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RESEARCH ARTICLE

Striatal Hand Deformities in Parkinson's Disease: Hand Surgical Perspectives

Elisabeth Brogren, MD, PhD,[1,2,*] *Lars B. Dahlin, MD, PhD,*[1,2,3] *Erika Franzen, PT, PhD,*[4,5] *and Beata Lindholm, PT, PhD*[6,7]

ABSTRACT: Background: The knowledge about striatal hand deformities (SHD) in Parkinson's disease (PD), has recently increased but need more attention due to their early impact on dexterity. The focus of clinical studies has been on the staging of SHD severity and neurological features. However, a hand surgical perspective has not been considered.

Objectives: Our purpose was to examine SHD in patients with PD using hand surgical assessment methods and the recommended staging of SHD.

Methods: In this observational study, a specialist in neurological physiotherapy examined 100 consecutive PD patients and identified 35 with suspected SHD, who were then examined by two hand surgeons. Their hands were clinically evaluated for severity of SHD, according to a previous proposed staging, focusing on metacarpophalangeal (MCP) joint flexion, presence of intrinsic and extrinsic tightness, as well as other hand deformities.

Results: Three kinds of deformities were identified among 35 included patients: surgical diagnoses unrelated to PD (n = 5), SHD (n = 23), and PD related hand deformities with increased extrinsic tightness (n = 10); three of these 10 patients had also contralateral SHD, thus are included in SHD group. In addition to previously described MCP joint flexion, swan neck deformity and z-thumb deformity, we found in most hands finger "clefting," abduction of the little finger and/or an increased intrinsic tightness, indicating pathology of intrinsic muscles of the hand involved in SHD.

Conclusions: SHD diagnosed with a modified staging method, including features of intrinsic and extrinsic hand deformities, should be considered in PD to implement early and more accurate treatment.

A striatal hand deformity (SHD) is recognized by flexion of the metacarpophalangeal (MCP) joints, sometimes in combination with hyperextension of the proximal interphalangeal (PIP) joints.[1–3] The term "striatal deformity" has been used since the 70's to describe various abnormal parkinsonian postures of the hand, foot and spine.[3] Today, we know that lesions in the striatum (caudate and putamen) cause dystonia, but evidence supporting that "striatal deformities" are related to the striatum are in fact scarce.[3] Thus, SHD in Parkinson's disease (PD), occurring in 8–10% of PD patients,[4,5] is still relatively unrecognized and not fully understood.[1,4,5] It seems as SHD is poorly responsive to medication effects,[2] is not action-induced[6] and is still present with the hands relaxed and during sleep,[1,6] indicating that a SHD shares features with rigidity, although dystonia has been discussed in relation to the origin of SHD.[3,4,7] Also, other PD related deformities, such as "clenched fist"[8] or wrist flexion while walking related to motor fluctuations,[9] are closely related to dystonia. Interestingly, mild stages of SHD can be one of the earliest parkinsonian signs in untreated patients and the laterality of SHD is highly specific for the worst affected side of PD.[2–4] Patients with SHD are younger, have an earlier onset of initial parkinsonian symptoms, and have a more severe disease compared to their counterparts without deformity.[4,5]

SHD may be part of the primary PD process and significantly adds to disability in persons already coping with the complexity

[1]*Department of Hand Surgery, Skåne University Hospital, Malmö, Sweden;* [2]*Department of Translational Medicine, Lund University, Malmö, Sweden;* [3]*Department of Biomedical and Clinical Sciences, Linköping University, Linköping, Sweden;* [4]*Division of Physiotherapy, Department of Neurobiology, Care Sciences and Society, Stockholm, Sweden;* [5]*Women's Health and Allied Health Professionals Theme, Medical Unit Occupational Therapy and Physical Therapy, Karolinska University Hospital, Stockholm, Sweden;* [6]*Department of Neurology, Rehabilitation Medicine, Memory Disorders and Geriatrics, Skåne University Hospital, Malmö, Sweden;* [7]*Department of Clinical Sciences, Clinical Memory Research Unit, Lund University, Malmö, Sweden*

***Correspondence to:** Dr. Elisabeth Brogren, Department of Hand Surgery, Skåne University Hospital, Jan Waldenströms gata 5, 205 02 Malmö, Sweden; E-mail: elisabeth.brogren@med.lu.se
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of PD symptoms, such as resting tremor, micrography and difficulties in handling objects due to brady−/hypokinesia.[1,4,10] Moreover, misdiagnoses with rheumatoid arthritis and other hand surgical diagnoses, such as osteoarthritis of the thumb base and Dupuytren's disease with contracture, are common, especially when deformities occur early and in the absence of cardinal parkinsonian signs,[1,11] which makes the judgment of the condition complex for an untrained eye.[1,4]

In hand surgical terms, the SHD posture refers to the intrinsic-plus hand, caused by muscle spasm and/or adhesions of the intrinsic muscles in the hand. An intrinsic-plus hand can also be seen in other upper motor neuron lesions, such as stroke, cerebral palsy or encephalitis.[12] Although the underlying pathogenesis differs, the intrinsic-plus posture can also develop after hand trauma or secondary to rheumatoid arthritis.[12] Inability to fully open the hand and affected gripping ability are commonly seen regardless underlying pathogenesis.[12]

