The Clinical Presentation of Parkinson's Disease and the Dyadic Relationship between Patients and Carers

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CAMBRIDGE
SCHOLARS
PUBLISHING

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This book first published 2013

Cambridge Scholars Publishing

12 Back Chapman Street, Newcastle upon Tyne, NE6 2XX, UK

British Library Cataloguing in Publication Data
A catalogue record for this book is available from the British Library

ISBN (10): 1-4438-4603-1, ISBN (13): 978-1-4438-4603-5

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FOREWORD

This book is an important and exceptional contribution to the neuropsychological literature on Parkinson's disease within the context of care-giving. It highlights the bond that emerges over the course of the disease between the person suffering from the condition and his/her carer, emphasising the significance of the dyadic relationship between couple in determining care-giving outcomes – a factor that has hitherto not been adequately appreciated. A novel approach taken by the author, building on these relationship factors, is in examining the carer's attitudes to the psychological changes occurring in the care recipient and demonstrating that these are of primary importance to determining care-giving outcomes.

In many countries there are increasing numbers of elderly people who are forming an ever greater proportion of the overall population. This poses many problems for society in general. One of these is that the prevalence of people suffering the health problems associated with ageing, is also increasing. In turn this leads to the problem of how to cope with a growing need to care for the elderly infirm. In the U.K. we are currently on the verge of introducing new legislation aimed at consolidating existing, somewhat piecemeal, laws to create a modern system for adult care and support, in which the disabled person and his/her family should be at the centre of service considerations.

Many relatives wish to care for their infirm or disabled family members themselves or with minimal contribution from the more formal methods of care provided by health and social agencies. Indeed these formal services would be unable to cope with the numbers of infirm elderly were it not for the often willing support provided by informal carers such as relatives, friends and neighbours.

There are two reasons why the problem of informal care needs to be addressed. The first, as indicated already, is that informal care is an essential feature of looking after the infirm elderly. It is important that relatives and others provide care in the best possible way. The second is that being a carer is not easy and creates problems for the carers themselves who may suffer a number of adverse consequences in carrying out their role. It is important that the problems experienced by carers are recognised and that, where necessary, addressed.

To date, the majority of work considering the problems experienced by informal carers of elderly people with infirmities has been based on dementia, not surprisingly because this is one of the most prevalent disorders. Here it has been found, for example, that carers experience various problems, particularly depression. Also depression in carers is more linked to the psychological changes in the person for whom they care rather than dealing with the physical disabilities or limitations of the sufferer.

Little attention has so far been given to the problems of caring for other disorders occurring in the elderly. This is especially true for Parkinson’s disease which is the focus of the author’s research. In the theoretical section of the book, the clinical presentation of Parkinson’s disease, including both the motor and non-motor symptoms, is covered thoroughly and the detail in which the author has discussed questions of particular interest is impressive. Her comments on methodology demonstrate a competent grasp of the important issues.

In reporting the research study, the author has presented a clear justification for her research and has raised and made progress in resolving a number of issues relating to the psychological consequences of Parkinson’s disease. These include the nature of depression in that disorder and the key factors causing distress in carers. She provides a clear summary of the findings with a competent discussion of their significance. In particular, she demonstrates an impressive ability to deal with a number of difficult methodological issues. To the best of my knowledge, it is the first sustained piece of research into this aspect of Parkinson’s disease and as such constitutes a significant contribution to this field. As such it merits wide attention.

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CHAPTER ONE

INTRODUCTION

1.1 Parkinson's disease: a neuropsychological disorder

The study of Parkinson's disease (PD) and caring for someone with the disorder that is presented in the following pages is undertaken from a clinical neuropsychological perspective. Much has been written in recent years, particularly in the field of dementia, about the problems of caring for someone with a chronic and intractable disease and this is reflected in the exponential growth in the academic literature on the subject over the last thirty years. Far fewer studies have focused on other neurodegenerative disorders, such as PD, and whilst this is an omission in its own right in need of repair, there is another reason, which from the neuropsychological perspective, highlights PD as a focus of interest. This is because PD has been aptly described as the "quintessential neuropsychiatric disorder" (Weintraub and Burn 2011). Although the condition is still primarily diagnosed from the perspective of a movement disorder, the high prevalence of accompanying non-motor symptoms in the form of neurocognitive and neuropsychiatric complications make it a unique model for the focus of research in neuropsychology.

Most attention to date has been devoted to the cognitive impairment characteristic of the condition and attempting to identify the neural substrate for dementia in PD. Cognitive impairments are recognised as a frequent accompaniment of the disorder affecting up to 25% of non-demented patients with PD (Aarsland et al. 2010). Dementia is considered to be a long-term complication, affecting up to 80% of patients as the disease evolves (Aarsland et al. 2003). Affective disorders on the other hand, in the form of depression or "melancholia", whilst having been recognised as a concomitant of the disorder since its earliest descriptions (Parkinson 1817), have generated a great deal of controversy and yet, as pointed out by Ponce and Lozano (2010), have received relatively little systematic attention. In recent years, agreement seems to have been reached that a more serious or "major" form of depression occurs with a frequency of 5-20%, but that milder forms (minor or subsyndromal depression) occur in an additional 35% of patients (Reijnders et al. 2008). However, there remains considerable debate surrounding the nature, profile and origin of the affective symptoms in PD. A main concern of the empirical study, which is described in later sections of this book, was to reach a better understanding of these features of the depressive disorder in PD and their relationship to the other symptoms of the condition.

