PSYCHOLOGICAL MORBIDITY FOLLOWING GENETIC COUNSELLING TO ESTABLISH DEGREE OF RISK FOR AUTOSOMAL DOMINANT DISORDERS: HUNTINGTON'S DISEASE AND MARFAN'S SYNDROME

AND RESEARCH PORTFOLIO

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Thesis submitted in partial fulfilment of the degree of Doctor of Clinical Psychology, in the Faculty of Medicine, University of Glasgow

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Psychological implications of predictive testing for autosomal dominant disorders: The case of Huntington's Disease

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Short title for running head: Psychological implications of predictive testing for Huntington's Disease.

Prepared in accordance with the submission requirements for the British Journal of Health Psychology (see appendix 1).

Psychological implications of predictive testing for autosomal dominant disorders: The case of Huntington's Disease.

Abstract

Advances in molecular genetics have led to the availability of predictive test programmes for some individuals who are at risk for genetic disorders. This paper reviews the psychological implications of giving predictive test information to individuals who are at risk for the autosomal dominant disorder, Huntington's Disease. The implications for those receiving both increased and decreased risk advice are considered. Studies to date have concluded that in the majority of cases the benefits of predictive testing for this disorder far outweigh the costs. The author suggests that such conclusions may be premature and highlights the need for further research in this area.

Introduction

Over the last decade significant advances in molecular genetics have enabled the implementation of predictive test programmes for some individuals who are at risk for genetic disorders. Predictive tests to establish degree of risk for Breast Cancer, Huntington's Disease, familial Alzheimer's Disease and many other disorders are now widely in use and readily available to those individuals who

seek them. Whilst on the one hand, such advances are seen to be beneficial, in that they give people the opportunity to plan their futures and families with new insight, concern has been voiced regarding the possible adverse psychological implications of giving people such information, especially in the case of conditions which promise severe disability and which will ultimately be fatal (Kessler, 1987; Tyler & Harper, 1983).

The aim of this paper is to review the literature relating to the psychological implications of predictive testing for the autosomal dominant disorder Huntington's Disease (HD). HD is a neurodegenerative disorder which manifests itself typically around the age of 35-42 years of age. It presents with a triad of symptoms - a choreiic movement disorder, cognitive impairment and psychiatric disturbance (Harper, 1991). The course of HD lasts approximately 15 years and it is ultimately fatal. All offspring of an affected individual start life with 50% chance of having inherited the gene for the disorder. The gene has complete penetrance, therefore, all those who have the gene will ultimately develop HD, assuming that they live to the age of onset.

In 1983 work by Gusella and colleagues identified a polymorphic marker linked to the HD gene on chromosome 4 (Gusella, Wexler, Conneally, Naylor, Anderson, Tanzi, Watkins, Ottina, Wallace, Sakaguchi, Young, Schoulson, Bonilla & Martin, 1983). This discovery enabled predictive testing for some family members, with approximately 95% accuracy. Further developments in 1993 led to the identification of the gene itself (Huntington's Disease Collaborative Research

Group, 1993). Since then predictive tests have been available to all individuals and results are considered 100% accurate. Following a predictive test an individual is informed that their risk has altered from 50% to either 0 or 100%. It is intuitively obvious that such a change in risk status will have psychological implications both for the individual and also for their relatives.

Prior to the development of the predictive test for HD there was considerable moral and ethical debate regarding whether or not a test should be made available for a condition for which there is as yet no treatment programme to halt the degenerative process. A number of authors anticipated catastrophic responses, particularly in those individuals given increased risk status (e.g. Mastromauro, Myers & Berkman, 1987; Meissen & Berchek, 1987). High suicide rates were predicted and Hayden (1991) advised that considerable psychological harm would be inflicted on individuals should the availability of the predictive test programme increase. Perry (1981) argued that in the light of a likely increased risk for suicide a predictive test for HD should not be made available at all. A similar view was held by Marsden (1981).

It is now nearly ten years since the first predictive test programmes came in to operation and a number of studies have investigated the psychosocial consequences for those individuals who have already received modified degree of risk information. Despite the initial pessimism surrounding the implementation of this predictive test, studies to date have suggested that overall the benefits of having a predictive test for HD far out weigh the costs and that the psychiatric

risk to an individual receiving a test result is minimal (e.g. Hayden, Bloch & Wiggins, 1995). The sections below examine the evidence for such conclusions and highlight the need for continued research in this area.

Psychological response to the receipt of increased risk status

As mentioned above, discussion surrounding the implementation of predictive testing for HD tended to be somewhat pessimistic. The majority of this pessimism was directed at the likely consequences for those individuals for whom the result was positive. It was anticipated that these were the people most likely to be at risk for psychiatric disturbance and suicide (e.g. Perry, 1981). Studies to date have largely concluded that such pessimism was somewhat premature. A study by Bloch, Adam, Wiggins, Huggins & Hayden (1992), for example, presents four case studies discussing the experience of those receiving increased risk results. Responses of 66 further patients are included in their discussion. They conclude that no candidate who has received an increased risk result has made a suicide attempt or required psychiatric hospitalisation. Similar findings are presented by Simpson, Besson, Alexander, Allan & Johnston (1992). Quaid (1993) suggests that the rate of psychiatric hospitalisation following predictive testing for HD is 2%. The figure is low and may not be higher than that observed in the at-risk population as a whole (Schoenfeld, Myers, Cupples, Sax & Clark, 1984). It is obviously not possible to rely solely on rates of suicide and psychiatric hospitalisation in order to reach conclusions regarding psychological aspects of

outcome since individuals may suffer adverse psychological reactions to predictive test results without falling in to either of these categories. If one looks closely at the Bloch et al. (1992) study it is implied that some of the participants in their testing programme did present with difficulties of a psychological nature. In this study participants were assessed using the Symptom Checklist 90 - Revised (SCL 90-R) and the Beck Depression Inventory (BDI). Assessments were carried out at baseline, at 1 week and at 2, 6, 12, 18 and 24 months post test result. The authors present scores for the four individual case studies but do not give details of the additional 66 participants. They report, however, that 'most' patients experienced significantly increased stress when dealing with the immediate impact and shock of receiving an increased risk result, with symptoms of anxiety and depression being highest in the first two months post test. This was most commonly followed within a year by adjustment and return of stress and features of depression (as measured by the BDI) to pretest levels. It is unclear how many patients constitutes 'most'. They also fail to identify how many of these patients reached diagnostic criteria for a depressive illness. The authors state that they selected the four case studies to represent common themes. They do not say what these themes are and unfortunately do not give any indication as to how representative the four cases are of the other participants in their predictive test programme. It is notable, however, that of the four presented cases only one would be considered psychologically distressed using the SCL 90-R and only two of the four would score above the cut-off point for caseness on the BDI (Beck, Ward, Mendelsohn, Mock & Erbaugh, 1961) at any stage in the predictive test programme. This is a finding similar to that observed by Meissen, Myers, Mastromauro, Klinger &

Farrer (1988) who observed that out of four individuals who had received an increased risk result two demonstrated features of depression when assessed three months after the receipt of their test result. Wiggins, Whyte, Huggins, Adam, Theilmann & Bloch (1992) also working with the Bloch et al. (1992) sample concluded that at one year follow-up BDI and SCL 90-R scores for those receiving increased risk results were no higher than at baseline and that for many individuals scores had actually decreased. This led the authors to conclude that for some individuals predictive testing was beneficial even if the results placed them in the increased risk group. It should be noted that in both the Wiggins et al. (1992) and the Bloch et al. (1992) papers the timing of the baseline assessments are not given. They are, however, presumably taken after the individuals have decided to seek information about the predictive test programme. Seeking this information alone could pose psychosocial consequences for some individuals. To date, no study has considered this possibility.

The few studies that have been carried out on this patient group give the impression that although the test programme can lead to distress for some participants the overall levels of distress are not severe. Further evidence for this comes from Brandt, Quaid, Folstein, Garber, Maestri, Abbott, Slavney, Franz, Kasch & Kazazian (1989) who noted that two years after testing the mean Global Severity Index of the SCL 90-R remained within normal limits for 12 people who had received an increased risk status. A later study carried out by Codori & Brandt (1994) investigated the views of 17 participants who had received increased risk results. Using open ended questions they found that 47% of this

sample felt that the information they received was an emotional burden for them and 43% felt guilty about the possibility of having passed on the gene. Having said that, 76% said that they spent less time worrying about getting HD and most test participants stated that given the choice they would have the test again. Codori & Brandt concluded that for these individuals the benefits of predictive testing for HD outweighed the costs. Their conclusion is subjective and it should be noted that the time since individuals received their test result varies in their sample between one and six years. It may be that individual responses to the questions posed vary with time. They may also vary with age of the participant and proximity in age to expected time of onset. Demographic data of this nature is not considered. One other point worth mentioning is that in their study Codori & Brandt interpret the reported reduction in worry regarding the development of HD as being beneficial. Tibben, Duivenvoorden, Vegter vanderVlis, Niermeijer, Frets, vandeKamp, Roos, Rooijmans & Verhage(1993), by contrast, have argued that decreased preoccupation with HD reflects denial which may in turn lead to psychopathology. An argument against this is that denial may in any case be quite adaptive since at-risk individuals are not ill and, therefore, no specific health related behaviours are required in relation to the information they receive.

The studies described have certainly given us information of importance. Sample sizes, however, are in most instances small and, as mentioned, some methodological issues need to be considered when interpreting conclusions that have been reached. As yet, no study adequately provides a systematic profile of psychological difficulties encountered by this patient group.

One final point which must be taken in to account when investigating psychosocial difficulties in those receiving an increased risk result is the fact that psychiatric disturbance is an early indication of HD in some patients (Harper, 1991). It is necessary to find a way of differentiating between individuals in early stages of the condition and asymptomatic individuals who are experiencing psychiatric symptoms as the result of the predictive test process.

Psychological response to the receipt of decreased risk status

Before predictive test programmes became widely available it was thought by some researchers that the receipt of decreased risk status for the HD gene would promise clear benefits (Crauford & Harris, 1986). Crauford & Harris (1986) believed in this so strongly they advocated that providing follow-up to those receiving such a result would not be justified. Others did not agree; Meissen & Berchek (1987), for example, anticipated that those individuals who were informed that they did not carry the HD gene would be at risk for experiencing survivor guilt, especially in cases where they had a sibling who was known to carry the gene. It was felt that survivor guilt would in turn lead to a range of psychological difficulties. Such an argument has been supported by the findings of Huggins, Bloch, Wiggins, Adam, Suchowersky, Trew, Klimek, Greenberg, Eleff, Thompson, Knight, MacLeod, Girard, Theilmann, Hedrick & Hayden (1992). They present 4 case studies and material based on a further 101 patients who have received decreased risk status. Participants described are taken from the same test

programme as those in the Bloch et al. (1992) study. Case A reported significant depression and guilt at having deserted her brother who was already affected. The discussion indicates that 10% of these decreased risk patients required professional intervention due to significant psychological difficulties coping with their change in risk status. It is unclear how representative the case studies are of the total patient group and details regarding the nature of the difficulties leading to the requirement for professional intervention are not given.

The Codori & Brandt (1994) study also investigated the perceived outcome for those individuals who had received decreased risk advice. 31% reported that they had experienced no negative outcomes to the test programme. 26% of those questioned reported feelings of guilt and this was most common in cases where a sibling had received an increased risk result.

As with the increased risk group there is as yet no adequate study which has provided a detailed profile of psychological deficits encountered by those who have received decreased risk results. A number of authors have, however, suggested reasons as to why one might expect deficits in this group, in addition to the presence of survivor guilt mentioned above. Huggins et al. (1992), for example, suggest that for many at-risk individuals their identity is developed with the assumption that they like other family members will develop HD at some stage in the future. When they are then told that they do not have the gene a large part of their identity is taken from them; as one of the Huggins et al. cases said, 'If I'm not at-risk then who am I?'. The authors suggest that psychological dysfunction

will be most common in cases where the decreased risk result is at odds with the individual's anticipated result and in cases where previous irreversible actions, such as sterilisation, have been carried out on the assumption that they would develop the condition. They also predict that some individuals who have high dependence needs may see the development of HD as being positive in the sense that it would lead to the caretaking they long for. The theory would appear to make sense although further research with large samples would be required to assess its validity.

Certainly, in the majority of cases it would appear that response to decreased risk advice is favourable. There are, however, some individuals for whom the change in risk status prompts psychological difficulties. Further research should aim to profile the nature of difficulties in this group and also to identify factors which would predict those individuals most vulnerable to developing them.