Hyperextension of the PIP joint and flexion posture of the distal interphalangeal (DIP) joint is often termed swan-neck deformity and develops secondary to imbalance between extrinsic and intrinsic muscles in forearm and the hand.[13] Swan-neck deformity in addition to flexed MCP joints is often seen in patients with an intrinsic-plus hand and has also been described in patients with more severe SHD, although a swan-neck deformity may develop for other reasons and irrespective of MCP joint flexion.[3] The z-thumb deformity has also been attributed to the SHD and refers to a MCP joint flexion and IP joint hyperextension and is a common finding in patients with rheumatoid arthritis.[14]

Based on increasing flexion of the MCP joints and co-occurrence of deformities, subluxations and contractions in other finger joints, SHD severity staging was just recently suggested.[3] Importantly, testing of intrinsic muscles tightness, commonly used in hand surgical assessment of intrinsic-plus hand,[15] is not included in the proposed SHD examination.[3] Our aim was to examine SHD in patients with PD using common hand surgical assessment methods and the recommended staging of SHD.[3]

Methods

Participants

All patients in this study were assessed during outpatient visits, which were scheduled at a time of day when the patient usually reported to feel at best (ie, in "ON" state). Thirty-five of 100 consecutive patients with PD, who visited one physical therapist with extensive experience of movement disorders (BL) at the Neurological Department at Skåne University Hospital in Malmö, Sweden, were identified as possibly suffering from SHD according to Wijemanne and Jankovic,[3] further so called "suspected SHD." PD was diagnosed according to the United Kingdom Parkinson's Disease Brain Bank criteria[16] by neurologist specialized in PD (BL).

Clinical Evaluation

The patients with suspected SHD were invited for clinical evaluation at the Department of Hand Surgery at Skåne University Hospital in Malmö, Sweden between June 1, 2019 through May 31, 2021. All 35 participants were evaluated by two hand surgeons: one sub-specialized in cerebral palsy and neurological disorders (LD) and one sub-specialized in rheumatoid disorders (EB).

Parkinsonian motor symptoms were assessed with the Unified PD Rating Scale (UPDRS) part III (motor examination)[17] and the severity of PD was determined with Hoehn and Yahr staging (H&Y) by an experienced physical therapist (BL).[18] Demographics and information on anti-Parkinsonian medications being taken were obtained from the participants' medical records. Daily levodopa equivalent (LDE) doses (mg/d) were calculated according to recommended conversion factors.[19] Both hands of the patients were assessed for signs of SHD and staged according to Wijemanne and Jankovic,[3] where stage 0 corresponds to normal MCP join position, while stage 1–4 indicate increasingly more flexed MCP joints with more severe finger deformities, joint subluxations and contractions[3] (Table S1). The patient was instructed to extend the arms out in front of his/her body with the palms down. The hands were held against gravity, but no other force was used to modify the natural posture. The hands were rated from lateral/medial with the palm down and the arms and hands extended anteriorly.[4] Intrinsic tightness was examined as described by Bunnell.[15] The test is an evaluation of PIP joint stiffness when the MCP joint is held in extension and then in flexion. If the PIP joint flexion is more restricted when the MCP joint is held in extension than in flexion, the test is considered positive.[12] In normal hands, as well as in cases of joint stiffness or paratonia, the resistance of the PIP joint should be invariant of the MCP joint position. Any z-thumb deformity of the thumb was evaluated by inspection.

The hands were also clinically screened for other PD related deformities, such as dystonic involuntary movements/abnormal positions at rest, and/or induced/aggravated by actions[20] such as rapid altered movement according to item 25 of motor examination[17] and gait.[7] Abnormalities were observed with respect to the intrinsic muscles (ie, the short muscles of the hand) and the extrinsic muscles of forearm (ie, the long flexor and/or extensor muscles) and their specific functions.

A deformity, that due to the contractures was no longer reducible by passive manipulation (passive range of movement [PROM] less than 50% of normal PROM), was stated as "fixed" in contrast to a "non-fixed" deformity.

Also, occurrence of other common hand disorders, such as osteoarthritis in the thumb basal joint, trigger finger or Dupuytren's contracture, was investigated. Standard radiographs of the affected hand were obtained to evaluate any joint involvement if an appropriate suspicion of any joint affection was found.

Statistical Analysis

Data were evaluated regarding underlying assumptions and described accordingly using IBM® SPSS® software version

27 (IBM Inc.). Normally distributed interval/ratio level variables were described using means and SDs. In other cases, medians (q1–q3) were used. Categorical variables were described using n (%).

Results

Characteristics of the 35 patients with suspected SHD are described in Table 1. Radiographs were obtained in 28/35 patients in which there was a suspected condition with relevance of performing an investigation of the bones and joints, for example, osteoarthritis or subluxation/luxation of the joints. After hand surgical assessment, five of these 35 patients were judged as stage 0 (ie, had normal flexion in MCP joints), but had other common hand surgical diagnoses, such as thumb basal joint osteoarthritis (n = 2), PIP joint osteoarthritis (n = 1), trigger finger (n = 1) or Game keepers' deformity of the thumb secondary to trauma (n = 1). None of these patients had positive intrinsic tightness test in any of the fingers (Fig. 1). Their mean age was 65 (SD, 10) yr. The median (q1–q3) PD duration was 0.3 (0.3–7) yrs and the UPDRS III score was 21(16–31).