Other clinically significant, neurobehavioural complications include anxiety symptoms, impulse control disorders, obsessionality, fatigue, disorders of sleep and wakefulness and apathy. The importance of these "non-motor" symptoms in PD is reflected in the growth of publications related to this topic over the last twenty-five years, primarily in the area of dementia and cognitive impairment (Weintraub and Burn 2011) and in the fact that it has increasingly been realised that improving the quality of life of people with PD and their carers demands extending clinical and treatment approaches from a primarily motor-oriented focus to include the non-motor features of the disorder (Cheng et al. 2010; Spica et al. 2013). It goes without saying that clinical neuropsychology has a clear contribution to make in the area of the cognitive and affective symptomatology of the condition in both the theoretical and clinically applied domains.

1.1.1 A brief note on terminology

A striking feature of neuropsychological approaches to distinct disorders is that they have traditionally tended to focus on cognitive impairments. Thus the neuropsychology of PD has largely been dominated by the study of attentional, perceptual, visuospatial, memory, language and more recently executive-type deficits (see chapter 4). Whilst behavioural disorders have also come within the realm of clinical psychology, disorders of emotion have traditionally been the sphere of psychiatry. As recognition has emerged that emotional disturbances, in particular disturbances of affect, are associated with neurological disorders such as PD, it has become increasingly common to refer to them as "neuropsychiatric" in origin. These divisions between neuropsychology and neuropsychiatry are somewhat artificial and this is reflected in the interchangeable use of the terms in the literature to account for cognitive, behavioural and affective symptoms. For example, Weintraub and Burn (2011) include cognitive deficits in their definition of the neuropsychiatric disorder in PD. Equally, neuropsychological approaches to the study of emotions are well-established (e.g. Damasio 1996; Grabowska 1999; Herzyk 1999, 2000a; Kądzielawa 1986; Konorski 1969; Luria 1976; Szepietowska and Misztal 1999; Watts 1993).

1.1.2 Clinical neuropsychological perspectives on Parkinson's disease

A widely accepted definition of neuropsychology is that it is the study of brain-behaviour relationships and traditionally, two main approaches have been discernable in this field as described by Miller (1984). The first has been concerned with delineating the kinds of functional disturbance that may occur as the result of lesions affecting specific parts of the brain in an attempt to specify the functions supported by defined anatomical substrates. Much of the early work in neuropsychology was of this kind, attempting to correlate focal lesions with observed deficits in higher cognitive functions. The second approach has had a more functional orientation, concerned with determining the kinds of change that may occur, for example in language or memory, following organic brain damage associated with specific disorders, such as Alzheimer's or Parkinson's disease. Here there has, until recently, been less emphasis on lesion location, largely because of technological limitations in identifying biochemical as opposed to anatomical lesions. Thus both anatomical and functional trends formed the background to the earlier understanding of brain-behaviour relationships in neuropsychology. However, over the last twenty years, dramatic advances in neuro-imaging techniques, especially those of a functional nature, have brought these trends together and are of considerable theoretical importance, both in furthering understanding of normal brain function, but also in providing explanatory models of how functions are integrated. PD has been at the forefront of research in these areas, as it has provided a unique model of a commonly encountered neurological disorder presenting with deficits in the motor, neuropsychological and neuropsychiatric fields of functioning, thereby enabling understanding of how functional processes are integrated and related to anatomical substrates in the human brain (e.g. Robbins 2010). However, despite the huge gains made in the theoretical understanding of conditions such as PD over recent years, this has not as yet, translated into significant improvements in pharmacological treatments or in advancing the possibility of a cure, which is understandably, the main desire of patients and is reflected in the key aims of patient organizations such as Parkinson's UK (2012a). This ultimate goal will doubtless be achieved in time, but in the interim, the majority of patients diagnosed with the condition must rely on their families for support and care in living with the consequences of this long-term neurodegenerative disease. A primary concern for clinical neuropsychology is to provide the empirical foundations for guiding practice in helping patients and their families to cope with the ensuing difficulties. This recognition provided the stimulus and motivation for the work reported in this volume, in the final part of the empirical study, which is reported in chapters 8-12.

It is self-evident that empirically guided, theoretical developments should lie at the heart of clinical practice. Without this core knowledge, proper diagnostic understanding and suitable treatment interventions are impossible. This has been demonstrated in the failure of a variety of allegedly useful interventions in PD, ranging from attempts to treat depression with pharmacological agents whose efficacy had not been adequately established in the condition to ill-fated surgical interventions ranging from thalamolysis to brain implants, which variously resulted in severe cognitive impairment and even premature death. Clinical neuropsychology, however, requires further elaboration and refinement of purely theoretical approaches. Here the questions raised demand answers that are of practical application, both in terms of helping to restore function in so far as this is possible, and to maintain the patient at optimal levels of functioning, preventing further deterioration within the limits imposed by the illness. This is usually seen as the task of neuropsychological rehabilitation and this field too, has developed significantly over the last thirty years (e.g. Pąchalska 2007; Sohlberg and Mateer 2001; Wilson et al. 2009). Clinical approaches in neuropsychology have been primarily patient focused and have gone a long way towards improving the quality of life of people with neuropsychological deficits in the wake of neurological disease and the disabilities consequent upon it.

