



Ten Steps for NPH Management: Advancements in Diagnosis and Treatment of Adult Hydrocephalus

Dez etapas para o manejo da NPH: avanços no diagnóstico e tratamento da hidrocefalia em adultos

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Abstract

Objectives The authors of the present study intend to describe a straightforward protocol for normal pressure hydrocephalus diagnosis and management, with the employment of a multidisciplinary team approach effort.

Methods Using a strict methodological approach for initial diagnosis, taking into consideration occupational therapy and physical therapy assessment, the authors have set out to elaborate a simple protocol for suspicion and, once diagnosed, treatment of normal pressure hydrocephalus. We have used the MoCA (Montreal Cognitive Assessment) and walking assessment that included speed, independence, and distance (SID), 10 m walk test, TUG (timed up and go) evaluation, 6-minute Walk Test, MiniBESTest, as the main factors for pre and post lumbar drainage assessment, after which, the alternatives were deliberated and followed, or not, by ventriculoperitoneal shunt insertion.

Results The authors have described a protocol, consisting of ten easy steps, which involves a multidisciplinary team, including occupational therapy and physical therapy professionals, as well as neurologists and neurosurgeons for improved and objective assessment prior to insertion of lumbar drain and, thereafter, detecting the population at most benefit for ventriculoperitoneal shunt insertion. We have described the Ten Step Approach for Normal Pressure Hydrocephalus management, including from initial clinical presentation and imaging, to pre and post lumbar drainage, for lastly deciding upon necessity for ventriculoperitoneal shunt insertion.

Conclusions A straightforward protocol for normal pressure hydrocephalus seems not only feasible, but simple to implement in most neurosurgical departments, with good accuracy of prediction of lumbar drainage assessment to shunting outcomes.

Keywords

- ▶ normal pressure hydrocephalus
- ▶ ventriculoperitoneal shunt
- ▶ idiopathic normal pressure hydrocephalus
- ▶ adult hydrocephalus

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Introduction

Normal Pressure Hydrocephalus (NPH), also known as Idiopathic Normal Pressure Hydrocephalus (iNPH), is a defective accumulation of cerebrospinal fluid (CSF) in the intracranial ventricles of the central nervous system. As the name suggests, pressure assessment varies within a normal limit range, at times with slight elevation only, although volume within intraventricular spaces seems to be significantly enlarged. Clinical presentation is commonly straightforward, with a triad at presentation being common, including gait dysfunction, sphincter control abnormalities and cognitive impairment, which has a tendency for progression if not treated.¹

Aside from the now notorious clinical presentation, imaging findings provide substantial information into the condition. Initially, the Evans Index (EI) larger than 0.3 in brain MRI had been described as reliable measurement for ventricular dilation, however being common to other subsets of ventricular enlargement, including ex-vacuo ventriculomegaly for instance. Further measurements, thereafter, have risen as promising tools, such as the measurement of the callosal angle (50–80°), which proved to be more reliable for iNPH.² Although an integral part of NPH assessment, imaging by itself cannot solely predict improvement in patients after cerebrospinal fluid diversion. Albeit, assessing modifications of more objective measures may provide better understanding on potential for clinical improvement after cerebrospinal fluid diversion procedures. It is therefore necessary to evaluate objective changes in cognitive and gait after temporary cerebrospinal fluid drainage to understand which subset of this population would clinically benefit of CSF diversion intervention.³

Although significantly prevalent, especially in an aging population worldwide, with clinically relevant deterioration of quality of life, there is no standardization for appropriate management of NPH. Much has been previously described in medical literature in regard to diagnosis, prevalence and treatment of NPH,^{4,5} however, there is no set protocol for easy to follow and lastly to replicate in everyday practice. Therefore, the authors of the present study, taking as background departmental protocol, set out to describe ten easy steps to follow for best decision-making into filling this gap of the literature.^{6–8}

Methods

The authors describe an institutional protocol for idiopathic normal pressure hydrocephalus, encompassing ten steps for clear and easy-to-follow schema. The gait assessment is regularly performed by trained physical therapists (PT), experienced with gait dysfunction in various neurological conditions. For the evaluation of cognitive status, occupational therapists (OT) accustomed to assessing patients with a variety of neurological conditions. This protocol, due to its objectivity and straightforwardness, may be replicated in distinct scenarios of the neurological and neurosurgical practice.