The need for further research

There are a number of reasons as to why further research in this area is necessary, in addition to those highlighted above. Firstly, it has been suggested that those individuals who have sought genetic testing to date do not represent the at risk group as a whole but instead represent a group of self selected individuals who have relatively good psychological functioning. This suggestion followed the observation that although the majority of at risk individuals expressed an interest

in taking a predictive test prior to its implementation few actually followed up this interest. In one study it was found that only 10% of those who had access to the test programme went ahead and took it (Meissen & Berchek, 1987). The issue of self selection was investigated by Codori, Hanson & Brandt (1994) who carried out a comparison study of 32 at risk individuals who had sought predictive testing with 32 individuals who had previously considered testing but who had later decided not to go ahead with it. They concluded that those who chose to take the test saw themselves as being able to cope with 'bad' news whereas those who did not take the test anticipated difficulties associated with their emotional reactions should the test result indicate that they carry the gene. Similar conclusions were reached by Decruyenaere, Evers-Kiebooms, Boogaerts, Cassiman, Cloostermans, Demyttenaere, Dom, Fryns & Van Den Berg (1995). They investigated the psychological profile of test applicants using the Minnesota Multiphasic Personality Inventory and concluded that those individuals who chose to have the test constituted a self selected sample who are better able to cope with stress and more adjusted psychologically than the general population.

If it is the case that those who have already gone through the predictive test programme represent only a specific group of the total at-risk population then we can not assume that the nature of this group will remain constant. A possible reason for this is that increasing demands from insurance companies and employers may lead to some individuals seeking genetic testing when they would not previously have done so. Another factor which may also be relevant has been put forward by Codori et al. (1994) who have suggested that at risk individuals

are already 'shopping round' for predictive test programmes which do not demand so much time. This may lead in the future to some individuals going ahead with the test when they would have decided not to under conditions of more extensive pre test counselling. These factors point to the fact that future predictive test participants may differ from those observed in previous studies. It will, therefore, be necessary to continue monitoring psychosocial outcome in future patient cohorts.

The second factor which indicates the need for future research is the observation that current knowledge in the area is largely based on work carried out in two localities. The Bloch et al. (1992) and the Huggins et al. (1992) studies were based on the Canadian Collaborative Test Programme which was developed in British Columbia. The conclusions reached by Codori & Brandt (1994) and Codori et al. (1994) are based on samples of test applicants in Baltimore. It is not possible to assume that conclusions reached in these studies will also hold for samples elsewhere. Results will clearly be influenced by the nature of the predictive test programme itself, including the quality of pre-test counselling and follow-up. In addition, cultural factors as yet undetermined may influence response to genetic risk advice. It is the authors intention to investigate response to the predictive test for Huntington's Disease in a sample from Scotland.

One final point which must be considered is that since the HD gene was identified in 1993 research has focused on the assessment of those individuals who are at risk for this particular disorder. There is a danger that conclusions reached

regarding the effects of genetic testing for Huntington's Disease could be over generalised and held to indicate a likelihood of similar responses to inheritance of other autosomal dominant disorders. There is clearly a case for identifying psychological responses to other autosomal conditions in order to identify if there are illness specific responses as well as responses influenced by the specific pattern of autosomal dominant inheritance. The author intends to compare a sample of patients who have sought genetic testing for HD with those who have been assessed for Marfan's syndrome, an autosomal dominant condition with a very different medical outcome.

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Research Proposal

Psychological morbidity following genetic counselling to establish degree of risk for autosomal dominant disorders: Huntington's Disease and Marfan's Syndrome

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The measures section of this proposal has been amended. Please see appendix 2 for details of amendments made.

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Title of the proposed project

Psychological morbidity following genetic counselling to establish degree of risk for autosomal dominant disorders: Huntington's Disease and Marfan's Syndrome.

Introduction

Over the last decade significant advances in medicine and in molecular genetics have enabled the implementation of predictive test programmes for some individuals who are at-risk for genetic disorders. Predictive tests to establish degree of risk for breast cancer, Marfan's Syndrome, Huntington's Disease and

many other disorders are now widely in use and readily available to individuals who seek them. Whilst, on the one hand, such advances are seen to be beneficial, in that they give people the opportunity to plan families and futures with new insight, concern has, nevertheless, been voiced regarding the possible adverse psychological implications of giving individuals such information, especially in the case of conditions which promise severe disability and which will ultimately be fatal (Kessler, 1987; Tyler & Harper, 1983).

The proposed project is designed to investigate symptoms of psychological disturbance following the receipt of genetic risk advice in the case of disorders which are transmitted by autosomal dominant inheritance. In these cases all offspring of affected individuals start life with a 50% risk of having inherited the disease in their family. If, following genetic counselling, they are then told that they carry this gene not only does this imply that they will develop the disorder, at some undefined time in the future, but, in addition to this, they will have the added burden of also knowing that they may have already transmitted the gene to 50% of their children. It is intuitively obvious that such a change in status from being at 50% risk to being at 0 or 100% risk can not fail to have psychological implications. Kessler, in 1987, warned that in the case of Huntington's disease a positive test result would lead to significant risk of psychiatric disorder or suicide. Others speculated that this would be the case and recommended extensive counselling for these individuals (Harper, Morris & Tyler, 1990).

Despite speculation and concern from both professionals and the public, very few studies have investigated the profile of psychological disturbance in those who have already received genetic risk information. Predictive test programmes follow strict protocols which include protocols for aftercare. Protocols for aftercare which are currently in use were designed without knowledge of likely psychological responses to predictive test programmes, a fact which now seems a little shortsighted.

To date, those studies which have attempted to investigate psychological disturbance in these patients groups have, for the most part, been inadequate, most frequently based on small sample sizes or use of inadequate measures (e.g. Huggins et al., 1992; Bridges et al., 1992). They have, however, provided information which has been of importance. Huggins et al. (1992) present five case histories of individuals who suffered psychological distress following receipt of information implying decreased risk for Huntington's Disease. Crauford & Harris(1986) suggested that follow-up provisions would not be required for this group, a suggestion clearly based on inadequate information. There is an obvious need to investigate the issue more fully if one is to provide a post-test service which is of optimum benefit to those who require it.

The present study aims to provide a profile of psychological disturbance in people who have sought genetic counselling in order to establish their degree of risk for Huntington's Disease or Marfan's Syndrome. Huntington's Disease is a neurodegenerative condition which manifests itself typically around the age of 35-

42 years of age (Folstein, Chase & Wahl, 1987). It presents with a triad of symptoms - a choreiic movement disorder, cognitive impairment and psychiatric disturbance. The course of the disorder lasts around 15 years and it is ultimately fatal. Marfan's syndrome is a disorder of connective tissue, in which there is an ever present threat of sudden death from ruptured aortic aneurysm. The immediate threat in Marfan's syndrome is, therefore, far greater than it is in Huntington's Disease.

Published studies regarding the psychological effects of predictive testing for Huntington's Disease and Marfan's Syndrome are, to date, inadequate. Huggins et al. (1992), as mentioned, presents five case histories of individuals receiving decreased risk advice for Huntington's Disease. Bloch et al. (1992) present 4 cases of individuals receiving increased risk advice. Samples are small. In the case of Marfan's Syndrome only one paper was identified. Bridges et al. (1992) investigate the responses of 37 family members seeking risk advice. The authors note that following receipt of this advice '41% of patients were more worried about their health and 48% more worried about their future.' Those given increased and decreased risk advice are not differentiated and the study does not investigate the issue more fully. The current study will enable a more detailed profile to be produced for each disorder and will also enable comparison of responses to two conditions which are both transmitted by autosomal dominant inheritance but which have very different medical outcomes. It is hoped to be of benefit to those involved in future service planning.

Aims and hypotheses to be tested

The study proposes to investigate psychological morbidity following genetic counselling to establish degree of risk for Huntington's Disease and Marfan's Syndrome. It aims to answer the following specific research questions:

1. Is there evidence of psychological distress in individuals who have undergone genetic counselling to establish degree of risk for either Huntington's Disease or Marfan's Syndrome? If so, what is the nature of this disturbance?

2. Are there differences in the nature, timing or duration of psychological disturbance following receipt of increased or decreased risk status?

3. Do people given increased or decreased risk status differ in their use of resources e.g. visits to G.P.s, further counselling or use of self help groups?

4. Is there a difference in reactions to knowledge of inheritance of Huntington's Disease and Marfan's Syndrome?

Josephine Green, argues that the role of psychology in predictive test programmes is of paramount importance. She argues that psychology currently has a role to play in describing and measuring the psychological effects of genetic screening and that later it will have a role to play in the prediction, anticipation and explanation of the behaviour of those individuals who have undergone such

screening (Green, 1995). The present study provides a starting point for such a process by providing a detailed profile of psychological symptoms experienced by individuals following genetic counselling to establish their degree of risk for Huntington's Disease or Marfan's Syndrome. The study will also enable assessment of whether this is dependent on the degree of risk identified.

It is predicted that individuals in both increased and decreased risk status groups will experience some form of psychological stress following genetic counselling which leads to a change in risk status. It is also predicted that the nature of disturbance will differ according to status group. The 'survivor syndrome' amongst those of decreased risk status is already documented although as yet it has not been adequately identified (Simpson et al., 1992). The survivor syndrome acknowledges that those given decreased risk status frequently experience feelings of guilt at having escaped a disorder prevalent in other family members. Such feelings of guilt may leave individuals prone to depression which, as Huggins et al. (1992) point out, may be exacerbated by previous irreversible actions based on a prior assumption that they, like other family members would develop the disorder. Those given increased risk status may also experience depression if they consider themselves helpless and unable to control their condition. They may also experience anxiety regarding both their own future and that of their children.

The study also compares response to genetic counselling for two conditions that have very different medical outcomes. It is expected that the two groups will differ since the immediate threat posed by diagnosis is far more severe in the case

of Marfan's syndrome. It is, therefore, predicted that symptoms of anxiety will be highly prevalent amongst those individuals receiving increased risk status for this condition. In addition, it is predicted that individuals in this group will place a far greater demand on available resources and require substantially more post-test support than will those at high risk for Huntington's Disease.

The proposed study can adequately assess the validity of these predictions.

Plan of Investigation

Recruitment of Volunteers

Forty volunteers who have sought counselling to establish their degree of risk for either Huntington's Disease or Marfan's Syndrome will be recruited to the study by Dr. Sheila Simpson and Dr. John Dean. 10 individuals will be recruited following increased risk advice and 10 recruited following decreased risk advice for each of the two conditions. All individuals recruited will have received their genetic risk advice a minimum of one year prior to their participation in the study.

Measures

Data for each individual will be collected during one semi-structured interview. It is envisaged that the duration of this interview will not exceed 1.5 hours. Interviews will be conducted at Aberdeen Royal Infirmary or at the individual's own home, dependant on their preference.

During the interview volunteers will be asked to complete the following self report questionnaires:

Beck Depression Inventory (Beck, Ward, Mendelson, Mock & Erbaugh, 1961)

Beck Anxiety Inventory (Beck, 1987)

General Health Questionnaire (Goldberg, 1981)

These questionnaires will provide a measurement of psychological disturbance at the time of interview. In addition, open ended questions will be used to derive self report data detailing the nature, timing and duration of any mood changes or sleep disturbance which have occurred since receiving genetic risk advice. Volunteers will also be asked to detail their use of resources (further counselling, use of self help groups and visits to G.P.s) during the six month periods before the onset of genetic counselling and after receiving degree of risk information. With the volunteers' consent G.P.s will be contacted and asked to complete a short questionnaire detailing the frequency and nature of visits during these periods.

Design and Procedure

Two main groups of patients who have sought genetic risk advice will be assessed; the first group will provide a sample of individuals who have sought genetic advice regarding risk for Huntington's Disease and the second group will provide a sample of individuals who have sought advice to establish degree of risk for Marfan's Syndrome. Each of these two main groups will be subdivided according to individuals' increased or decreased risk status. There will, therefore, be four groups in total. Comparisons will be made between the two main groups in order to compare the psychological morbidity which follows genetic counselling for the two conditions. Within each main group comparison will also be made of individuals of increased and decreased risk status. From these comparisons it will be possible to derive answers to the proposed research questions.

Management and Analysis of Data

All information collected in the course of the study will be treated as confidential and stored in the manner of medical records at the Department of Psychological Medicine, University of Glasgow.

Data will be analysed using SPSS-X, available on Unix at Glasgow University. Since the data collected will be categorical non-parametric statistical tests will be employed.

Practical Applications

The identification of a profile of psychological disturbance in these patient groups will be of benefit to future resource allocation and will be helpful in the planning stages of future post-test aftercare protocols. In addition, it may be possible in the course of the study to identify factors which make individuals vulnerable to psychological disturbance following the receipt of genetic risk advice. Investigation of the resources which people find most helpful and which they use most frequently will also provide useful information which will be of value in assisting and advising people who seek genetic advice in the future.

Timescales

It is envisaged that data will be collected between 1.9.95 and 29.2.96 and that data analysis and preparation of a research paper will take an additional two months. It is anticipated that the project will be completed by 1.5.96.

Ethical Committee Approval

Ethical Committee Approval for the proposed project was granted by the Joint Ethical Committee of Grampian Health Board and the University of Aberdeen on 30th March 1995.

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Psychological morbidity following genetic counselling to establish degree of risk for autosomal dominant disorders: Huntington's Disease and Marfan's Syndrome

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Prepared in accordance with the submission requirements for Psychiatric Genetics (see appendix 3).