1.2 Carers of people with Parkinson's disease

An underlying principle of the present work, in keeping with current concerns in health policy in the U.K. (Department of Health 2012a,b,c) and other member states of the European Union (e.g. Harkin 2011) is the notion that clinical neuropsychology not only has an obligation to the patient suffering from the condition that has brought him/her to the attention of the clinician, but also to his/her carer. The majority of neurological conditions involving neuropsychological assessment and intervention are long-term, intractable or progressive disorders, such as PD, in which the patient ultimately comes to depend on the care of a family member in order to be able to sustain functioning in his/her own home in the community. As a result, the carer becomes almost inextricably linked to the patient, embarking upon and sharing his/her journey with the disease, through all of its stages. It is inevitably a journey that has not been planned, heralds undesirable changes in the lives of both the patient and the carer and follows a singularly unpredictable course. Above all, the illness brings about an irreversible disruption, not only in the life of the patient, but also in that of his/her carer. For some way along their common course, they may be brought closer together by the illness experience, as they seek to adapt to its mercurial demands affecting both their lives, with the patient becoming increasingly dependent on the carer. In the main however, the changes wrought by the illness, especially those presenting in the form of neuropsychological and neuropsychiatric deficits, mean that from the carer's point of view, apart from increased problems with mobility, the patient may become more forgetful, disinterested and withdrawn and cease to be the source of support that s/he may have been in the past. As a result, the carer experiences a sense of loss and increasing isolation, and the couples' relationship comes under increasing pressure. This is further exacerbated by the patient's frequent unwillingness or inability to maintain social activities.

Not surprisingly, in view of the impact that caring for someone with a long term progressive illness has on the carer, a major theme running through the literature on care-giving is the emphasis on the highly stressful nature of the task. Models of care-giving based on stress (process) theories have predominated (e.g. Pearlin et al. 1990) with the consequent result that attention has focused on concepts such as the "burden" involved in the caring process (Zarit, Orr and Zarit 1985). For more than two decades, elaboration and refinement of this concept was the main preoccupation of the care-giving literature, with the consequent proliferation of instruments to measure burden. Many of these treated burden as a unidimensional construct in keeping with early conceptualizations (e.g. Zarit, Reever and Bach-Peterson 1980). Whilst the notion of burden was a useful concept from the point of view of providing a unifying approach to care-giving in diverse groups of older people, increasing experience and sophistication in this field of research has revealed its many limitations. First and foremost, researchers have achieved a better appreciation of the complexities of care-giving which are evident in the diversity of processes and outcomes in different groups of carers and care recipients. For example, it is becoming increasingly apparent that caring for someone with dementia differs in some important ways from caring for someone with other types of neurodegenerative disorder, such as PD (Hooker et al. 2000). However, the notion of burden has suffered not only from conceptual limitations in being too narrow, but was also found to lack empirical support. Only limited success was achieved in demonstrating that high levels of carer distress were linked to the "objective" demands of the care-giving situation, such as the physical tasks, time demands and financial responsibilities encompassed in the notion of burden. The idea of "subjective" burden (Hoenig and Hamilton 1966) was thus developed to account for the carer's appraisal of the situation and his/her perceived strain, which was conceptualised as the stress of care-giving and considered to be an outcome measure of the difficulties faced by carers. These approaches served to accentuate the perception of care-giving as a predominantly negative experience. In the late 1990s, the discovery that not only physical morbidity (Schulz et al. 1995; Vitaliano, Zhang and Scanlan 2003), but mortality rates were increased among carers (Schulz and Beach 1999) appeared to bring irrefutable validation to the growing concern that caring placed family members under intolerable levels of stress and required systematic and effective intervention procedures for the relief of carers. Alongside the relatively slow progression of research in this area towards finding successful remedial solutions, carers themselves formed support groups which were extremely effective in lobbying governments to recognise their role in caring for elderly and disabled relatives and to adopt policies that went some way towards meeting their needs. Nevertheless, the legislation relating to carers, which has emerged in somewhat piecemeal fashion over the last sixty years, is generally recognised as being ".... inadequate, often incomprehensible and outdated" (Law Commission 2012) and the funding of the care system as ".... not fit for purpose" and in need of "urgent and lasting reform" (Dilnot 2011). It is recognised that reforms, especially those involving substantial amounts of government funding will need to be carried out over a number of years, but at the time of writing a consultation process is being completed in relation to the draft Care and Support Bill (Department of Health 2012) which has proposals for making some significant and long-promised changes to help older and disabled people in need of care and support and their carers. A synopsis of this and other U.K. legislation in relation to carers is provided in Appendix A.

In spite of the stressful nature of caring, the majority of older people with a chronic and incapacitating condition are cared for at home, usually by their family. Families are willing carers, and their contribution is reflected in the not inconsiderable number of individuals who take on the role: over 6 million in the U.K. (NHS Carers Direct 2012a) and 65.7 million in the U.S.A. (National Alliance for Caregiving 2009). These numbers are likely to rise in the foreseeable future as developed countries experience a demographic shift, in which older people are living longer, but with the increased likelihood of suffering from poor health. One of the reasons for the success of carers' efforts probably lies in the individualized and high quality of care that they provide and their commitment to providing it. Yet this commitment finds itself in stark dissonance to the emphasis on the hardships of care-giving which have, to a large extent, been underlined by the stress models that have dominated the field of care-giving research. As Schulz and Williamson (1997, 123) pointed out over a decade ago: "Relatively little attention has been paid to the assessment of positive aspects of caregiving, the quality of care delivered by caregivers and the impact of caregiving transitions." Whilst a few attempts have been made to redress the bias in the presentation of caring as a wholly negative experience for carers (e.g. Nolan, Grant and Keady 1996) by drawing attention to positive aspects such as the satisfaction and recognition that some carers experience in their role, the negative focus has nonetheless predominated.