A total of 23 of the remaining 30 patients with suspected SHD (24 hands) fulfilled criteria for SHD stage 1–4,[3] that is, all had MCP joint flexion and some additional joint deformities in at least one hand (Fig. 1). 20 of these 23 patients presented with slight or mild SHD; stage 1 (n = 13) and stage 2 (n = 7). Two patients had SHD stage 3 and one patient stage 4. Further details are presented in Table 2. If the patient had bilateral deformities, the worse affected hand was reported in the table.

Four of the patients with SHD (n = 23) had bilateral deformities; stage 2 in both hands (n = 1), stage 2–3 in one hand and other PD related deformities in the contralateral hand (n = 3). These are described below and reported in both Tables 2 and 3.

Except from MCP joint flexion, swan-neck deformity (n = 1) and z-thumb deformity (n = 6), we found "clefting" of the third and fourth fingers (n = 2) and abduction of the fifth finger (n = 6) in patients with SHD Fig. 2. Bunnell's test[15] was not applicable in one patient with SHD (stage 2) due to severe Dupuytrens contracture. Of the 22 remaining patients (SHD stage 1–4), (82%) had positive test, indicating intrinsic tightness in at least one finger. Further details are presented in Table 2.

TABLE 1 *Sample characteristics of 35 patients with Parkinson's disease and suspected striatal hand deformity (SHD)*

Age (years)	64 (11; 44–82)
Female gender	13 (37)
PD–duration (years)	1 (0.5–5; 0.3–19)
Motor symptoms (UPDRS III)	21 (14–35; 7–83)
Severity of disease (H&Y)	II (II–III; I–IV)
Daily total levodopa equivalent (LDE) dose (mg)	300 (300–500; 0–802)[a]

[a]Derived according to.[19]
Values are mean (SD; min–max), n (%), or median (q1–q3; min–max).
Abbreviations: H&Y, Hoehn & Yahr stage; PD, Parkinson's Disease; UPDRS III, part III of the Unified PD Scale.

Three of all patients with SHD (n = 23) had co-existence of other hand surgical diagnoses: ipsilateral Dupuytrens contracture (n = 1), contralateral ulnar nerve entrapment at the elbow and carpal tunnel syndrome (n = 1) and ipsilateral, mild osteoarthritis of the MCP joint of the index finger, in multiple DIP joints, PIP joints and the thumb basal joint (n = 1). There were no other radiological findings.

A total of 10 of the 30 patients (11 hands) had other PD related deformities not covered by the suggested SHD staging (Fig. 1). Three of these patients had SHD in one hand and other PD related deformity in the contralateral hand. These included involuntary movements and/or abnormal positions in wrist or fingers, that were induced or aggravated by actions in the extrinsic muscles. Seven of these 10 patients presented with non-fixed deformities and three were action-induced. Typical involvement of the extrinsic flexor muscles was clenching of the fist and flexion of the wrist joint. Typical involvement of extrinsic extensor muscles was MCP joint hyperextension in one finger, with or without simultaneous IP joint flexion or isolated swan-neck deformity without MCP joint flexion. Three patients (four hands) presented fixed deformities due to severe contractures. These contractures manifested as a combination of involvement of both the long flexors and the long extensors muscles, manifesting in a "shooting position," with extension of the index finger, clenching of the third, fourth and fifth fingers and flexion of the IP joint of the thumb, or as flexion of all fingers, manifesting as a "clenched fist." Intrinsic tightness was not applicable in one patient but confirmed in six of the other nine patients. Details are presented in Table 3. If the patient had bilateral deformities, the worse affected hand was reported.

None of these 10 patients had any co-existence of other hand surgical diagnoses.

Discussion

In this study, we clinically and radiologically examined PD patients with suspected SHD and found that, except from the previously defined stages of SHD (stage 1–4), such as MCP joint flexion, "z-thumb" deformity and swan-neck deformity, also other clinical features, like finger "clefting," abduction of the little finger and increased intrinsic tightness, were common findings in the patients with SHD. In our case-series, we also observed other abnormal hand postures, ranging from involving a single finger with MCP joint hyperextension and simultaneous PIP and DIP joint flexion, swan-neck deformity or rigidity in a single extensor muscle, to clenched fist and "shooting position," severely affecting the entire hand function. These symptoms probably descend from dystonia and extrinsic muscle hyperactivity. We propose that the described changes also should be incorporated in a modified SHD staging system.

