCA is noted to be

0.6 to 2.5 points per year, whereas it was shown in most studies how training would effectively reduce SARA at least by one point, displaying clinical importance of these interventions [5].

3.2 Virtual reality and exercise games

It could be acknowledged that maintaining the provided gains from exercise is of importance in degenerative disorder [2, 6]. Several recent studies had revealed how exergames (exercise games) and immersive virtual reality would be able to fill up this shortfall in conventional therapy [8, 9]. Exergames here are considered adjuvant to the traditional physiotherapy, and could never replace their roles in ataxia rehabilitation [9]. An important feature in exergames that should be highlighted, is the fact that there are rapidly changing environment, thus demands an accurate anticipation from the ataxic cases, providing excellent gains to the sensorimotor system [9]. These anticipation were shown to correlate with real life situations, and could effectively maintain exercise effects throughout longer period [8, 9].

Proper choosing of modalities would benefit SCA subjects in different stages, where early stages could follow high demand competitive sporting exergames such as ping-pong, badminton and squash [8]. These exergames and virtual reality should be performed on elastic carpet, as it's shown to give additional benefits in improving coordination and postural control through proprioceptive feedback. More severe ataxia would not allow them to play on competitive exergames, and would obtain greater advantage from good postural control. Games such as tightrope walk, which requires the user to maintain a specific position while still advancing forward, had been reported to effectively enhance both static and dynamic balance [8]. On the other hand, mild to moderate stages would benefit from conventional coordinative physiotherapy and severe stages though have no clear guidelines yet [9]. Hence it's shown how exergames would play its best role in early stages, and also to maintain the gains accrued from conventional physiotherapy. As stated previously that studies had shown how mobility learning mechanisms may still be preserved in SCA cases, these newer therapies would then be targeted to hone these adaptive skills and apply them to their daily situations.

3.3 Trunk and intra-limb control

While many studies had shown how trunk control could have many exercise options, improving limb control has been shown as a big challenge in SCA subjects. It was also reported that good intra limb control is best seen in walking analysis, observing their coordination in performing effective transition from single and double leg stance [19]. Several studies had shown that static cycling would be an effective intervention to improve intra limb control [20, 21]. A controlled trial comprising of four week long cycling exercise was reported to restore the ability in modulating H-reflex inhibition, and is also correlated with better functional performances [40]. Although the impact was not as major as their healthy control counterpart, it could be seen that cycling would present itself as a potential exercise option for improving coordination in SCA subjects [40]. Added effects of endurance and strength gains through stationary cycling has also been reported, especially in mobility disorder patients such as cerebral palsy [41].

Postural exercise approach are generally based on "re-learning" strategies of destabilizing responses, that anticipatory movements are trained in various environments, as well as honing of sensorimotor reflexes in the light of preserved plasticity [9, 13]. Postural instability would also lead to chronic low back pain, and thus stretching must also be given in order to alleviate these before and after

training sessions [9]. One voxel based study had shown 2 weeks of postural training would lead to improvement of balance, which was maintained for 3 months after training. Additionally, gray matter volume would also increase, and interestingly it is on the non-affected areas, meaning to say that targeted plasticity lies in the cerebral areas to compensate their cerebellar loss [28]. The study had mentioned that dorsal premotor cortex obtained the most compelling change, as they project to primary motor cortex and cerebellum, all of which are involved in movement planning and motor learning. Both patients and controls demonstrated an increase in gray matter volume in temporal association areas, this may be due to the requirement of performing sequential actions, which would in turn stimulates procedural memories in both hippocampus and basal ganglia. Cerebellar changes post exercise are not seen in cerebellar degenerative disorders as expected, but is significantly seen in healthy controls, with parallel increase in visuospatial and temporal inputs. These would then show how interventions towards premotor cortex growth should be the main goal of exercise in SCA subjects in preserving mobility [28].

3.4 Occupational therapy interventions

Besides physiotherapy interventions, occupational therapy is another important modality within the attempts of improving quality of life in SCA subjects [9, 42]. As mentioned previously, occupational therapy is usually incorporated with physiotherapy courses [18]. In the big picture, they should be given after warm up stretches to obtain better postural control during the specific exercises. In the published studies, mainly occupational therapy intervention would have a one hour duration, and this addition have been proven effective in improving both SARA and Functional Independence Measure (FIM) scores. The program itself consists of basic activity of daily living exercises, which are essentially a part of the FIM and Barthel Index Scoring sheet. Some of the examples include dressing activities such as tying shoelaces, buttoning shirts; tabletop instrumental activities for instance inserting sewing needles, drawing, cutting paper figures, using keyboards; and finally communication activities like reading texts out loud, commenting and interpreting verbal and textual information [18].

