

Caring for a Child with a Degenerative Disorder

A Parent's Perspective

*A Summary
of the
Skills, Knowledge and Understanding
needed to look after a
Child with a
Genetic, Degenerative Disorder.*

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See also:-

Andrew: a journey – a parent's perspective. D. Wray and S. Wray.

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Executive Summary

Parents are the front line carers who carry the main burden of care. They will be looking after their ill child for the greater part of the 24 hour period, seven days a week.

If their child has a static condition, the task of understanding the illness and organising support is largely a one-off exercise, given that there will be changes as the child matures.

For the parents of a child with a degenerative disorder, no sooner have they adjusted to the existing situation, then they have to start again, organising services, finding equipment, improving their understanding of the condition and discovering how to deal with new symptoms. A major problem for them is gaining access to information.

Much of the information they are given is provided in a haphazard manner, on a need-to-know basis and is therefore in a fragmented and limited form. This is not conducive to good learning and will not result in the desired outcome of a better understanding of the care processes.

Community planning seems to assume a large professional training and care input. The reality is that input is limited and the parents carry much of burden relying on intuition, creativity and common sense.

From the perspective of a professional specialist, with their extensive field of knowledge, it may appear that the proportion of information required by parents to care for their child is quite limited. However, this overlooks the fact that the parent carers are generalists; they are hands-on practitioners with their own body of knowledge consisting of elements drawn from many different disciplines. This does not make it any less academically valid or less arduous to acquire; it is an essential body of knowledge without which they cannot function effectively.

Much of this information already exists in different forms. It simply requires putting together as one information source set within an agreed set of parameters. It is extraordinary that such a simple editing exercise, which would make such a huge difference to the lives of carers and sick children, has not been done before!

Perhaps this paper might be a starting point?

The areas of knowledge which might be covered could be:-

- Carer's issues.
- Carer Training.
- Care of the Ill Child.
- Service Provision.
- Equipment.
- Quality of Life.
- Siblings.
- Bereavement.
- Reference Sources.

Key Recommendation

That a handbook be produced covering the care issues that are most commonly met by parents caring for a child with a life limiting, degenerative disorder.

Preface

For parents whose child has a static condition, the task of understanding the illness and organising support is largely a one-off exercise, given that there will be changes as the child matures. For us, as the parents of a child with a degenerative disorder we found that no sooner had we adjusted to the existing situation, then we had to start again, organising services, finding equipment, improving our understanding of the condition and discovering how to deal with new symptoms. A major problem was gaining access to information. We are aware of other parents for whom the situation was the same.

Discussing this with Prof. Tricia Sloper, (Social Policy Research Unit, University of York) she suggested that we list, in general terms, the new skills, knowledge and expertise that we had to acquire in order to manage our son's illness. This could then be the starting point for a handbook which could give guidance and basic information to the parents of children with degenerative conditions.

This we have endeavoured to do and the paper is the result. Even though we are the authors of the work, we are surprised at the range of skills and knowledge needed by parents to adequately care for their child under these circumstances.

David and Sally Wray

To Andrew

This paper is dedicated to our son Andrew, who died, aged nine, on the 29th August 2000, from the rare, degenerative, genetic disorder Niemann-Pick [Type C].

He is the real author: our part has been merely to record what he taught us.

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Field of Knowledge or Body of Knowledge?

Viewed from the perspective of a professional specialist, with their extensive field of knowledge on a very focussed topic, it may appear that the proportion of information required by a parent to function effectively as a carer quite is limited. However, this overlooks the fact that the parent carers are in fact generalists; hands-on practitioners with their own “specialised” body of knowledge which they need to care for their very ill child at home. The fact that it contains elements drawn from many different disciplines does not make it any less academically valid or less arduous to acquire. It is an essential body of knowledge; without it they cannot function.