One explanation for this may be that it is natural to focus on difficulties in order to try to find solutions to the problems identified. Another, is that in attempting to drive policy decisions, a case has to made regarding the hardships involved. However, sometimes the impression given in the literature is that the strain on carers is such, that family care generally, is on the verge of breaking down. Apart from being misleading, this kind of stance is unhelpful in gaining a better understanding of care-giving. For example, an observation that has long been apparent to researchers in this field, is that given relatively similar care-giving situations, no two carers seem to manage and deal with the problems involved in the same way, and hence the outcomes they experience may vary considerably. Within the context of the stress process model, this observation elicited the search for intervening variables which might modify the care-giving situation and thus determine carer outcomes. These included differences in the coping mechanisms used by carers (Hooker et al. 2000) and varying degrees of social support available (Edwards and Scheetz 2002; Lai and Thomson 2011; Monahan and Hooker 1997), which were posited as being essential to modifying the outcome in terms of carers' well-being or contributing to the distress that they experienced. Nevertheless, factors that have traditionally been used to explain differences in stress processes have had only limited success in accounting for the difficulties of care-giving. For example, in one of the first studies to investigate the problems faced by carers of people with PD, Miller, Berrios and Politynska (1996) did not find any beneficial effect of social support on carer well-being.

Furthermore, the assumption at the heart of stress models of care-giving has been that the physical and mental disability of older care recipients, is the main determinant of deterioration in family carers' psychological well-being. As already mentioned, this has proved to have only partial justification and has led researchers in recent years to undertake a close re-evaluation of the problem and the way in which it has been conceptualised. An examination of the literature on care-giving carried out for the purposes of the present research highlighted an empirical observation that has not received a great deal of attention, despite being identified from some of the earliest studies in this area. This finding points to the fact that carer (and not predominantly care recipient) variables appear to be better in predicting psychological morbidity and deteriorating well-being in carers over time (Hooker et al. 1992; Hooker, Frazier and Monahan 1994; Hooker et al. 1998; Hooker et al. 2000; The Resource Implications Study Group of the Medical Research Council study of cognitive function and ageing [RIS MRC CFAS] 2000). This is an important finding which requires careful analysis and elaboration if it is not to be trivialised to the detriment of carers. Given its potential to characterise an essential aspect of care-giving, it formed an important background to the present study and determined an important line in the investigation of care-giving in PD that is described here.

It is important to understand carer characteristics as a complex set of interdependent variables that are only partially antecedent to taking on the care-giving task. Moreover, in the context of care-giving demands, it would seem unlikely that they should remain fixed or stable, as by its very nature, the care-giving situation requires adjustment and flexibility. However, the literature that has recognised the importance of carer variables to the outcomes of care-giving has tended to conceptualize them in terms of relatively constant characteristics such as personality variables (e.g. Hooker et al. 1998) and as reflecting the previous quality of the relationship with the care recipient (Murray and Livingstone 1998). Applied within the stress process model, these characteristics may be seen as determining different coping styles for dealing with the task of care-giving (Hooker et al. 2000). However, other potentially more important dimensions in relation to the way in which the carer is able to adapt and reconstruct his/her relationship with the care recipient in the light of the disability and its demands have largely been overlooked. In the early stages of PD, adjustment to the condition is likely to be a reciprocal task in which both the carer and the care recipient participate, albeit in different ways, through the process of coming to terms with any limitations imposed by the illness and undertaking adaptations that are perceived to be necessary. These processes are described in chapter 5 of the present study. However, at different stages over the course of the illness, the ability of both the carer and the care recipient to maintain these efforts may be challenged by evolving symptoms, especially in the area of affective and cognitive functioning, which diminish not only the patient’s capacity to adapt his/her behaviour accordingly, but also alter his/her reactivity to situational needs. Hence the carer is likely to require a sensitivity to changes in the care recipient’s possibilities for performing previously held roles and tasks, an ability to initiate changes in life-style in response to the needs of the situation and to come to terms with new responsibilities and limitations imposed by the care-giving situation. These adaptations, of course, are not one-way, as even passivity on the part of the care recipient may be an adaptive response within the resources that remain available. Thus a crucial dynamic involving both the carer and the care-recipient develops which is illustrative of the care-giving situation. This aspect of care-giving, namely that involving the interaction between the carer and the care recipient, has received relatively little attention, especially in the context of PD, but would appear crucial to achieving a proper understanding of care relationships in long-term, neurodegenerative illness. It requires a dyadic approach to care-giving (Lyons et al. 2002), which meets the long-expressed demand of both carers and care recipients for recognition, that is reflected both in the academic literature and more recently, has found recognition in U.K. government policies (Department of Health 2012b).

1.2.1 Dyadic approaches to care-giving

It is important to recognise, however, that dyadic approaches to care-giving require rather more than simply recognising the existence of carers. This, successive governments in the U.K. have done over the last decade or so, by introducing legislation expressly aimed at meeting the needs of carers, granting them the right to an annual assessment, introducing services for carers in their own right, together with direct payments in lieu of services that have been identified as lacking, and latterly involving them in decisions regarding the planning of local services and provision of individualised care packages, along with greater opportunities for respite care (see Appendix A). These measures form an indispensible and long-awaited foundation to establishing provisions for meeting the needs of carers, but they represent a mere starting point in terms of understanding the complexity of care-giving needs. This is reflected in the fact that attempts to support carers through a variety of intervention studies have frequently been disappointing in their results (Schulz, Martire and Klinger 2005; Torti et al. 2004). One reason for this is that they have been based, as indeed has the legislation, on a response to carers’ appraisals of their own needs, but have been shown to lack support on empirical verification.

