INVESTIGATION OF THE POTENTIAL RELATIONSHIP BETWEEN STEREOTYPICAL BEHAVIOUR AND RESPIRATORY DYSRHYTHMIA IN RETT DISORDER

and

RESEARCH PORTFOLIO

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Prepared in accordance with guidelines for submission to Clinical Psychology Forum

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Has Opt-In Worked? - Evaluation of an Opt-In System

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Has Opt-In Worked? - Evaluation of an Opt-In System.

Introduction

The mismatch between the demand for psychology services and the available resources to meet that demand is a common phenomenon faced by departments of clinical psychology in the NHS. The undesirable result is long waiting lists. The solution is more trained clinical psychologists. This, however, is a long-term solution and cannot deal with the current waiting list problem . Psychologists have had to explore other ways of reducing waiting lists in the short-term. Recent organisational changes in the NHS, particularly the introduction of GP fund-holding and a pseudo-market-economy, have brought an increased 'external pressure' (Anderson and White, 1996) to reduce waiting times.

Various strategies have been suggested to reduce waiting times (Newnes, 1993 and Startup, 1994). These include: introducing evening clinical sessions, increased group work, more consultancy, restricting service access, brief therapeutic approaches and opt-in systems. These roughly fall into three general approaches: increasing resources, avoiding the problem and increasing efficiency (Startup, 1994). Opt-in systems have been identified as a means of reducing the amount of wasted clinical and administrative time, with a resultant improved efficiency.

Failure to attend appointments and the early termination of treatment have long been recognised as sources of inefficiency. Non-attendance at a booked appointment clearly wastes both clinical and administrative time. If it were possible to allocate these failed appointments instead to clients who would attend, this would help to reduce the waiting time. Early termination of treatment can lead to longer waiting lists, if clients are subsequently likely to be re-referred for the same problem. If the number of clients who terminate their own treatment at an early stage, without benefit, were to be reduced, waiting times for clients who would benefit would also be reduced.

Typically, did-not-attend (DNA) rates for first appointments of 20%-30% in departments of clinical psychology are reported (Madden & Hinks, 1987; Munro & Blakey, 1988; Spector, 1988; Green & Giblin, 1988; Anderson & White, 1996). Requiring patients to opt-in to treatment has been shown in a number of studies to reduce first appointment DNA rates. An increase in attendance of between 21% and 25% has been reported (Green & Giblin, 1988; Anderson & White, 1996). Spector (1988) provided information leaflets to patients prior to opting-in and reported similar improvement in attendance. Webster (1992), reported that the provision of an information leaflet increased attendance. In their study, Markman & Beeney (1990), reported that the use of an opt-in system had no effect on first appointment DNA rates but reduced drop-out rates by almost half. The inclusion of an information leaflet had no effect.

The results from previous studies, having indicated that opt-in systems were effective in reducing wasted time, led psychologists working in the 'Riverside group Service Locality' at the Lansdowne Clinic (Gartnavel Royal Hospital, Glasgow) to introduce an opt-in system for out-patient referrals, in 1995, as part of their waiting list strategy.

Examination of appointment attendance patterns had indicated that DNA's were most likely to occur at the first appointment. The introduction of the opt-in system aimed to reduce the first appointment DNA rate and maximise the use of clinical time. The purpose of this present study was to evaluate the effectiveness of the opt-in system in the year following implementation.

The Opt-in System.

All referrals received are initially assessed for urgency. Urgent referrals are then dealt with within two to three weeks. Non-urgent referrals are placed on the waiting list. At the same time the referred client is notified that they have been placed on a waiting list and are advised of the approximate length of wait. The referring agent is similarly informed. Three to four weeks before the patient is due to be seen, the client is sent an opt-in letter with a stamped addressed envelope for them to reply. The letter explains that the patient is now at the top of the waiting list and asks them to indicate whether or not they still require an appointment. An information leaflet about the clinical psychology service accompanies the opt-in letter. Patients who fail to reply or indicate that an appointment is not required are discharged and a letter of explanation is sent to the referring agent. Those who request an appointment are sent one within two weeks.

Research Questions.

How many people chose to opt-in?

Does the opt-in system result in lower first appointment DNA rates?

Under what circumstances do psychologists choose not to use the opt-in system?

Method

All 'Riverside Group' Locality out-patients due to be offered a first appointment between 1/1/96 and 31/7/96 were identified. Those assessed as being urgent referrals and given early appointments were excluded as were those who were incorporated into the waiting list initiative evening appointment system. Of the 207 remaining referrals, the opt-in system was used in 141 cases. In 66 cases, patients had been automatically sent an appointment when they reached the top of the waiting list. The high number of cases where the opt-in system was not in use was the result of new members of staff not using opt-in initially until a working caseload had been built up and one psychologist who had not implemented opt-in until May 1996.

Each patient sent an opt-in form was categorised as either appointment requested or appointment not required/no reply. It was not possible to differentiate between no reply and a negative response. For each patient offered an appointment it was ascertained

whether or not they attended or failed to attend their initial appointment. The waiting time for each patient offered an appointment was calculated.

Each psychologist in the 'Riverside Group' team was consulted and asked to indicate under what circumstances they did not routinely use the opt-in system.

Results

Opt-in rate:

opt-in letters sent	appointment required	appointment not required
141	118 (84%)	23 (26%)

DNA rate for first appointment:

	DNA 1st appointment	Attend 1st appointment
opt-in system	18 (15%)	100 (85%)
automatic appointment	17 (26%)	49 (74%)

Chi Square: X $(1, n = 184) = 2.96, p>0.05$ (not significant)

Waiting times

The mean waiting time between referral and date of first appointment offered was 80 days. There was no difference in waiting time between the opt-in system and the automatic appointments system nor any difference in waiting time between first appointment DNA's and attenders.

Use of opt-in

Once psychologists had implemented the opt-in system it was used for the majority of referrals received. The exceptions were a) urgent referrals and b) special circumstances, eg. where the partner of a referred patient is already being seen and a couples approach is to be adopted.

It would have been useful to have made a comparison between first appointment DNA rates pre- and post opt-in introduction. A number of difficulties prevented this. There was no clear information as to when individual psychologists introduced opt-in to their own practice. Secondly, centrally collected data is recorded under clinic location rather than locality served. In addition, until recently there was no differentiation between DNA and cancellation by patient in the centrally gathered data. Most, although not all patients seen at the Lansdowne clinic are in the Riverside locality. In the months April to July 1996, the first appointment DNA rate for the Lansdowne clinic was 20%. If first appointment cancellations are included, the first appointment 'non-attendance' rate is 36%. This is similar to a first appointment 'non-attendance' rate for the same period in 1995 of 34%.

Discussion

The opt-in rate in this study is similar to the 85% opt-in rate reported by Anderson and White (1996). The 15% of patients who did not opt-in probably comprise people who no longer require an appointment because their difficulties have got better during the time they have been waiting and those who were initially ambivalent to being referred and who given a free choice, would be unlikely to seek referral. Under the automatic appointment system, this group of patients would all have been sent an initial appointment. Some would have cancelled this appointment, others would have failed to attend their appointment, some would have attended with no further appointments being required and some are likely to have dropped out of treatment or have obtained little benefit due to their ambivalence about attending. In each case, this represents wasted clinical and administrative time, and likely to have the effect of maintaining lengthy waiting lists.

The comparison of first appointment DNA rate between opt-in and automatic appointment systems shows a tendency towards fewer wasted first appointments with the opt-in system, although this fails to reach significance. The non significant result in this case may reflect the power of the chi square statistic to detect a significant difference with the sample sizes used. If the percentage difference were to be maintained in larger samples, there would be a clear difference in first appointment DNA rates between opt-in and the routine sending of appointments. Although in this study there was a tendency towards improved first appointment attendance rates using the opt-in system, the degree of improvement is not as great as some other studies have found.

Opt-in systems seem likely to result in lower DNA rates either because difficulties improve without psychological intervention or the patient is able to make an informed decision whether or not to engage in a psychological intervention (Startup, 1994; Seager, in press). It would be useful to identify those referrals where there is an appropriate outcome without a psychological intervention. This may help to assess the appropriateness of requests for clinical psychology involvement and help identify those referrals which might be best dealt with by other professionals, eg. counsellors.

The patient's involvement in deciding whether or not a referral is necessary or likely to be beneficial, appears to be a central aspect in the success of opt-in systems. This has raised the question of when in the referral process should the patient be overtly involved in this decision. Seager (in press) has evaluated a new referral system. One of the aims of the new approach has been to 'psychologise' the referral process from the beginning. Patients are given information about clinical psychology services by their GP prior to referral and subsequently make a decision to opt-in, ie. proceed with the referral, or not at that point in the process. This system aims to give more responsibility for initial referral to the patient. Results reported show that the implementation of this new referral system led to a further reduction in DNA rates from 16% to 8%. An interesting finding reported by Seager is that three times as many patients dropped-out of the waiting list under the 'old' opt-in system as did using the revised opt-in system. It may be that patients who are engaged into a psychological process at an early stage, involving them taking more responsibility for referral, are more likely to remain committed to a psychological approach (Seager, 1994). This early-stage opt-in, however, may discriminate against those patients who are not initially motivated to seek

psychological help but do engage with their therapist and arrive at a satisfactory outcome. However, do such patients attend or do they currently opt-out or DNA?

Conclusions.

The opt-in system evaluated in this study has shown a tendency towards an improved first appointment DNA rate, although not a statistically significant difference. This, along with a proportion of patients who choose not to opt -in, represents a saving of both clinical and administrative time. The improved efficiency is likely to assist in maintaining shorter waiting lists, although in this study it was not possible to examine the effect on waiting times. It would be worthwhile investigating whether patients who opt-out or fail to attend their first appointment are subsequently re-referred. The level of early termination of treatment using opt-in should be ascertained. Another question worth addressing is whether the provision of information about clinical psychology services at an early stage, prior to referral, has any effect on commitment to a psychological approach and, therefore, subsequent opt-in and DNA rates.

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Literature Review

Rett Disorder: A Review of Clinical Aspects, Behavioural Characteristics and Breathing Irregularities

Prepared in accordance with guidelines for submission to the Journal of Intellectual Disability Research (Appendix 2.1)

Rett Disorder: A Review of Clinical Aspects, Behavioural Characteristics and Breathing Irregularities

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Abstract

Rett syndrome is a developmental disorder associated with intellectual disability, loss of purposeful hand movement, development of behavioural stereotypical behaviour and breathing irregularities. The course of the disorder follows through four stages. EEG abnormalities are typically present and there is evidence of a dysfunctional autonomic system. Stereotypical behaviour may have some relationship to episodes of breathing dysrhythmia. Behaviour modification techniques demonstrate some success in altering stereotyped movement, thus indicating that these movements are not entirely beyond voluntary control. Further study of stereotyped behaviour, autonomic functioning and breathing irregularities, focusing on their relationship to each other will assist understanding of the disorder

.