First, a trained and experienced neurologist assesses patient upon first medical appointment, and scrutinizes his/her clinical presentation, with a thorough medical history and physical examination, focusing on a commonly described triad, which includes gait disturbance, sphincter issues (more commonly presenting with urinary incontinence) and memory deficits. For the clinical diagnosis, as previously described, it is not necessary that all of these findings be present or reported, but the presence of the common triad reinforces the probability of the clinical presentation of iNPH.

Second, appropriate imaging, including more importantly for NPH, brain MRI, with measurements of the Evans Index and of the Callosal Angle. An Evans Index greater than 0.3 and a Callosal Angle of 50 to 80° although not pathognomonic, associated with the above clinical findings, is significantly suggestive of NPH. The presence of trans ependymal transudation is not a common finding of iNPH, although its presence might be indicative of more recently decompensated hydrocephalus.¹¹

Third, when the diagnosis of iNPH is suspected and before deliberating on CSF drainage (be it lumbar puncture of lumbar drain insertion), subjective questionnaires and objective assessments are done. To begin, we start with a questionnaire, which involves 11 questions (NPH Log scale – see addendum A) broadly covering physical, cognitive and sphincter domains. The same questions are interviewed to the patient and closest caretaker, separately. The scoring is done as; strongly agree, agree, unsure, disagree and strongly disagree. Thereafter, occupational, and physical therapy teams are involved for objective cognitive and physical assessments respectively. Occupational therapy is responsible of evaluating patients' cognitive status using MoCA (Montreal Cognitive Assessment). Other cognitive testes may be applicable, such as the MMSE (Mini Mental Status Exam), however, in our institution MoCA is more widely applicable for the purpose of iNPH evaluation, as is also the case in previous literature reports.¹²

Fourth, unlike on the 10-m Walk Test described below, when patient starts standing up and initiates to ambulate once the evaluator instructs in that sense, for this evaluation, TUG (Timed Up and Go), patient initiates assessment in a sitting position and is instructed to stand up and start walking. Patient is assessed for balance for standing up before ambulating, and then walk ten meters, make a 180° turn, and walk back ten meters. Patient is assessed for timing of execution, as well as imbalance that may occur more commonly during standing up from the sitting position and when turning around 180°. Above three assessments can be done; in tandem with each other. If patients can complete above test satisfactorily, patients proceed to ambulation endurance test.

Fifth, physical therapy professionals start with more objective gait evaluation with the 10-m Walk Test. The 10-m Walk Test aims to assess for gait velocity. Patients are instructed to ambulate at normal speed, and then as quickly as safely possible, being assisted all the while (from a safe distance, but which does not impact directly on the evaluation). This is

performed for two trials and averages are taken, being recorded for the number of steps and time achieved after ambulating for the monitored ten meters, recording the average speed in meters per second.

Sixth, the 6-minute Walk Test (6MWT) is employed in the sense of endurance measurement, instead of velocity, as is the case in the 10-m Walk Test. Patients are instructed to walk at his/her normal speed, as far as he or she can walk within six minutes, and the results are reported objectively in distance in meters walked.

Seventh, still with physical therapy, the Mini Balance Evaluation System Test (MiniBESTest) is performed. Patients are objectively evaluated on 14 different items, with scores rated from 0 to 2, with a maximal of 28 points, with higher scores meaning better overall balance (see addendum B). On Timed Up and Go part of the test, the recorded information previously obtained may be used in the score for documentation and comparison purposes.

Eighth, although it has been previously described lumbar puncture as a means for effective CSF drainage, with drainage of different CSF volumes, usually at least 50 mL, our institution routinely employs a lumbar drain, as it seems more effective for continuous CSF drainage and being able to drain for more prolonged periods of time and larger volumes of CSF, which may ultimately influence more significantly the post drainage assessment. A total time of drainage as minimum as 12 hours, but more commonly 24 to 48 hours is routine. Lumbar drain is inserted in sterile settings, by experienced neurosurgeons, in the operating theater and under local anesthesia. CSF is aimed to be drained from 5–10mL per hour, which is a total of 120 to 240 mL in 24 hours, a volume estimated to produce consistent changes within the central nervous system to effectively be assessed in the post lumbar drain removal reassessment.^{12–14}

Ninth, after removal of lumbar drain, usually 30–60 minutes to allow patient to feel comfortable to carry out all the necessary steps, occupational and physical therapy are again involved to perform all the above-mentioned assessments, including repeating subjective questionnaire, which is re-interviewed with patient and same caregiver, at 12, 24 and 48 hours. MoCA, 10-m Walk Test, Timed Up and GO (TUG), 6-minute Walk Test (6MWT), Mini Balance Evaluation System Test (MiniBESTest) are all repeated in the same sequence previously performed. The results of reassessment are carefully described and compared with the pre-CSF drainage assessment and added to electronic medical records for documentation purposes and keeping objectivity.