A classic example of this is the often cited need for respite care, prioritised once again in the recent legislative proposals. Although there can be no doubt that the restrictions and pressures of the care-giving task mean that carers frequently report detrimental effects on their health in the form of excessive tiredness, increased irritability and depression (Health and Social Care Information Centre 2012) and fulfil both objective and subjective criteria in requiring a break from care-giving, respite care is not taken up by carers in the numbers that might be expected to make use of this facility from an analysis of their expressed needs. The reasons for the failure to use these opportunities when they are made available are not adequately understood, but they seem to be related to the inflexibility and standards of the care on offer, which do not match carers' expectations of alternative care for their relatives (Lamura et al. 2006). However, clinical experience with patients and carers of people with PD also suggests that their desire for respite is a desire for respite from the disease itself, a yearning for the health status that preceded the onset of the disease, if only briefly. At the same time they realise that such expectations are unrealistic, and sometimes a break from their usual routine only serves as a reminder of this painful fact. It is not unusual for carers to report, that a period of respite care for their relative did not provide them with the restorative experience they had anticipated and that they missed the care recipient's presence, a fact that is reflected in their maintaining frequent visits to the patient in the respite care facility. The complexities of the situation can only be understood if a dyadic perspective is applied to care-giving.

Thus, in order to make a difference to the conditions and circumstances in which the patient and carer find themselves, it is essential not only to reach an understanding of their individual strengths and the difficulties that they face, but also to gain an appreciation of how they function in relation to each other. This relational approach advocating consideration of the mutuality of influences on care-giving from both the parties involved, allowing for greater flexibility and taking account of the multidimensional nature of the task, was first proposed by Kahana and Young (1990, 76) as a way of expanding on the "prevalent conceptual orientations to the study of caregiving". Research over the last twenty years has been slow to follow in the direction indicated. Indeed, most care-giving studies, especially those in areas that have received relatively little attention to date (such as PD), are unfortunately also failing to learn from the experience of research in related fields and remain fixed in the static, unidirectional outcome model, where focus remains on the carer, and, in the words of Lyons et al. (2002), the care recipient is reduced to little more than an independent variable and viewed as the source of stress to the carer. Apart from the limited empirical utility of this approach which has now been established (and which is discussed in greater detail in chapter 6), the conceptual framework for care-giving research, rooted as it has been in stress models, has emphasised the negative aspects of providing care for a dependent, elderly relative at home, and as mentioned above, virtually ignoring the fact that most families do and are happy to be engaged in caring for a loved one, finding satisfaction and fulfilment in a commitment they consider to be a natural part of family life (Nolan et al. 1996). This does not mean however, that they do not require support: the question that has still not been answered satisfactorily is what kind of support is likely to be most effective and how should it be delivered?

The task undertaken in the present study represents an attempt to use the applied methodology of clinical neuropsychology to understand the situation of both the patient and the carer in an effort to improve the care of people with long-term neurological illness. It is hoped that the dual focus adopted in accounting for both carer and care recipient outcomes will help to generate a meaningful conceptual framework within which to evaluate the various factors shaping care-giving in PD. This is a relatively new conceptualisation of the task in terms of a neuropsychological approach, where traditionally, assessment of the patient has been at the forefront of clinical concerns and work with the carer has essentially been of a supportive nature. Assessment of the patient's neuropsychological and neuropsychiatric performance continues to occupy a central place in any evaluation, but the purposes to which it is put, extend beyond that of diagnosis *per se*. It is also recognised that an evaluation of the carer's needs must be a prerequisite to any formulation of the care-giving situation. Depending on the circumstances, this may involve assessment of the symptoms of psychological distress, subjective feelings of self-efficacy and ability to cope, and attitudes (optimistic versus pessimistic) towards the future (Lyons et al. 2004). Whilst these components of the carer's perspective have received some attention in the literature, there is a further dimension, that appears as yet, not to have been considered, but which may be of importance as it may help to shape the characteristics mentioned. This relates to the way in which the carer perceives and subsequently interprets the care recipient's functioning in a variety of domains, including physical abilities, neuropsychological and neuropsychiatric performance. The latter two are of particular interest to the present study as there is evidence that the psychological symptoms of PD, in particular disturbances of affect, are of primary importance in influencing carer outcomes (Miller et al. 1996).

1.2.2 Carers' attitudes to the care-giving situation

Carers' attitudes to the care-giving situation are likely to be shaped by both emotional and cognitive factors, or indeed by an interaction between the two. There have been numerous explorations of care-giving based on theories focusing on emotional explanations, including attachment and familism (as described in chapter 7). Far less interest has been shown in cognitive factors, although it would be reasonable to expect that carers' perceptions of different aspects of the care-giving situation should influence their attitudes to caring and the way in which they cope. For example, if their perceptions of the physical and psychological changes in the care recipient are predominantly bleak, it is to be expected that frustration and depression might result. The same might be true concerning their relationship with the care recipient. Their attributions regarding these changes are equally important. Potential "cognitive biases" in the way in which the care recipient's physical ability, mood and cognitive performance are perceived, triggered by pre-existing cognitive schemata (Beck et al. 1979) would be expected to colour the carer's attitude to both the present and future, determining patterns of adjustment to the illness and feelings of self-efficacy.