SHD represents a specific entity of parkinsonian hand abnormalities that are less described and probably frequently

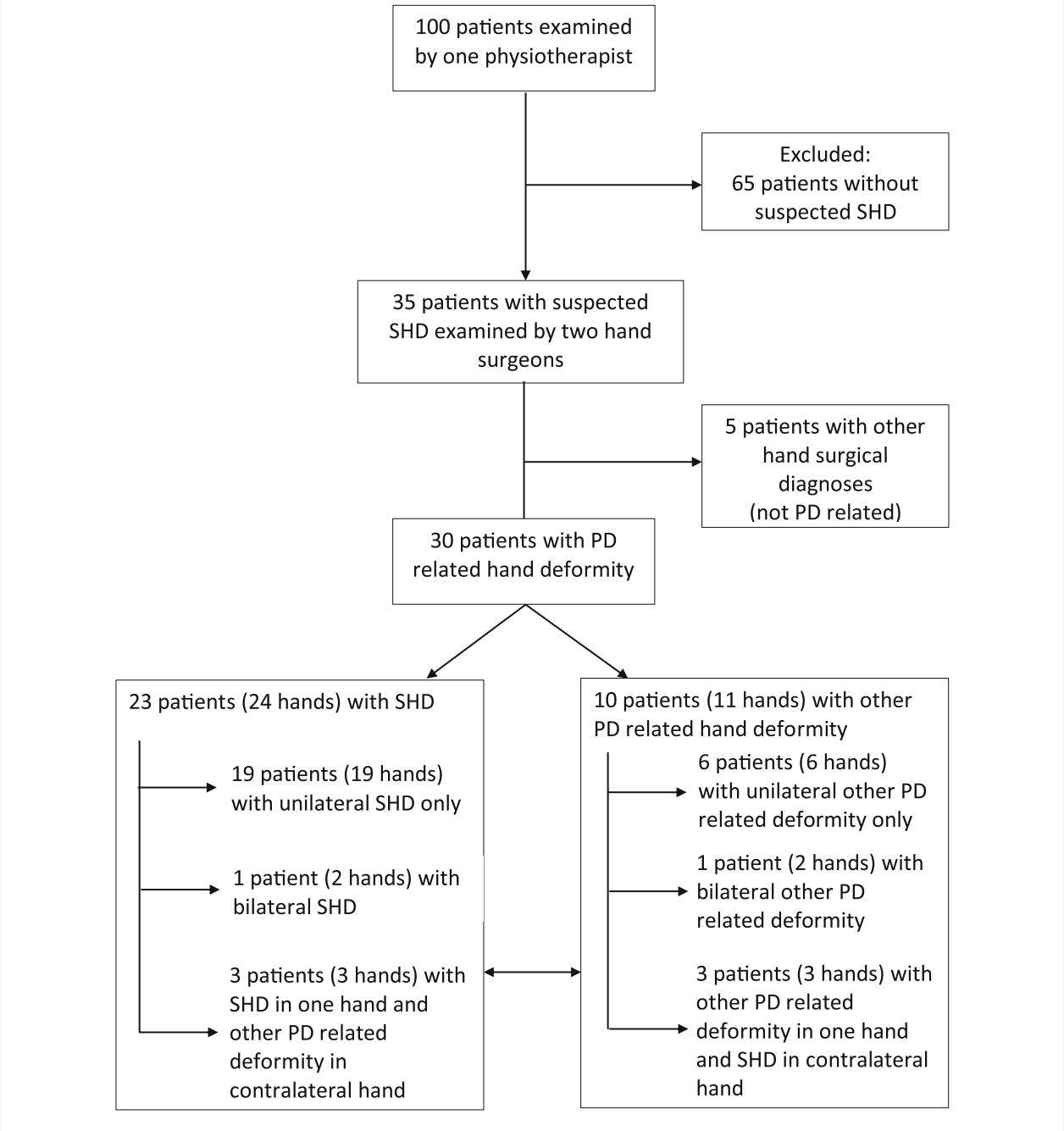

FIG. 1. Flow chart of included and excluded patients with Parkinson's disease (PD) and diagnosed with striatal hand deformity (SHD).

misdiagnosed.[1,21] In recent years, knowledge about SHD has increased through several observational studies, focusing on classification, demographics, prevalence, and neurological features.[3–6] By adding a hand surgical perspective, we intended to further improve our knowledge with the suggested modification of the SHD staging. Most of the present patients were in the early stages of PD with milder SHD stages (1, 2). Patients with longer PD duration and more parkinsonian symptoms tended to have more advanced stages of striatal hand. The majority of the SHD patients had positive intrinsic tightness test in at least one finger of the affected hand(s). Although this was not pathognomonic, since Bunnell's test was positive also in patients with other PD related deformities and negative in a few patients with SHD, a positive Bunnell's test is an expected finding in the classical intrinsic-plus hand.[12] This may indicate a need for treatment with botulinum toxin or, in severe cases, even surgical

TABLE 2 *Clinical assessment of 23 patients with striatal hand deformity (SHD)*

Stage[a]	No. of patients	Age (years)	PD duration (years)	UPDRS III	Intrinsic tightness	Other SHD related deformities
1	13	63 (13)	0.7 (0.5–1.0)	16 (12–28)	10 (77)	2 cleft between long and ring fingers 2 abduction of little finger
2	7	61 (11)	1.0 (0.5–4.0)	16 (14–29)	6 (86) 1 (14) NA	5 z-thumb deformity 3 abduction of little finger
3	2	74 (11)	5.0 (4.0–NA)	62 (52–NA)	1 (50)	1 z-thumb deformity 1 abduction of little finger 1 swan-neck deformity index and long finger
4	1	70 (NA)	19 (NA)	67 (NA)	1 (100)	1 subluxation of extensor tendons at MCP joint level with inability to extend fingers[b]