It is therefore important that parent carers are trained and skilled effectively and provided with both verbal *and* written information from as few a sources as possible. The current situation of the fragmented and largely verbal transmission of information is ineffective and most unsatisfactory. It is well recognised that the greater part of what is heard for the first time is forgotten within twenty four hours. It is clearly more efficient and effective to provide parents with the opportunity, if required, of refreshing the details of a briefing or training session from pre-prepared, written information which they hold.

Much of this information already exists in different forms. It simply requires putting together as one information source set within an agreed set of parameters. It is extraordinary that such a simple editing exercise, which would make such a huge difference to the lives of carers and sick children, has not been done before!

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Contents

Section 1.	Introduction: Page 6
Section 2.	The Carers: Page 7
Section 3.	Carer Training: Page 12
Section 4.	Care of the Ill Child: Page 19
Section 5.	Service Provision: Page 28
Section 6.	Equipment: Page 33
Section 7.	Quality of Life: Page 38
Section 8.	Siblings: Page 48
Section 9.	Bereavement: Page 50
Section 10.	Conclusion: Page 54
Section 11.	References: Page 55
Section 12.	Useful Reference Books: Page 56
Section 13.	Appendices: Page 57

The layout of each section

**The issues within each section are arranged in alphabetical order.
Each topic has a summary of the main points.
Where there is additional background information, it is written in italics.**

Section 1. Introduction

Many people were involved in Andrew's care and we are grateful to them for the skills and knowledge they either knowingly or unknowingly passed on to us. Without them we could not have done what we did. (*See Appendices 4, 5 and 6: Observations on Good Practice*). Despite this however we still needed to do a considerable amount of research for ourselves, from a wide range of resources, in order to find the solutions to the many problems we faced. In addition, much of what is written here is the result of practical experience; a case of the necessity of the moment demanding a successful solution.

For this reason, it is difficult to attribute our knowledge to any one source. However, certain reference books we found to be particularly useful throughout the whole of Andrew's illness. These books we have listed in the Useful Reference Books section.

We are particularly indebted to "The British Medical Association's Guide to Medicines and Drugs" for much of the background information we needed to acquire about Andrew's medication.

Our hope is that through reading this paper others may benefit from what we learned.

Section 2. The Carers

This section is divided into five parts.

The part on the Consequences of Caring is further subdivided.

- **Carer versus Career**
- **Carer Identity**
- **The Consequences of Caring**
- **Respite**
- **Visualisation**

Part 1: Carer versus Career

Many parents of very ill children are faced with making a choice between continuing with a successful and worthwhile career or abandoning it to become a full time carer. Making a decision is not easy. The main issue is the balance between financial security (and reduced time with the ill child) or loans and debt repayment (but the once only opportunity to care for and spend quality time with the ill child).

There is no easy solution to this problem and unless the difficulties facing parents of children who have life limiting illnesses are recognised by government and adequate financial support provided, full-time parent carers will continue to be disadvantaged, whatever choice they make.

This is set against a background of:-

- *Increased expenditure on equipment and adaptations (budget implications).*
- *Huge physical and emotional care demands in a situation of inadequate rest and relaxation.*
- *Role conflict, (ill child's carer versus sibling's parent) and the consequent physical, emotional, and organisational demands.*
- *Expert (at work) versus novice nurse/carers (at home) and the need to quickly learn new skills.*
- *The experiential split that can develop between the carer and the wage earner (through living in two different worlds) and the effect on family relationships.*

Clearly a decision of this nature requires in-depth discussion by all the parties involved and careful consideration of the outcomes associated with each option.

Part 2: Carer Identity

Carers can suffer from stereotyping and this can result in difficulties, particularly in the areas of communication and training.