Rett syndrome is a pervasive developmental disorder, was first described in 1966 by Dr. Andreas Rett but has only become widely recognised and researched since the mid 1980's. The disorder is characterised by the onset of a number of specific deficits after a period of apparently normal development following birth and is associated with intellectual deficiency. Classically it is reported only in females and incidence is estimated at 1 in 10 000. Until recently, aetiology was unknown, but a genetic cause has now been confirmed (Willard and Hendrich, 1999).. Diagnoses is based on clinical criteria and until recently, no biological markers had been identified. However, Kerr et al (1995), have reported finding short fourth toes in 20% - 28% of Rett disorder cases.

Current DSM IV diagnostic criteria require the following: apparent normal prenatal and perinatal development, apparent normal psychomotor development during the first five months, and normal head circumference at birth. Subsequently, between the age of 5 and 48 months, head growth decelerates and between 5 and 30 months previously learned hand skills are replaced by characteristic stereotypical hand movements, such as 'hand-wringing'. Severe impairment of expressive and receptive language development occurs, with severe psychomotor retardation and a loss of interest in the social environment. Difficulty in maintaining co-ordinated gait or trunk movements becomes apparent. Hagberg and Witt-Engerstrom (1986), proposed four distinct stages through which the disorder progresses: early onset stagnation, rapid destructive stage, pseudostationary stage and late motor deterioration. Kerr (1995), describes a similar four stage progression of the disorder moving through pre-regression, regression, early post-regression and late post-regression. Kerr (1995), also highlights four sub-types: hypotonic, mildly hypotonic, dystonic and rigid.

Stage one, having onset between 6-18 months, is characterised by developmental stagnation, deceleration of head growth, diminished interest in play, hypotonia, the development of involuntary movement and incoordination. Kerr (1995) comments that during this stage the 'developmental ceiling' is reached, typically around 9 - 12 months. During the second stage developmental and cognitive deterioration occurs, there is social withdrawal, loss of acquired skills and communication, loss of purposeful hand movement, the development of stereotypical hand movement, episodes of screaming, irregular breathing, seizures and the onset of intellectual deficiency. The third stage reveals continued breathing dysrhythmia, hand stereotypy, seizures and intellectual deficiency. There is some recovery of communication and improved emotional contact. Gross motor dysfunction, postural difficulty and early scoliosis is apparent. Characteristics of stage four include progressive scoliosis, muscle wasting, rigidity and deteriorating mobility. Communication and emotional contact improves and the frequency of seizures is reduced. Dystonia and episodes of a staring gaze are observed.

Prior to regression, EEG recordings are normal (Kerr, 1995) but thereafter are abnormal, although apparently having no specific pattern (Jelligner et al, 1992). CT scans are normal in the early years but reveal some degree of generalised cortical atrophy as the disorder progresses (Jelligner et al, 1992). SPECT scans have revealed hypoperfusion of the basal ganglia and cerebellar vermis (Watters, G. et al, 1991) and MRI indicates globally smaller than normal brains and progressive cerebellar atrophy with advancing age (Murakami et al., 1991). Sleep disturbance is common in Rett syndrome, with lower than normal periods of REM (Jelligner et al, 1992).

For some time there has been speculation of genetic cause in Rett disorder but it is only recently that evidence of several mutations in the gene MeCP2 at Xq28 have been identified in a proportion of Rett disorder patients (Willard & Hendrich, 1999). Twin studies reveal concordance in monozygotic twins and discordance in dizygotic twins (Zoghbi, 1988; Braddock et al., 1993). There is also a greater reported incidence of familial cases of Rett syndrome than in the general population (Zoghbi, 1988 There is some evidence that there may be reduced levels of biogenic amines in those with Rett Disorder (Budden et al., 1990; Zoghbi et al., 1989).

Whilst it is agreed that intellectual deficiency is part of the Rett syndrome, there has been some suggestion of continued progressive cognitive deterioration leading to dementia following regression (Budden et al., 1990; Elian & Rudolf, 1989). Perry et al. (1991), administered the Cattell Infant Intelligence and the Vineland Adaptive Behaviour Scales to 28 girls, aged 2-19, years having diagnoses of Rett disorder. There was no correlation between age of onset and the Cattell Mental Age and there was a negative correlation between Cattell Mental Age and chronological age. Only the Daily Living Skills domain of the Vineland correlated with chronological age and age of onset. The results may be consistent with the notion of a progressive cognitive deterioration, although the authors point to methodological difficulties and alternative interpretations. These results do not demonstrate the presence of dementia in Rett's although they highlight the profound nature of the intellectual deficiency present. Cognitive assessment in ten Rett disorder patients, reported by Del Priorie (1986), similarly provides no support for any progressive cognitive deficit and Kerr et al. (1987), propose that the use of term 'dementia'should not be applied to Rett disorder without clear supporting evidence.

Stereotypical hand movements are considered a characteristic sign of Rett syndrome. These include hand-wringing, hand-mouthing and hand-biting (Paisley et al., 1993) Initially the hand movements involve contact with the mouth although this diminishes later, as does the extent of movement (Lindberg, 1991). Lindberg (1991) has observed that the hand movement in each individual Rett's girl follows a certain pattern repeated in a rhythmic cycle. These stereotypical hand movements follow loss of purposeful hand movement and appear to constrain normal use of the hands (Lindberg, 1991). Kerr (1987), has noted that, when interested and strongly motivated, some intentional hand use was possible, although this remained clumsy and poorly coordinated. These hand movements may become intensified when agitated (Lindberg, 1991) or with increased alertness (Kerr et al., 1990). Such movements may become reduced in intensity or cease when the person distracted or engaged in attending to a stimulus in the environment (Lindberg, 1991). They are not present during sleep (Lindberg, 1991). Elian & Rudolf (1989), report that stereotypical hand movements are more prominent when "bored, alone, excited or in anticipation" and they suggest that they may represent a form of communication.

Whilst stereotypical movements follow loss of purposeful hand movement during regression, Kerr et al. (1987), report evidence of abnormal movement prior to regression. Forty classic Rett disorder participants were studied. Family video recordings of 4 pre-regression girls, along with detailed histories of all, were analysed along with video films of all post-regression cases. The pre-regression films revealed patting and waving actions with jerky movement. There was also some evidence of abnormal repetitive opening and closing of hands and twisting of upper limbs, pre-

regression. Reports of the child's cleverest hand use suggested early difficulty and failure to develop beyond the 10-12 month developmental stage (Kerr et al., 1987).

There have been some behavioural interventions aimed at helping the Rett disorder person gain greater control over movement reported, although these are single case studies. Bat-Haee (1994) reported the successful use of shaping, graduated guidance and regulating of hand use in their participants subsequent development of self-feeding and drinking skills and improvement in ambulation. These changes had been maintained at 6 month follow-up. Sullivan et al. (1995), demonstrated the use of a contingency intervention programme to promote learning and discrimination amongst various environmental contingencies. At 18 month follow-up the authors reported that hand use appeared purposeful during training with no hand waving observed. Their participant showed an ability to appropriately modify behaviour in response to environmental contingencies and they conclude that the opportunity to exercise control over the environment promotes attention to stimuli in the environment. There was also evidence of generalisation of behavioural changes from the training session to the classroom. Paisley et al. (1993), used a behavioural treatment intervention for self-injurious behaviour associated with stereotypical hand movement. Using graduated guidance, reinforcement and contingency-response interruption, they obtained some success in promoting functional hand use and the virtual elimination of hand to mouth self-injurious behaviour.

Irregular breathing patterns are a prominent feature of Rett disorder, with episodes of hyperventilation, breath-holding and apnoea being apparent. Breathing irregularities in Rett disorder are only observed in the awake state ,with normal breathing patterns being

present during sleep. Breathing is controlled by two control systems: the automatic system during sleep and the behavioural system when awake, (Jelligner et al, 1992). The irregular breathing in Rett syndrome when awake seems likely to involve the behavioural control system (Lugaresi et al., 1985; Kerr, 1992; Glaze et al., 1986). This characteristic breathing pattern becomes apparent early in the course of the disorder but improves later as the disease stabilises (Lugaresi et al., 1985). Kerr et al. (1987), in a study of 21 Rett syndrome girls observed onset of respiratory disturbance about age four or end of regression, with the greatest disturbance being between the ages 5 -15 years.

In a study of 18 Rett syndrome participants and 12 controls, Southall et al. (1988), report three distinct patterns of breathing irregularity in those with Rett disorder. Group 1 (n=10) displayed episodes of hyperventilation and apnoea, with valsalva manoeuvres (voluntary forced expiration against a closed glottis) apparent, whilst awake. Hyperventilation was accompanied by increased heart rate, an exacerbation of movement and increased muscle tone. There was no evidence of breathing dysrhythmia during sleep. Those in the second group (n=4) did not hyperventilate but had a history of hyperventilation. During observation, episodes of apnoea and valsalva manoeuvres were apparent, accompanied by agitation. The third group (n=4) had no history of hyperventilation but displayed apnoea and valsalva episodes, although less so than group 2. Thus, in this study, 56% of the Rett Disorder participants displayed hyperventilation and in 22% there was evidence of past hyperventilation (Southall et al., 1988). Those in group 2 had an average age 6 years greater than those in group 1, suggesting improvement in breathing irregularity with age. Glaze et al. (1986), has suggested that hyperventilation in Rett syndrome follows periods of disorganised breathing, a response aimed at compensating for hypoxaemia. Kerr (1992) and

Southhall et al. (1988), however, report no evidence of hypoxaemia prior to hyperventilation. Kerr (1992), reports that the abnormal EEG pattern associated with Rett disorder seems associated with normal breathing and becomes normal in appearance with the onset of hyperventilation.

There are observable differences in autonomic functioning between 'normal' persons and those with Rett disorder (Kerr et al., 1997). The sympathetic and parasympathetic systems are part of the autonomic system. The sympathetic system responds to demands by increasing the heart rate and blood pressure. The parasympathetic system responds by increasing vagal tone in order to moderate sympathetic changes and maintain homeostasis. At rest, the Rett disorder person has a lower than normal vagal tone (Kerr et al., 1997). Normally, when a person hyperventilates, there is a resultant increase in heart rate and blood pressure as the sympathetic system acts. This is quickly followed by an increase in vagal tone as the parasympathetic system responds. However, in the Rett disorder person although vagal tone begins to rise following hyperventilation, it then begins to reduce again, with a resultant imbalance between sympathetic and parasympathetic systems (Kerr et al., 1997). Similarly, with normal breath holding, blood pressure and heart rate increase then gradually reduce as normal breathing returns but in Rett syndrome, wide fluctuations in blood pressure and heart rate are observed (Kerr et al. 1997). These observations suggest poor autonomic control and the dilation of pupils, and cooling of both hands and feet observed during hyperventilation are signs of sympathetic over-stimulation, (Kerr et al. 1997).