Values are mean (SD), n (%), or median (q1–q3).
[a]Stage of intrinsic SHD deformity; 0 (no MCP joint flexion), 1 (slight MCP joint flexion), 2 (mild MCP joint flexion), 3 (moderate MCP joint flexion), 4 (severe MCP joint flexion).
[b]Because of severe deformity with subluxated tendons, this patient was further investigated with anti-CCP antibodies, rheumatoid factor and ultrasound of MCP joints and extensor tendons, all turned out negative for rheumatoid arthritis.
Abbreviations: MCP, metacarpophalangeal; NA, not applicable; PD, Parkinson's Disease; PIP, proximal interphalangeal; SHD, Striatal Hand Deformity; UPDRS III, part III (motor score) of the Unified PD Rating Scale.

interventions.[22] The normal function of the ulnar nerve innervated interosseous muscles (intrinsic muscles), is flexion of MCP joints and extension of IP joints. When the intrinsic-plus hand develops after trauma, contractures of the intrinsic muscles are caused by adhesions and fibrosis secondary to edema or compartment syndrome.[12] In patients with rheumatoid arthritis, the intrinsic-plus hand is caused by joint inflammation with subsequent adhesions and muscle spasm, often exacerbated by tendon dislocation and MCP joint subluxation.[12] Table 4 describes differences in clinical and radiological features of patients with SHD and the rheumatoid hand deformity. The etiology of the intrinsic-plus hand seen in PD is less understood but may be caused by local muscle rigidity and/or dystonia.[3,4,7] Gortvai recognized the significance of the intrinsic muscles and its innervation, and demonstrated that SHD in PD patients could be reversed by blocking the ulnar nerve at wrist level with local anesthesia.[23]

In some of our patients with SHD, we also noted an abduction of the little finger or cleft between two adjoining fingers, the latter in fact also an abduction of fingers. These deformities

TABLE 3 *Clinical assessment of 10 patients with Parkinson's disease related hand deformities, not covered by SHD staging*

Type	No. of patients	Age (years)	PD duration (years)	UPDRS III	Intrinsic tightness	PD related deformities
Non-fixed	7	61 (12)	1.0 (0.5–3.0)	25 (18–33)	5 (71)	1 hyperextension of MCP joint of long finger 1 hyperextension of MCP joint and flexion of IP joints of little finger 1 swan neck deformity of long finger without MCP joint flexion 3 action-induced clenched fist 1 wrist flexion
Fixed	3	71 (10)	4.0 (6–15)	71 (52–83)	1 (33) 1 (33) NA	1 "shooting position" characterized by extension of digitorum II–III and flexion of digitorum IV–V at MCP and PIP joints and flexion of IP joint of thumb 1 "shooting position" and wrist flexion contracture 1 clenched fist

Values are mean (SD), n (%), or median (q1–q3).
Abbreviations: MCP, metacarpophalangeal; NA, not applicable; PD, Parkinson's Disease; PIP, proximal interphalangeal; SHD, Striatal Hand Deformity; UPDRS III, part III (motor score) of the Unified PD Rating Scale.

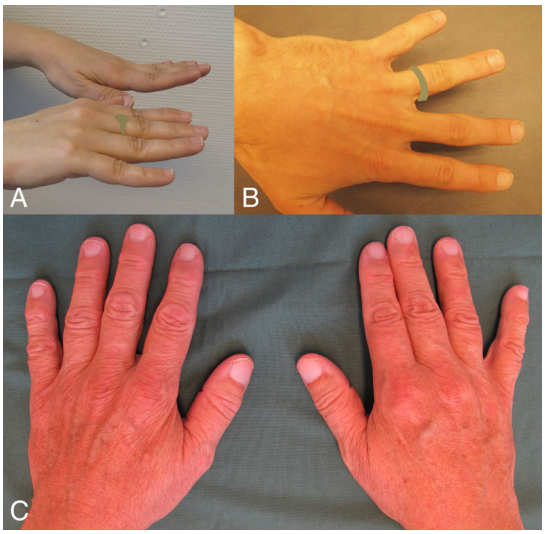

FIG. 2. Images showing three examples of the striatal hand deformity (SHD) seen in Parkinson's disease with flexion of the mcp joints in the right hand (**a**), indicating intrinsic tightness, a slight cleft deformity between the long and ring fingers in the left hand (with a tendency to abducted little finger) (**b**) and an abducted little finger in the right hand (**c**). Please note the camoflage of the subjects rings in a and b masking the identity of the subject.

are novel findings not previously considered or described in the earlier staging system, although additional analysis of photos published by Wijemanne et al and Ashour et al indicates that at least clefting may be a common phenomenon in SHD.[1,3] Clefting and abduction of the little finger was observed in early stages of SHD in our material and may be included already in stage 1, although larger studies are needed in order to confirm this. The intrinsic muscles, besides earlier described function, are also responsible for ab- and adduction of the fingers. The previously suggested "z-thumb" deformity was common, mostly observed in SHD stage 2, which is consistent with previously observations.[3] In patients with rheumatoid arthritis, the "z-thumb" deformity is caused secondary to synovitis of the MCP joint, weakening of the extensor pollicis brevis tendon insertion and subluxation of the extensor pollicis longus tendon.[14] However, the deformity could by speculation also develop secondary to tightness in the median and ulnar nerve innervated intrinsic two heads of the flexor pollicis brevis muscle.