The assumption should not be made that the child's carer is either:-

- *One person only, on the basis that the other parent carer works. Both parents may be carers.*
- *Two people fulfilling interchangeable roles. It is unlikely that each carer is a duplicate of the other in terms of experience, skills, knowledge, aptitudes and also style and nature of interaction with the child.*
- *Female, which can overlook the stress for fathers resulting from a complete change of life-style.*
- *An adult, which results in the role of children as carers being frequently overlooked.*

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Part 3: The Consequences of Caring

Parent carers are well aware of their poor quality of life, but because of the immediacy of the situation they are unable to obtain a detached view in order to identify causal factors. In addition, because of the huge care demands they have little time to reflect on the problems and devise solutions. Many parents are reassured to discover that their experiences are common to most families caring for ill children and are helped greatly by gaining some understanding what might be causing their difficulties. These factors might include **Anger, Anxiety, Depersonalisation**, changes in **Family Dynamics** and **Stress**.

(1) Anger

Parent carers should be aware of a range of appropriate and effective strategies for dealing with anger and its causes.

Parent carers have to cope with a range of powerful emotions at a time when they are also tired and generally highly stressed. Anger is often the result of the frustration of dealing with an insensitive service provider; the individual feels that their opinions and needs have not been recognised, respected or valued. If unresolved, anger can be particularly destructive, adversely affecting both the physical and mental health of the individual and also their relationships with others, whether family, friends or professionals. In addition, it blurs rational thinking and can result in hasty and unfortunate actions which serve only to worsen the situation.

However, if the individual is able to adopt appropriate strategies, then this will both neutralise the anger and produce positive results. These might include:-

- *Distraction: Engaging in another, preferably peaceful, activity to allow the emotions to settle, knowing that the matter can be attended to later.*
- *Dissipation: Recording on paper (or some other medium) all the thoughts, ideas, facts, insults, needs and emotions which are swirling round in the individual's mind. Once stored in this way, they can be left and forgotten for*

the moment, to be returned to later. This overcomes the problem of the thoughts being continually recycled, thereby maintaining a high emotional state. Returning to the records at least a day later allows the facts to be extracted (without the emotions) and a logical, reasoned argument produced. It may be that by highlighting the individual's needs and hinting at possible solutions, the professionals are discretely provided with a set of targets, which, if acted upon, would resolve the difficulties.

- *Relaxation: This might include breathing exercises, controlled muscle relaxation, gentle physical movement, focussed mental activity and the use of music or natural sounds.*

(2) Anxiety

Parent carers need to be aware of:-

- The debilitating affect of anxiety on all the family members.
- What strategies to adopt to ameliorate the difficulties.
- Where and how to seek support from skilled and experienced counsellors.

Anxiety is an issue for all the family:

- *For the ill child, their whole life experience is anxiety producing: Living in a contracting and increasingly unpredictable world, loosing independence, experiencing unpleasant, disorientating and painful symptoms, having to tolerate increasing levels of intervention and technological support, steadily loosing their ability to communicate through orthodox means and having to rely increasingly on the abilities of others to interpret and satisfy all their needs.*
- *For the parents, there is the uncertainty about coping with future unknowns; unforeseen circumstances, distressing situations, not knowing what level of skill, knowledge or survival ability is required to support the ill child, the siblings, their partner and themselves: All this at a time when each family member may be close to being overwhelmed by their experiences.*
- *For the siblings, there are empathetic concerns for their ill brother or sister, worries about their tired, preoccupied and troubled parents and understandable concerns about their own future; their own emotional, physical and mental health, their social robustness within peer group situations and their general well being within a changed and possibly destabilised family.*
- *Linked to all the above are issues of seeking and receiving appropriate group and individual support, either within the family itself, or from outside agencies.*

(3) Depersonalisation

The caring situation results in varying degrees of depersonalisation, brought about by:-

- Lack of sleep.

- Loss of privacy and personal space.
- Loss of the opportunity to be creative and reflective.
- Loss of freedom of choice in personal and family matters.
- Loss of leisure activities, including holidays.
- Social isolation.

Although a lot could be said about any of the problems listed above, perhaps the most difficult to cope with is the combination of disturbed sleep patterns and inadequate sleep. It is not surprising that there is a sense of unreality about the world in which carers live.
(2004 Blackwell Publishing: D. and S. Wray)

(4) Family Dynamics

As the child becomes increasingly unwell and the care demands increase, the dynamics within the family gradually change, often with unfortunate results for the family.