Stereotypical movements apparent in Rett disorder following regression appear to become exacerbated during episodes of breathing dysrhythmia. In a study of 14 girls

with Rett syndrome, Kerr et al. (1990), considered the relationship between movement, breathing irregularities and non-seizure EEG activity. EEG recordings and respiratory measures were taken, along with a video recording of each participant during observation. Following an analysis of the video material, movement was categorised as ++, + or - . All measures were synchronised in time and divided into 4 second bands for analysis. The ++ category represented energetic movement with hand-wringing or clapping. The + category included intermediate movement with tense posturing of lips. Absent or minimal activity, with fingertip movement reflected the - category. The authors report a significant difference in the amount of ++ activity between episodes of breathing dysrhythmia and normal breathing in the awake state. The change from normal to dysrythmic breathing was observed on awakening and when stimulated. Non-seizure paroxysmal abnormality of EEG was associated with periods of normal breathing. Increasing age revealed a tendency for a reduced hyperventilation with movement and EEG changes becoming less episodic (Kerr et al., 1990). The authors suggest that the association of exacerbated bursts of movement associated with breathing dysrhythmia reflect a common trigger for both, beyond voluntary control but possibly associated with alerting. Elian and Rudolf (1989), suggest, however, that breathing dysrhythmia may be more noticeable when Rett disorder persons are under-stimulated, excited or anticipating activity. They posit that stereotypical movement may be interchangeable with breathing dysrhythmia which may become apparent when hand movements are restricted. They suggest that both movement and irregular breathing is under voluntary control and that both may be a form of stereotyped communication.

There are a number of theories which attempt to explain the basis of stereotyped behaviour. The homeostatic theory posits that stereotypy serves to modulate the level of

arousal in an organism to maintain optimal arousal (Leuba, 1955). Thus, when under-aroused stereotypy may serve to increase stimulation and when over-aroused, for example by frustration or anxiety, stereotyped behaviour may act to reduce stimulation (Leuba, 1955; Goodall & Corbett, 1982; Guess & Carr, 1991). Developmental theorists have argued that stereotyped behaviours are an exaggeration of normal behaviours from an earlier developmental stage, which become fixed when the child fails to develop more advanced behavioural skills (Jones et al., 1995). Organic theories adopt the position that brain damage underlies the presence of stereotypy. Evidence for a relationship between stereotypy and low IQ, syndromes (e.g. autism), brain lesions and neurochemistry provide some support for the theory (Jones et al., 1995; Lewis et al., 1987). Learning theorists view stereotyped behaviour as learned responses, maintained by reinforcement (Jones et al., 1995). None of these theories on their own are likely to be able to account for all the factors influencing the emergence and maintenance of behavioural stereotypy in all cases and conceptual models draw on different theories.

Lovaas et al. (1987) adopt a behavioural position in advancing their perceptual model of stereotypy. They argue that stereotyped behaviours are operant responses, reinforced by the perceptual stimuli produced as a consequence of the stereotypy itself and that the perceptual reinforcers are controlled by the individual and not mediated by environmental stimuli.

Guess & Carr (1991) have proposed a tri-dimensional model of stereotypy. The model draws on developmental, homeostatic and operant theories. Level 1 represents behaviour that is rhythmic, internally regulated and potentially adaptive. It is biologically determined and resistant to environmental influence, similar to the rhythmic

movements apparent in infancy. Level 2 is a transitional stage between the internally regulated patterns of movement and later use of stereotypy to control others. Stereotypical behaviours at level 2 are adaptive and are seen as a mechanism for homeostatic modulating of arousal as environmental stimulation varies. It is the emergence of a "controlling function" (Guess & Carr, 1991) at level 2 which paves the way for stereotyped behaviour being used to control others. At level 3 stereotypy is viewed as an operant responding to external reinforcement. Guess & Carr (1991) state that transition between levels "occur when variables change to more closely reflect the behavioural and organismic conditions associated with that next level" (p 310).

Jones et al. (1995) have commented that different models of behavioural stereotypy place different emphasis on particular hypotheses without any evidence that one is any more correct than another. Miller et al. (1994) have proposed a new model of stereotyped behaviour, termed the 'interactive treatment model of stereotypy'. The model acknowledges that stereotypy is most common among learning disability populations and most often seen in people with some kind of organic impairment (Jones et al., 1995). The model allows for non-organic factors, such as operant factors, environmental stimulation, trauma and psychiatric condition to be a cause of stereotypy. Organic factors may lead directly to stereotyped behaviour. Syndromes such as Lesch-Nyhan and Rett disorder, where stereotypy is invariably present, may fall in to this category. However, the direct organic link is tentative and it may be the case that organic factors are mediated by non-organic factors. Negative outcomes of stereotypy, such as reduced opportunity to learn new skills, may also influence the presence of stereotyped behaviour.

In Rett disorder, it is not yet clear to what extent, if any, behavioural stereotypies and breathing irregularities in Rett disorder are under voluntary control. The demonstration of some success in reducing stereotyped movement and increasing functional hand use with behavioural interventions indicates some capacity to control movement. Given that EEG recordings tend to normalise during hyperventilation, it may be possible that the hyperventilation is a voluntary attempt to alter some perceived internal state. However, it seems that abnormality of the autonomic system is present and the Rett disorder person's ability to exercise control is likely to be limited. Kerr et al. (1990), have demonstrated a correlation between breathing dysrhythmia and exacerbated stereotypical movement, although this finding related specifically to paroxysmal episodes of stereotypy. Further study of the relationship between these features may reveal whether the presence of a particular behaviour is typically associated with any specific breathing rhythm and thus may be a useful behavioural marker of respiratory dysrhythmia or autonomic dysfunction in Rett disorder. It may be important to obtain a clear understanding of the baseline relationship between behavioural events and respiration before undertaking specific experimental interventions or manipulations. A clearer understanding of factors influencing and related to stereotypical behaviour in Rett disorder may inform theories and models of stereotypical behaviour

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Major Research Project Proposal

Investigation of the Potential Relationship between Stereotypical Behaviour and Breathing Dysrhythmia in Rett Disorder

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2. Title

A Study of the potential relationship between observed stereotypical behaviour and respiratory disturbance in Rett disorder.

3. Summary

People with Rett Disorder display dysregulation of autonomic functioning as evidenced by respiratory disturbance and poorly controlled vagal tone. Further they also show irregular EEG traces. Whilst stereotyped movements and respiratory dysrhythmia are two key characteristics of the disorder the precise nature of any relationship between these variables is unclear. The aim of this project is to investigate the possible relationship between observed stereotypical behaviours and respiratory disturbance. An investigation of behavioural manifestations and respiratory functioning in participants with Rett's Disorder, who have previously undergone autonomic assessment will be carried out by means of analysis of video recordings and temporally synchronised measures of respiration.

4. Introduction

Rett disorder is a progressive disorder, reported only in females and having onset in infancy. A genetic cause has long been suspected but it is only recently been confirmed (Willard and Hendrich, 1999). Rett disorder is characterised by the onset of numerous specific progressive impairments following a period of apparent normal development. Diagnostic criteria (American Psychiatric Association, 1994) require the following: apparently normal prenatal and perinatal development, normal psychomotor development during the first five months, and normal head circumference at birth. Subsequently, between the age of 5 and 48 months head growth decelerates and between 5 and 30 months previously learned hand skills are replaced by characteristic stereotyped movements, such as 'hand-wringing'. There is severe impairment of

expressive and receptive language development, with severe psychomotor retardation and a loss of interest in the social environment. Difficulty in maintaining co-ordinated gait or trunk movements is observed (American Psychiatric Association, 1994). The clinical course of Rett's disorder follows four stages (Hagberg et al. 1986). Treatment is focused on the clinical management of the individual.

CT scans in Rett's disorder patients appear normal whilst young but reveal mild to moderate generalised atrophy in later stages (Jellinger et al., 1992). Although there is no particular EEG pattern specific to Rett's disorder, abnormal EEG recordings are evident. In common with many learning disability conditions, abnormal sleep patterns are observed with lower than normal REM (Jellinger et al., 1992), reflecting severity of brain damage. Whilst awake Rett's disorder patients display disorganised breathing but during sleep regular continuous breathing is evident and may be indicative of a disordered voluntary/behavioural respiratory control system (Jellinger et al.,1992, Glaze et al., 1987 & Nomura et al.,1986). In the awake state, normal breathing is observed but this is interrupted by respiratory dysrhythmias. Irregular breathing patterns observed in Rett disorder include: breath holding, regular breath holds (succession of breath holds), apnoea, hyperventilation, shallow breathing, rapid shallow breathing, deep breathing, tachypnoea, protracted inspiration, cheyne-strokes (periodic breathing), Biots breathing (periodic abrupt apnoea and abrupt regular breathing), valsalva's manoeuvre and atypical breathing (Julu et al., 1997; Kerr et al., 1998). The respiratory dysrhythmia in Rett disorder appears to be due to dysregulation of the autonomic system, as a result of brain stem immaturity (Julu et al.,1997).

Stereotyped behaviour comprises repetitive motor responses having no apparent purpose or adaptive significance and considered pathological due to their excessive or inappropriate nature (Baumeister 1978, Barton & Broughton 1980). There are a number of theories which attempt to explain the basis of stereotyped behaviour. The homeostatic theory posits that stereotyped behaviour serves to increase the level of arousal when the environment is under-stimulating but does not account for the presence of stereotypy as a consequence of increased arousal (Jones et al.,1995). Developmental theorists have argued that stereotypy is an exaggeration of normal behaviour from an early developmental stage, which may become fixed when a child fails to develop more advanced behavioural skills (Jones et al.,1995). However, in Rett disorder there is an apparent loss of purposeful movement and developmental regression. Organic theories adopt the position that brain damage underlies the presence of stereotypy. Evidence for a relationship between stereotypy and IQ, syndromes, brain lesions and neurochemistry provide support for the theory (Jones et al.,1995) Learning theories view stereotyped behaviour as learned responses which are reinforced (Jones et al.,1995). None of these theories on their own are likely to be able to account for the factors influencing the emergence and maintenance of stereotyped behaviour and conceptual models draw on the different theories. Miller et al. (1995) present an interactive treatment model of stereotyped behaviour. The model allows for a direct relationship between stereotypy and organic factors, a direct relationship between non-organic factors (psychological factors and environmental factors) and stereotypy, or that that non-organic factors act as mediating variables between organic factors and stereotyped behaviour.

In Rett disorder behavioural stereotypy is a characteristic feature of the disorder and although there exists some variation between individuals in topography of stereotypy, there is also a striking similarity, particularly repetitive movement involving the hands

(See appendix 3.3 for a list of stereotypical behaviour reported in the literature). This may reflect a common underlying organic cause with minimal influence from non-organic variables. There is clear evidence of organic involvement in Rett disorder (Julu et al., 1997; Kerr et al., 1998), which amongst other symptoms gives rise to autonomic dysregulation. Respiratory dysrhythmia is one aspect of the autonomic dysfunction.

The precise nature of any relationship between respiratory dysrhythmia and stereotyped behaviour in Rett disorder is not clear. However, stereotypy appears to become exacerbated during periods of breathing dysrhythmia and a common trigger for both, beyond voluntary control and associated with alerting has been suggested (Kerr et al., 1990). Elian & Rudolf, (1996), following telephone interviewing of carers concluded that in 72% of Rett disorder subjects the intensity of hand movements varied directly or inversely with the pattern of respiration. Thus, stereotypy and breathing irregularity are two characteristics of Rett disorder, both likely to have an underlying organic contribution but may be influenced by environmental contingencies.