Tenth, if results are significant for post CSF drainage improvement of at least 30% in comparison to pre-CSF drainage assessment, ventriculoperitoneal shunt (VPS) is indicated and performed as routine for other indications of hydrocephalus, with the use or not of neuronavigational system, depending on patients' specificities and availability. In our institution, the authors routinely use laparoscopic abdominal insertion of the distal portion of the VPS system, in order for direct visualization and possibly decreasing postoperative complications.¹⁵ A head CT is performed on postoperative day 1 for confirmation of appropriate proximal

position and to rule out any complication, and the patient is then discharged, with routine follow-up in two weeks for postoperative assessment.

Results

Upon deliberating on necessity for permanent CSF deviation procedure for NPH treatment, patient needs appropriate clinical assessment, with temporary CSF drainage and pre and post evaluation for final decision-making.^{9,10} An overall improvement of 30% of symptoms after drainage, is consistent with long-term and permanent improvement of daily life activities, supporting decision for an intervention that obviously carries risks, especially in regards to infection and obstruction, with, not uncommonly, necessitating for revision surgery in the future. This ten-step flowchart, therefore, is an easy-to-follow method for decision-making of such intricate and limited comprehended condition.

The below 10-step-list is presented for an easily reproducible protocol, which may be performed in different level complexity scenarios.

- 1–Clinical Presentation Assessment – experience neurologists to assess for clinical presentation suggesting NPH, usually looking for the triad findings of gait instability, urinary changes, and cognitive decline.
- 2–Imaging – imaging showing evidence of dilatation of intracranial ventricles, with Evans Index of > 0.3 and callosal angle of 50–80° is suggestive of NPH, although not conclusive (see image below – ►Fig. 1 – showing a callosal angle at the level of anterior commissure, of a callosal angle of ~65°).
- 3–MoCA and NPH Log Questionnaire – occupational therapy professional, with experience in various neurological conditions assess patients and documents scoring based on MoCA and NPH Log Questionnaires (addendum 1), which will be used as baseline for comparison after CSF drainage.
- 4–TUG (Timed Up and Go) – this step precedes the other walking tests, as this will evaluate for feasibility of standing up from a sitting position before ambulating. The test has shown evidence of predictive value for balance, velocity, and endurance of gait, which carries by itself great worth for this subset of neurological patients.
- 5–10 m walk test – physical therapists will evaluate for gait velocity, requesting patients to initially walk in his or her normal speed and later to walk as quickly as safely possible.
- 6–6-Minute Walk Test – this test, performed by PT professional experienced with gait assessment in neurological patients, evaluates endurance, measuring in meters, distance walked in six minutes.
- 7–MiniBESTest – this more complex test, which also measures for balance, endurance, and velocity of gait, is performed after TUG, 10 m walk test and 6-minute walk test since it is more thorough and complex, demanding more of the patient, meaning that it might be halted in certain cases, needing to be repeated in a