Altogether, this supports the hypothesis that the intrinsic muscles are involved in the pathogenesis of SHD, where rigidity and/or dystonia cause the observed positions and deformities, such as intrinsic-plus hand, little finger abduction, cleft position, and z-thumb deformities. This is further supported by our clinical observations of improved posture and function after treatment with botulinum toxin injections in the interosseus muscles of the hand, sometimes with additional injections in extrinsic flexor tendon muscles. Since botulinum toxin treatment has an effect

TABLE 4 *Clinical features of striatal hand deformity and rheumatoid hand deformity*

	Striatal hand deformity	Rheumatoid hand deformity
Clinical features		
Joints	Typically painless. No arthrosynovitis.	Joints are painful, swollen (arthrosynovitis) and often stiff.
Symmetry	Usually asymmetric involvement	Usually symmetric involvement
MCP joint flexion	Primary caused by intrinsic muscle tightness. Extensor tendons normally aligned at the top of the joint, unless in very severe cases.	Primary caused by ulnar subluxation of the extensor tendons and/or volar MCP joint subluxation (due to inflammation).
PIP joint hyperextension	Primary caused by unbalance between intrinsic and extrinsic muscles.	Often caused by a combination of unbalance between intrinsic and extrinsic muscles secondary to subluxation of the MCP joint and/or weakening of volar structures in the PIP joint and/or weakening of terminal tendon to the DIP joint, all secondary to arthrosynovitis.
Ulnar deviation of fingers	More subtle ulnar deviation of one or more fingers. Clefting between two adjacent fingers. Ulnar deviation of little finger alone.	Often an obvious ulnar deviation of all four fingers.
Other features	Rigidity. Bradykinesia. Tremor. Dystonia.	Rheumatoid noduli, often wrist involvement with arthrosynovits and typical rheumatoid wrist deformity.
Radiological features		
MCP joints	Normal, even in late stages	Normal in very early stages, but then subluxation and/or typical arthritic destructions

Abbreviations: DIP, distal interphalangeal joint; MCP, metacarpophalangeal; PIP, proximal interphalangeal.

on the intrinsic muscles, we interpret the mechanism as not being a myopathy, but rather central mechanisms affecting the signal pattern in these very sensitive muscles.

Swan-neck deformity of fingers, described as one of the features in SHD,[3,6,21] was relatively uncommon in our case-series. Swan-neck deformity was found in one patient with SHD stage 3 and in one patient with hyperactivity in the extrinsic extensor muscle of the third finger. The deformity develops from either loss of extension of the DIP joint (ie, traumatic mallet finger) or from overpull of the extensor mechanism of the PIP joint; the latter caused either by hyperactivity in the extrinsic extensor muscles, or by increased tightness in the intrinsic muscles, often in combination with a congenital joint laxity.[13]

The most severe SHD (stage 4) includes severe MCP flexion and "advanced deformities with subluxation and contractures."[3] However, in addition to contractures and subluxations, severe and often fixed flexion contractures of both MCP and IP joints, with nails digging into the palm (i.e., clenched fist), are often being mentioned as late-stage SHD features.[1,3,24] We found three patients with such severe and fixed deformities that included abnormal positions and descended from extrinsic muscles (the flexor and/or extensor muscles in the forearm) rather than from the intrinsic muscles, resulting in clenched fist or "shooting position." Therefore, we suggest that SHD related to MCP flexion caused by shortening of the intrinsic muscle of the hand, should be distinguished from deformities and contractures caused by extrinsic muscles of the forearm. A contracture of the intrinsic muscles, however, severe it may be, never leads to a clenched fist or a shooting position.

We only found one patient that fulfilled criteria of stage 4 definition according to Wijemanne and Jankovic.[3] This patient presented with severe flexion of the MCP joints and inability to actively, or passively, extend the MCP joints due to ulnar subluxation of the extensor tendons at the level of the joints. Nevertheless, intrinsic tightness could be present along with other types of contractures and may be revealed first after corrections of severe extrinsic flexion- or extension contractures.[22]