Despite the promising effects, studies focusing on occupational therapy as an individual therapy is still lacking due to the progressive nature of the disease, thus could only be shown as an additive effect to the proven effective physiotherapy. A study had shown that occupational therapy would improve Hamilton scores for depression in SCA3, and this improvement was independent from its confounders [42]. So far there are no studies yet on occupational therapy being a home program, but it could be speculated that more frequent practices would eventually trigger better quality synapses in the brain, leading to a more superior functional improvement. Therefore, aside from improving the functional abilities, it could also be inferred that occupational therapy would enhance self-confidence, alleviates depressive mood, and thus forecasts better participation [9, 42].

3.5 Speech therapy interventions

In order to fully complement the comprehensive care of SCA patients, speech and language pathology should be addressed to achieve well-being [23]. However, there are very few studies that discusses this, since only few types of ataxia that has bulbar involvement and thus results in dysphagia due to excess salivation [43]. A study had shown and compared how dysphagia is more severe in SCA3, whereas mild dysphagia is seen in SCA6 [37]. Possible treatment options would depend on

related problems, but mostly rehabilitation targets is to increase willingness and independence [23, 43]. At the same time, swallowing exercise would also aim in cueing patients to gain self-recognition in their swallowing process, thus triggering anticipatory self-evaluation [23, 43]. Since cognition is also an identified problem in SCA, this could also be identified by the speech and language pathologist, and self-corrections cueing prove to be effective [23]. In a more severe cases, safe swallowing practice along with appropriate dietary modification may be done in order to prevent aspiration [23]. Despite the scarcity of studies, a Cochrane review on speech disorder treatment for hereditary ataxia revealed that all the rehabilitative interventions have been reported as safe, and hence should be recommended in the comprehensive care [43].

3.6 Neuroplasticity and therapy duration

Clinicians should always remember that neuroplasticity plays a big role in alleviating SCA cerebellar signs, as proven in SCA2 subjects [10]. Unfortunately, there are still no reported significant effect on non-ataxia signs [28]. It also appears that SCA2 subjects may have more progressive disorders, and thus 24 weeks of therapy was suggested, whereas for other types such as SCA6 and SCA31, 4 weeks of training may already show better SARA score improvements [27]. Thus, the extent of affected area in the cerebellum would correlate directly to the progressiveness, hence would warrant different sets of rehabilitation strategy. Recognizing the neuroplasticity potential of SCA individuals would assist clinicians in identifying therapy focus, as well as motivating patients and family members to improve exercise adherence.

3.7 Essential points in rehabilitation strategies

Although there is no general guideline on this, rehabilitation strategies would adhere to functional disorder basis, that each strategy is given only when the disorder is identified [1, 6, 9]. Due to the progressive nature of the disease though, it is also plausible to administer the intervention even when the disorder have not emerge, knowing the fact that it may alleviate functional deterioration in the future. Two things that should be remembered are that fatigue may be one of the limitation in performing all the available strategies, and secondly, maintenance in quality of life must always be upheld [30]. Despite not many studies focused on quality of life, clinical experiences showed that progressive disease rehabilitation interventions should emphasize on giving life to the rest of the years, rather than adding years to the remaining life.

4. Rehabilitation goals and expected outcome measures in spinocerebellar ataxia

Aligned with their natural progression of disease, rehabilitation goals in degenerative ataxias would differ significantly from acquired ataxias. It was shown that acquired ataxias such as in stroke cases, would come with focal ischemia, ergo a better prognosis as compared to the diffuse lesion in SCA [1, 44]. Additionally, the degenerative process of SCA is of the highest concern, therefore it must be addressed and evaluated with valid measuring tools. Several subjects that were focused in prior studies include functional abilities, mobility function, balance, endurance, and quality of life [30].