Whereas in an unaffected family the interaction of the adults is between themselves and their children, in an affected family, owing to the pressures of caring, the pattern of the interaction changes; the adults focus less on each other and more on the ill child; there are separate lines of interaction for the ill child and the unaffected siblings, who are expected to defer to the demands of the ill child.

After the death of the affected child, it is difficult for family to make the necessary re-adjustment and the family can become dysfunctional. Also, because the experience has been so traumatic, the broken emotional links between the adults can be difficult to re-instate, particularly at a time when memories are still fresh and everyone is in a state of post traumatic stress. This can lead to the fragmentation of the family.

(5) Stress

The carers will evidence stress. This will not arise solely from the physical act of caring. Other, hidden factors will also add to the difficulty of the situation, possibly leading to the eventual break-up of the relationship.

Examples of these are:-

- *Financial difficulties.*
- *Unsatisfactory dealings with the service providers resulting in:-*
 - a) A high level of unresolved stress.*
 - b) Strong negative feelings which can adversely influence domestic, social and work situations.*
- *Inappropriate and unnecessary secrecy, which result in:-*
 - a) A difficulty in accessing information (barriers).*
 - b) Information not being freely available (restrictions).*
- *Hospitalisation of the home.*
There is a strong feeling of a "loss of ownership" as the home is adapted to

fulfil care needs. Also, whereas professional carers have the opportunity to leave the care area at the end of their shift and go home, no such opportunity exists for parent carers.

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Part 4: Respite

Respite from the task of caring does not necessarily mean the complete detachment of the parents from the patient. Simply allowing the parents to move from the front line of care to a position of standby is to provide respite. This is also reassuring to the anxious child who knows that although the parents are not in the same room, they are still nearby.

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Many parents feel that children with profound disabilities, requiring complex medical and nursing care, cannot be left for long periods in the care of others. A constant, parental presence is necessary to mediate between the child and the environment otherwise they may become highly stressed.

There is also an expectation/perception (perhaps motivated by propriety) that parent carers must be together when they are given time-off for recreation or leisure. This overlooks the fact that if they are both at home working together as carers for long periods, they may actually welcome a short break from each other.

In addition, many carers benefit from being in the company of people whose circumstance are more “normal” than theirs. It acts as a temporary distraction and counteracts the sense of unreality that the situation creates.

Part 5: Visualisation

Some professionals avoid revealing future situations to parent carers on the basis that they are too terrifying to consider. It may well be that it is ignorance and not knowledge that creates a problem. It is important that parents are able to rehearse in their own minds an impending situation so that they can consider coping strategies well before the event occurs. It cannot be right that the professionals are mentally prepared but not the parents.

It can help, when considering these situations, to think of the events happening to a third party so that the focus of the discussion is away from the child. The detachment can then allow a difficult and distressing issue to be discussed and prepared for.

Section 3. Carer Training

Although they may have acquired some expertise as carers, parents are without formal training and do not have the benefit of previous experience or easy access to academic reference materials, good practice handbooks, or informed colleagues. Much of the information they are given is provided in a haphazard manner, on a need-to-know basis and is therefore in a fragmented and limited form. This is not conducive to good learning and will not result in the desired outcome of a better understanding of the care processes.

Community planning seems to assume a large professional training and care input. The reality is that input is limited and the parents carry much of burden relying on intuition, creativity and common sense.

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Issues that might be covered under the heading of **Carer Training** are:-

- (1) Abbreviations**
- (2) Bed Care**
- (3) Care Issues**
- (4) Health and Safety**
- (5) Medicines**
- (6) Mobility**
- (7) Nutrition**

(1) Abbreviations

There are many abbreviations in common use, particularly in the medical world and in patient's notes. For the parent, who is the main carer of the child and is often making critical judgements on care issues, this coded information is unhelpful and can impact on their understanding of the situation. A glossary of common terms and abbreviations that they are likely to come across would help them to be better informed and therefore more able carers.