Psychological morbidity following genetic counselling to establish degree of risk for autosomal dominant disorders: Huntington's Disease and Marfan's Syndrome

Abstract

This study presents a profile of psychological disturbance and psychiatric diagnosis in thirty-nine volunteers who sought genetic counselling to establish their degree of risk for either Huntington's Disease or Marfan's Syndrome. 10 individuals were assessed following increased risk advice for each of the two conditions, together with 10 who had received decreased risk advice for Huntington's Disease and 9 who had received decreased risk advice for Marfan's Syndrome. These groups were compared on measures of anxiety and depression and psychiatric diagnoses using the Schedule for Affective Disorders and Schizophrenia. The effects of the receipt of genetic risk advice on relationships with family and friends and number of visits to G.P.s were also considered. The results indicated that a proportion of volunteers in all four groups experienced psychological distress of sufficient severity to reach diagnostic criteria for either Major Depression, Generalised Anxiety or Panic Disorder. Between group differences were insignificant.

Keywords

Huntington's, Marfan's, predictive, at-risk, psychological, depression, anxiety, counselling

Introduction

Over the last decade significant advances in medicine and in molecular genetics have enabled the implementation of predictive test programmes for some individuals who are at-risk for genetic disorders. Predictive tests to establish degree of risk for breast cancer, Marfan's Syndrome, Huntington's Disease and many other disorders are now readily available to individuals who seek them. Whilst such advances are seen to be beneficial, in that they give people the opportunity to plan their futures with new insight, concern has been voiced regarding the possible adverse psychological implications of giving individuals such information, especially in the case of conditions which promise severe disability and which will ultimately be fatal (Kessler, 1987; Tyler and Harper, 1983).

This paper presents a study which investigated the symptoms of psychological disturbance experienced by individuals following the receipt of genetic risk advice for Huntington's Disease (HD) or Marfan's Syndrome (MSyn). HD is a neurodegenerative condition which manifests itself typically around the age of 35-

42 years of age (Folstein, Chase and Wahl, 1987). It presents with a triad of symptoms - a choreiic movement disorder, cognitive impairment and psychiatric disturbance (Harper, 1991). The course of the disorder lasts around 15 years and it is ultimately fatal. MSyn is a disorder of connective tissue, in which there is a threat of sudden death from ruptured aortic aneurysm. Both disorders are transmitted by autosomal dominant inheritance. This means that all offspring of affected individuals start life with a 50% risk of having inherited the condition. Genetic counselling to establish degree of risk can provide the knowledge to change this risk to virtually 0 or 100%. It is intuitively obvious that such a change in risk status will have psychological implications. Kessler, in 1987, warned that in the case of HD a positive test result would lead to significant risk of psychiatric disorder or suicide. Others speculated that this would be the case and recommended extensive counselling for these individuals (Harper, Morris and Tyler, 1990).

Despite such speculation few studies have investigated psychological disturbance in those who have already received genetic risk information. Blackmore (1996) reviews the literature relating to HD and notes that studies, to date, have concluded that in the majority of cases the benefits of predictive testing outweigh the costs. Blackmore (1996) suggests, however, that such a conclusion is premature and gives a number of reasons as to why this is the case. To begin with the sample sizes in previous studies have been small; Huggins, Bloch, Wiggins, Adam, Suchowersky, Trew, Klimek, Greenberg, Thompson, Knight, MacLeod, Girard, Theilmann, Hedrick and Hayden (1992), for example, presented five case

histories of individuals receiving decreased risk advice for HD. Bloch, Adam, Wiggins, Huggins and Hayden (1992) presented 4 cases of individuals receiving increased risk advice. In both these papers cases were selected from a larger sample. It is unclear how representative they are of the total patient group. Previous studies have also reached conclusions based on inadequate methodology. Bloch et al. (1992), for example, note that no participant in their predictive test programme has yet committed suicide or required psychiatric hospitalisation. They use this information to support the argument that overall the outcome of predictive test programmes is good. It would, however, be possible for a person to reach diagnostic criteria for a psychiatric illness without falling in to either of these categories. No study, to date, has adequately assessed whether or not those who receive genetic risk advice regarding risk for HD actually reach diagnostic criteria for a psychiatric illness after having done so. A number of studies have considered specific illnesses but they have used inadequate measures to assess their prevalence. For example, Wiggins, Whyte, Huggins, Adam, Theilmann and Block (1992) reached conclusions regarding the prevalence of depression in those that have under gone a predictive test using the Beck Depression Inventory (BDI: Beck, Ward, Mendelsohn, Mock and Erbaugh, 1961). Such an instrument does not provide evidence of diagnostic criteria and can only be considered a measure of depressive symptoms.

The decision to include both the HD and the MSyn patient groups in the current study was taken for two main reasons. Firstly, it was noted that since the gene for HD was identified in 1993 (Huntington's Disease Collaborative Research Group,

1993) the vast majority of research investigating the effects of predictive test programmes has focused on this particular disorder (Blackmore, 1996). It was felt that there was a danger that conclusions reached regarding the effects of genetic testing for HD could be generalized and held to indicate a likelihood of similar response to inheritance of other autosomal dominant disorders. The author felt that it was necessary to compare response to inheritance of HD with that for other autosomal dominant disorders in order to identify whether or not there are responses which are illness specific as well as responses which are influenced by the specific pattern of inheritance itself. MSyn with its very different medical outcome allows a good comparison for this purpose. The paucity of research with this particular group of patients also suggested that it was a population worth investigating in its own right. The author could only identify one study which had considered the psychological effects of genetic screening for this condition. Bridges, Faed, Boxer, Gray, Bundy and Murray (1992) investigated the responses of 37 family members seeking risk advice for MSyn. They found that following receipt of risk advice '41% of patients were more worried about their health and 48% more worried about their future.' Those given increased and decreased risk advice are not differentiated and the study does not investigate the issue more fully. The other main reason for selecting MSyn as a comparison group was that it is a condition for which psychiatric disturbance is not a symptom. Since psychiatric difficulties are a symptom in HD it is unclear whether difficulties of this nature in those who have the gene represent a response to the receipt of risk advice or whether they are an early sign of the disorder. With MSyn this difficulty does not arise.

The study presents a profile of psychological disturbance experienced by a Scottish sample of patients who had received genetic risk advice regarding inheritance for HD or MSyn. This profile was determined by psychological measures of symptoms of anxiety and depression and psychiatric diagnosis using the Schedule for Affective Disorders and Schizophrenia (SADS: Spitzer and Endicott, 1978). The study was designed to answer the following research questions:

1. Is there evidence of psychological and psychiatric disturbance in individuals who have undergone genetic counselling to establish degree of risk for HD or MSyn? If so, what is the nature of this?

2. Is there a difference in the nature of psychological and psychiatric disturbance following receipt of increased or decreased risk status?

3. Is there a difference in reactions to knowledge of inheritance of HD and MSyn?

The author predicted that individuals in both increased and decreased risk status groups would experience some form of psychological distress as a result of change in risk status. It was also expected that the nature of response to the change in risk status would differ according to the condition. The threat posed by MSyn is potentially more immediate than it is in HD and it was, therefore, predicted that symptoms of anxiety would be more prevalent amongst those

individuals receiving increased risk status for this condition. Depression, on the other hand, was not expected to differ in the two increased risk groups.

Method

Recruitment of Volunteers

Thirty-nine volunteers who sought counselling to establish their degree of risk for either HD or MSyn were recruited to the study. 10 individuals were recruited following increased risk advice for each of the two conditions, together with 10 who had received decreased risk advice for HD and 9 who had received decreased risk advice for MSyn. All individuals recruited received their genetic risk advice a minimum of six months prior to their participation in the study.

Genetic Screening

Genetic screening to establish degree of risk was carried out at Aberdeen Royal Infirmary by or under the supervision of consultants from the Department of Medical Genetics. Because of the very slight possibility of error in screening techniques results are referred to in terms of decreased or increased risk. Individuals who undergo such procedures are aware that these correspond to virtually 0 or 100% chance of having inherited the condition in question.

Psychological Measures

Psychological data for each individual were collected during one semi-structured interview. All interviews were carried out by Dr. Louise Blackmore (Psychologist). Interviews were conducted at Aberdeen Royal Infirmary or at the volunteer's own home, dependant on their preference.

The SADS is divided in to two sections which provide information detailing current and life time psychiatric diagnoses. For the present study the sections relating to current and lifetime episodes of depressive illness, generalised anxiety and panic disorder were included. Sections relating to schizophrenia were thought to be inappropriate in this case and were, therefore, excluded from the study out of sensitivity to the volunteers who took part. The researcher noted whether psychiatric diagnoses pre or post dated genetic risk advice.

Volunteers were also asked to complete the Beck Depression Inventory (BDI: Beck et al, 1961) and the Beck Anxiety Inventory (BAI: Beck, 1987). These questionnaires provided an additional measurement of psychological disturbance at the time of interview. Open ended questions were also used to derive self report data detailing the nature of any changes in relationships with family and friends since receiving genetic risk advice.

To obtain further information a questionnaire was sent to volunteers' G.P.s asking four questions. These were related to the number of times the volunteer had

visited their G.P. in the six month periods before and after receiving risk information. Questions were phrased in the following format:

a. Please indicate how many appointments the above patient had with a member of your practice between (date supplied) and (date supplied).

b. Of these how many would you consider to be due to stress or other psychological reasons?

Results

Demographic Variables

Table I presents demographic variables for the four groups. Time refers to the mean number of months since genetic risk advice was received. Analysis of variance for age indicated a significant between group difference ($F= 2.95$, $df= 3$, $p= .0459$). A post hoc Tukey test indicates that the mean age for the Marfan's Syndrome Increased Risk group (MSynI) is significantly greater than those for the other three groups ($p < .05$). Differences in the mean ages of the Huntington's Disease Increased Risk (HDI), Huntington's Disease Decreased Risk (HDD) and Marfan's Syndrome Decreased Risk (MSynD) groups are not significant. Analysis of variance also indicated a significant between group difference for Time ($F = 8.06$, $df= 3$, $p= .0003$). A post hoc Tukey test shows that the mean for the MSynI

group is significantly greater than those for the other three groups ($p < .05$). It is acknowledged that it would have been preferable to have an MSynI group with a mean time since assessment which was more comparable to the other groups. The rarity of the condition, however, did not allow for this. Differences in the mean times of the HDI, HDD and MSynD groups are not significant.

Table I. Demographic Variables in HDI, HDD, MSynI and MSynD Groups

Group	n	Age			Sex		Time	
		Mean	(SD)	Range	m	f	Mean	(SD)
HDI	10	42.0	(10.94)	25-55	2	8	44.0	(24.09)
HDD	10	39.6	(5.99)	32-49	2	8	55.2	(25.75)
MSynI	10	47.4	(16.59)	27-73	4	6	144.4	(117.42)
MSynD	9	30.9	(13.55)	17-56	3	6	11.9	(8.12)

Key: HDI - Huntington's Disease risk increased; HDD - Huntington's Disease risk decreased; MSynI - Marfan's Syndrome risk increased; MSynD- Marfan's Syndrome risk decreased; Time - Mean number of months since genetic risk advice was received

Schedule for Affective Disorders and Schizophrenia - Diagnoses following the receipt of genetic risk advice and at the time of assessment interview

Table II details the number of volunteers in each of the four groups who reached Research Diagnostic Criteria for Major Depression or Generalised Anxiety and/or Panic Disorder following the receipt of genetic risk advice and at the time of assessment interview. The Generalised Anxiety and Panic Disorder categories were combined owing to high co-morbidity between the two diagnoses. Fisher's exact tests were used to compare group frequencies. The difference between those reaching diagnostic criteria for Major Depression in the HDI and the MSynI groups following the receipt of genetic risk advice approached significance ($p = .065$). All other between group differences were insignificant.

Table II. No. of volunteers in HDI, HDD, MSynI and MSynD groups reaching diagnostic criteria for Major Depression, Generalized Anxiety and/or Panic Disorder following the receipt of genetic risk advice and at the time of assessment interview

	Major Depression		**Generalized Anxiety and/or Panic Disorder**	
	Post Advice	(diagnosis at interview)	Post Advice	(diagnosis at interview)
HDI n=10	5	(1)	3	(3)
HDD n=10	3*	(2)	2	(2)
MSynI n=10	1	(1)	3	(3)
MSynD n=9	0	(0)	2*	(2)

Key: HDI - Huntington's Disease risk increased; HDD - Huntington's Disease risk decreased; MSynI - Marfan's Syndrome risk increased; MSynD- Marfan's Syndrome risk decreased

* one case predates genetic risk assessment

Beck Depression Inventory and Beck Anxiety Inventory Scores

Table III summarises scores on the BDI and the BAI. Analyses of variance indicated no between group differences for either measure (BDI: F= .47, df= 3, ns; BAI: F= .89, df= 3, ns). Pearson Correlation Coefficients were used to correlate BDI and BAI scores with time since receipt of risk advice for each of the four groups. Correlations ranged from -.0897 (p= .527) to -.3756 (p= .285) indicating no significant influence of time over these variables.