4.1 Disease related tools

Ataxia specific tools such as SARA and Inventory of Non-Ataxia Symptoms (INAS) are most commonly utilized in many studies [45, 46]. The SARA score is designed to assess cerebellar symptoms semiquantitatively, and exclusively only SCA subjects were tested during the validation process. The SARA score ranges from 0 to 40, higher number showing more severe ataxia, the values then reflect eight physical examination items each with specific numeric scores. Physical examination of gait, stationary standing, and sitting position are observed, with a cut off of maintaining 10 second stationary position without difficulty as sufficient. One common bulbar component of SCA being speech production is also evaluated, and will be scored worst if the subject could only do unintelligible speech during normal conversation. Last but not least, performance of coordination tests namely finger chase, nose to finger test, dysdiadochokinesia, and heel shin test are graded with a score of 4 as the worst performance. As could be seen in these scored items, all of these are included in the general rehabilitation examination of cerebellar symptoms, and therefore this score is very much applicable in daily practice. Consistently it was shown that SARA score would correlate closely to symptom severity, and thus could be practically used to evaluate the efficacy of rehabilitation program [45].

With that being said, the main limitation of SARA would be that other scorings are required in the light of addressing extracerebellar symptoms. Therefore, the same research group had devised INAS score which could quantify the presence and severity of non-ataxia neurological symptoms [46]. The inventory consists of 30 items that is divided into two main section, the first spans widely from addressing cerebellar oculomotor signs, spinal reflexes, upper and lower motor neuron signs through physical examination. The second section on the other hand, lists the possible symptoms that the patient might bring about, such as double vision, dysphagia, urinary dysfunction, cognitive impairment, and other related findings that have not been listed. Similarly, the INAS scoring was also validated by utilizing SCA subjects with varying types, namely SCA1, SCA2, SCA3, and SCA6. Among these SCA types, it was reported that SCA1 and SCA2 presents extracerebellar symptoms along with the baseline ataxia, thus they are good candidates for the INAS, while SCA6 being purely cerebellar would play its role as control. The summation of the score is called INAS count, in which they have concluded that both INAS and INAS count shows good reproducibility, but unsatisfactory responsiveness over extended period due to the wide variation of measurement [46]. However it was clearly shown that INAS is an excellent supplement to the SARA score for SCA subjects.

4.2 Functional measurement tools

Other studies had also shown that in very early ataxia stages, both SARA and INAS are ineffective in prodromal stage [18]. Functional test alternatives such as tandem gait for 5 meter test was suggested to be used, as it is very sensitive to changes post rehabilitation [18]. The tandem gait itself is a complex task which may not be performed well by all SCA subjects, therefore traditional balance assessments such as Berg Balance Scale (BBS) [17, 25, 44]. The BBS consist of 14 item list, with an ordinal scale of 0 to 4, higher number meaning better balance function. The main categories in the item list revolves around maintenance of stationary position, transfer, and change of position while performing simple activities. Summation of all the scores for less than 45 would indicate greater risks of falling [47]. In cases that the BBS is not used in total, the components could also be used individually to monitor a specific progression within therapy evaluation.

Another study had also utilized the timed up and go (TUG) test to evaluate balance and function in degenerative ataxia subjects [20, 21]. In several rehabilitation trials, TUG test are well preferred due to their ease of examination, quantification of results in seconds, and finally their best representation to daily living tasks. However in cases of SCA, probably the complete TUG test might not always be performable due to high risk of fall. Several studies had also utilized expanded TUG test, which divides the full TUG test into segments measured by milliseconds, namely sit to stand, gait 1, turning, gait 2, and stand to sit [48, 49]. By separating these components, physiatrists would have a better view on which component are hindering the subject in achieving good TUG performance, and at the same time, would be able to assess improvements more accurately. Although the expanded TUG have not been utilized in SCA studies, it has been commonly used in other chronic neurological cases such as stroke, and hence should be recommended for future studies on degenerative ataxia [48].

Aside from TUG test, a more comprehensive functional test tool such as functional independence measure (FIM) are commonly used in SCA studies [25]. The utilization of FIM had expanded the view on functional activities and illustrate their level of independence in those activities. The FIM tool comprise of several components such as bowel, bladder control, transfer, locomotion, social participation, communication and also self-care activities [50, 51]. These components will then be graded from 1 to 7, when value of 6 and above shows complete dependence, scores 3 to 5 shows moderate dependence, and lastly below 3 shows full dependence. Therefore, this tool would be best used when the subjects are not fully independent, and other individuals such as caregivers are involved. Naturally FIM would have a ceiling effect when the patient is fully independent, and there are no additional scoring for the performance quality.

In SCA subjects it was reported that reduction of 1 point in FIM score would be significantly reflected in 4.49 point decrease in the physical functioning of Short Form 36 (SF-36) score [25]. In the light of SF-36, it is the most commonly used tool to assess quality of life in SCA subjects. As the name implies, this tool has 36 questions which covers eight domains of health, for instance limitations in physical activities, social activities, role function, pain, emotional problem, mental health, fatigue, and finally general health perceptions [52]. Various ordinal options in each questions should firstly be calculated through a formula to obtain domain scores. This finding then reveals how FIM could also be used to assess overtime changes that would complement the changes in other ataxia specific tools, in which better mobility correlates with greater quality of life [25, 53].