This also raises the issue of patient's and carer's access to information. Should a body of information about the patient be restricted to professional carers only, access to which for parents can require permission to be sought from managers? Parents might be forgiven for perceiving in this an element of outdated attitudes associated with prestige, mystique and control.

(2) Bed Care

Parents need to be aware of a range of bed care techniques which:-

- Allow the child's needs to be attended to with a minimum of disturbance to the ill child.

- Allow the child to be positioned, moved and handled in a manner which does not cause distress to the child and is safe for the carers.
- Allow the child to be placed in a number of comfortable positions.
- Improve the general comfort and well-being of the child.

For example:-

- *Use circular pads to avoid pressure sores on the ears.*
- *As well as using matchets or wedges, use rolled towels and small sleeping-bag pillows to provide support for limbs and trunk. This avoids the problems of over-flexion of joints (which can result in inflammation and discomfort) and bruising caused by one limb resting on another.*
- *Use a co-ordinated strategy whereby several carers lift the ill child vertically, very gently and slowly, while two others replace the sheets under the child.*
- *In conjunction with the above, use a mirror to check underneath the child for pressure sores rather than disturb them by rolling them onto their side or back.*

(3) Care Issues

The enormous range of skills required by parents to effectively care for a very ill child at home is often overlooked by those outside the family. A list (in alphabetical order) of day to day care skills used by parent carers might include:-

1. Acquiring new skills and knowledge on a wide range of topics.
2. Being sensitive to the patient's perceptions of the professional carers and monitor whether or not the child is happy and content with them.
3. Becoming communication sensitive and developing an increased awareness of cues used by the child to communicate needs and emotional states.
4. Being at all times mindful of the anxiety produced by the child's loss of speech and their inability to seek clarification.
5. Being aware of the child's reversion to thinking mainly in concrete terms.
6. Being aware of the difficulties caused by the child's visual impairment and adopting procedures to minimise anxiety and distress.
7. Being aware of the great effort involved in coughing and the possibility that pain may result.
8. Being aware that liver function may be affected and the need for care with medications.
9. Carefully planning the lay-out of the room(s), noting the relative positions of (and distances between) equipment, zones and services to avoid the problems noted below in 15.
10. Developing strategies to avoid or minimise the problems noted below in 15 and ensure their communication to and implementation with all who are caring for the child.
11. Developing proactive planning strategies with regard to the monitoring, ordering, collection or delivery of resources and consumables, e.g. medication (may be more than ten different items used at different rates), gastrostomy feeds, gastrostomy giving-sets, incontinence pads, wipes, syringes, aspirator catheters, oxygen cylinders and concentrator servicing.

12. Endeavouring to understand the perceptual problems associated with the disorder and developing strategies to reassure the child and thereby reduce their level of anxiety, fear and disorientation.
13. Ensuring that the child feels secure and comfortable with their surroundings.
14. Helping the child cope with memory loss.
15. Identifying the causes of and being sensitive to the distress associated with the child being moved. These may result from perceptual distortions, insecurity, anxiety, pain, a wish not to be disturbed at that moment, an inability to keep pace with rapid changes of events, a lack of prior warning (to give the child sufficient time to understand what is about to happen) or an invasion of the child's personal territory without prior warning or permission being sought.
16. Recognising the possible need for all the family (including the ill child) to obtain emotional and psychological support.
17. Recognising and identifying the many complex, interactive and largely unpredictable symptoms and taking appropriate action to minimise their effects.
18. Recognising the symptoms of allergies (some of which might be less common such as photo-sensitivity or cold injury), adopting avoidance procedures and initiating treatments.
19. Recognising the symptoms of reflux, responding to the severe symptoms and adopting strategies to reduce them.
20. Recognising the various "forms" of epilepsy (absences, unconsciousness, tonic-clonics, hallucinations, anxiety attacks, etc) and adopting strategies to ensure the welfare of the child.
21. Responding appropriately to respiratory difficulties caused by asthma, loss of muscle tone, infection, secretions in the lungs, inhalation of saliva, throat spasm, spasms of the chest muscles, over-full stomach or poor positioning resulting from an inability to maintain a particular position.
22. Understanding that there needs to be the opportunity (and equipment) for individuals to be able to change their positions at frequent intervals throughout the day; standing, sitting upright, reclining, prone, or lying sideways.
23. Understanding that neurological deterioration will affect basic functions such as temperature control, respiration, peristaltic action, digestion, heart rate and circulation.
24. Understanding the support technology, maintaining peripheral equipment, anticipating and resolving problems that might arise.
25. Working as part of a team.