The present study aims to investigate whether there is an association between observed stereotypical behaviours and respiratory dysrhythmia. This will be done by analysis of video recordings of Rett disorder patients which are temporally synchronised with concurrent measures of respiration. The amount of normal/abnormal breathing present when stereotypy is observed and absent will be determined. If observed stereotypy is found to be associated with abnormal breathing, the question of association with specific types of breathing irregularity (e.g. 'energetic breathing', comprising hyperventilation, deep breathing, tachypnoea or valsava's manoeuvre (Kerr and Julu, 1999; Julu, in press).

5. Research question

The following research question will be addressed:

Is there a relationship between behavioural manifestations of stereotypy and respiratory disturbance in Rett Disorder?

Specifically, it is hypothesised that the presence of behavioural stereotypy is associated with:

a) an increase in the presence of respiratory abnormality.

b) specific type(s) of respiratory abnormality, e.g. 'energetic breathing'.

6. Plan of Investigation

Participants

The project will involve the analysis of existing data obtained during previous autonomic assessment of people with a diagnosis of Rett Disorder. Video recording and associated time synchronised respiratory data will be analysed. Six sets of video and respiratory data, each of 30 minutes duration, will be selected for analysis. The video samples will be chosen on the basis that each features stereotyped hand movement, which is a hallmark characteristic and diagnostic of Rett disorder (other stereotypy may also be present) and have minimal external interference, such as direct presentation of

stimuli (e.g. toys) or lengthy periods when the hand is out of view of the camera. Video material of the 6 participants in a setting where movement is unrestricted is available.

Observations of Behavioural Stereotypy

Preliminary viewing of the autonomic assessment video tapes to identify the range of stereotypical behaviour present has been carried out and is detailed below. Each of the stereotypical behaviours identified appears typical of stereotypy in Rett disorder (see appendix 3.3 for list of stereotypical behaviours in Rett disorder reported in the literature).

Case 1 -	Repetitive movement of the fingers
	Hand-mouthing
	Tonge protrusion/chewing
Case 2	Repetitive movement of the fingers
	Body rocking
	Head shaking

Case 3	Repetitive movement of the fingers
	Repetitive hand movement
	Hand-mouthing
	Chin tapping
	Body Rocking
	Head shaking
Case 4	Repetitive movement of the fingers
	Body Rocking
	Arm waving
Case 5	Repetitive movement of the fingers
	Arm waving
Case 6	Repetitive movement of the fingers
	Hand - mouthing i) thumb ii) fingers

Validity of behavioural measures

In the video recordings to be used, those people with Rett Disorder undergoing autonomic analysis have one arm gently restrained (by holding of the hand), to ensure that a blood pressure monitor placed on the finger does not become detached. This places some restriction on freedom of movement, preventing the presence of behavioural stereotypy involving both hands (e.g. hand wringing), although the unrestrained hand is still able to make contact with the restrained hand. Thus, in this study, stereotypical hand movement is restricted to observations of one hand only. To ensure that the stereotypical behaviours observed whilst the movement of one arm is restricted are representative of stereotypical behaviour in Rett Disorder, samples of video tape of the six people with Rett Disorder in an unrestricted setting will be examined to establish whether or not the particular stereotypical behaviours present in the main analysis tapes are also present in the unrestricted movement setting tapes. A second observer will be asked to judge whether or not the target stereotypical behaviours are present in the unrestricted setting video tapes, after viewing an example of each individual stereotypy from the main analysis tapes.

Measurement of Behavioural Stereotypy

Once the validity of the measures of behavioural stereotypy under study has been established, each of the 6 video recordings will be viewed and the onset, duration and termination of bouts of behavioural stereotypy will be recorded. Periods from each video where stereotypy is absent will also be identified. Inter-rater reliability of the

classification of observed stereotypy, as measured using Cohen's Kappa, will be established for a 20% random sample.

Measurement of Respiratory State

Respiratory movement data obtained through the use of a chest plethysmograph are represented by a computer generated analogue waveform. Julu's definitions of the breathing movements in childhood will be used to classify the breathing movement represented by the waveform (see Julu, 1998, for a brief description of these respiratory patterns). Thus the presence of normal and various categories (n=13) of abnormal breathing (e.g. central apnoea, breath holding, and hyperventilation) can be identified. Dr Julu has provided training in classification of respiratory pattern based on his definitions. Inter-rater reliability of coded respiratory state classifications will be established, using Cohen's Kappa, for a 20% sample of respiratory data.

Data Analysis

a) For each case, the ratio of normal to abnormal breathing during periods when each behavioural stereotypy is both present and absent will be determined and the resultant data analysed for association using a contingency coefficient statistic.

b) For each case, the specific dysfunctional respiratory pattern present during the periods of abnormal breathing will also be described and analysed using a contingency coefficient statistic to determine whether any particular abnormal respiratory state is associated with periods when behavioural stereotypy is present.

7. Practical application

This study aims to assist in the understanding of the relationship between stereotypical behaviours and respiratory disturbance in Rett disorder. Stereotypical behaviour and breathing irregularities are both features of the disorder. There is clear evidence of an organic basis underlying the respiratory irregularity, however, it is not entirely clear that stereotypical behaviour is the result of a direct organic causal influence. There appears to be some relationship between stereotyped movements and respiratory dysrhythmia, although individual variations are reported. The present study aims to provide a better understanding of the relationship between behavioural stereotypy and respiratory dysregulation in Rett Disorder and the study may contribute towards establishing whether or not stereotypy in Rett disorder is a behavioural marker of autonomic dysregulation. Further, findings be useful in guiding further research of a more

experimental nature which may help address the question of the utility of psychological intervention in aspects of clinical management.

8. Ethical approval

The data used in this project has been gathered during clinical assessment of autonomic functioning in each of the participants, for which ethical approval was not required.

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Major Research Project

Investigation of the Potential Relationship between Stereotypical Behaviour and Respiratory Dysrhythmia in Rett Disorder

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Investigation of the potential Relationship between Stereotypical Behaviour and Respiratory Dysrhythmia in Rett Disorder

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An Exploration of the Potential Relationship between Stereotyped Behaviour and Respiratory Dysrhythmia in Rett Disorder - A Single Case Series.

Abstract

Rett disorder is a developmental disorder affecting 1 in 10 000 females. There is apparently normal development in the first six months after birth, followed by developmental stagnation and regression. Respiratory dysrhythmia and stereotypical behaviour are classic features of the disorder. The study is a single case series investigating the possible relationship between stereotyped behaviour and respiratory dysrhythmia. Results show some modest relationship between these variables in some cases.

Introduction

Rett disorder is a developmental disorder, affecting 1 in 10 000 females (Kerr, 1995). After a period of apparently normal development, there is emergence of multiple specific deficits, such as : deceleration of head growth, poor co-ordination of gait and trunk movement, severe impairment of expressive and receptive language development and development of hand stereotypy (American Psychiatric Association, 1994). By around the end of the first year, developmental stagnation is apparent (Braddock et al.,1993; Kerr, 1995). Sometime after there is rapid developmental regression, including loss of acquired purposeful hand use and the emergence of stereotypical behaviour, particularly involving the hands (Braddock et al., 1993). Severe or profound learning disability is associated with the disorder (American Psychiatric Association, 1994), as well as EEG abnormalities, seizures, respiratory dysrhythmia and physical deformity (see Wilson, this volume for a review).

Irregular breathing is a central feature of Rett disorder, with those having the disorder displaying a number of respiratory dysrhythmias (Julu et al., 1997). Kerr et al. (1998) have identified over 12 types of respiratory dysrhythmia associated with Rett disorder, namely: hyperventilation, apnoea, breath-holding, regular breath holds, deep breathing, protracted inspiration, rapid shallow breathing, shallow breathing, tachypnoea, cheyne stokes, biot's breathing, atypical breathing and valsava's manoeuvre. These dysrhythmias may be grouped into five categories (Julu, in press).. Apneustic respiration (breath-holding, regular breath-holding and protracted inspiration) and feeble respiration patterns (apnoea, shallow breathing and rapid shallow breathing) may lead to inadequate ventilation (Julu, in press). Forceful breathing (hyperventilation,

tachypnoea, deep breathing and valsava's manoeuvre) and periodic breathing (cheyne-strokes and Biot's breathing) may cause excessive ventilation (Julu, in press). Atypical breathing represents respiratory rhythms that cannot be classified.

Respiratory rhythm in Rett disorder is unstable (Kerr & Julu, 1999) and Julu et al. (1997) found it was unusual for people with Rett disorder to sustain any specific dysrhythmic breathing pattern for longer than 2 minutes before changing to another pattern of respiratory dysrhythmia. Kerr (1998) following autonomic assessment of 7 people with Rett disorder reported that each switched between an average of 4 abnormal respiratory rhythms. Irregular respiration in Rett disorder is considered to be underpinned by immaturity in the brain stem, with evidence emerging of an imbalance of the brain stem serotonergic system (Kerr & Julu, 1997). Irregular respiratory patterns are only evident in the awake state and not during sleep (Lugaresi et al., 1985), leading to the suggestion of impaired voluntary / behavioural control of the respiratory system (Lugaresi et al., 1985; Glaze et al., 1987). Kerr (1992) has suggested that the breathing / movement disorder associated with Rett disorder may be linked to a homeostatic alerting response provoked by cortical inactivity. Woodyatt & Murdoch (1996) comment that cortical arousal may lead to disruption of the cyclic respiration pattern.

Stereotyped behaviour comprises repetitive motor responses having no apparent purpose or adaptive significance and considered pathological due to their excessive or inappropriate nature (Baumeister, 1978; Barton & Broughton, 1980). In Rett disorder, stereotyped behaviour, especially hand stereotypy, is a classic feature of the disorder post-regression, with evidence of subtle abnormalities pre-regression (Kerr et al., 1987). Stereotyped behaviour commonly associated with Rett disorder includes: hand-

wringing, hand-mouthing, hand-biting, repetitive opening and closing of hands, hand clasping, tapping, rubbing, squeezing involving hands, skeletal movement involving the face, trunk and limbs (Kerr et al., 1987; Kerr et al., 1990; Paisley et al., 1983; Sullivan et al., 1995; Wehmeyer et al., 1993; Wylie, 1996). Kerr (1987, 2000) comments that each hand performs it's own typically invariable routine, with each hand following a distinctly different pattern from it's partner. Although typically the hands come together, hand stereotypy is often performed with hands apart in teenage years or older (Kerr, 1987).