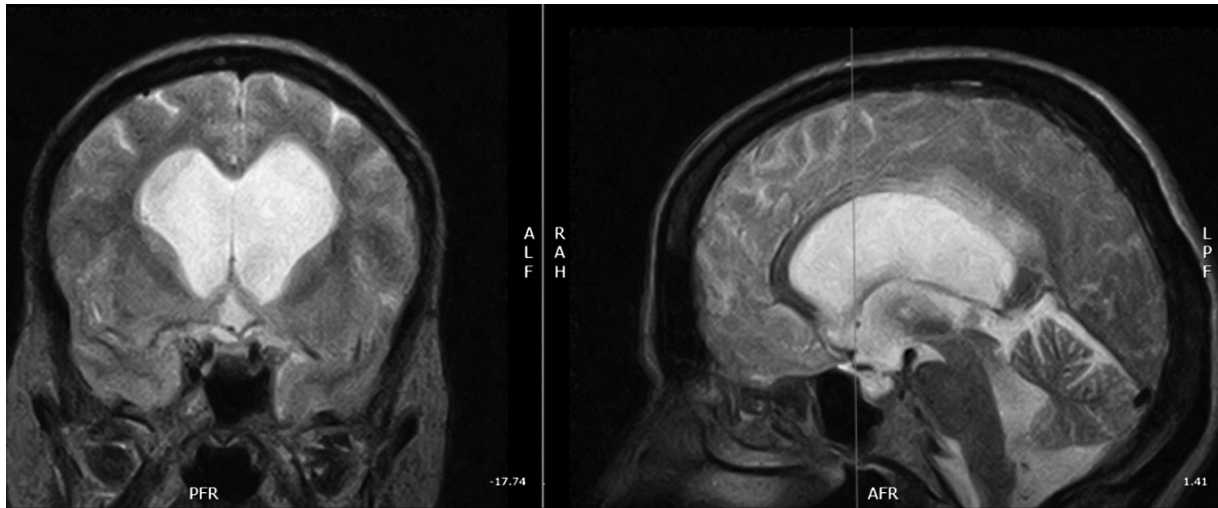


Fig. 1 Dilation of intracranial ventricular system, with an Evan’s Index of >0.3 and Callosal Angle of ~65°.

subsequent time. The score which ranges from 0 to 28, encompassing 14 questions scored 0–2, may use the TUG information to avoid repeating same tests and become too demanding for some patients.

- 8–Lumbar Drainage (assessment at 6–12–18–24 hours) – lumbar drainage is performed by experience neurosurgeons in operating theater setting, under sterile conditions and local anesthesia. In cases of difficult execution, due to previous lumbar surgical interventions or further anatomical nuances, the procedure may use fluoroscopy for improved outcome.
- 9–Post-CSF Drainage Assessment (Repeat PT and OT evaluation) – the cognitive and physical assessments are

performed in the same sequence that were performed before CSF drainage, usually waiting at least 30 minutes after lumbar drain removal. The results are recorded and then compared with the pre-CSF drainage assessment. An improvement that is deemed of more than 30% is viewed as positive and decision for VPS is made, with patient’s and/or family’s consent.

- 10–Ventriculoperitoneal Shunt (yes or no) – after decision for VPS is made and consent is obtained, procedure is performed in routine surgical standards, in our department with use of laparoscopic insertion of distal part of VPS catheter and under navigation for proximal catheter. – **Fig. 2**