The carer's view of the care recipient's functional performance may be important for two main reasons. Firstly, it may have a critical effect in shaping the process by which the care-giving situation evolves and determining the outcomes for both of the protagonists involved. For example, it has been observed that the amount of help that carers provide with the tasks of daily living, such as dressing, bathing, feeding etc. is relatively independent of the functional disability in PD and is more a feature of the carer's attitudes (Davies et al. 1999). This may result from differences in carers' perceptions of the extent of the disability in the care recipient, from a well-intentioned desire to be helpful and to provide assistance as an expression of affection for the patient or because they feel under pressure to complete the task at hand as quickly and efficiently as possible. In the latter case, this may be because of external pressures such as those of work or the need to keep to a specified timetable, or it may reflect impatience and intolerance of less than perfect performance. Whatever the cause, the consequences may be somewhat unpredictable (from the carer's point of view) and worse still, undesirable. An illustration of this comes from institutional settings, where it has long been recognised that certain patterns of behaviour on the part of care staff serve to reinforce dependency and result in learned helplessness in care recipients (M. Baltes and P. Baltes 1986). It is equally possible that family carers may activate a cycle in which care recipient dependency is potentiated, through behaviours which, while being well-meaning, result in the patient not attempting to do certain things which s/he is still capable of, because they are done for him/her. As a result, care recipient helplessness is reinforced and carer burden increased.

Secondly, needless to say, carer perceptions of care recipient functioning are also likely to influence the carer's ability to act as a proxy informant on his/her behalf, as may be required in certain circumstances for the purposes medical and needs assessments for care and support. It is important to understand that carers' judgements are likely to be influenced by a complex interplay between emotional and cognitive factors at different stages in the care-giving process.

The (mis)interpretation of care recipient psychological symptoms may take a variety of forms. Deficits in memory may be frustrating and annoying for the carer or may be "normalised" in terms of explanations of advancing age. More severe deficits are often poorly understood and may raise anxieties surrounding dementia. Some of the specific deficits associated with PD, such as apathy, withdrawal, depression, deterioration in communication or forms of impulse control disorders may be interpreted as "personality change" or as a rejection of the carer and his/her efforts to help. Carers are concerned at the lack of interest, refusal to see people, reluctance to engage in conversation, and even the failure to complain, that is characteristic of so many patients with PD. Occasionally they are startled by uncharacteristically hurtful comments or unusual behaviour. There is evidence to suggest that depression in the patient is particularly problematic for carers (Miller et al. 1996). One of the possible reasons for this, explored in the study, is that the misinterpretation of psychological symptoms, or their apparently brutal transparency in the minds of carers, wrecks havoc even with long established relationships, contributing further to the hardship of caring.

In view of the opportunities that exist for distortions in the perception of the care-giving situation, a priority of the present study was to examine carers' perceptions of the cognitive and affective state of their spouses, for whom they were caring. The primary purpose was to determine the extent to which carers' perceptions were in keeping with the formal neuropsychological and neuropsychiatric assessment of the care recipient. Establishing the extent to which there was concurrence between the two evaluations was deemed to be important for two reasons. The first was the possibility that carer perceptions of the care recipient's functioning (rather than the results of the neuropsychological assessment) might be more strongly associated with their own psychological well-being or lack of it. The second reason was related to the reliability of using proxy reports from carers in order to assess aspects of the care-giving situation. This is particularly important in view of the U.K. government's proposals to introduce new legislation in the form of the Care and Support Bill in 2013 (Department of Health 2012b), in which a commitment has been made to acknowledge carers as expert partners with formal services, ensuring that they have access to the integrated and personalised services they need to support them in their caring role. This recognition of carers answers a long-established charge from researchers in the field, calling for an acknowledgement of the dyadic nature of care-giving and the need to extend the focus of care services to include the carer, the wider family and formal care systems, whilst keeping the needs of the patient at the forefront of concerns (Kahana and Young 1990; Nolan et al. 2003; Zarit, Pearlin and Schaie 1993).

However, if acknowledging carers as "expert care partners" is to remain little more than a political slogan, there is a need for the care services offered to take an evidence based approach to the way in which goals are implemented for the fulfilment of this aim. There can be no doubt that carers have a unique perspective on the care-giving task, that cannot be matched in its richness of experience by other viewpoints. At the same time it has to be borne in mind, that theirs is the "insider" perspective of direct involvement, influenced by the emotional and perceptual factors referred to above. This involvement, whilst giving carers their unique expertise and invaluable insight into the complexities of caring for a disabled relative, also means that their perceptions are shaped by the actual experience of care-giving. Hence, they are very much part of the situation, involved in its evolution and the way in which it is constructed and relayed to others. Indeed, acknowledgement of this fact is the essence of the dyadic approach to caring that has been argued in favour of in the preceding section. However, it is in contrast to the way in which carers have typically been represented in the academic literature, that is, rather more as (albeit participant) observers, suffering the ill-effects and hardships of the toll exerted by the care-giving role, but essentially ignoring their ability to influence the process of caring and its outcomes at different stages, except through their practical efforts. It has rarely been acknowledged that their judgements and decisions are almost certainly influenced by their emotional, as well as cognitive state, sometimes leading to the observed disparities between their views and those of the person they help to care for. This is evidenced in the fact that carers frequently rate their care recipient's disabilities higher and their quality of life as much lower than does the patient him/herself (Fleming et al. 2005). Very little appears to be known about the way in which carers evaluate their care recipients' affective and cognitive functioning, still less about how these judgements may affect their own mental state. These considerations provide a salient reminder of the necessity for a comprehensive and evidenced based approach to understanding the caring process, in order to fully account for its multi-dimensional nature. Only then will it be possible to construct rational intervention procedures which have improved chances of success, as they are more likely to address the real issues at stake in care-giving. An attempt to understand how carers' perceptions of the mental state of their spouses with PD influenced care-giving outcomes formed an innovative part of the empirical work presented in this volume, providing an elaboration of the dyadic approach to care-giving.