Non-fixed deformities without flexion of MCP joints, but rather a hyperactivity of the extrinsic muscles, are an interesting finding. Action-induced clenched fist or wrist flexion are well recognized dystonic signs in PD[7] and hyperextension of single MCP joins may be a very early stage of the "shooting position" deformity observed in severe cases described before, probably also descended from dystonia. Thus, different pathology (ie, rigidity and dystonia) and distinctive muscle groups seem to be involved in the development of PD related deformities already at early stages of the disease. However, disease specific methods for evaluation of dystonia in PD are lacking.[7] Current scales for grading of idiopathic dystonia may be useful, but there is also a need to consider the complexity of PD related dystonia[25,26] and disease specific symptoms, such as rigidity, hypokinesia and impaired dexterity,[27] as well as its long-term effects on small hand muscles during long curse of disease.[1,4] Therefore, it should be agreed that early and repeated clinical investigation of hand status is important to work proactively. Responsiveness to systemic antiparkinsonian drugs is usually modest and not sustained.[3] However, botulinum toxin injection(s), in combination with systemic drugs, and use of orthosis at nights may be an option in patients with hand dysfuntion.[1] Also stretching of joints and range of motion exercises are recommended as part of standard care, particularly in conjunction with botulinum toxin injections and use of orthosis and should be continued over a longer time period as described for patients with cerebral palsy with the purpose of preventing impairment.[3] If conventional, non-operative treatments are unsuccessful, hand surgical interventions treatments can be attempted.[1,3,22]

In the radiographic examination of our cohort, we found co-existence of mild radiological osteoarthritis in two patients with SHD, but no other arthropathies were seen. Finger joints seem to stay unaffected by SHD even in advanced stages[3] and radiographs may not be useful in diagnosing classical SHD, except to exclude other hand surgical diagnoses. In fact, misdiagnoses of SHD are common and can be mistaken for rheumatoid arthritis, Dupuytren's disease or trigger finger.[1,3,11] The latter a sometimes painful condition, where one or more fingers, or the thumb, are locking or catching when flexed or extended, caused by tightness at the A1 pulley of the flexor tendon. Two patients in our original cohort had thumb basal joint osteoarthritis, with adduction contracture of the affected joint, leading to a swan-neck deformity of the thumb, which could be misinterpreted as a z-thumb. We also found several patients with SHD and other hand surgical conditions in co-existence. Thus, clinical assessment by a hand surgeon or orthopedic surgeon may be considered in some cases.

The strength of this study was the co-assessment of patients, including hand surgical expertise, which may increase the understanding of the clinical features in SHD. A limitation is that we did not use validated methods to grade SHD, since such are lacking. Nor did we examine the patients independently and have consequently not performed any inter-rater agreement analysis of our findings, which could be seen as a limitation. However, the patients were carefully co-examined, alternatively, by two experienced hand surgeons with different sub-specialties (rheumatoid arthritis and cerebral palsy/neurological disorders), thus using a consensus approach. Our cohort of PD patients had various disease severity and were examined on one occasion only; thus, we do not know whether intrinsic tightness fluctuate over time. To better understand the development of SHD and whether the deformity follows disease progression, longitudinal studies with larger study samples and control groups, are needed.

Conclusion

Hand deformities occur in early stages of PD with involvement of intrinsic and extrinsic muscles, causing a variety of deformities. The SHD involves pathology of the intrinsic muscle function of the hand and the useful current SHD staging[3] could be extended by testing intrinsic tightness. Evaluation should also include additional deformities, such as abduction of the little finger or cleft between two adjoining fingers. Other PD related hand postures, probably descended from dystonia and extrinsic muscles pathology, should be identified and if suspected, prevented from development into contractures.

Author Roles

(1) Research Project: A. Conception, B. Organization, C. Execution; (2) Statistical Analysis: A. Design, B. Execution, C. Review and Critique; (3) Manuscript Preparation: A. Writing of the First Draft, B. Review and Critique.

E.B.: 1B, 1C, 2B, 2C, 3A, 3B.

L.D.: 1B, 1C, 2C, 3B.

E.F.: 2C, 3B.

B.L.: 1A, 1B, 1C, 2A, 2B, 2C, 3B.

Disclosures

Ethical Compliance Statement: The Regional Ethical Review Board in Lund approved the study (Dnr 2020-01830). The participants included in this study (n = 35) provided written informed consent after oral and written information. We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this work is consistent with those guidelines.