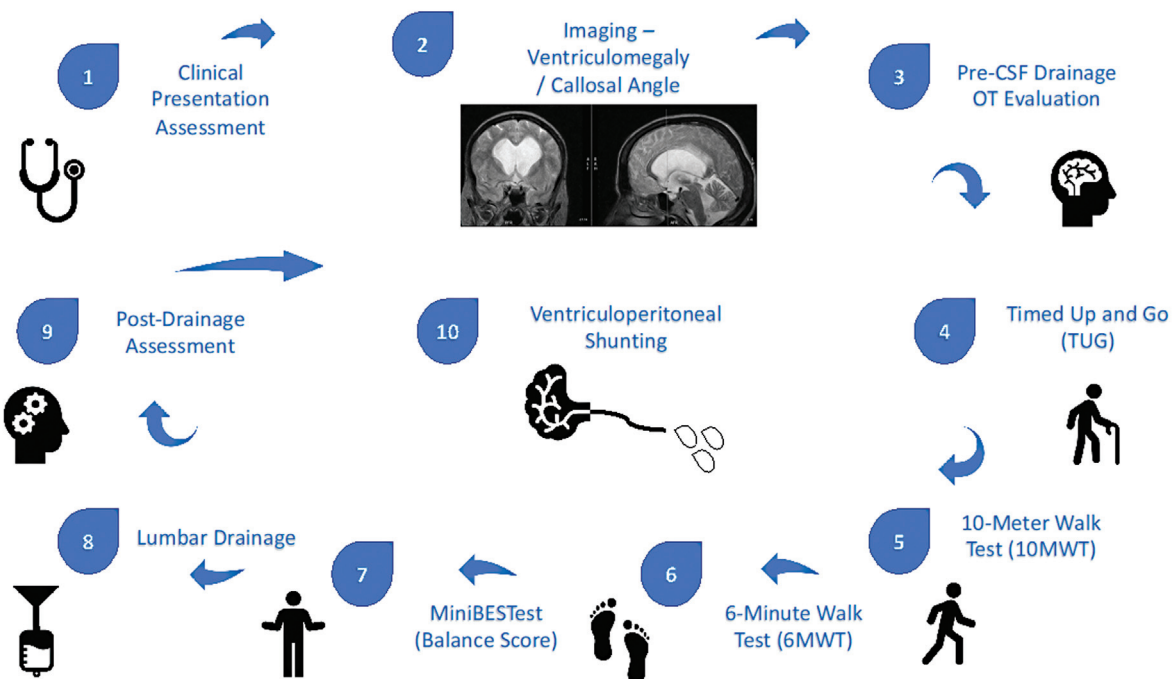


Fig. 2 10 Step for NPH Flowchart.

In our experience, the ten-step protocol has shown to be effective and reliable, for not only establishing the population at most benefit for surgical intervention with ventriculoperitoneal shunt (VPS), but also, and perhaps more importantly, those that would ultimately fail any intervention, halting any deliberation of potential risk averse procedures in such patients. As previously shown, the Timed Up and Go (TUG) assessment before and after lumbar drain insertion, has been decisive in many cases, especially for cases which the initial clinical presentation might have been borderline. The objectivity of pre and post temporary CSF drainage has been shown to support decision-making with potential to reassure patient and patient's family on indication, or lack of, ventriculoperitoneal shunt insertion, significantly decreasing, therefore, procedure-related complications, as well as those attributed to not managing normal pressure hydrocephalus in a timely fashion.

Discussion

Normal Pressure Hydrocephalus (NPH) or Idiopathic Normal Pressure Hydrocephalus (iNPH) is a known entity which courses with a triad clinical presentation of gait dysfunction, sphincter issues and memory impairment. Its diagnosis and treatment have been amply discussed previously, encompassing CSF drainage and CSF diversion, respectively. In the present study, the authors present an institutional protocol for best standard of management for NPH, with an easy to follow and apply Ten-Step Protocol for NPH, which may be employed in distinct level scenarios.

Each of the ten steps makes diagnosis, using clinical findings and imaging evidence,¹⁶ as well as cognitive assessment and gait evaluation, with velocity, endurance, and balance as its tripod basis, in two distinct phases, pre and post CSF drainage through lumbar drain,^{17,18} a simple manner for managing this complex array of patients, with the best possible outcomes.

When evaluating before and after cerebrospinal fluid drainage, it is of the utmost importance to keep objectivity, as patients' and relatives' information are commonly only a subjective account of patients' status, although not to be overlooked. That is the reason that the use of established measurements tools, such as the ones here described, including MoCA, 10-m Walk Test, TUG, 6-minute Walk Test, MiniBESTest, performed before and after CSF drainage by the same professionals and utilizing same environmental infrastructure, provides a controlled surrounding to minimize any unknown bias. Also, the use of at least 30% improvement from baseline assessment, while limiting several patients whose improvement may lay below this cutoff point, brings a more clinically significant outcome after permanent CSF diversion, with little room for subjective improvement observed in other scenarios.

This descriptive narrative thoroughly details each of the ten steps, from clinical presentation assessed by a trained and experienced neurologist, with corroborating imaging in brain MRI showing specifics for probable NPH, through evaluations of occupational therapy and physical therapy

professionals, before and after CSF drainage, for lastly supporting a conscious decision-making for final ventriculoperitoneal shunting, which should be the permanent treatment for the condition.²⁰⁻²²

Conclusion

Idiopathic Normal Pressure Hydrocephalus is a prevalent neurological condition, with significant influence on a large population's quality of life. Establishing an easy-to-apply protocol, such as the one herein described Ten-Step Protocol for NPH, may prove to significantly facilitate decision-making with potential great impact on this subset of neurological population.

Previous Presentations

The contents of this manuscript have not been copyrighted or published previously.

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Conflict of Interest

None.