Kerr et al. (1990) reported that energetic stereotyped behaviour was more likely to be present during respiratory dysrhythmia than normal breathing. The stereotyped behaviours were "sequences of skeletal movement affecting trunk, face and limbs, identical on repeated occasions" Kerr et al. (1990, p 63). Levels of activity were defined as ++, +, or -. The difference between the levels of stereotypy and breathing pattern only reflected the ++ category, for 5 cases where the movement was described as paroxysmal (i.e. occurring in sudden outbursts). The measurement of stereotypy appears to be a global rating which may have comprised different topographical stereotypic movements co-occurring and does not identify hand stereotypy, the most commonly reported stereotypy in Rett disorder. Woodyatt & Murdoch (1996) studied the effect of visual and auditory stimuli on respiratory dysrhythmia in Rett disorder, in two cases. They reported that the presentation of visual and auditory stimuli led to increased disorganisation of habitual respiratory patterns. They also noted that hand wringing movement ceased during apnoea in one case and in the other became still or clasped during "extended disrupted apnoeic pauses" (Woodyatt & Murdoch, 1996).

However, they provide no details of method of measuring the stereotyped hand movement or whether there was any systematic comparison with normal respiration.

Different theories of stereotyped behaviour include: the homeostatic theory, developmental theory, organic theories and learning theories. Conceptual models tend to draw on these theories. Guess & Carr (1991) propose a three level theory. Level 1 draws on developmental theories and stereotypy is conceived as being internally regulated rhythms associated with early development. At Level 2, stereotypy serves a homeostatic function and reflects some degree of control. At level 3, stereotypy is seen as an operant response and can be used to control others in the persons social environment. It is unlikely that all stereotypy is caused by a single factor and models of stereotyped behaviour need to be able to account for the influence of several potential causes. Rett disorder is particularly interesting as there is also respiratory dysrhythmia, with evidence of organic involvement, in which the brain stem is implicated. However, as mentioned above, respiratory dysrhythmia may be triggered by cortical arousal interrupting cyclic breathing rhythms and consequently may be influenced by environmental stimuli. Kerr et al. (1990) have suggested that stereotypical movement and respiratory dysrhythmia may share a common trigger linked to alerting. If so, it may be reasonable to assume that the presence of stereotyped behaviour in Rett disorder is associated with abnormal breathing or a particular respiratory dysrhythmia.

The aim of the present study was to identify behavioural stereotypy in people with Rett disorder and measure the association between respiratory dysrhythmia and any observed stereotypy. Since there appears to be topographical variability between Rett disorder subjects, a single case series design was used. It was hypothesised that the presence of

stereotyped behaviour would be associated with abnormal breathing or a specific type of respiratory dysrhythmia.

Method

Participants

The project involved the analysis of existing data obtained during autonomic assessment of people with a diagnosis of Rett Disorder. The participants attended the autonomic assessment clinic and were accompanied by family members or carers, who remained present throughout the assessment. The equipment used during the autonomic assessment (i.e. chest plethysmograph, blood pressure monitor and ECG) requires that the person with Rett Disorder remain seated throughout the assessment. Video recording, providing a view of the upper body, from the waist and associated time synchronised respiratory data were available. Six sets of video and respiratory data, each of 30 minutes duration, were selected for analysis. The video samples were chosen on the basis that each featured stereotyped hand movement, which is a hallmark characteristic and a diagnostic feature of Rett disorder (other stereotypy may also be present) and had minimal external interference, such as direct presentation of stimuli (e.g. toys) or lengthy periods when the hand was out of view of the video camera. Video material of the 6 participants in a setting where movement was unrestricted was also available and viewed. Three participants in this study were aged between 4years and 11 years and three were aged between 20 years and 29 years. None of the participants had verbal ability and all were judged to have a severe learning disability.

Materials

A standard VHS video cassette recorder and television was used to view the video recordings of the participants. Respiratory measurement was originally obtained by use of a stretch sensitive chest plethysmograph and chest movement subsequently represented by a computer generated analogue waveform. During the assessment of

autonomic function, beat to beat heart rate was monitored using an ECG and beat to beat systolic and diastolic blood pressure was measured using a Finapres™ blood pressure monitor (Ohmeda, Eagleswood, USA). This data, represented by computer generated waveforms was available for each participant. For a detailed description of the apparatus and methods used in the assessment of autonomic function, see Julu (1997).

Procedure

Observations of Behavioural Stereotypy

Preliminary viewing of the autonomic assessment video tapes to identify the range of stereotypical behaviour present was carried out and is detailed in appendix 3.4. Each of the stereotypical behaviours identified appeared typical of stereotypy reported in Rett disorder. (see appendix 3.3 for list of stereotypical behaviours in Rett disorder reported in the literature).

Validity of behavioural measures

In the video recordings used, those people with Rett Disorder undergoing autonomic analysis had one arm gently restrained (by holding of the hand), to ensure that a blood pressure monitor placed on the finger did not become detached. This placed some restriction on freedom of movement, preventing the potential presence of behavioural stereotypy involving both hands (e.g. hand wringing), although the unrestrained hand was still able to make contact with the restrained hand. Thus, in this study, stereotypical hand movement is restricted to observations of one hand only. To ensure that the stereotypical behaviours observed whilst the movement of one arm was restricted were representative of stereotypical behaviour in Rett Disorder, samples of video tape of the six people with Rett Disorder in an unrestricted setting was examined to establish the

presence of the same stereotypical movements across settings. This was carried out by establishing whether or not the particular stereotypical behaviours present in the main analysis tapes were also present in the unrestricted movement setting tapes. A second observer (a clinical psychologist) was asked to judge whether or not the target stereotypical behaviours were present in the unrestricted setting video tapes, after viewing an example of each individual stereotypy from the main analysis tapes. Only those stereotypical behaviours which both raters agreed were present in the video samples of unrestricted movement were included in the analysis, with the exception of the hand-mouthing behaviours in case 6. Although examples of the hand-mouthing stereotypy were not evident in the unrestricted setting video, they were particularly prevalent in the main analysis video tape and typical of stereotypy in Rett disorder, and so were retained for analysis.

Measurement of Behavioural Stereotypy

Each of the six video recordings were viewed and the onset, duration and termination of bouts of behavioural stereotypy was recorded. Periods from each video where stereotypy was absent were also identified. Inter-rater reliability of the classification of observed stereotypy was established. This was carried out by asking a second rater (a clinical psychologist) to View a random 20% sample of video tape for each case and record the onset, termination and duration of each target stereotypical behaviour. This data was then entered into a spreadsheet (SPSS for Windows version 7.5) with a code used to indicate the presence (1) or absence (0) of the behaviour, each second during the sample period (i.e. each spreadsheet cell represented a 1 second duration). The observations of the stereotypical behaviours for each case were entered into one column consecutively, such that the observations of all stereotypical behaviours during the sample time period, for all 6 cases were contained in a single column. Observations of

stereotypical behaviour carried out by the main rater, for the same sample time periods was similarly entered in to a second column on the spreadsheet. Thus, inter-rater reliability of the classification of all observed behaviours in all cases was then established using a single measurement. The Cohen's Kappa statistic was used, with the inter-rater reliability being 0.89, for observations of all stereotyped behaviours accross the 6 cases.

Measurement of Respiratory State

Respiratory movement data obtained through the use of a chest plethysmograph and represented by a computer generated analogue waveform were viewed. Julu's definitions of the breathing movements in childhood (see appendix 3.2) were used to classify the breathing movement represented by the waveform (see Julu, 1998, for a brief description of these respiratory patterns). Whilst it is possible to accurately determine the frequency of the respiratory waveforms, amplitude is measured in arbitrary units (see Julu, in press for a discussion of this issue). In order to establish average depth of breathing for each individual, the trough to peak depth of inspiration for 5 normal breaths was measured and the average inspiration depth ascertained. This value ±50% provided the range considered to be the average depth of breathing for that person. In addition, inspiration is easily determined from the waveform but it is not always clear when expiration has occurred for example during protracted inspiration. Therefore, in order to ensure consistency of measurement, expiration was considered to have occurred if an apparent expiration had a depth of >25% of the inspiration depth. The presence of normal and various categories (n=13) of abnormal breathing (e.g. central apnoea, breath holding, and hyperventilation) were identified. The classifications were then grouped into one of 5 respiratory categories. these were a) normal breathing, b) apneustic breathing (breath holding, regular breath holds,

protracted inspiration, rapid shallow breathing and shallow breathing) c) forceful breathing (hyperventilation, tachypnoea, deep breathing and valsava's manoeuvre) d) periodic breathing (cheyne-strokes and biot's breathing) e) Atypical breathing (respiratory rhythms that cannot be classified). Inter-rater reliability for coded respiratory state classifications (6 categories) was 0.74, using Cohen's Kappa, for a 20% sample of respiratory data. This was calculated using the same proceedure described above for observations of behavioural stereotypy. Thus, the inter-rater reliability figure represents the measured reliability for all respiratory categories across the 6 cases.

Data Analysis

The data was analysed using SPSS for Windows version 7.5. Respiratory data was entered into the spreadsheet with one column used for each of the 6 respiratory states and a code used to indicate the presence (1) or absence (0) of the breathing pattern. Similarly, one column represented each stereotypical behaviour under assessment, again with a code being used to indicate the presence (1) or absence (0) of the stereotyped behaviour. All measurements were synchronised in time. For each case, the association between each of the 6 respiratory states and the presence or absence of each stereotyped behaviour under observation was measured using Cohen's Kappa.

Results

The percentage of normal and abnormal respiration observed and the proportion of time spent exhibiting stereotyped behaviour for each participant during analysis is presented in Table 1. It can be seen that there is a large amount of variability, between cases, in the amount of abnormal respiration and stereotypy present. Case 1 has the lowest amount of irregular respiration, with respiratory dysrhythmia present for less than half the duration of assessment. In case 6 there is almost always abnormal respiration apparent. None of the six cases displayed periodic respiration and, therefore, abnormal respiration comprises forceful respiration, apneustic respiration, feeble respiration and atypical respiration patterns. Case 5 displayed stereotypy for 13% of the time and respiratory dysrhythmia during 79.9% of the assessment period. Case 3 engaged in both stereotyped behaviour (88.7%) and respiratory dysrhythmia (74.6%) for a substantial amount of time during assessment

---

insert Table 1 here

As can be seen from Table 2, in case 1 repetitive hand movement showed a low positive concordance with normal respiration. A low negative concordance between hand-mouth stereotypy and normal respiration was also evident. There was no relationship between head shaking, chin tapping or repetitive finger movement and any respiration type; the magnitude of these concordances being almost negligible. The graphs in figures 1-5 show the distribution of stereotyped behaviour across respiratory type for case 1.

---

insert Table 2 here

---

insert Figures 1-5 here

Results for case 2 are shown in Table 3. All measured associations between observed stereotyped behaviour (finger movement and hand-mouth stereotypy) and respiratory type are slight, with the strongest association between hand-mouth stereotypy and forceful breathing falling just below 0.2. The distribution of finger movement and hand-mouth stereotypies across respiratory type is illustrated in figures 6-7.

---

insert Table 3 here

---

insert Figures 6-7 here

The concordance measurement results for case 3 are contained in Table 4. A low positive concordance exists between normal respiration and both repetitive finger movement and hand -mouth (thumb) stereotypy. Both Hand-mouth (thumb) and hand-mouth (fingers) stereotypy also have concordance at a low level with forceful respiration. Figures 8-10 contain graphs showing the distribution of stereotyped behaviours across respiratory type.