Palliative care requires a special type of nursing from a special type of nurse; they need to be skilful, empathetic and resourceful, have a good understanding of the issues involved and be experienced in children's nursing.

Each day, at work, they are faced with supporting and caring for a very ill and vulnerable child who may be:-

- *Disabled.*
- *Experiencing neurological deterioration.*
- *Experiencing periods of considerable pain and discomfort.*
- *Experiencing physical changes to their body.*

- *Experiencing respiratory, circulatory, digestive, sensory and temperature-control problems as well as having difficulty with perception and cognition.*
- *Finding being moved and handled painful, disorientating and distressing.*
- *Having high levels of anxiety.*
- *Having communication problems.*
- *Having learning difficulties.*
- *Ill with a complex mix of unpleasant and interacting symptoms.*
- *Technology dependent.*

Even for trained and experienced professionals, caring for a child under these circumstances would be a daunting task but for parent-carers, new to the situation, the challenge would seem to be insuperable. For the care to be most effective, they must learn to work as part of a team, where each member recognises the expertise of the others. In addition, their role is different to the paid professionals; they are not at work where they might employ professional detachment, but are at home with their partner, their children and their totally dependent ill child. They are continually shifting from a position of parent and authority on their child to “colleague” and learner of new skills and knowledge. They have quickly to become very skilled or, by default, inadvertently cause distress to their child. Learning under these circumstances can sometimes be very difficult.

(4) Health and Safety

As the disorder develops the child may become unsteady and suffer visual problems which could lead to falls and impact injury. The majority of accidents can be avoided if the parents and the carers take precautions to ensure that the environment (both inside and outside the home) is safe. This requires constant vigilance and a high level of anticipatory planning. It may be necessary to have gates in a variety of locations about the house and also have bedsides long after the time when they would normally be used. *(See also Equipment: Health and Safety on page 35)*

In addition, when away from the home an awareness of where to seek medical help in the locality is not only reassuring but essential for the survival of the child if they suddenly become very ill.

Many parents also avoid activities which may temporarily incapacitate them, knowing that at any time they may be required to respond immediately to an emergency situation in a clear headed and decisive way.

Particular care should be taken when visiting unfamiliar buildings or public areas as these may not have been adapted and are likely to have unexpected hazards. Car parks and shops all present their own particular difficulties with suddenly opening car doors, reversing vehicles, projecting wing mirrors, shopping trolleys, or new stock stacked in the aisle, awaiting transfer to the supermarket’s shelves.

Day trips to recreational areas and holiday accommodation also requires careful pre-use checking and careful supervision as there is the additional factor of the unpredictable behaviour of third parties.

Clearly every care needs to be taken to protect the child from:-

- *Impact with sharp corners or projecting surfaces.*
- *Losing their balance on uneven or sloping floors.*
- *Tripping over objects left in their way.*
- *Falling from elevated areas.*
- *Heat sources.*
- *Hazards such as roads, rivers, ponds, moving machinery.*
- *Electric shock.*
- *Reversing or passing vehicles.*

Later, when the child loses their mobility, issues of lifting and handling come to the fore. Adequate training in correct lifting techniques and the safe use of slings and hoists is required.