1.3 Statistical approaches to the research

A specific feature of the research presented in this volume is in the use of multivariate statistical methods to identify relationships among the data. This has been identified by Weintraub and Burn (2011) as a priority for research in the area of PD, and was undertaken in the present work in an effort to capture the multidimensional nature of both the disorder and the complexities of the care-giving situation. The use of univariate statistical approaches to data still predominates in the field and it is sobering to be reminded that these problems of analysis were remarked upon by Alexander Luria in 1976 (85-86; author's own translation from Polish):

> It is necessary to mention the final direction in which neuropsychological research is developing, which in this case is more technical in nature. During our many years of research we have collected numerous observations and facts which form the basis of neuropsychological theory. These however need to be specified with greater precision, require further deliberation and in certain cases should be presented in the form of quantitative indicators, which would ensure their credibility.
>
> We are far from endorsing the view that measurement alone is the true criterion of science. The ordinary methods of medical statistics cannot be rated particularly highly, as they are able to do little more than to indicate the number of cases highlighting a particular symptom. In neuropsychology, as in other scientific disciplines in which the amount of material available for examination is relatively small, the centre of interest ought to be transferred to intercorrelations among symptoms and it is for these purposes that special mathematical methods need to be developed.
>
> There can be no doubt that this task can be realised by a single generation of scientists; indicating the necessary directions in which

developments ought to occur may help to undertake the necessary research activities.

In a field that is characterized by complex interdependencies among the many factors which combine to determine the well-being of both the person with PD and his/her carer, it is impossible to reach an understanding of the care-giving situation and crucial influences upon it, without adopting a more sophisticated approach to data analysis allowing for the covariance between key variables to be taken into account. These considerations were fundamental to the study conducted in three stages which is described in this volume.

1.4 Overview of the study

The model of PD as a neurodegenerative disorder was chosen for this study because the illness presents with symptoms in three distinct areas of functioning: physical, affective and cognitive, which provides the opportunity for examining how the three symptom domains relate to one another. It also allows a controlled way of determining how the effects wrought by the disabilities emerging from the impairment in these specific areas may affect not only the person suffering from the condition, but also his/her nearest relatives. In the case of the present study, the latter is the patient’s spouse, and with the progression of the illness, s/he becomes increasingly involved in assisting with the day to day tasks of everyday living. The couple become entwined with the illness and both are affected by it, albeit in different ways. The resultant process, one of care-giving and care-receiving, is two-way and is based on a dyadic relationship between the patient and the spouse/carer.

The aim of the present work is to attempt a systematic approach to achieving an understanding of the various situational, clinical and relational interactions shaping different aspects of care-giving in PD, utilising a neuropsychological approach to the analysis. In order to do so, it is necessary to start by becoming acquainted with the current state of knowledge with regard to the three main symptom domains of PD. Hence chapter 2 provides a brief account of the nature of PD describing the key motor aspects of the disorder, its clinical presentation, and approaches to treatment of the condition. The two chapters that follow are concerned with the non-motor symptoms of PD: chapter 3 adopts a neuropsychiatric perspective, focusing mainly on the affective disorder which is the most common of these complications, but also providing a brief description of other psychiatric disturbances frequently present in PD. Chapter 4 provides an overview of the neuropsychological (cognitive) dysfunctions associated with PD. Both chapters 3 and 4 examine the mechanisms underlying the respective disorders and evaluate their aetiological status as arising from a common neurodegenerative processes or as comorbid conditions. In chapter 5 the effects of these three symptom areas on the disability in PD are considered along with the psychosocial adaptation that patients undergo in coming to terms with the condition. Consideration of functional adaptation to neurodegenerative disease processes forges a link between the patient and those closest to him/her, who through being involved in their relative's care, are also intimately involved in the experience of living with the condition from the perspective of carers. This is the subject matter of chapter 6, which provides the context to care-giving and critically examines different approaches to conceptualising care-giving outcomes before presenting the main theoretical models that have been applied to caring. A review of the research relating to care-giving in PD is presented in chapter 7 with a focus on identifying factors which lead to poorer outcomes in carers. Circumstances which may help to remediate some of the adverse effects of long-term care-giving commitments, with particular reference to the way in which adult attachment styles may affect the carer-care recipient relationship and help to promote relationship satisfaction, thus contributing to buffering the negative effects of care-giving are also considered in chapter 7. Other protective mechanisms reviewed include cultural expectations and family relationships. Chapters 2-7 form the theoretical basis for the research study, described in the later chapters. The empirical study is divided into three parts, parts I and II being concerned with testing specific hypotheses concerning the relationships among symptoms in PD. Affective symptoms, which as mentioned above, have hitherto received relatively little attention, were of special interest to the research, in view of the emerging evidence that they have particular significance for carer outcomes. Part III of the study focused on evaluating a model of the care-giving process, specifically taking into account carer perceptions of the patient's symptom presentation in the area of psychological functioning and the quality of the dyadic relationship between the carer and the care recipient. The justification for this approach is presented in chapter 8. Chapters 9 and 10 respectively, provide a description of the methods used and of the results obtained. Chapter 11 contains a discussion of the findings of stages I and II of the research which focused on the neuropsychological presentation of PD and chapter 12 presents a discussion of the empirical study of the dyadic relationship between the patient with PD and his/her carer, carried out in stage III of the research with reflections on the implications for clinical practice.

CHAPTER TWO

PARKINSON'S DISEASE: THE NATURE OF THE DISORDER

2.1 Introduction

The diagnosis of PD is made primarily on the basis of the clinical manifestation of the characteristic motor symptoms of the disorder. It is a progressive condition, with insidious onset, and the motor symptoms eventually give rise to the disabilities that seriously impair activities of daily living and lead to the need for care. This chapter provides an overview of the clinical presentation of PD in terms of the main symptoms and their clinical course, together with a classification of the disorder according to syndromes. There follows a discussion of the neural mechanisms underlying the disorder and a summary of the evidence regarding its possible aetiology. Finally the main forms of treatment, pharmacological and surgical, are presented.