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Addendum A

Normal Pressure Hydrocephalus (NPH) Log

Before performing the LP/Lumbar Drain and at 12, 24 and 48 hours after Lumbar Drain, the patients and caregivers were routinely asked to measure the patient's baseline performance status using questions in the LP log assessment form. The *patients and caregivers* were asked to state if they "*strongly agreed, agreed, were unsure, disagreed, or strongly disagreed*" with 11 statements describing patient's function regarding activities of daily living.

Pre LD (patient)	Pre LD (caretaker)	Questions	Post Lumbar Drainage	12 hours (Patient)	12 hours (Caretaker)	24 hours (Patient)	24 hours (Caretaker)	48 hours (Patient)	48 hours (Caretaker)
		1. I feel balanced							
		2. I feel confident walking inside and outside							
		3. I can stand up and sit down with ease.							
		4. I can walk up and down stairs and or hills with ease.							
		5. I have energy each day to complete my daily tasks.							
		6. I am easily able to make plans, problem solve, and move from one task to the next							
		7. I am easily able to pay close and continuous attention to tasks.							
		8. I have the motivation to perform daily chores, errands, and call or see my family and friends							
		9. I enjoy listening to music.							
		10. I have issues with my urinary urgency							
		11. In the past 3 months, I feel that I can process questions/commands/ requests that are made to me and react appropriately to them without delay or needing of repetition.							

Addendum B

Mini-BESTest: Balance Evaluation Systems Test

1. SIT TO STAND

Instruction: "Cross your arms across your chest. Try not to use your hands unless you must. Do not let your legs lean against the back of the chair when you stand. Please stand up now."

(2) Normal: Comes to stand without use of hands and stabilizes independently.

(1) Moderate: Comes to stand WITH use of hands on first attempt.

(0) Severe: Unable to stand up from chair without assistance OR needs several attempts with use of hands.

2. RISE TO TOES

Instruction: "Place your feet shoulder width apart. Place your hands on your hips. Try to rise as high as you can onto your toes. I will count out loud to 3 seconds. Try to hold this pose for at least 3 seconds. Look straight ahead. Rise now."

(2) Normal: Stable for 3 second with maximum height.

(1) Moderate: Heels up, but not full range (smaller than when holding hands), OR noticeable instability for 3 second

(0) Severe: < 3 second

3. STAND ON ONE LEG

Instruction: "Look straight ahead. Keep your hands on your hips. Lift your leg off of the ground behind you without touching or resting your raised leg upon your other standing leg. Stay standing on one leg as long as you can. Look straight ahead. Lift now."

Left: Time in Seconds Trial 1: _____ Trial 2: _____

(2) Normal: 20 second

(1) Moderate: < 20 second

(0) Severe: Unable.

To score each side separately use the trial with the longest time.

To calculate the sub-score and total score use the side [left or right] with the lowest numerical score [i.e., the worse side].

Right: Time in Seconds Trial 1: _____ Trial 2: _____ (2) Normal: 20 second

(1) Moderate: < 20 second

(0) Severe: Unable

R _ E _ A _ C _ T _ I _ V _ E _ P _ O _ S _ T _ U _ R _ A _ L _ C _ O _ N _ T _ R _ O _ L _____ -

S _ U _ B _ S _ C _ O _ R _ E _ : _____ / _ 6 _

4. COMPENSATORY STEPPING CORRECTION- FORWARD

Instruction: "Stand with your feet shoulder width apart, arms at your sides. Lean forward against my hands beyond your forward limits. When I let go, do whatever is necessary, including taking a step, to avoid a fall."

(2) Normal: Recovers independently with a single, large step (second realignment step is allowed). (1) Moderate: More than one step used to recover equilibrium. (0) Severe: No step, OR would fall if not caught, OR falls spontaneously.

5. COMPENSATORY STEPPING CORRECTION- BACKWARD

Instruction: "Stand with your feet shoulder width apart, arms at your sides. Lean backward against my hands beyond your backward limits. When I let go, do whatever is necessary, including taking a step, to avoid a fall."

(2) Normal: Recovers independently with a single, large step.

(1) Moderate: More than one step used to recover equilibrium.

(0) Severe: No step, OR would fall if not caught, OR falls spontaneously.

6. COMPENSATORY STEPPING CORRECTION- LATERAL

Instruction: "Stand with your feet together, arms down at your sides. Lean into my hand beyond your sideways limit. When I let go, do whatever is necessary, including taking a step, to avoid a fall."