---

insert Table 4 here

---

insert Figures 8-10 here

In case 4 there was a low negative concordance between repetitive finger movement and normal respiration and feeble respiration as can be seen from Table 5. There is also a low positive concordance between repetitive finger movement and apneustic respiration. Figure 11 shows the distribution of repetitive finger movement across respiration type.

---

insert Table 5 here

---

insert Figure 11 here

In case 5, all concordance between repetitive finger movement and respiration type were slight, almost negligible suggesting no relationship between the stereotypy and respiration type (Table 6). The graph in figure 12 illustrates the percentage time that finger movement stereotypy was observed for each respiratory type.

---

insert Table 6 here

---

insert Figure 12 here

Similarly, the results for case 6 also reveal no evidence for a relationship between repetitive finger movement and respiration type (see Table 7). The distribution of repetitive finger movement across respiration type is shown in figure 13.

---

insert Table 7 here

---

insert Figure 13 here

Discussion

Results from this study reveal that in cases 1, 3 and 4, when each case is considered individually, there is evidence for a definite, moderate concordance between some stereotypical behaviours and respiratory types. The nature of these associations differs between cases. For example, repetitive finger movement in case 3 has a low positive concordance with normal respiration, whereas in case 4, a low negative concordance between repetitive finger movement and normal respiration is seen. In cases 2, 5 and 6 there were no clear associations between stereotypy and respiration, and the magnitude of any measured associations were slight or almost negligible.

The apparent between case variability in the measured associations between stereotyped behaviour and respiratory type casts some doubt on the notion that stereotypy in Rett disorder is directly caused by underlying organic factors, at least those associated with respiratory function (e.g. imbalance of a common neurotransmitter), without any other influence. That is to say, although all the cases share the same diagnoses, similar symptoms, clinical course and purported similarity of cerebral impairment, the nature of the relationships between stereotypy and respiratory type are dissimilar between cases.

It is possible that this variation reflects differing levels of developmental ceiling in each subject. The cases where there was evidence of moderate concordance between some stereotyped behaviour and respiratory dysrhythmia were all older subjects. Thus, it may be that in the three cases where there was no apparent relationship between respiration and stereotypy, this reflects a developmental ceiling within level 1 of the Guess & Carr (1991) model of stereotypy, i.e. representing internally regulated rhythms associated with early development. Again, with reference to Guess & Carr (1991), it is

conceivable that the associations between stereotypy and respiration apparent in cases 1, 3 and 4, reflect a greater degree of individual control over stereotypy (e.g. the use of stereotypy to serve a homeostatic maintenance function), reflecting a relatively more advanced developmental status.

Results from this study suggest that a great deal of the behavioural stereotypy in Rett disorder is present during both normal breathing and respiratory dysrhythmia. This finding is not inconsistent with the observations of Kerr et al. (1990),who found that paroxysmal energetic stereotypy (rated ++) was associated with respiratory dysrhythmia but also that less energetic stereotypy (rated +) was present during a substantial proportion of both normal breathing and respiratory dysrhythmia.

Although the single case series design, chosen because of apparent between subject variability in topography of stereotypical behaviour, allowed an in-depth analysis of stereotypy and respiration in each case, findings must be accepted with some caution due to small overall number of subjects included in the study. In addition, the analysis carried out in this study did not address the possibility of a particular temporal or sequential relationship between stereotypy and respiratory dysrhythmia, although the modest concordance levels where some association was apparent may mean that a strong temporal or sequential relationship is unlikely.

The variability between subjects underlines the importance of establishing baseline relationship between stereotypy and respiration prior to any experimental or treatment interventions. Kerr et al. (1998) have reported a case of improved respiratory functioning following treatment with a agonist for 1A serotonin receptors. A study to

examine any effect of such pharmacological treatment for respiratory function on stereotypy would be useful.

Due to the retrospective analysis of assessment data, it was not possible to introduce structured control over environmental events and this would be an important consideration in future research. A reversal design (i.e. ABAB) may be the most appropriate method of controlling the presence or absence of external stimuli , allowing analysis of any effect. Ambulatory monitoring would be a useful means of assessing respiratory and behavioural function allowing greater freedom of movement. Studies of the relationship between stereotypy and autonomic dysfunction may be more informative if EEG measurements are included.

Acknowledgement

Permission was given by the autonomic research team to use data included in a larger study. Advice was also received from Dr. Peter Julu, Dr. Stig Hansen and Dr. Alison Kerr from the research team. I am grateful for their help and input.

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	Normal respiration	Abnormal respiration	Stereotypy present	stereotypy absent	Missing data (stereotypy)
Case 1	59.6%	40.4%	47.5%	52.5%	0%
Case 2	40.2%	59.8%	41.7%	58.3%	13.2%
Case 3	25.4%	74.6%	88.7%	11.3%	0.3%
Case 4	36.3%	63.7%	29.1%	70.9%	14.4%
Case 5	20.1%	79.9%	13.0%	87.0%	7.8%
Case 6	4.9%	95.1%	63.5%	36.5%	3.4%

Table 1 *Percentage normal abnormal respiration and percentage stereotypy present/absent for each case.*

	Repetitive finger movement	Chin tapping	Repetitive hand movement	Hand - mouth	Head shaking
Normal respiration	-0.044 (0.000)	-0.035 (0.001)	0.348 (0.000)	-0.288 (0.000)	0.013 (0.312)
Apneustic respiration	-0.044 (0.046)	-0.072 (0.000)	-0.049 (0.000)	0.100 (0.000)	0.002 (0.916)
Feeble respiration	0.163 (0.000)	-0.155 (0.000)	-0.187 (0.000)	0.106 (0.000)	0.011 (0.608)
Forceful respiration	-0.045 (0.015)	-0.044 (0.052)	-0.094 (0.018)	0.161 (0.000)	-0.055 (0.018)
Atypical respiration	-0.019 (0.371)	-0.023 (0.163)	-0.011 (0.030)	0.013 (0.070)	0.010 (0.458)

Table 2 *Cohen's Kappa results (p value in brackets) for each stereotyped behaviour and respiration type in case in case 1.*

	Repetitive finger movement	Hand - mouth
Normal respiration	-0.111 (0.000)	-0.128 (0.000)
Apneustic respiration	-0.037 (0.106)	-0.135 (0.000)
Feeble respiration	-0.055 (0.023)	-0.171 (0.000)
Forceful respiration	-0.045 (0.036)	0.196 (0.000)

Table 3 *Cohen's Kappa results (p value in brackets) for each stereotyped behaviour and respiration type in case in case 2.*

	Repetitive finger movement	Hand - mouth fingers	Hand - mouth thumb
Normal respiration	0.200 (0.000)	0.176 (0.000)	0.358 (0.000)
Apneustic respiration	-0.062 (0.000)	-0.045 (0.034)	0.148 (0.000)
Feeble respiration	0.162 (0.000)	-0.003 (0.881)	-0.190 (0.000)
Forceful respiration	0.072 (0.002)	0.208 (0.000)	0.252 (0.000)
Atypical respiration	0.022 (0.349)	0.073 (0.002)	0.043 (0.063)

Table 4 *Cohen's Kappa results (p value in brackets) for each stereotyped behaviour and respiration type in case in case 3.*

	Repetitive finger movement
Normal respiration	-0.220 (0.000)
Apneustic respiration	0.318 (0.000)
Feeble respiration	-0.239 (000)
Forceful respiration	0.109 (0.000)
Atypical respiration	0.049 (0.000)

Table 5 *Cohen's Kappa results (p value in brackets) for repetitive finger movement and respiration type in case in case 4.*

	Repetitive finger movement
Normal respiration	-0.123 (0.000)
Apneustic respiration	-0.171 (0.000)
Feeble respiration	-0.005 (0.828)
Forceful respiration	0.176 (0.015)
Atypical respiration	0.087 (0.371)

Table 6 *Cohen's Kappa results (p value in brackets) for repetitive finger movement and respiration type in case in case 5.*

	Repetitive finger movement
Normal respiration	-0.003 (0.708)
Apneustic respiration	0.004 (0.831)
Feeble respiration	-0.039 (0.004)
Forceful respiration	0.003 (0.886)
Atypical respiration	0.034 (0.000)

Table 7 *Cohen's Kappa results (p value in brackets) for repetitive finger movement and respiration type in case in case 6.*

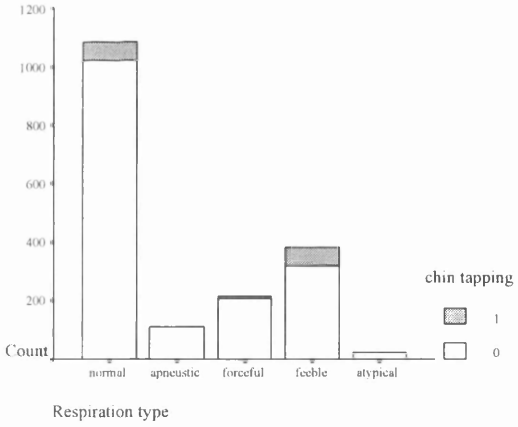

Figure 1 *Graph showing the distribution of chin tapping stereotypy across respiratory type in case1*

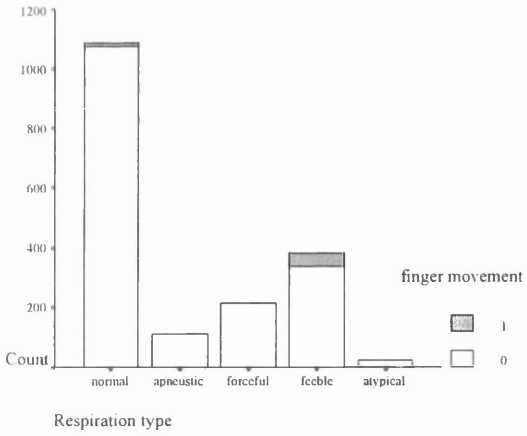

Figure 2 *Graph showing the distribution of finger movement stereotypy across respiratory type in case 1*

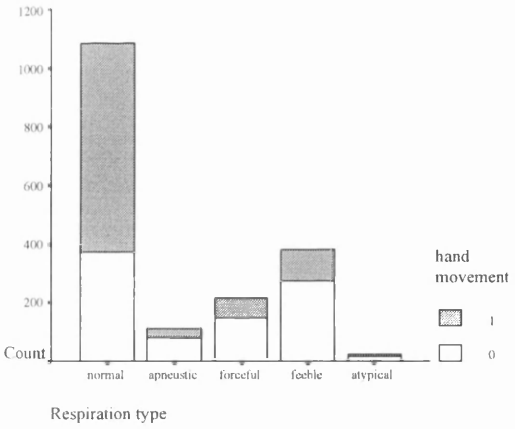

Figure 3 *Graph showing the distribution of hand movement stereotypy across respiratory type in case 1*