Table III. Mean BDI and BAI scores for HDI, HDD, MSynI and MSynD groups

	BDI			BAI		
	Mean	(SD)	Range	Mean	(SD)	Range
HDI n=10	2.9	(4.9)	0-16	3.5	(4.5)	0-14
HDD n=10	4.0	(7.4)	0-24	3.4	(3.5)	0-13
MSynI n=10	3.9	(4.1)	0-11	6.0	(6.0)	0-17
MSynD n=9	1.4	(1.7)	0-5	7.3	(10.0)	0-32

Key: HDI - Huntington's Disease risk increased; HDD - Huntington's Disease risk decreased; MSynI - Marfan's Syndrome risk increased; MSynD- Marfan's Syndrome risk decreased

Self report data regarding influence of genetic risk advice on relationships with family and friends

As indicated above, open questions were used to derive self report data relating to the influence of receiving genetic risk advice on relationships with family and friends. Responses were categorised according to whether relationships had got better or worse or if there had been no change. Results are presented in tables IV and V. For statistical analysis the Relationships Better and the No Change categories were combined. Fisher's exact tests were then used to compare group frequencies. No between group differences were significant for either the Friends or Family variables.

Table IV. Self report data relating to influence of receipt of genetic risk advice on relationships with family members

	Relationships Better	Relationships Worse	No Change
HDI n=10	0	2	8
HDD n=10	1	2	7
MSynI n=10	2	1	7
MSynD n=9	0	0	9

Key: HDI - Huntington's Disease risk increased; HDD - Huntington's Disease risk decreased; MSynI - Marfan's Syndrome risk increased; MSynD- Marfan's Syndrome risk decreased

Table V. Self report data relating to influence of receipt of genetic risk advice on relationships with friends

	Relationships Better	Relationships Worse	No Change
HDI n=10	0	0	10
HDD n=10	1	0	9
MSynI n=10	0	1	9
MSynD n=9	0	0	9

Key: HDI - Huntington's Disease risk increased; HDD - Huntington's Disease risk decreased; MSynI - Marfan's Syndrome risk increased; MSynD- Marfan's Syndrome risk decreased

Results from G.P. Questionnaire

Table VI details the results obtained from the questionnaire sent to G.P.s, providing information regarding the number of times volunteers visited their G.P. surgeries in the six month periods before and after receiving degree of risk information. The number of occasions the G.P. considered to be due to stress or other psychological reasons is also given. Two volunteers requested that their G.P.s should not be contacted and four questionnaires were not returned. Analyses of variance for pre and post risk advice visits indicated no significant

between group differences. Paired t-tests were used to compare pre and post test G.P. visits for each of the four groups. Pre and post test comparisons were insignificant for all groups for the total number of visits and for the number of visits that were considered to be stress related.

Table VI. Results from G.P. Questionnaire

	Pre Risk Advice		Post Risk Advice	
	Total Visits	**Stress Related**	**Total Visits**	**Stress Related**
	Mean (SD)	Mean (SD)	Mean (SD)	Mean (SD)
HDI n=9	1.7 (1.73)	0.1 (0.33)	2.6 (2.19)	0.1 (0.33)
HDD n=10	3.5 (5.42)	0.5 (1.58)	4.6 (5.21)	0.9 (1.73)
MSynI n=9	2.9 (3.10)	0.1 (0.33)	3.4 (3.61)	0.1 (0.33)
MSynD n=5	1.2 (1.30)	0.0 (0.00)	2.8 (3.90)	1.0 (2.24)

Key: HDI - Huntington's Disease risk increased; HDD - Huntington's Disease risk decreased; MSynI - Marfan's Syndrome risk increased; MSynD- Marfan's Syndrome risk decreased

Discussion

The results from the SADS have demonstrated that a proportion of individuals in all four of the research groups experienced psychological distress of sufficient severity to reach diagnostic criteria for either Major Depression, Generalised Anxiety or Panic Disorder following a change in genetic risk status. Differences between the four samples were insignificant for all three diagnoses going against the hypothesis that symptoms of anxiety would be more prevalent in the MSynI group. BAI scores also suggested that symptoms of anxiety were not differentially greater in this group. The majority of volunteers in the MSynI group stated that they had received very good genetic counselling and information regarding their condition. They also took advantage of regular check ups to assess the strength of their aorta. It is possible that these factors helped to prevent an escalation of anxiety of sufficient severity to reach diagnostic criteria.

The lack of significant difference between those reaching diagnostic criteria following the receipt of increased or decreased risk status was also noted for both conditions. This is an important finding since a number of previous authors have suggested that those in decreased risk status groups would not require post risk advice counselling (e.g. Crauford and Harris, 1986). The current findings would suggest that such counselling should be offered to all patient groups regardless of outcome.

In absolute terms it is notable that Depression was most prevalent amongst the two HD groups; 50% of those in the HDI group experienced an episode of Major Depression following their change in risk status. In four out of the five cases in the HDI group these episodes of depression had resolved by the time of interview. The author proposes that a model similar to that in bereavement can account for this pattern, whereby a person experiences feelings of helplessness and loss in initial stages following their change in risk status. These feelings leave them prone to symptoms of depression. At later stages as the individual adjusts to their new status these feelings diminish and more realistic anxieties regarding the condition, either for oneself in the case of the HDI group or one's relatives in the HDD group, take over. The one case of Major Depression in the HDI group which had not resolved was that of a volunteer who had received a change in risk status twelve months before the interview. A bereavement style model would suggest that this individual had yet to reach the stage where depressive symptoms had diminished. All cases who had experienced an episode of Major Depression which had resolved had received their change in risk status between 24 and 60 months prior to the current study. The fact that such depression did resolve would also suggest that its presence in these individuals was due to external factors as opposed to being an early symptom of the condition. The lower prevalence of depression amongst the MSynI group would suggest that in most cases a bereavement process is not in operation for these individuals. The author suggests that this may be, in part, due to the nature of the condition. With HD there are no pre-symptomatic signs which give an indication of status group (Harper, 1991; Blackmore, Simpson and Crawford, 1995). By contrast with MSyn visible

characteristics such as having longer fingers than other family members may lead individuals to the premature conclusion that they are in the MSynI group. If this is the case then any bereavement type response to this conclusion may well have occurred and reached resolution prior to genetic assessment.

The data relating to relationships with family and friends proved to be somewhat encouraging. Only 5 of the 39 participants in the study felt that their relationships with their families had got worse since receiving genetic risk advice. The equivalent figure for relationships with friends was only one. The results are subjective, however, and it is suggested that further research is required to investigate the issue more fully.

The results from the G.P. questionnaire were also of interest and will be so to a range of parties including those responsible for healthcare purchase. The results indicated that providing genetic risk advice did not increase the number of times volunteers visited their G.P.s. Given the prevalence of psychiatric diagnoses this is perhaps a little surprising. It should, however, be noted that the G.P. questionnaire applied only to the six month periods before and after the receipt of risk advice. In the six month period after receiving risk advice post advice counselling would be at its most intense. It is possible that this provides a support mechanism which lessens the need to visit a G.P. It is suggested that all G.P.s are given information regarding the possible psychological consequences of receiving risk information since it is clearly necessary to ensure that individuals reaching

diagnostic criteria for a psychiatric illness after the receipt of risk advice do not go undetected.

The present study has provided information of interest. There is, however, still scope for considerable research in this area. The sample sizes in the present study are small and research with larger samples is required to further elucidate findings. There is also a need for a longitudinal study charting change in psychological status from the time of initial contact with genetic services to stages in the post test period. Unfortunately the time available for the present study did not allow for this. Findings should also be replicated with future patient cohorts. A number of studies (e.g. Codori, Hanson and Brandt, 1994) have suggested that those who present for genetic risk assessment represent a group of self selected individuals who are better able to cope with stress than are at-risk populations as a whole. It is not possible to assume that future patient cohorts will hold the same characteristics.

Ethical Approval

Ethical Approval for the study was granted by the Joint Ethical Committee of Grampian Health Board and the University of Aberdeen. All volunteers who took part in the study were asked to sign a consent form before doing so.

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Psychiatric input following attempted suicide in children and adolescents: implications for future resource management

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Psychiatric input following attempted suicide in children and adolescents: implications for future resource management

Abstract

This paper presents a descriptive analysis of patients referred to a child and family psychiatric service following attempted suicide. All patients referred by accident and emergency within a five year period are included. The age, sex, nature of the suicide attempt, previous contact with the service, psychiatric diagnosis, primary factor contributing to the suicide attempt and further use of psychiatric resources are recorded for each patient and the implications for the future management of this patient group are discussed.

Introduction

Although completed suicide amongst children and adolescents is, fortunately, rare it has been recognised for some time that the incidence of attempted suicide is less so (Otto, 1972). Birleson (1980), on the basis of a study carried out in Scotland, estimated that the incidence of attempted suicide amongst those in the 15 - 19 year age group is 700 per 100,000 of the total population for girls and 350 per 100,000 for boys. The management of these individuals places considerable

demand on limited psychiatric resources and consequently represents a huge problem for the health services.

It is a legal requirement that all individuals who have attempted suicide should be assessed, in order to establish their degree of risk for future self harm. Most of these assessments are currently carried out by psychiatric services. In developing management strategies for those who have attempted suicide there has, in the past, been little differentiation between services provided to adolescents and children, and services provided to adults. Initial management has tended to be the same with the proviso that, where possible, risk for future self harm in the case of the former group is assessed by a member of personnel with expertise in child psychiatry.

In order to provide an adequate assessment of risk the psychiatrist must travel. Most often this will be to the accident and emergency department where the individual's acute medical need has initially been met. They must then carry out a psychiatric interview lasting approximately one hour and, finally of course, deal with all the necessary paper work that such an assessment entails, for example providing a letter to the patient's G.P. summarising the nature of the suicide attempt, the predicted likelihood of future self harm and plans, if any, for future psychiatric input in the particular patient's case. All in all dealing with one case of attempted suicide can be a very time consuming process. In addition, high rates of failure to attend subsequent appointments when follow-up is offered have led to a further drain on psychiatric service time. This, coupled with the fact that many

psychiatrists subjectively feel that in a substantial proportion of these cases no psychiatric disturbance can be identified, has bought the whole management of this patient group in to question.

In order to plan management strategies in any patient group, the group has to be clearly defined and described in detail, to enable prediction of which particular services members of that group are likely to require. To date, the vast majority of studies investigating attempted suicide have been based on adult populations with relatively few studies investigating attempted suicide amongst children and adolescents. Those studies that have been carried out have largely been concerned with linking suicide attempts with particular difficulties e.g. family break-up (Garfinkel, Froese & Hood, 1982), stress at school (Gispert, Davis, Marsh & Wheeler, 1987) and relationship or other interpersonal difficulties (Pfeffer, Newcorn, Kaplan, Mizruchi & Plutchik, 1988). Whilst such studies have given us information of considerable import they have not given us an adequate description of the patient group from which to base future management decisions and policy. To date the most detailed description of suicidal behaviour in adolescents is that carried out by Juon, Nam and Ensminger (1994). They present a study describing the epidemiology of suicidal behaviour amongst school children in Korea. It is not possible to assume that the descriptive factors they present also hold for samples within the UK.

The present study was requested by a psychiatric service in Scotland and aims to provide a description of children and adolescents who attempted suicide over a

five year period. The study also considers the percentage up-take of further psychiatric input when offered.

Implications for future management of this patient group are discussed.

Procedure

Sample

The sample is comprised of all patients referred to the child psychiatry service at Hawkhead Child and Family Centre, following suspicion of attempted suicide, between 1990 and 1994, inclusive. In all cases referrals derived from physicians in the Accident and Emergency Department of the Royal Alexandria Hospital. It is their routine practise to refer children of school age to psychiatry at Hawkhead Child and Family Centre following suicide attempts.

Method

Cases of children who had been referred following suspected suicide attempts were initially identified from the weekly lists of psychiatric referrals. Retrospective data were then derived from the child's medical records which are held at Hawkhead Child and Family Centre. For each case the following information was recorded:

Age

Sex

Nature of the suicide attempt e.g. overdose, lacerations etc.

Previous contact with child psychiatry services

Number of appointments attended following referral

Number of failures to attend appointments offered

Number of suicide attempts per patient within the five year period 1990-1994

Psychiatric Diagnosis (as indicated by medical notes/ letters to G.P.)

Primary factors contributing to the suicide attempt (as indicated by medical notes/ letters to G.P.)

In assessing the primary factors contributing to the suicide attempt cases were classified in to seven groups - family, stress at school (e.g. exam pressure, bullying), relationships, other social factors, adjustment, substance abuse and other. The adjustment group consisted of those cases who had made a suicide attempt following a specific change such as death of a close friend or relative or

diagnosis of a medical condition. Placement in a group was determined by information derived from medical notes. This information was interpreted by the current author and cases allocated to a group accordingly. It was not possible to involve the psychiatry service in these decisions, firstly, because many of the cases were from five years prior to the present study being carried out and, secondly, because a number of the psychiatrists had left the service within this five year period.

Data was recorded on coding sheets with identifying features of each case (e.g. name, address etc.) excluded.

Results

Total number of cases classified as referral due to attempted suicide

156 (an average of 31 cases per annum)

Records for 9 of these were untraceable at the time the current study was carried out. The following results are, therefore, representative of the remaining 147 cases.