Left

(2)

SENSORY

ORIENTATION

SUB

SCORE:

/

6

7. STANCE (FEET TOGETHER); EYES OPEN, FIRM SURFACE

Instruction: "Place your hands on your hips. Place your feet together until almost touching. Look straight ahead. Be as stable and still as possible, until I say stop."

Time in seconds: _____

(2) Normal: 30 second

(1) Moderate: < 30 second (0) Severe: Unable.

Normal: Recovers independently with 1 step (crossover or lateral OK). Moderate: Several steps to recover equilibrium. Right

(2) Normal: Recovers independently with 1 step (crossover or lateral OK).

(1)

(0)

Use the side with the lowest score to calculate sub-score and total score.

Severe: Falls or cannot step.

(1) Moderate: Several steps to recover equilibrium. (0) Severe: Falls or cannot step.

8. STANCE (FEET TOGETHER); EYES CLOSED, FOAM SURFACE

Instruction: "Step onto the foam. Place your hands on your hips. Place your feet together until almost touching. Be as stable and still as possible, until I say stop. I will start timing when you close your eyes."

Time in seconds: _____

(2) Normal: 30 second

(1) Moderate: < 30 second (0) Severe: Unable.

9. INCLINE- EYES CLOSED

Instruction: "Step onto the incline ramp. Please stand on the incline ramp with your toes toward the top. Place your feet shoulder width apart and have your arms down at your sides. I will start timing when you close your eyes."

Time in seconds: _____

(2) Normal: Stands independently 30 second and aligns with gravity.

(1) Moderate: Stands independently <30 second OR aligns with surface. (0) Severe: Unable.

DYNAMIC

GAIT

SUB

SCORE:

/

10

10. CHANGE IN GAIT SPEED

Instruction: "Begin walking at your normal speed, when I tell you 'Fast', walk as fast as you can. When I say 'slow', walk very slowly."

(2) Normal: Significantly changes walking speed without imbalance.

(1) Moderate: Unable to change walking speed or signs of imbalance.

(0) Severe: Unable to achieve significant change in walking speed AND signs of imbalance.

11. WALK WITH HEAD TURNS – HORIZONTAL

Instruction: "Begin walking at your normal speed, when I say "right," turn your head and look to the right. When I say "left" turn your head and look to the left. Try to keep yourself walking in a straight line."

(2) Normal: performs head turns with no change in gait speed and good balance. (1) Moderate: performs head turns with reduction in gait speed.

(0) Severe: performs head turns with imbalance.

12. WALK WITH PIVOT TURNS

Instruction: "Begin walking at your normal speed. When I tell you to 'turn and stop', turn as quickly as you can, face the opposite direction, and stop. After the turn, your feet should be close together."

(2) Normal: Turns with feet close FAST (< 3 steps) with good balance. (1) Moderate: Turns with feet close SLOW (>4 steps) with good balance. (0) Severe: Cannot turn with feet close at any speed without imbalance.

13. STEP OVER OBSTACLES

Instruction: "Begin walking at your normal speed. When you get to the box, step over it, not around it and keep walking."

(2) Normal: Able to step over box with minimal change of gait speed and with good balance. (1) Moderate: Steps over box but touches box OR displays cautious behavior by slowing gait. (0) Severe: Unable to step over box OR steps around box.

14. TIMED UP & GO WITH DUAL TASK [3 METER WALK]

Instruction TUG: "When I say 'Go', stand up from chair, walk at your normal speed across the tape on the floor, turn around, and come back to sit in the chair."

Instruction TUG with Dual Task: "Count backwards by threes starting at _____. When I say 'Go', stand up from chair, walk at your normal speed across the tape on the floor, turn around, and come back to sit in the chair. Continue counting backwards the entire time."

TUG: _____seconds; Dual Task TUG: _____seconds

(2) Normal: No noticeable change in sitting, standing, or walking while backward counting when compared with TUG without Dual Task.

(1) Moderate: Dual Task affects either counting OR walking (>10%) when compared with the TUG without Dual Task.

(0) Severe: Stops counting while walking OR stops walking while counting.

When scoring item 14, if subject's gait speed slows more than 10% between the TUG without and with a Dual Task the score should be decreased by a point.

TOTAL SCORE: _____/_2_8