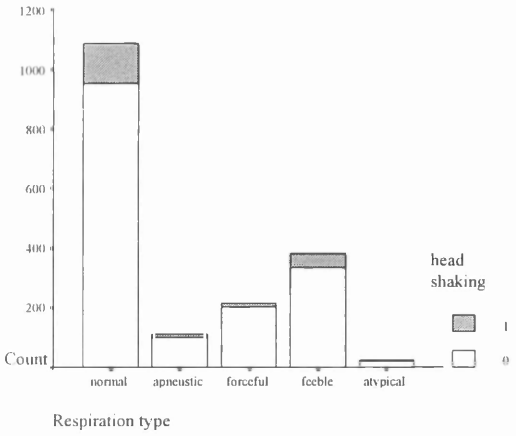

Figure 4 *Graph showing the distribution of hand-mouth stereotypy across respiratory type in case 1*

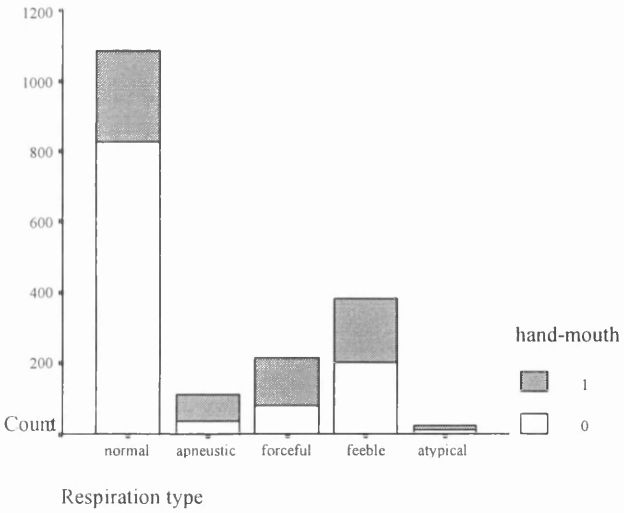

Figure 5 *Graph showing the distribution of head shaking stereotypy across respiratory type in case 1*

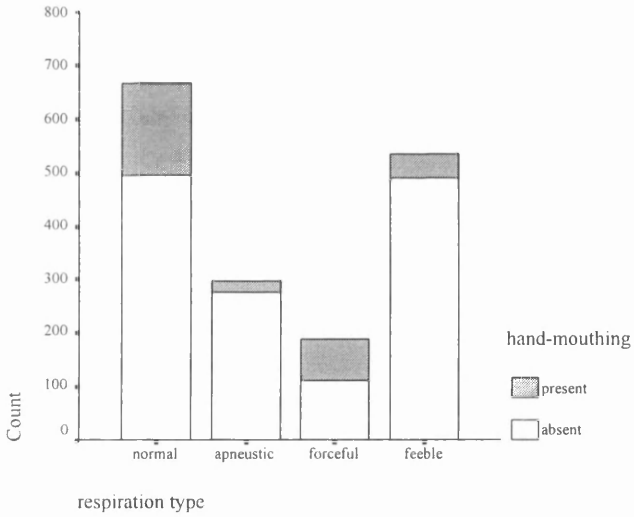

Figure 6 *Graph showing the distribution of hand-mouth stereotypy across respiratory type in case2*

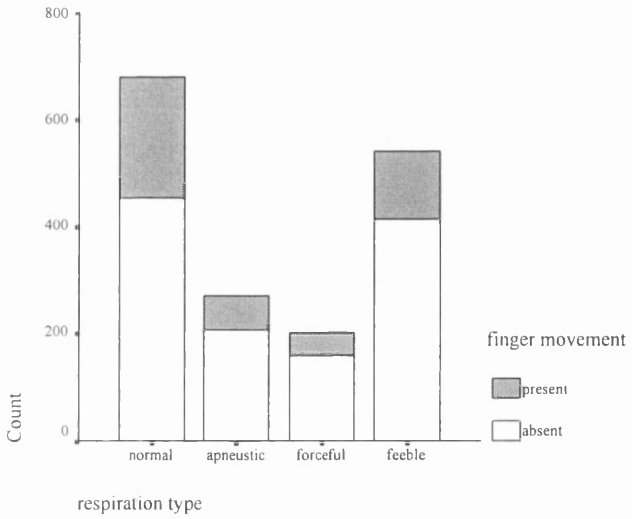

Figure 7 *Graph showing the distribution of finger movement stereotypy across respiratory type in case 2*

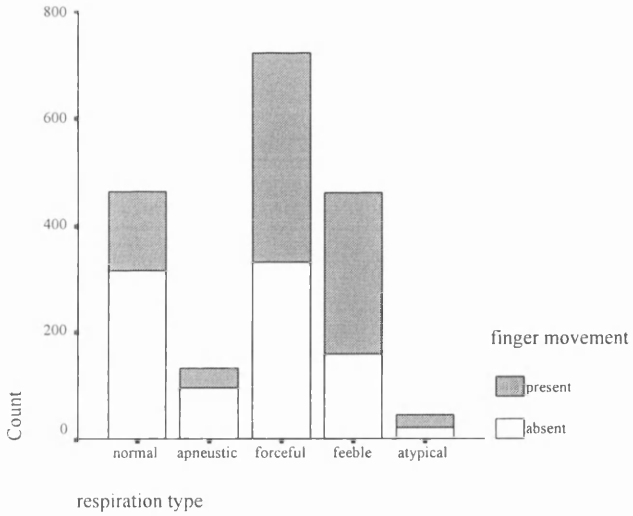

Figure 8 *Graph showing the distribution of finger movement stereotypy across respiratory type in case 3*

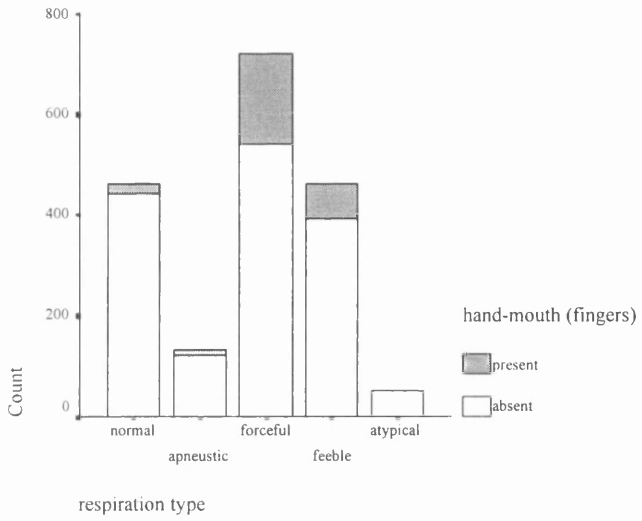

Figure 9 *Graph showing the distribution of hand-mouth (fingers) stereotypy across respiratory type in case 3*

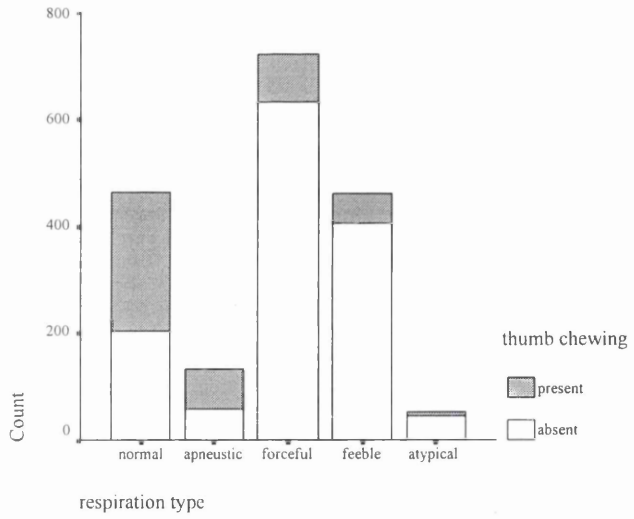

Figure 10 *Graph showing the distribution of hand-mouth (thumb chewing) stereotypy across respiratory type in case 3*

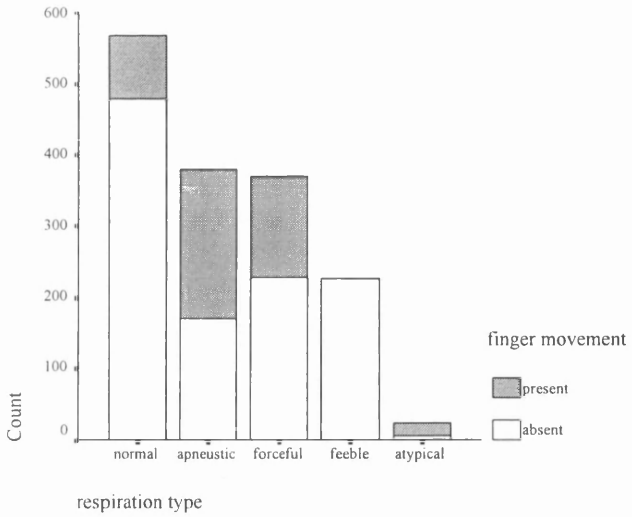

Figure 11 *Graph showing the distribution of finger movement stereotypy across respiratory type in case 4*

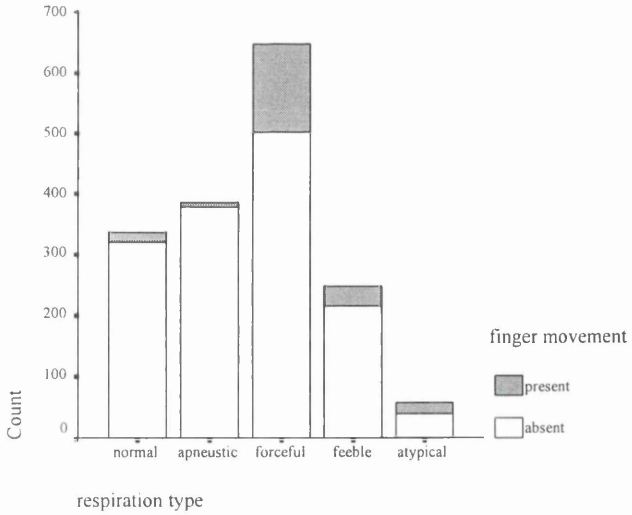

Figure 12 *Graph showing the distribution of finger movement stereotypy across respiratory type in case 5*

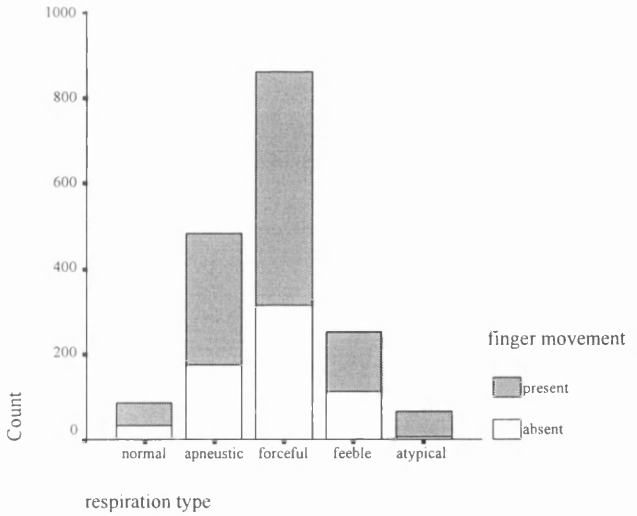

Figure 13 *Graph showing the distribution of finger movement stereotypy across respiratory type in case 6*

SINGLE CASE RESEARCH STUDY

A Comparison between the Effect of Familiar Music, Non-Familiar Music and No Music on Heart Rate, Blood Pressure and Leg Kicking in a 10 Year Old Girl with Rett Disorder

Abstract

Rett disorder is a developmental disorder of unknown aetiology, affecting 1 in 10000 females. Autonomic dysfunction and stereotypical movement are features of the disorder. Anecdotal evidence suggests that those with Rett Disorder are particularly responsive to music and that music has a calming effect. This single case study describes an experiment during which the influence of music on heart rate, blood pressure and a behavioural measures (leg kicking and vocalisation) was examined.