Sex

Males	34 (23.13%)
Females	113 (76.87%)

The ratio of males to females is in line with that of other studies which have suggested that although the rate of completed suicides is higher in males the rate of attempted suicides is higher in females. This is a stable finding regardless of age (McClure, 1984; Allen, 1987).

Age of Patients Referred

	<12	12	13	14	15	16	17	>17	Total
Male	1	-	5	14	8	5	1	-	34
Female	-	4	17	35	39	14	2	2	113
Total	1	4	22	49	47	19	3	2	147

As these results indicate, 65% of Children referred to child psychiatry services following attempted suicide fall within the 14-15 year age group. Comparatively low figures for those above the age of 16 are accounted for by some patients

within this range being appropriately referred to the adult service within the locality. Again, these findings are in line with other studies which have suggested that suicide attempts are rare prior to adolescence (Evans, 1982).

Nature of Suicide Attempt

Drug Overdose	138
Lacerations	3
Ingestion of mushrooms	1
No information	5

As indicated, by far the most common method of attempted suicide by this patient sample was by drug overdose.

Previous contact with child psychiatric service

Yes 24 (16.3%)

No 123 (83.7%)

This would suggest that in the majority of cases difficulties prior to the suicide attempt had either not been identified or were not considered to be of sufficient severity to warrant input from child psychiatry services.

Number of referrals per patient following attempted suicide (1990-1994)

1	128 (87.1%)
2	15 (10.2%)
3	3 (2.0%)
4	1 (0.7%)

As these results show approximately 13% of the total sample made more than one suicide attempt within the specified 5 year period. Obviously, it has to be noted that this can not give a true indication of repetition rates as those who made attempts towards the end of the five year period had a decreased time in which to repeat. Follow-up studies will be required if true repetition rates over a five year period are to be assessed.

The chance of repetition is quite clearly a serious concern following any case of attempted suicide. A number of studies have concluded that approximately 1% of those who attempt suicide die by suicide within one year of the attempt (e.g. Hawton & Fagg, 1988; Buglass & Horton, 1974). Hawton and Catalan (1987) have suggested that with the adult population 40-50% of those referred following suicide attempts will have made previous attempts and that 12-16% will have a history of 5 or more previous suicidal acts. The equivalent figures for the child and adolescent population would be expected to be less since, by virtue of their age, children have had less time in which to make attempts.

Number of appointments with psychiatric services following referral

Average number of appointments 1.95 (range 0 - 10)

0	3 (2.1%)
1	72 (49.0%)
2	38 (25.9%)
3	16 (10.9%)
4	11 (7.5%)
5	5 (3.4%)
>5	2 (1.4%)

As shown just over half of all cases were seen on 1 occasion or not at all. 57 cases (38.8%) were discharged after the initial appointment in the Accident and Emergency Department. Of these, 3 were transferred to other services (e.g. adult services within the locality). In 36.7% of cases no further psychiatric input was thought to be required.

Of those offered follow-up appointments at Hawkhead Child and Family Centre 18 out of 87 (20.7%) failed to attend. As highlighted above, such failures to attend represent an inefficient use of limited psychiatric resources.

Psychiatric Diagnosis

In 5 cases a full psychiatric assessment was not possible - reasons given for this in notes included drowsiness, failure to co-operate and in one case a child's youth and learning disability. Of the remaining 142 cases letters to G.P.s suggested the following diagnoses:

Depression	6 cases
Anxiety	2 cases
Schizoid Personality	2 cases
Hysterical Dissociative State	1 case

Of the remaining 131 cases the majority of letters to G.P.s and medical notes stated that 'there was no evidence of depression or other psychiatric disturbance'. In these cases difficulties tended to be referred to in more general terms referring, for example, to difficult social circumstances, relationship problems, family difficulties or history of substance abuse. The results contrast with those in the Korean sample (Juon et al., 1994). They concluded that depression is the strongest predictor of suicidal behaviour in adolescents. The above results can not support this.

Primary factor contributing to suicide attempt

Family Difficulties	53 (36.1%)
School	27 (18.4%)
Substance Abuse	24 (16.3%)
Relationships	9 (6.1%)
Other social reasons	12 (8.2%)
Adjustment to new circumstance	9 (6.1%)
Other	8 (5.4%)
No information	5 (3.4%)

As indicated in the introduction, placement in each of these groups was determined by the author who allocated cases to a group according to the most prominent area mentioned in case notes/ letters to G.P.s. The results above can not, therefore, be considered truly objective. It is, nevertheless, quite clear that difficulties in the family and in school, and substance abuse contributed to a substantial proportion of the child/ adolescent suicide attempts included in the current study.

Looking at case notes in more detail, 23.1% of letters to G.P.s specifically stated that the primary aim of the act was not suicidal in nature. In other words, approximately one fifth of patients did not wish to die when the suicidal act was carried out. Presumably, the aim for the majority of these patients was to bring

about some kind of change in their present circumstances. It could be argued that such patients should not be included in the current study since they do not represent cases of true suicide attempt. They do, however, initially have to be treated and managed in the same manner as those who do present with suicidal ideation. It is, therefore, appropriate that they too should be included in the current study and, furthermore, included in future management plans since even in the absence of suicidal intent, any action with mimics an attempted suicide must be treated as a serious symptom and managed accordingly.

Discussion

The above results have provided a detailed description of patients who have been referred to the child psychiatric service at Hawkhead Child and Family Centre following a suicidal act. Many of the findings regarding the profile of this patient group, including age, sex and primary factors contributing to the suicide attempt are, as mentioned above, in accordance with the findings of other studies.

It is not the authors remit to be involved with management or policy making decisions. It is, however, the opinion of the author that some of the findings in this study will be of interest to those who are involved with future planning regarding the management of this patient group. Findings are discussed, from this viewpoint, below.

Perhaps, one of the most striking findings in the current study which will be of interest to those involved in policy making is the apparent lack of psychiatric disturbance within the patient group. Letters to G.P.s indicated psychiatric diagnoses in only 7.4% of cases. Of course, it could be argued that psychiatrists may be wary of giving a diagnosis in a particular case because they do not wish the individual to be 'labelled'. There is also the fact that retrospective studies of this nature are necessarily limited by the information that is documented; perhaps the psychiatrist did have a diagnosis in mind in a particular individual's case but, for whatever reason, it was excluded from the case notes. Even if one takes such arguments in to consideration, however, the current study has clearly demonstrated that in 36.7% of cases psychiatric input is confined to the initial assessment with further input thought to be unnecessary. This in itself suggests that a substantial proportion of this group of patients are not presenting with psychiatric disorder. Psychiatric assessment of such a high percentage of patients who do not indicate features of psychiatric disturbance may be considered an inefficient use of service time. Inefficiency is further increased by 20.7% of those patients who are offered subsequent appointments failing to attend.

From the policy making point of view these findings would suggest two areas worthy of further investigation; firstly, a method of screening patients prior to referral to a psychiatric service may be considered. Previous studies have indicated that other personnel, such as Junior Doctors, Nurses and Social Workers are equally as effective at assessing risk in those who have made suicide attempts (Owens, 1990; Newson-Smith & Hirsch, 1979). To implicate such a

policy would clearly involve investing money in further research and in the training of staff who are to be involved in this type of decision. The second area which needs to be taken in to consideration is in improving the attendance rates of those for whom further psychiatric input is thought to be of benefit. Amongst the adult population, Greer and Bagley (1971) and Motto (1976) suggest that prognosis is poorer for those who lapse from contact. Further research investigating the reasons why people fail to attend follow-up appointments is clearly indicated if one's aim is to improve attendance rates and subsequently reduce future risk.

Those involved in decisions regarding the management of this patient group clearly have a role to play in improving efficiency of the service provided to individual patients, from the moment of their reception in to an accident and emergency or paramedical service. The findings of the current study may, however, also be of interest from a public health stance. Juon et al. (1994) have suggested that preventative strategies may be the way forward. They have recommended in-service training in schools and workshops aimed at educating parents in the identification of signals indicating risk for suicidal acts. A study comparing populations with and without such programmes would be of great interest in Britain. Investigating the efficacy of preventative strategies provides clear opportunities for much needed research. Working at a preventative level aimed at the identification and diffusion of difficulties prior to an individual reaching the crisis of a suicidal act would also result in reduced distress, not only for the individuals concerned but for their families and for the professionals who

are involved in their treatment. The use of preventative strategies would also be in line with the British government's aim of reducing the number of suicides per annum by the year 2000.

Summary

In summary, the present study has provided a detailed description of children and adolescents treated in an accident and emergency department and subsequently referred to a psychiatric service. The results suggest that current management of this group of patients is not efficient in that a substantial amount of psychiatric service time is being allocated to individuals who do not indicate psychiatric disturbance. Service time is further reduced by 20.7% of those patients who are offered follow-up appointments failing to attend.

It is suggested that there is a need for further research and training to investigate the efficacy of using other personnel to screen patients prior to a referral to psychiatric services. In addition, it is recommended that the study of preventative strategies should be considered an area of priority for future research.

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Cognitive deficits in myotonic dystrophy and difficulties associated with differential diagnoses - a single case illustration

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Cognitive deficits in myotonic dystrophy and difficulties associated with differential diagnoses - a single case illustration

Abstract

This paper presents a neuropsychological assessment of a 54 year old lady with myotonic dystrophy and a history of alcohol abuse. Results indicate impairment of general intellectual functioning, mnestic, executive and visuospatial skills. The report illustrates the process of testing hypotheses in neuropsychological settings and demonstrates the difficulties associated with differentiating between the effects of alcohol abuse and the effects of hypoxia relating to myotonic dystrophy. Further research in the area is recommended.

Key Words: Myotonic, dystrophy, alcohol, memory, diagnosis, intelligence

Introduction

Myotonic dystrophy is an inherited disorder which results in widespread systemic effects including production of myopathy, cataract formation, endocrine disorders, cardiomyopathy and impaired pulmonary ventilation. Most cases are diagnosed between the ages of twenty and thirty years when muscle weakness is

commonly the first recognisable sign. The course of the disorder lasts around fifteen to twenty years with death usually being due to respiratory infection or cardiac failure (Harper, 1989).

In recent years a number of studies have demonstrated a link between myotonic dystrophy and cognitive deficits. This would be expected given the possibility of hypoxic brain damage resulting from secondary cardiac involvement and pulmonary dysfunction. Swash and Oxbury (1991) link myotonic dystrophy with mental apathy, personality change and depression. Van Spaendonck, Ter Bruggen, Weyn Banningh, Van de Biezenbos & Gabreels (1995) have demonstrated that patients with myotonic dystrophy are significantly impaired on Stroop Colour Word Test performance when compared to matched controls. Colombo, Perini, Miotti & Armani (1992) have noted significantly impaired Wechsler Adult Intelligence Scale - Revised performance in patients in advance stages of the condition.

Studies that have investigated cognitive deficits in patients with myotonic dystrophy have, to date, considered only limited areas of functioning. Those deficits that have been identified are not specific to myotonic dystrophy. They are also characteristic of a range of other disorders and syndromes. Executive and general intellectual impairments which appear to be characteristic of some patients with myotonic dystrophy, for example, are also characteristic of some patients who have a history of long term alcohol abuse (Parsons & Farr, 1981) and some patients with traumatic brain injury (Walsh, 1994). On a practical level

this makes it difficult to discount differential diagnoses when attempting to account for any cognitive deficits observed in myotonic dystrophy patients. To illustrate this difficulty a case is presented of a 54 year old lady with myotonic dystrophy and a history of alcohol abuse. The case demonstrates how various hypotheses were considered prior to reaching a conclusion regarding the most likely origin of her deficits.

Case report

History

MCh was a 54 year old woman who had myotonic dystrophy and a history of alcohol abuse. She left school at the age of 15 and had no formal qualifications. She had been previously employed in a variety of clerical jobs but had been unemployed for 4 years prior to assessment. MCh was diagnosed with muscular dystrophy at the age of thirty. She was referred for assessment after an incident two months previously when she had collapsed on her stairs and had been taken to her local general hospital. No cause for her collapse had been identified. Whilst she was in hospital, however, staff suspected that her memory was impaired and requested neuropsychological assessment to investigate this.

Aims of Assessment

Prior to assessment it was clear that there would be a necessity to determine whether any deficits in cognition observed were due to the primary effects of myotonic dystrophy, the secondary effects of organ failure and consequent hypoxia due to progression of the disease, the influence of previous alcohol abuse or the effect of collapsing on the stairs. It was also possible that deficits could be an indication of pseudo-dementia resulting from depression. Assessment aimed to differentiate between these alternatives. The following information was, therefore, required:

1. An estimation of MCh's current stage of myotonic dystrophy
2. An accurate account of MCh's alcohol history and dietary intake
3. An assessment of depression
4. Further information regarding her recent fall on the stairs
5. A comprehensive assessment of her current cognitive functioning and if possible an indication of the time of onset of any deficits observed

To obtain this information MCh was seen for clinical and psychometric assessment on two occasions. In addition, her sister was interviewed. Her sister visited her daily and was, therefore, able to provide corroborative information regarding MCh's alcohol and dietary history. Her sister was also able to provide an estimation of the onset of her cognitive deficits.