4.3 Fatigue and endurance measurement tools

In relationship to quality of life, fatigue is pretty much prevalent and thus is essential to note. Aside from SF-36 that touches on the fatigue concept, the fatigue severity scale (FSS) is a specific 9 item scale which measures fatigue in a 1 to 7 scale, 7 being strongly agree with the fatigue item being stated [31]. Accomplishing the FSS requires only 5 minutes, but the questions would not accurately direct the underlying functional disorder beneath, especially in chronic cases where fatigue is evident. Therefore, both cardiovascular and respiratory specific tools must be administered separately in order to evaluate through time. There are a selected number of studies that discuss the changes of cardiorespiratory attributes through evaluation of maximum oxygen consumption (VO2 max), six minute walk test (6MWT), peak expiratory flow (PEF), and maximum inspiratory pressure (MIP) [21, 38, 54].

Evaluation of VO2 max is done by performing ramped ergometer exercise stress testing, while 6MWT could be performed with assistance if the subjects are unable

to [21]. With all these limitations, it could be possible that evaluation of cardiovascular function will not be optimal owing to the natural progression of the disease and obstacles in maintaining stationary position. On the contrary, respiratory function has shorter examination time, allowing better examination compliance [54]. A recent study had shown how examination of both PEF and MIP are safe to be performed in SCA2 subjects, when better respiratory function seemed to correlate well with Activity of Daily Living scales, and ataxia specific SARA scales [54]. Additionally, the study had also reported that one third of the subjects complained of dyspnea, with interpretation of restrictive pulmonary disease. It is speculated that the restriction may be caused by the lack of coordination of the respiratory muscles, ultimately resulting in reduced chest expansion [54]. On the other point of view, postural control exercises would be able to improve diaphragmatic excursion, and thus provide attempts in correcting respiratory dysfunction [55–57]. Therefore, it is only possible that physiotherapy interventions to manage ataxia related symptoms also alleviate respiratory symptoms, thus very mild respiratory dysfunction that could be reported.

5. Conclusion

Being a progressive degenerative disease, physical medicine and rehabilitation have an important role in alleviating symptoms as well as improving quality of life in Spinocerebellar Ataxia [4]. Initial analysis of SCA begins with identifying the ICF concept, in which body structure being cerebellum, and several body function problems such as postural control and intra-limb control should firstly be addressed [8]. These underlying disorders would lead to below par daily living performance, further leading to restriction in participation, and might also result in depression [25, 30]. Nevertheless it should be remembered that there is a natural progression of functional decline, which is unavoidable in SCA [1]. Therefore, rehabilitation goals are generally focused in maintaining functional capacity, as well as improving social participation and quality of life [4].

Achieving the aforementioned rehabilitation goals could be done through several interventions, but it was shown that physiotherapy exercise sessions focused in improving posture, balance, and gait had proven to be the most effective. Duration of session generally lasts for 1 hour or more, but it should be preceded with stretching to ease pain and provide better proprioceptive feedback [18]. In order to enhance retaining of the exercise gains, proper choices of virtual reality and exergames could be done [8, 9]. Most studies have also incorporated occupational therapy that rehearses daily living activities, and it's seen to correlate well with quality of life [42]. Simultaneously, speech therapy would also play its role in SCA by managing communication and swallowing disorders that are present in several types of SCA [43]. Several valid outcome measuring tools have been shown effective to monitor changes over time, and it should be remembered that these measures could assess specific components that are being trained [22, 30]. In conclusion, effective rehabilitation approach should comprise of all the previously mentioned components, while always being validated by specific outcome measure tools. In addition to that, further studies should devise a guideline for general rehabilitation of SCA through validated trials.

Conflict of interest

The authors declare no conflict of interest.

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Author details

Kevin Triangto[1*], Steven Setiono[2] and Herdiman Bernard Purba[2]

1 Cardiac Prevention and Rehabilitation Department, National Cardiovascular Center Harapan Kita, West Jakarta, Indonesia

2 Physical Medicine and Rehabilitation Department, Cipto Mangunkusumo Hospital University of Indonesia, Central Jakarta, Indonesia

*Address all correspondence to: kevintriangto14@gmail.com

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