2.2 Parkinson's disease – symptom presentation and clinical course of the disorder

PD was described for the first time by James Parkinson, a British physician, in 1817, and was originally known as paralysis agitans or the "shaking palsy". It is a sporadic (or idiopathic), degenerative disorder, once thought to affect only the central nervous system, but now known to be associated also with pathology of the peripheral and enteric nervous systems (Braak and Del Tredici 2008). It has thus increasingly been recognized as a multisystemic disorder, in which there is preferential obliteration of dopaminergic neurons in the nigrostriatal pathways of the basal ganglia. Clinically, the disorder is primarily associated with late age at onset and relatively slow progression of symptoms (Fahn 2003). There is general agreement that the condition presents with four cardinal symptoms, which include (i) a resting tremor and shaking of limbs, (ii) muscular rigidity, (iii) slowness and poverty of movement and (iv) postural instability (Caslake et al. 2008; Fahn 2003; Probst, Bloch and Tolnay 2008). The cause of the condition remains unknown, although some forms of the disorder show a slight familial tendency in patterns of occurrence (Lang and Lozano 1998) and a number of genetic mutations have been implicated, albeit in a minority of cases (Davie 2008; Weintraub, Comella and Horn 2008). The disorder stems from the degeneration of nuclei in the dopaminergic neurons of the brainstem, leading to depletion of the neurotransmitter dopamine in these regions. When dopamine production is reduced, the motor system nerves are unable to control movement and coordination. Patients suffering from PD are known to have lost 70%-80% of their dopamine-producing cells by the time that symptoms first appear (Kuran 2002; Martin 1998; Youdim and Riederer 1997). The discovery that PD was associated with dopamine depletion by Hornykiewicz in the 1960s led to the successful treatment of the disorder with the drug levodopa (L-dopa) (Górska and Majczyński 1997; Hornykiewicz 2006).

PD is the second most common neurodegenerative disorder, after Alzheimer's disease (DAT; Choonara et al. 2009; de Lau and Breteler 2006). The age at onset is typically in the sixth decade of life (mean age at onset is 60 years) with approximately 1-2% of the world's population in this age group affected by the disorder (Olanow, Stern and Sethi 2009; Post, Speelman and de Haan 2008). The condition is rare in people below 30 years of age, and risk for the disease increases with age. It is estimated that 5-10% of patients who have PD, experience symptoms before the age of 40 (Golbe 1991; Muthane et al. 1994). PD is common in the elderly and affects more than 3% of the population aged more than 75 years and 4% of those over the age of 80 (de Lau and Breteler 2006). The overall incidence rate, adjusted for age and gender, is estimated at 13.4 per 100,000, with a higher prevalence among males (19.0 per 100,000) than females (9.9 per 100,000) (Ben-Shlomo 1997; Van Den Eeden et al. 2003). Hence the relative risk for men of developing the condition appears to be approximately twice that for women, and other studies are broadly in keeping with these estimates e.g. Wooten et al. (2004) found a relative risk for men of 1.5 times greater than in women. There has been considerable controversy regarding variations in PD associated with race or ethnicity (Friedman 2006; Stern and Lees 1982), although in one American study, PD appeared to be higher among Hispanics than Caucasians, but less common among Asians and African-Americans (Van Den Eeden et al. 2003). There also appear to be geographical variations in the occurrence of PD: the frequency is greater in industrialized than in non-industrialized regions, and higher in Europe than in North America (Choonara et al.

2009), although rates in some countries (e.g. Poland) appear to be disproportionately low in comparison to other European countries (Friedman 2006). The disorder carries a risk of earlier mortality compared with the general population, especially for people with a young age at onset of the condition (Ishihara et al. 2007), although there is some evidence to suggest that the death rates among both men and women with PD have fallen in recent years in the U.K. (Mylne et al. 2009). There is also an increased risk of morbidity in persons with PD, which is associated with complications of inactivity, including poor nutrition, cardiovascular disease and pneumonia (Martin 1998).

2.2.1 The clinical presentation of Parkinson's disease

The main symptoms of PD usually develop over time; however patients do not have to present with the full symptom complex of bradykinesia, muscular rigidity, resting tremor and postural instability in order to be diagnosed with the disorder and indeed most patients do not develop the full range of symptoms that are associated with the condition (Zigmond and Burke 2002). Symptoms are usually lateralized at the start of the illness, but over time, gradually begin to affect both sides of the body. The rate of symptom progression is extremely variable; some patients become profoundly disabled whilst others continue to function relatively well, for up to ten years after the initial diagnosis. The symptoms of PD often show a great deal of fluctuation, often from day to day, but even from one moment to another. The reasons for this are often unclear, but are thought to be attributable to the disease process or to medication. Such fluctuations, when they occur, are difficult to manage clinically and are particularly problematic for patients.

2.2.2 Primary symptoms

The primary symptoms of PD, considered as being characteristic of the disorder, are tremor, rigidity, bradykinesia and postural instability (Caslake et al. 2008; Probst et al. 2008; Zigmond and Burke 2002). The description that follows is partly based on a classic and beautifully written text by Stern and Lees (1982), authorities in the field of PD, which is aimed at providing patients and their families with information about the condition.

The sign most frequently associated with PD, probably because it is so conspicuous, is that of tremor. It affects about 70% of patients with PD (Hoehn and Yahr 1967) and because it is perceived as being such an