Psychometric Assessment Measures

Psychometric assessment aimed at providing a comprehensive overview of general intellectual and mnestic abilities. The psychometric assessments detailed below were administered:

The Wechsler Adult Intelligence Scale - Revised (WAIS-R: Wechsler, 1981; Lea, 1986)

The Wechsler Memory Scale - Revised (WMS-R: Wechsler, 1987)

The National Adult Reading Test: Second Edition (NART-R: Nelson, 1992)

The Modified Card Sorting Test (Nelson, 1976)

Controlled Oral Word Association Test (Benton, 1968)

Cognitive Estimates Test (Shallice & Evans, 1978)

The WAIS-R and WMS-R were selected, in the first instance, to provide an overview of intellectual and mnestic skills respectively. Both measures have extremely well established and adequate reliabilities and validities and both cover a broad range of functioning (Lezak, 1995). The NART-R is a measure of present ability which is largely resistant to neurological impairment (Nelson, 1992; Crawford, Besson & Parker, 1988). This was used to give an estimate of premorbid intellectual functioning.

MCh's performance on the WAIS-R suggested the possibility of frontal lobe dysfunction. This was indicated by poor performance on the Picture Arrangement

subtest which provides a measure of sequential thinking, largely dependent on frontal lobe integrity. Impaired performance on the Block Design subtest also suggested this possibility. For this reason it was decided that further measures of frontal lobe functioning should be included in the assessment. The Modified Card Sorting Test, the Controlled Oral Word Association Test and the Cognitive Estimates Test were selected because of their known sensitivity to frontal lobe impairment (Lezak, 1995).

Results of assessment and additional information obtained

Estimation of current stage of myotonic dystrophy

MCh's muscle weakness at the time of assessment was clearly severe. She was confined to her flat which was up three flights of stairs. She was also experiencing hair loss and breathlessness. These factors indicated that she was in advanced stages of the disorder, with cardiac and pulmonary involvement.

Clinical impressions

MCh's behaviour during assessment was co-operative and relaxed throughout both test sessions. She was fully orientated in person, time and place. For the

majority of the testing procedure her attention/ concentration appeared to be good. She showed insight in to the difficulties she had with the psychometric tests. In addition, her general conversation skills were good and she showed no signs of confabulation.

Overall, MCh came across as being a pleasant lady who was contented with her life. There was no evidence of anxiety or depression.

Her performance indicated that she was left handed.

Alcohol History

MCh stated that for the three years prior to her recent collapse she had consumed an average of one bottle of whisky every three days. Since her collapse she had not consumed any further alcohol. At the time of assessment she had not consumed alcohol for two months. This information was confirmed by her sister and was considered accurate. Her sister also confirmed that MCh had a good diet. Meals were provided for her and examples of menus indicated that vitamin deficiency, commonly seen in conjunction with alcohol abuse, was unlikely in her case.

MCh's length of abstinence from alcohol of two months is important to note since in a person of her age you would expect most improvements in cognitive

functioning resulting from detoxification to have already occurred within this time period (Ryan & Butters, 1986). It is reasonable to assume that at the time of assessment MCh's level of cognitive functioning was stable with respect to her previous alcohol intake.

Assessment of Depression

Depression was assessed via clinical judgement and use of the Hospital Anxiety and Depression Scale (HAD: Zigmond & Snaith, 1983). The HAD was selected as a suitable measure for depressive symptoms in MCh's case since it's results are thought to be unaffected by the presence of bodily illness. As mentioned above, MCh did not appear to be clinically depressed. This was confirmed by her response to questions on the HAD. She scored 3 for both anxiety and depression. These scores do not meet the required level of 'caseness' for either measure.

Information regarding MCh's recent collapse

Medical notes indicated that no cause for her recent collapse could be identified. Her collapse was, therefore, contributed to muscular weakness. There was no suggestion that she had hit her head in the course of falling.

Psychometric Test Results

National Adult Reading Test - Revised (NART-R)

NART Error Score 29

Estimated premorbid full scale WAIS-R IQ	95 (score obtained = 79)
Estimated premorbid verbal WAIS-R IQ	94 (score obtained = 83)
Estimated premorbid performance WAIS-R IQ	96 (score obtained = 76)

MCh's performance indicates that her estimated level of premorbid intellectual functioning is within the average range. The discrepancies between predicted and obtained IQ scores are indicative of moderate intellectual deterioration.

The Wechsler Adult Intelligence Scale - Revised (WAIS-R)

Full Scale IQ	79
Verbal IQ	83
Performance IQ	76

Age graded subtest scores are given in Table 1. In each case the normal mean and standard deviations for a person in MCh's age group are 10 and 3 respectively.

Table 1. Age graded WAIS-R subtest scores

Verbal Scale	Score	Performance Scale	Score
Information	10	Picture Completion	7
Digit Span	8	Picture Arrangement	3
Vocabulary	7	Block Design	5
Arithmetic	6	Object Assembly	8
Comprehension	3	Digit Symbol	4
Similarities	6		

As Table 1 indicates, MCh's performance on the Wechsler Adult Intelligence Scale - Revised was below average on 8 of the 11 subtests. There is no significant discrepancy between verbal and performance IQ.

The individual subtest scores, which are markedly low for Block Design, Picture Arrangement, Comprehension and Digit Symbol, suggest impairment of visuospatial organization, poor sequential thinking, impaired verbal reasoning and deficits in psychomotor performance. Impaired Picture Arrangement and Block Design Subtests are also suggestive of frontal lobe dysfunction. Her performance on the Picture Arrangement, Block Design and Digit Symbol subtests which are timed tasks may have been compromised by muscle weakness. Of note, however, is the fact that her performance on the Object Assembly

subtest was within normal limits. If performance had been influenced by muscle weakness then you would also expect impaired performance on this task.

The Wechsler Memory Scale - Revised (WMS-R)

The following memory quotients were obtained. They are essentially comparable with IQ, having a mean of 100 and a standard deviation of 15.

Verbal Memory	76
Visual Memory	77
General Memory	70
Attention/Concentration	74
Delayed Recall	62

These measures suggest significant impairment of both short and long term memory, with performance one or two standard deviations below the mean in each case. They represent a discrepancy based on estimated premorbid intellectual level, but are comparable to current intellectual performance.

Individual subtest scores are given in table 2.

Table 2. WMS-R Subtest Scores

Subtest	Normal Mean and S.D. For MCh's age group	Score
Mental Control	4.9 (1.2)	2 (below average)
Figural Memory	6.2 (1.4)	7 (average)
Logical Memory 1	22.5 (6.3)	14 (below average)
Visual Paired Associates 1	11.2 (4.3)	11 (average)
Verbal Paired Associates 1	18.0 (3.2)	13 (below average)
Visual Reproduction 1	29.0 (5.2)	19 (below average)
Digit Span	14.9 (3.3)	10 (below average)
Visual Memory Span	15.0 (3.0)	13 (average)
Logical Memory 2	18.1 (6.0)	9 (below average)
Visual Paired Associates 2	4.6 (1.8)	1 (below average)
Verbal Paired Associates 2	6.9 (1.2)	4 (below average)
Visual Reproduction 2	25.4 (7.2)	8 (below average)

As table 2 indicates MCh's performance on the Wechsler Memory Scale - Revised was below average on 9 of the 12 subtest measurements. It is notable that MCh's ability to recall verbal information from short term memory is impaired whilst her ability to recall non-verbal information from short term memory is within the normal range. This is indicated by the below average scores for the Logical Memory 1, Verbal Paired Associates 1 and Digit Span subtests together with

average scores for the Figural Memory, Visual Paired Associates and Visual Memory Span subtests. This dissociation does not hold for long term memory where performance indicates that memory for both verbal and non verbal material is compromised.

Assessment of Frontal Lobe Dysfunction

a. The Modified Card Sorting Test

		Normal Mean (S.D.)
Categories Completed	2	5.0 (1.6)
Total Errors	17	9.2 (8.5)
Perseverative Errors	3	5.2 (3.9)

b. Controlled Oral Word Association Test

Letter	No. words generated within 1 minute
F	4
A	4
S	4
Total	12

MCh's performance on this task is in the percentile range of 1-3, indicating severe impairment.

c. Cognitive Estimates

Patient's Score = 9*

Normal mean and standard deviation = 3.6 (1.92)

* Higher scores are indicative of poorer performance.

MCh's performance on the above three tests is indicative of impairment which is usually associated with frontal lobe dysfunction. This is characterised by impaired ability to shift cognitive sets, deficits in planning and sequential thinking, decreased verbal fluency and impaired ability to make cognitive estimates.

Additional information regarding the onset of cognitive deficits - as estimated by MCh and her sister

MCh's sister estimated that MCh first began to experience difficulties with cognitive functioning four years prior to her current assessment. This coincided with the time she became unemployed. MCh also felt that she had begun to experience difficulties with memory at around this time.

Discussion

Results from the psychometric assessments detailed above are indicative of significant cognitive impairment and intellectual deterioration from a previous level of functioning. Impaired mnestic ability, general intelligence, visuospatial skills and frontal lobe functioning are clearly demonstrated and there is ample evidence to suggest that these deficits are organic in origin. The organic mechanism underlying the deficits observed, however, is less clear. As mentioned above, the possible hypotheses regarding the cause of deficits observed were the effects of long term alcohol abuse, the primary or secondary effects of myotonic dystrophy, pseudo-dementia resulting from depression and the effect of MCh's collapse on the stairs. Information obtained suggested that the latter two factors could be discounted. There was no evidence that MCh was depressed. Pseudo-dementia resulting from low mood was, therefore, unlikely. Information regarding her fall on the stairs suggested that the most likely cause of the fall was muscular weakness. She did not hit her head. In addition, both MCh and her sister felt that the onset of her difficulties with cognitive functioning pre-dated the collapse. It was, therefore, apparent that the collapse could not account for the difficulties observed.

In order to assess the relative influence of the effects of alcohol abuse on MCh's cognitive functioning it is necessary to consider the relevant literature. Patients who have cognitive deficits associated with alcohol abuse largely fall in to two separate but closely related groups - these are patients with Korsakoff's syndrome

and patients with alcoholic dementia. Korsakoff's syndrome is an organic brain disease associated with long term alcohol abuse together with thiamine deficiency. Cognitive difficulties typically include a severe anterograde amnesia, a retrograde amnesia which covers most of the individual's adult life, visuospatial deficits, deficits in executive functioning, poor insight in to difficulties and, frequently, confabulation (Butters & Cermak, 1980). Typically, such difficulties present in the context of a normal IQ and onset is fairly sudden. Such a picture does not fit the profile of deficits observed in MCh. Although she does present with difficulty learning new information and impairment of executive and visuospatial functioning she also demonstrates good insight and no evidence of confabulation. She also has impaired general intellectual ability. In addition, as mentioned above, evidence from her sister suggested that her diet was adequate. It is, therefore, unlikely that her deficits could be attributed to a vitamin deficiency. Deficits observed in patients with alcoholic dementia tend to be more global and are associated with a gradual decline (Lezak, 1995). As with Korsakoff's syndrome, there is impairment in executive, visuospatial and mnestic skills. In alcoholic dementia there is also an impairment of general intellectual functioning (Alerdice, McGuinness & Brown, 1994). This would appear to fit more closely with the picture observed in MCh. If one looks at the literature in detail, however, there appear to be subtle differences between the profile of deficits observed in MCh and the deficits typically observed in alcoholic dementia patients. Butters & Granholm (1987), for example, found that patients with alcoholic dementia commonly show a discrepancy between verbal and performance IQ as measured by the WAIS-R, with significantly greater

impairment shown on the performance scale. MCh did not show this despite the fact that one might have expected such a discrepancy simply by virtue of her muscular weakness which could have impaired her ability to carry out performance scale tasks. Similarly, MCh's performance on mnestic tasks is not consistent with that shown in the alcoholic dementia literature. Ryan & Butters (1986), for example, have shown that patients with alcoholic dementia perform poorly on tasks requiring short term memory for visual material whilst their memory for verbal material is often intact. With MCh this dissociation is reversed. Whilst it is not possible to discount the influence of alcohol abuse entirely the pattern of deficits MCh shows is not typical of patients who have histories of alcohol abuse. This would suggest that a factor unrelated to alcohol is contributing to her difficulties. Additional support of this argument, of course, would be the fact that both MCh and her sister estimated that the onset of her cognitive difficulties preceded the onset of her alcohol abuse.

As noted earlier, a further hypothesis regarding the origin of observed deficits would be that they are related to the myotonic dystrophy itself. MCh's presentation demonstrated that she was in advanced stages of the condition with cardiac and pulmonary involvement. Since this is the case hypoxic damage is likely. In general hypoxic injury tends to be most profound in the hippocampus (Lezak, 1995). Diffuse injury elsewhere, however, can lead to disruption of a broad range of cognitive functions, including impairment in general intellectual ability, impaired mnestic ability and compromised executive and visuospatial functioning (Prigatano, Parsons, Wright, Levin & Hawryluk, 1988). Patients with

hypoxic damage tend to retain good insight in to the nature of their deficits and typically they will show no signs of confabulation on memory tasks. These latter points are, in general, features of patients with temporal lobe damage. The profile fits that seen in MCh and would support the hypothesis that hypoxia due to secondary organ failure in myotonic dystrophy is the most likely cause of deficits observed. One might speculate that in her case the hypoxic damage to her left temporal lobe is greater than that shown in her right. This would account for the discrepancy between her short term memory for verbal and non-verbal material. The other possibility, of course, is that her difficulties are associated with the primary effect of muscular dystrophy. This seems unlikely since MCh was diagnosed as having myotonic dystrophy at the age of thirty whist cognitive deficits became apparent approximately 20 years later, perhaps coinciding with the decline in organ functioning and the beginnings of more pronounced hypoxic damage.