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References

1. Ashour R, Tintner R, Jankovic J. Striatal deformities of the hand and foot in Parkinson's disease. *Lancet Neurol* 2005;4(7):423–431. https://doi.org/10.1016/S1474-4422(05)70119-8.
2. Spagnolo F, Fichera M, Bucello S, et al. Striatal hand in Parkinson's disease: The re-evaluation of an old clinical sign. *J Neurol* 2014;261(1):117–120. https://doi.org/10.1007/s00415-013-7165-y.
3. Wijemanne S, Jankovic J. Hand, foot, and spine deformities in parkinsonian disorders. *J Neural Transm (Vienna)* 2019;126(3):253–264. https://doi.org/10.1007/s00702-019-01986-1.
4. Ashour R, Jankovic J. Joint and skeletal deformities in Parkinson's disease, multiple system atrophy, and progressive supranuclear palsy. *Mov Disord* 2006;21(11):1856–1863. https://doi.org/10.1002/mds.21058.
5. Cervantes-Arriaga A, Rodriguez-Violante M, Morales-Briceno H, et al. Frequency and clinical correlates of postural and striatal deformities in Parkinson's disease. *Clin Neurol Neurosurg* 2016;142:140–144. https://doi.org/10.1016/j.clineuro.2016.02.003.
6. Baizabal-Carvallo JF, Alonso-Juarez M, Murillo Ortiz B, Fekete R. Clinical correlations of striatal hand deformities in Parkinson's disease. *Acta Neurol Scand* 2019;140(2):162–166. https://doi.org/10.1111/ane.13117.
7. Jost WH. Use of botulinum neurotoxin in Parkinson's disease: A critical appraisal. *Toxins (Basel)* 2021;13(2):87. https://doi.org/10.3390/toxins13020087.
8. Cordivari C, Misra VP, Catania S, Lees AJ. Treatment of dystonic clenched fist with botulinum toxin. *Mov Disord* 2001;16(5):907–913. https://doi.org/10.1002/mds.1186.
9. Jankovic J. Motor fluctuations and dyskinesias in Parkinson's disease: Clinical manifestations. *Mov Disord* 2005;20(11):S11–S16. https://doi.org/10.1002/mds.20458.
10. Keus S, Munneke M, Graziano M, et al. *European physiotherapy guideline for Parkinson's disease*. the Netherlands: KNGF/ParkinsonNet; 2014.
11. Shu X, Wang G, Lu X, Xie Y. Rheumatoid-like deformities in Parkinson's disease with 1-year follow-up: Case report and literature review. *Rheumatol Int* 2010;30(11):1493–1497. https://doi.org/10.1007/s00296-009-1094-1.
12. Paksima N, Besh BR. Intrinsic contractures of the hand. *Hand Clin* 2012;28(1):81–86. https://doi.org/10.1016/j.hcl.2011.10.001.
13. Lane R, Nallamothu SV. *Swan-neck deformity*. Treasure Island (FL): StatPearls Treasure Island; 2021.
14. Hara T, Yoneda H, Kurimoto S, Yamamoto M, Iwatsuki K, Hirata H. Thumb boutonniere deformity without rheumatoid arthritis or trauma. *J Hand Surg Eur* 2018;43(3):324–330. https://doi.org/10.1177/1753193417704610.
15. Bunnell S. Ischaemic contracture, local, in the hand. *J Bone Joint Surg Am* 1953;35(1):88–101.
16. Hughes AJ, Daniel SE, Kilford L, Lees AJ. Accuracy of clinical diagnosis of idiopathic Parkinson's disease: A clinico-pathological study of 100 cases. *J Neurol Neurosurg Psychiatry* 1992;55(3):181–184. https://doi.org/10.1136/jnnp.55.3.181.
17. Fahn S, Elton R. Unified Parkinson's disease rating scale. In: Fahn S, Marsden CD, Calne D, et al., eds. *Recent Developments in Parkinson's Disease*. Florham Park, NJ: McMillan Healthcare Information; 1987:153–163.
18. Hoehn MM, Yahr MD. Parkinsonism: Onset, progression, and mortality. 1967. *Neurology* 2001;57(10 Suppl 3):S11–S26.
19. Tomlinson CL, Stowe R, Patel S, Rick C, Gray R, Clarke CE. Systematic review of levodopa dose equivalency reporting in Parkinson's disease. *Mov Disord* 2010;25(15):2649–2653. https://doi.org/10.1002/mds.23429.
20. Albanese A, Bhatia K, Bressman SB, et al. Phenomenology and classification of dystonia: A consensus update. *Mov Disord* 2013;28(7):863–873. https://doi.org/10.1002/mds.25475.
21. Baizabal-Carvallo JF, Alonso-Juarez M, Fekete R. The frequency and diagnostic accuracy of hand deformities in Parkinson's disease. *J Neural Transm (Vienna)* 2018;125(12):1813–1817. https://doi.org/10.1007/s00702-018-1937-6.
22. Tosti R, Thoder JJ, Ilyas AM. Intrinsic contracture of the hand: Diagnosis and management. *J Am Acad Orthop Surg* 2013;21(10):581–591. https://doi.org/10.5435/JAAOS-21-10-581.
23. Gortvai P. Deformities of the hands and feet in parkinsonism and their reversibility by operation. *J Neurol Neurosurg Psychiatry* 1963;26:33–36. https://doi.org/10.1136/jnnp.26.1.33.
24. Kyriakides T, Hewer RL. Hand contractures in Parkinson's disease. *J Neurol Neurosurg Psychiatry* 1988;51(9):1221–1223. https://doi.org/10.1136/jnnp.51.9.1221.
25. Calabresi P, Standaert DG. Dystonia and levodopa-induced dyskinesias in Parkinson's disease: Is there a connection? *Neurobiol Dis* 2019;132:104579. https://doi.org/10.1016/j.nbd.2019.104579.
26. Tolosa E, Compta Y. Dystonia in Parkinson's disease. *J Neurol* 2006;253(7):vii7–vii13. https://doi.org/10.1007/s00415-006-7003-6.
27. Gebhardt A, Vanbellingen T, Baronti F, Kersten B, Bohlhalter S. Poor dopaminergic response of impaired dexterity in Parkinson's disease: Bradykinesia or limb kinetic apraxia? *Mov Disord* 2008;23(12):1701–1706. https://doi.org/10.1002/mds.22199.

Supporting Information

Supporting information may be found in the online version of this article.

Table S1. Striatal hand staging according to Wijemanne and Jankovic.