SINGLE CASE RESEARCH STUDY

Does soccer play a role in increasing risk for dementia in later life? - A single case neuropsychological assessment.

Abstract

Recently there has been growing interest in potential long-term neuropsychological effects of playing soccer, particularly with regard to the question of increased risk for developing dementia in later life. This is a single case study detailing the neuropsychological assessment of a retired professional soccer player who displayed symptoms of dementia and discussion of these in relation to soccer and other potential aetiological factors

SINGLE CASE RESEARCH STUDY

Psychological intervention in self-injurious behavior in a residential setting - A single case study

Abstract

The prevalence of self-injurious behaviour is highest amongst those living in a residential setting on a long-term basis. Presented is a single case study detailing a psychological intervention in a 31 year old man displaying self-injurious behaviour in a residential setting. Details of assessment, intervention and outcome are discussed, along with possible reasons why initial change was not maintained. The role of staff behaviour on client behaviour is considered.

Appendix 1

1.1 *Notes for Contributors "Clinical Psychology Forum"*

Clinical Psychology Forum

ical Psychology Forum is produced by the Division of Clinical Psychology of The British hological Society. It is edited by Steve Baldwin, Lorraine Bell, Jonathan Calder, Lesley Cohen, Simon thorpe, Laura Golding, Helen Jones, Craig Newnes, Mark Rapley and Arlene Vetere, and circulated ll members of the Division monthly. It is designed to serve as a discussion forum for any issues of rance to clinical psychologists. The editorial collective welcomes brief articles, reports of events, espondence, book reviews and announcements.

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Appendix II

2.1 *Notes for Contributors "Journal of Intellectual and Disability Research"*

Information for contributors

Papers (in English) should be sent to the Editor, *Journal of Intellectual Disability Research*, University of Wales College of Medicine, Meridian Court, North Road, Cardiff CF4 3BL, Wales, UK. Papers are accepted on the understanding that they have not been and will not be published elsewhere. The original and three copies should be submitted to aid refereeing and these should be typed (with a wide margin), double spaced, on one side of standard paper (A4—30 × 21 cm). A title page should contain the author's name(s), place of work, address for correspondence, full title and short running title. Authors should retain one copy of the text, tables and illustrations as the editor cannot accept responsibility for damage or loss of manuscripts.

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A structured summary should be given at the beginning of each article, incorporating the following headings: Background, Method, Results, Conclusions. These should outline the questions investigated, the design, essential findings and main conclusions of the study.

The text should proceed through sections of Abstract, Introduction, Materials and Methods, Results and Discussion. Tables and figures should be submitted on separate sheets and referred to in the text together with an indication of their approximate position recorded in the text margin. The reference list should be in alphabetical order thus:

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Journal titles should be in full. References in text with more than two authors should be abbreviated to (Brown *et al.* 1977). Authors are responsible for the accuracy of their references.

Spelling should conform to *The Concise Oxford Dictionary of Current English* and units of measurements, symbols and abbreviations with those in *Units, Symbols and Abbreviations* (1977) published and supplied by the Royal Society of Medicine, 1 Wimpole Street, London W1M 8AE. This specifies the use of S.I. units. Illustrations should be labelled with the figure number and author's name in soft pencil on the back identifying the top edge. Photographs should be glossy bromide prints of good contrast and well matched, preferably with a transparent overlay for protection. Colour photographs will be allowed only in special circumstances and the author will be asked to contribute towards the cost of reproduction. Line diagrams should be drawn with black ink on tracing paper or white card, or supplied as glossy prints. Papers may be judged to require extra-rapid publication by the Editor and referees.

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Royal Society for Mentally Handicapped Children and Adults (MENCAP)

The Royal Society for Mentally Handicapped Children and Adults is the largest national organization exclusively concerned with people with intellectual disability and their families. The primary objective of the Society is to secure for intellectually disabled people provision commensurate with their needs. To this end, the Society aims to increase public knowledge and awareness of the problems faced by intellectually disabled people and their families, and thus create a sympathetic climate of public opinion as a necessary prérequisite of their acceptance into the community.

The Royal Society for Mentally Handicapped Children and Adults provides:

- through a network of Local Societies and Regional Offices support in all parts of the country;
- funds and support for research;
- specialist advisory and information services for the lay public and for professional workers;
- books and literature and, bi-monthly, the *Journal of Intellectual Disability Research, Parents Voice* and *Viewpoint*, MENCAP's new newspaper;
- an ongoing programme to facilitate the sharing of knowledge by means of symposia, conferences and information exchange;
- residential facilities for further education and for care and holidays;
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RSMHC & A is a registered charity, supported entirely by voluntary contributions. Applications for membership, or information, are invited by the Secretary General.

Appendix III

Information for contributors

Papers (in English) should be sent to the Editor, *Journal of Intellectual Disability Research*, University of Wales College of Medicine, Meridian Court, North Road, Cardiff CF4 3BL, Wales, UK. Papers are accepted on the understanding that they have not been and will not be published elsewhere. The original and three copies should be submitted to aid refereeing and these should be typed (with a wide margin), double spaced, on one side of standard paper (A4—30 × 21 cm). A title page should contain the author's name(s), place of work, address for correspondence, full title and short running title. Authors should retain one copy of the text, tables and illustrations as the editor cannot accept responsibility for damage or loss of manuscripts.

Page proofs must be returned to the Publisher within three days of receipt. Typographical errors and essential changes can be made at this stage. Major text alterations cannot be accepted. One free copy of the relevant issue will be distributed by the corresponding author to each co-author. Offprints may be purchased at prices determined by the Publisher by returning the form enclosed with page proofs.

The author should provide up to six keywords to aid indexing. Please note that 'intellectual disability', as used in JIDR, includes those conditions labelled mental deficiency, mental handicap, learning disability and mental retardation in some locales or disciplines.

Full reports of 1500–3000 words are suitable for major studies, integrative reviews and presentation of related research projects or longitudinal enquiry of major theoretical and/or empirical conditions. *Brief reports* of 500–1500 words are encouraged, especially for replication studies, methodological research and technical contributions.

A structured summary should be given at the beginning of each article, incorporating the following headings: Background, Method, Results, Conclusions. These should outline the questions investigated, the design, essential findings and main conclusions of the study.

The text should proceed through sections of Abstract, Introduction, Materials and Methods, Results and Discussion. Tables and figures should be submitted on separate sheets and referred to in the text together with an indication of their approximate position recorded in the text margin. The reference list should be in alphabetical order thus:

Giblett E.R. (1969) *Genetic markers in Human Blood.* Blackwell Scientific Publications, Oxford.

Moss T.J. & Austin G.E. (1980) Pre-atherosclerotic lesions in Down's syndrome. *Journal of Mental Deficiency Research* 24, 137–41.

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Definitions of the breathing movements in childhood

Dr Peter O.O. Julu

1) **Normal breathing:** Ramp inspiration terminated immediately by double phased expiration.

   rate: below 35 breaths/min

   depth: average depth according to the individual

2) **Breath hold:** A single full inspiration (achieved fast) followed by a delayed expiration (also achieved fast) with intervening period of lack of respiratory movement. The breath hold must not raise intrathoracic pressure enough to cause measurable blood pressure and heart rate changes characteristic of reduced venous return.

3) **Central apnoea:** Cessation of breathing movement at the end of expiration.

4) **Rapid and shallow breathing:** Episode of shallow inspiration followed immediately by equally shallow expiration.

   rate: above 35 breaths/min

   depth: below average according to the individual

5) **Hyperventilation:** Episode of exaggerated inspirations followed immediately by equally exaggerated expirations contributing directly to a central apnoea as the end-point.

6) **Tachypnoea:** Episode of rapid inspiration followed immediately by expiration without causing central apnoea.

   rate: 35-45 breaths/min

   depth: average or above average depths according to the individual.

7) **Deep breathing:** Episode of exaggerated inspiration followed immediately by equally exaggerated expiration without causing central apnoea.

   rate: below 35 breaths/min

   depth: must be well above average according to the individual.

8) **Biot's breathing:** Abrupt apneoa followed by equally abrupt regular breathing in which both the apnoea and regular breathing have variable lengths.

9) **Cheyne-Stokes breathing:** Periodic breathing interrupted by central apnoea during which the breathing movements gradually increase in amplitude and then decay again into apnoea.

10) **Regular breath holds:** Breath holds in successions. Definition of breath hold is given above.

11) **Protracted inspiration:** A prolonged and continuous inspiration ended abruptly by full expiration (achieved fast, often forcefully). This must not raise intrathoracic pressure enough to cause measurable blood pressure and heart rate changes characteristic of reduced venous return.

12) **Atypical breathing:** Episode of inspirations and expirations of variable patterns.

rate: below 35 breaths/min

depths: average according to the individual.

13) **Shallow breathing:** Episode of shallow inspiration followed immediately by equally shallow expiration.

rate: below 35 breaths/min

depths: must be below average according to the individual.

14) **Valsalva's manoeuvre:** Breath holds or protracted inspirations capable of raising intrathoracic pressure enough in magnitude and duration to reduce venous return and cause characteristic blood pressure and heart rate changes (saw-toothed responses with rebounds).

Stereotypical behaviour in Rett Disorder reported in the literature:

Hand wringing

hand washing

Hand clapping

Hand mouthing

Hand biting

Rhythmic hand movement

Repetitive opening and closing of the hand

Hand waving

Hand patting

Idiosyncratic hand movement

Tapping

Rubbing

Clasping

Squeezing

Head banging

Hair pulling

Cheek Patting

Repetitive movement of the tongue

Skeletal movement affecting the trunk, face and limbs

Other hand and limb stereotypy

Atypical stereotypy

Appendix - *Stereotypical behaviour identified during preliminary viewing of video tapes.*

Case 1 -	Repetitive movement of the fingers
	Hand-mouthing
	Tongue protrusion/chewing
Case 2	Repetitive movement of the fingers
	Body rocking
	Head shaking
Case 3	Repetitive movement of the fingers
	Repetitive hand movement
	Hand-mouthing
	Chin tapping
	Body Rocking
	Head shaking
Case 4	Repetitive movement of the fingers
	Body Rocking
	Arm waving

Case 5	Repetitive movement of the fingers
	Arm waving
Case 6	Repetitive movement of the fingers
	Hand - mouthing i) thumb ii) fingers