In conclusion, assessment of MCh has demonstrated deficits in general intellectual ability, mnestic and executive skills and visuospatial functioning. Her case has shown how various hypotheses as to the likely cause of her deficits were used to guide the course of her assessment and the collation of information required. Analysis of the information obtained at assessment led to the conclusion that hypoxia due to the secondary effects of myotonic dystrophy is the most likely cause of deficits observed. The conclusion, however, is tentative and the effects of the alcohol abuse can not be ruled out. The case has illustrated how difficult it is to differentiate between the effects of alcohol and the effects of

hypoxia relating to myotonic dystrophy. Part of this difficulty is related to the fact that as yet no study has adequately assessed the profile of cognitive deficits in myotonic dystrophy patients. Further research of this nature would clearly improve our understanding of the neuropsychological mechanisms of the disease as well as our ability to assess deficits in the single case.

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Managing self harm during the treatment of survivors of childhood sexual abuse - a single case illustration

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Managing self harm during the treatment of survivors of childhood sexual abuse - a single case illustration

Summary

This paper presents the case of a 27 year old woman who sought psychological treatment because she was experiencing difficulties coping with memories of sexual abuse. She presented with a major depressive illness, suicidal ideation and a history of self harm. The case illustrates how self injury was managed in her case and how it became a focus of the early stages of therapy.

Introduction

The link between self-injury and childhood sexual abuse is now well established (e.g. Romans, Martin, Anderson, Herbison and Mullen, 1995; Boudewyn and Liem, 1995) and a number of authors have presented theories which may be used to explain why this should be the case. Walsh and Rosen (1988), for example, have suggested that self-injury brings a relief from tension resulting from painful thoughts and feelings. Wise (1989) suggests that self injury represents a way of coping with betrayal. Favassa (1988) suggests that it is a way of venting anger and of establishing control. The underlying assumption with all these view-points is that self injury serves some kind of purpose. It helps the individual to cope

with painful feelings and emotions that they find hard to tolerate. As Favassa (1989) concludes, self injury is 'a purposeful, if morbid, act of self help'.

In the early stages of treating survivors of childhood sexual abuse levels of distress can increase (Hall and Lloyd, 1989). This may occur as the individual begins to acknowledge the reality of the abuse they have experienced and the pain which it has caused them. Self injury may serve as a way of coping with the resulting intense emotions. To optimise patient safety it is of paramount importance that such self injurious behaviours are well managed.

This paper presents the case of a 27 year old woman who sought psychological treatment to help her deal with distressing feelings associated with childhood sexual abuse. For reasons discussed below she was considered to be at high risk for self injury. The case illustrates how self injury was managed and how it became a focus of treatment in the early stages of therapy. The report considers the initial assessment and the first ten therapeutic sessions.

Case Report

JL is a twenty seven year old woman who was referred for psychological treatment after an event at a party had triggered feelings associated with childhood sexual abuse. This event involved a friend touching her in a way that made her feel uncomfortable. The onset of her abuse as a child was unclear. JL

believed, however, that it started when she was six years old. The abuse took the form of her father touching her breasts and placing his fingers inside her vagina. He would also ask her to stimulate his genitals. The abuse stopped when JL was fourteen.

JL had a long history of depressive illness and had experienced episodes of Major Depression since the age of 15. She also had a history of self harm and had taken two overdoses seven years prior to the current referral. At the time of initial assessment she was taking lophepramine as prescribed by her G.P. She had had no previous treatment from either psychological or psychiatric services. There was no medical history of note regarding physical illness. At the time of treatment she lived with her partner whom she had been with for the previous six years.

Presenting Problem

At interview JL came across as being a pleasant and articulate woman who was keen to seek help for her difficulties. She was tearful and it was clear that she was clinically depressed. She obtained a Beck Depression Inventory (BDI: Beck, Ward, Mendelson, Mock and Erbaugh, 1961) score of 35 which reflected a severe level of depressive symptoms. Consistent with this she reported that she had lost enjoyment in activities; her sleep was disturbed with sleep onset insomnia and early wakening; her appetite was impaired and she had low libido.

She also had very low self esteem. She reported that she was finding it difficult coping with her feelings regarding her childhood experiences of abuse. She also mentioned that she felt uncomfortable with most men and that, in general, she found it difficult to trust people. She expressed suicidal ideation but did not feel that she would carry this out. She also reported a considerable degree of self destructive behaviour. She would physically harm herself by beating her stomach, arms and legs. In the past she had gone out for walks with the intention of finding someone who would stab her. She had also placed herself in dangerous situations such as in the local park at night. Incidents of self harm would happen approximately twice a week. She could not initially identify what triggered them. She thought, however, that they may be related to feelings of anger. She had noticed that she sometimes punched herself in order to 'let feelings of anger out'. At the time of initial presentation she had been absent from work for three months. This was due to depression.

Formulation

At interview JL's presentation was consistent with a DSM-IV diagnosis of Major Depression (American Psychiatric Association, 1994). In formulating her case it was clear that her experiences of having been abused as a child had had a considerable effect on how she perceived and experienced her world as an adult. She was having difficulty coming to terms with conflicting feelings. She was confused by the fact that her father who gave her the attention that she liked as a

child was also the person who abused her sexually. She felt betrayed by him and this feeling of betrayal had subsequently influenced her relationships with other people. She was afraid to trust other people in case they also let her down. She also felt very guilty about the abuse. She felt different from other people and she was very isolated. These factors made her prone to depression. In addition, she was harming herself. It was believed that this was a way of coping with the distress that she felt. These acts of self harm, however, were thought to further lower her self esteem. It was thought that this led to increased depression and in turn to further self harm.

Treatment Programme

In devising a treatment programme for JL it was clear that an essential element of treatment needed to address the issue of her personal safety. She was engaging in self injurious behaviours and had a history of suicide attempts. In addition to this, JL's self injurious behaviours served the purpose of helping her cope with intolerable feelings. There was a danger that such feelings would escalate in the early stages of therapy and that this would increase the incidence of self injurious behaviour and suicidal thoughts, further compromising her safety. For these reasons, from the first therapy session following the initial assessment interview time was spent focusing on the self injurious behaviour itself. Taking the same stance as Wise (1989) and Burstow (1992) therapy started with the assumption that self injurious behaviour serves some purpose. Therapeutic intervention

aimed at discovering the nature of this purpose and seeking alternative behaviours that would serve the same purpose whilst causing less harm. It was hoped that if self injurious behaviours could be reduced then this would help raise JL's self esteem and, therefore, also assist in raising her mood. In the first therapy session time was also spent negotiating a plan of action with JL for times she felt that suicidal thoughts were overwhelming and that her life was in danger. Arrangement was also made for psychiatric back-up should this be required.

Once JL's personal safety was more established it was possible to begin work on her feelings associated with the sexual abuse she had experienced as a child. The approach that was taken with regards to this largely followed elements of the treatment programmes for survivors of childhood sexual abuse as advocated and found to be effective by Hall and Lloyd (1989) and Draucker (1994). These authors start from the viewpoint that survivors of childhood sexual abuse who experience difficulties in adulthood do so because of unresolved feelings which stem from the abuse they have experienced. These feelings alter the way they think about themselves and the world, causing them further distress. In therapy the survivor is encouraged to re-experience feelings in a safe therapeutic setting. In doing so they learn to tolerate these feelings. Once they can do so satisfactorily the feelings become less powerful and intrusive and less influential in the survivor's adult life. The survivor is also helped to reinterpret the abuse experience from an adult point of view and to correct any distorted beliefs about it.

Overall, treatment aimed to reduce JL's self injurious behaviours and to alleviate her depressive symptoms. It was also hoped that by the end of treatment she would be able to cope with feelings stemming from her past experiences without feeling overwhelmed and without feeling the need to resort to self injury in order to cope with them.

Measures of Assessment

To provide a measure of JL's level of depressive symptomatology the BDI was administered at the initial assessment interview and again in the tenth therapeutic session. In addition she was asked to record times when she harmed herself or had thoughts relating to a desire to do so. This provided a measure of the incidence of self injurious behaviours and associated thoughts.

Treatment Sessions

JL attended for 10 therapy sessions arranged on a weekly basis. Sessions 1 and 2 focused solely on her self injurious thoughts and behaviours. From session 3 onwards her feelings associated with having been sexually abused also became a target of treatment.

An outline of treatment sessions is given below.

Assessment Interview

Clinical assessment and history taken.

Therapeutic goals set in collaboration with JL aimed to reduce depression and self injurious behaviours and to enable JL to cope more adaptively with the painful thoughts and feelings associated with her past experiences of abuse. It was agreed that she should attend for ten therapy sessions in the first instance.

Therapy session 1

Sessions 1 and 2 focused on JL's self injurious behaviours. In session 1 it was emphasised that these behaviours served a purpose and that the role of therapy was to discover the nature of this purpose and find an alternative less harmful behaviour that would serve the same purpose. JL was asked to keep a diary detailing all incidents of self injurious thoughts or behaviours. For each incident she was asked to note the preceding situation, the thoughts and feelings she was experiencing, the nature of her behaviour and the consequences of it. In discussion, JL suggested that if she did not injure herself to relieve her feelings then the feelings would escalate and lead to her taking an overdose. She agreed that should this occur she would take herself to casualty or telephone for an ambulance.

Therapy session 2

Session 2 reviewed JL's self injurious behaviour diary from the previous week. She had recorded two incidents of self injury. In response to feelings of anger towards her father and towards her partner she had hit herself. She suggested that in preference to this she should try to hit her teddy bear. It was agreed that she should assess the efficacy of using this approach in the following week if self injurious thoughts relating to anger occurred. There was no evidence of suicidal ideation at this session.

Therapy session 3

A review of JL's diary from the previous week demonstrated that the strategy of hitting her teddy bear had not been effective in alleviating anger. An alternative approach of JL writing down her feelings was suggested and she was asked to try this the following week if the opportunity arose. No suicidal ideation was evident.

The second part of session 3 began to focus on the abuse experience. JL was asked to describe her memories of abuse up until the age of eleven. Therapy aimed at experiencing and naming associated feelings. It was clear in her description that JL was blaming herself for the abuse and was consequently harbouring associated guilt.

Therapy Session 4

JL's diary indicated that the strategy of writing down her feelings associated with anger had been successful in alleviating the desire to harm herself. Two further incidents, however, suggested that self injury also served a purpose of relieving feelings of disappointment. JL had been going for a drive with her partner. The car was likely to break down so they had turned back. JL recalled feeling very disappointed and hitting herself. It was suggested that she should also try the strategy of writing down her feelings at times when she found disappointment overwhelming. Again, no suicidal ideation was evident in this session.

The session also went on to consider abuse from the age of eleven. Around this age JL was having sex education at school and it was then that she realised that the abuse she was experiencing was inappropriate. In describing the abuse she felt very angry and frightened.

Therapy Session 5

One incident was recorded in JL's diary. This related to feelings of disappointment. JL managed to avoid harming herself by writing down the thoughts and feelings she was experiencing. This indicated that the strategy was also effective in dealing with this emotion. There was no evidence of suicidal ideation.

The session also focused on reinterpreting JL's abuse experience from an adult's point of view. In doing so JL began to recognise that she would have been very small and vulnerable and that the abuse was not her fault but the fault of her father who was much stronger and more powerful than she was.

JL decided to go back to work and did so the following week.

Therapy Session 6

From session 6 onwards no further self injurious thoughts or behaviours were reported. JL was advised to record relevant details if they recurred in the future.

Session 6 focused mainly on JL's feelings towards her father. She could identify a variety of emotions, including fear, anger and what she called an 'intense hatred'. This hatred was the most difficult feeling for her to cope with. She felt that hatred was a terrible thing because if you hated someone then you wanted to retaliate and if you wanted to retaliate then that meant you were as bad as the person you hated. To help her reframe this belief she was asked to imagine talking to someone else who had had the same experiences of having been abused as herself. She recognised through doing this that if she hated the abuser then that did not necessarily mean that she herself was bad.

Therapy Session 7

In session 7 JL said that she wanted to confront her father. The session focused on her reasons for wanting to do this. The necessity to avoid unrealistic expectations of the outcome of such a confrontation was emphasized. JL decided that for her own safety she would not confront her father at that time.

Therapy Sessions 8-10

Sessions 8-10 continued to focus on the abuse experience paying particular attention to the context of the abuse in terms of other family members. By session 9 it was clear that JL's mood was lifting considerably. A BDI score of 10 in session 10 confirmed this. She reported that although she knew she would always have memories of having been abused she no longer found these memories as painful. Five further sessions were arranged with the aim of focusing on her current life situation and establishing any changes she desired within it.

Outcome of treatment

Treatment had aimed to reduce JL's depression and self injurious behaviours. Her BDI of 35 at the assessment interview was indicative of a severe level of depressive symptoms. At session 10 her BDI score of 10 indicated that her depressive symptoms were mild. Her level of depressive symptoms had, therefore, decreased. Her self injurious thoughts and behaviours also decreased as indicated by her daily recordings. Taking her self reported data as a baseline, prior to treatment she was engaging in approximately two episodes of self injury a week. In the weeks preceding therapy sessions her reports of self injury reduced as shown in table 1. By session 6 she was no longer using self injury as a way of coping with her emotions. In addition, in session 10 she reported that she was able to cope with the feelings associated with having been abused without feeling overwhelmed.

Table 1. Incidence of self injurious thoughts and acts as indicated by weekly recordings

Week Preceding	Self Injurious Thoughts*	Self Injurious Acts*
Session 2	2	2
Session 3	1	1
Session 4	3	2
Session 5	1	0
Session 6	0	0
Session 7	0	0
Session 8	0	0
Session 9	0	0
Session 10	0	0

*All self injurious acts and thoughts involved JL punching her stomach, arms or legs or experiencing the desire to do so.

Discussion

The therapeutic approach to JL's difficulties clearly proved to be of benefit. It alleviated her depressive symptoms and her self injurious behaviours. In addition, by session 10 she felt more in control and able to cope with her feelings associated with having been abused.

In reviewing the treatment programme there are certain elements within it which seem to have been of crucial importance. Firstly, the decision to consider self injurious behaviour in its own right was essential. Had therapy simply aimed at focusing on JL's memories of abuse the resulting increase in distress may have increased her degree of self injury to the point of compromising safety. Secondly, emphasising that self injury served a purpose demonstrated to JL that she had the ability to adapt to her difficulties. This increased her belief in self efficacy and so helped to raise her self esteem. This was also an important factor in alleviating her symptoms of depression. A third factor that seemed to be of paramount importance in JL's case was the time taken in the first therapy session making the agreement that JL would take herself to casualty should she feel that her life was endangered by self injurious or suicidal behaviours. JL had stated that she was afraid that if she did not engage in self injurious acts to relieve overwhelming feelings then these feelings would escalate and lead to her taking an overdose. Setting up an agreement as to what she would do in such a situation lowered the fear associated with it. This lessened the necessity for self injury that she had previously perceived. It is notable that from the time the agreement was made

suicidal ideation was no longer evident. Reducing the associated fear appeared also to reduce the ideation.

Once JL began to use alternatives to self injurious behaviours it was possible to focus on a more direct approach to treatment for difficulties associated with childhood sexual abuse. Experiencing and naming feelings helped her to recognise that whilst her feelings were sometimes unpleasant they were not intolerable. This again emphasised that she could experience intense emotion without resorting to self harm. Reframing the abuse experience from an adult point of view also enabled her to see that she had no responsibility for the abuse. This helped to lessen her guilt and so lift her mood.

At initial assessment JL had come across as being extremely vulnerable. She expressed suicidal ideation. She had a history of previous suicide attempts and she reported substantial self injury. Her case illustrates how treatment for self injurious behaviour became an integrated part of her treatment programme. The approach taken enabled therapeutic exploration of distressing memories and feelings without compromising her safety. It is suggested that the integrated treatment described may be of benefit to other patients who present with similar difficulties.

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Cognitive-behavioural treatment of sleep related anxiety in childhood - a single case study

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Cognitive-behavioural treatment of sleep related anxiety in childhood - a single case study

Summary

The use of cognitive behavioural techniques for the treatment of adult insomnia is well known and has proven efficacy (Espie, 1992). This paper presents a case study in which these techniques are applied to an eleven year old girl. Within five one hour sessions anxiety regarding her inability to get off to sleep reduced markedly and sleep efficiency increased. It is suggested that cognitive behavioural techniques may be useful in the treatment of some childhood insomnias.

Introduction

In recent years, psychological treatments of adult insomnia have focused on cognitive behavioural techniques. These techniques are largely based on the underlying assumption that insomnia is caused by a complex interaction of physiological, cognitive and behavioural factors, and often maintained by a classically conditioned response through which going to bed becomes associated with alertness, frustration and sleeplessness (Espie, 1991). The typical treatment approach combines an educational component, giving information about the

nature of sleep and sleep hygiene, together with attempts to establish appropriate stimulus control (Espie, 1993; Bootzin, Epstein and Wood, 1991). Lacks and Morin (1992) present a review which clearly demonstrates the efficacy of using this approach in the adult population.

Ferber (1990) has suggested that over estimating the requirement for sleep, poor sleep hygiene and irregular sleep schedules lead to inappropriate stimulus control and insomnia in children in middle childhood and adolescence. If this is the case then it is reasonable to assume that the cognitive behavioural techniques which have shown to be of benefit to adults may also be of benefit to them. This paper investigates the issue by presenting a case in which cognitive behavioural techniques were used to treat sleep related anxiety in an eleven year old girl.

Case Report

HJ is an eleven year old girl who was referred to a child and family psychology service for the treatment of insomnia. The referral letter stated that this had been a long-standing difficulty, that she denied any specific anxieties and that she had no notable previous medical history. She was not on any medication and she had had no previous treatments for sleep disorder.

Presenting Problem

At initial assessment interview both HJ and her parents reported that her major difficulty was in getting off to sleep. They could not pin-point the onset of this difficulty. They suggested that this had always been the case and that she had never slept really well. Her parents stated that their reason for presenting her for treatment at that time was because she was due to be starting secondary school. They were concerned that inadequate sleep would affect her educational performance.

Typically HJ would go to bed at 9.30 p.m., lie awake until 11 p.m. and then sleep until 7.30 a.m. She did not generally waken during this period. The pattern was consistent and remained the same regardless of what she was due to be doing the next day. She was, therefore, regularly achieving sleep efficiency (hrs asleep/ hrs in bed x 100) of 85%. 85% is usually considered to be the cut off point for sleep disorder, however, HJ was not tired during the day and she did not report a lack of energy. For this reason she could not be considered to have insomnia per se since the diagnostic criteria given in DSM-III-R specifically state that a diagnosis of insomnia can only be given if there is 'significant daytime fatigue or impaired functioning' (American Psychiatric Association; DSM-III-R, 1987).

There was no evidence of separation anxiety and no life events of note which could account for H.J.'s difficulties. In formulating the case, therefore, it was felt that the over-riding difficulty for HJ was her specific anxiety relating to her

difficulty getting to sleep. It was also felt that given the long standing nature of HJ's difficulties it was likely that classical conditioning had a role to play in their maintenance. It seemed likely that, over time, bedtime for HJ had become automatically associated with heightened arousal and anxiety regarding whether or not she would be able to sleep.

Treatment

Treatment aimed primarily at reducing HJ's anxieties regarding her difficulty getting to sleep, since it was felt that this anxiety exacerbated sleep onset latency. With this in mind, from the first session, considerable time was spent reassuring both HJ and her parents that the total amount of sleep she was receiving was adequate for a girl of her age and that although she was currently not getting to sleep as quickly as they would expect, relaxing in bed was just as good. In addition, treatment tried to re-frame the situation in a more positive light by suggesting that requiring slightly less sleep than her peers was something that HJ might find useful later on.

In order to promote a sense of control for HJ she was introduced to techniques for managing insomnia, using a modified version of those developed by Espie (1993). This aspect of treatment covered giving information about the nature of sleep and sleep hygiene, attempts to re-establish appropriate stimulus control and teaching HJ ways of dealing with tension and intrusive thoughts. Giving

information about sleep was thought to be of prime importance since a number of authors, including Espie (1992) have suggested that information can ameliorate feelings of being out of control and, therefore, feelings of helplessness. Discussion with HJ also indicated that she tended to be very tense during the period leading up to her bedtime and whilst she was attempting to sleep. Teaching her relaxation also seemed appropriate since tension is clearly incompatible with sleep.

In order to re-establish stimulus control and move away from the association of her bed with tension and anxious thoughts regarding whether or not she would be able to get off to sleep, HJ was advised to go to her bedroom at 9.30 p.m. as usual but not to actually go to bed until she felt a 'sleepy tired' feeling. She was told that she should then go to bed but that it was all right to get up again if the 'sleepy tired' feeling went away or if she experienced persistent anxiety or tension that she was unable to combat.

Treatment Sessions

HJ attended for a total of five sessions. These were arranged on a fortnightly basis and each session lasted one hour. A brief outline of treatment sessions is given below.

Session 1

Clinical assessment and history was taken and time was spent reassuring the family that HJ was receiving adequate sleep for a girl of her age. HJ was asked to begin recording approximate onset of sleep and wakening times.

Sessions 2-3

These sessions introduced HJ to information about sleep and sleep hygiene. She was advised to go to bed only when she felt the 'sleepy tired' feeling and told that it was permissible, and in fact useful, to get up again if feelings of tension or anxious thoughts became unmanageable. HJ was also given a relaxation exercise to practise in which she was required to imagine herself as a puppet with strings, first when the strings were tense and then when the strings were cut.

Session 4

Session 4 reviewed techniques introduced previously and also introduced methods for managing anxious thoughts relating to sleep, including the use of distraction. It was also emphasized during this session that the techniques HJ was beginning to use could be used on occasions when she was staying away from home. This was attempted on two occasions.

Session 5

Session 5 reviewed sleep information and the techniques that had been introduced. Progress at this point suggested that no further treatment was indicated.

Measures of assessment

After the first interview HJ was asked to keep a sleep diary, recording her estimated times of going to bed, sleep onset and waking up. She was advised to do this as soon as possible after wakening and within a maximum of one hour. From these recordings sleep efficiency could then be recorded for the previous fortnight.

In addition to recording sleep data, a short 6 item questionnaire was devised and administered to HJ in subsequent sessions. This questionnaire was designed to provide a measure of HJ's degree of worry regarding getting to sleep both at home and in other peoples' houses. It comprised the following items:

1. How worried do you feel when you can't get to sleep at home?

2. How worried do you feel when you think you are the only person left awake at home?

3. When you are getting ready for bed at home how worried do you feel about the possibility of not being able to go to sleep?

4. How worried do you feel when you can't get to sleep at a friend's house?

5. How worried do you feel when you think you are the only person left awake at a friend's house?

6. When you are staying at a friend's house and you are getting ready for bed how worried do you feel about the possibility of not being able to go to sleep?

HJ was required to rate each item on a five point scale ranging from 1 (no worry) to 5 (the most worried I could feel).

Treatment outcome

By the end of session 5 HJ's anxieties relating to sleep both at home and when staying away had reduced substantially. This was suggested by self reports and by her responses to the six item questionnaire. Her responses to items in the questionnaire are given in table 1. Mean sleep efficiency also improved, as detailed in table 2. This had not been the primary aim of treatment, the primary aim being to reduce anxiety. Improving sleep efficiency was, however, an additional benefit.

Table 1. Responses to the six item questionnaire administered in sessions 2-5

	Question No.					
	1.	**2.**	**3.**	**4.**	**5.**	**6.**
Session 2	4	4	5	5	5	5
Session 3	4	4	3	5	5	5
Session 4	2	3	2	3	3	3
Session 5	1	1	1	1	2	1

Figures given refer to HJ's responses to the questions detailed above. These responses were given on a five point scale ranging from 1 (no worry) to 5 (the most worried I could feel).

Table 2. Mean sleep efficiency (SE) in two week period prior to each session

Session	**SE**
Session 2	85%
Session 3	85%
Session 4	90%
Session 5	90%

SE = (time spent asleep/ time spent in bed) x 100

Discussion

This case has shown that a cognitive behavioural approach was affective in the treatment of sleep related anxiety in an eleven year old girl. In considering why the approach proved to be beneficial in this case it is necessary to take in to account the following factors. On initial assessment it appeared that both HJ and her parents were overestimating the total amount of time asleep she required. Prior to treatment she was going to bed at 9.30 p.m., before she was physiologically ready to do so. Lying in bed unable to sleep caused her anxiety which in turn led to the classically conditioned response of bed being associated with frustration and corresponding sleeplessness. Telling HJ that she did not need so much sleep and instructing her not to go to bed until she felt a 'sleepy-tired' feeling reduced the performance related anxiety regarding her ability to get to sleep. This combined with her newly acquired relaxation skills weakened the classically conditioned response and consequently improved her sleep efficiency. The approach that was taken indirectly led to a delay in HJ's bedtime. Piazza and Fisher (1991) have suggested that this alone is beneficial in overcoming sleep disorders in some children. Given that HJ's anxiety primarily centred around her ability to fall sleep it is unlikely that simply delaying her bedtime would have had a beneficial effect in her case.

It should be noted that HJ was highly motivated and very compliant with the treatment programme. Her parents were also very supportive of it. Had HJ not

been so motivated then the success of the intervention would not have been so great.

In conclusion, this case has shown that cognitive behavioural techniques can be used effectively in the treatment of sleep related anxiety in children. It is likely that the techniques may also prove to be beneficial in the treatment of some childhood insomnias.

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