(5) Medicines

As the disorder progresses and the child becomes increasingly unwell with a variety of unpleasant symptoms, the range of drugs administered can increase considerably. In addition, because the parents are the front line carers, it falls to them to record the date, time and dosage of every drug administered, monitor carefully their effectiveness, note any side effects and then accurately report back to the nurses and doctors. In order to do this they need to have an understanding of the basic issues concerned with medication use. This might include:-

- Antibiotics; common types, rotation, longer courses of treatment.
- Being aware of reaction and absorption times, side effects, interactions, adverse reactions, therapeutic range and (particularly in the later stages of the illness) drug tolerance and half-life times (so that a pattern of the child always being unwell at particular times between medications can be linked to the reduction over time in the effectiveness of the drug) and offsetting administering times (so that the effects of all the medications do not wear off at the same time, resulting in problems of symptom interpretation).
- Common abbreviations associated with medications and their administration.
- Dilutions (and its impact on the dosage).
- Drug forms (tablet, capsule, liquid, mixture, elixir, emulsion, syrup, cream, ointment, lotion, injection, suppository, drops, spray, inhaler).
- Ending or changing treatments.
- General classification of drugs. (Specific names, chemical similarities, biological effects, and later in the illness, legal classification).
- Long term monitoring.
- Methods of administering and training in the use and maintenance of any equipment used.
- Record keeping.
- Safe storage.
- Special risk groups.
- Understanding the intended effects.

Linked to the above is training (if required) in the:-

- Use of established methods of pain assessment and pain assessment record-charts.
- Use and interpretation of charts and graphs.

It is usual for the patient's records to be for professional use only. This means parents having to devise and use their own charts to record what may be up to twenty medication events in a twenty four hour period. Clearly when powerful drugs are being used it is essential that these records are accurate. What would be helpful would be to have a parent's section within the child's medical notes with a pro forma which has been developed by parent-carer groups.

In addition as parents are the key carers and are frequently required to report daily care, medication and symptom patterns to other carers and health staff, the use of charts (as opposed to written records) can greatly simplify the task.

For example, using the written epilepsy diary records as a source (which might also include descriptions of the different types of episodes), simple charts (frequency against type) can be plotted which can be most informative by revealing and highlighting patterns of incidence over periods of a day, a week or several months. [See Appendices]

The above raises the important issues of:-

- *How can parents obtain (or be given) the information they require?*
- *How can they receive the practical training they need?*
- *What agency might be responsible for organising that provision?*

(6) Mobility

When a child is beginning to lose their ability to walk independently, they will require support in order to continue to be ambulant. It is important that the carer who is assisting them supports them in a way that does not impede this process. The child should be supported on the body, under the arms or on the hips, but should not be expected to walk with their arms stretched above their heads, a position which may be convenient for the carer but only adds to the difficulties of walking for the child, thrusting them into an unnatural walking posture.

Held in the correct way, the child can focus on keeping their legs locked in an extended position and be assisted in flicking their legs forward in turn as they move, like a pair of dividers, across the floor.

When ascending stairs, the child can be supported on the body under the armpits and can be nudged at the back of the knee by the carer's knee, to encourage the leg to be bent, lifted and placed on the next level together with the child's body being gently rocked the opposite way to aid the movement of the leg.

(7) Nutrition

Maintaining a healthy diet is important even for healthy unaffected individuals; for children who are ill, it assumes a far greater significance. This fact however is set against a background of difficulties created by the symptoms of the condition which can adversely affect the nutrient intake. Some factors, in themselves, may seem minor but collectively they can have an enormous impact.

The difficulties may include:-

- An increasing lack of mobility.
- Diminished peristaltic action.
- Loss of fine motor skills and an increasing dependence on others for feeding.
- Loss of speech and therefore an increasing dependence on others for interpretation of needs.
- Swallowing difficulties and the need for more time to consume foods (initially) and then liquids.
- Aspiration problems and associated fear of eating and drinking.
- Poor liver function.
- Malabsorption and intolerance to certain food types.
- A general loss of energy and an increasing susceptibility to infection caused by the reduced calorie intake.
- Constipation and the need for medication.

Under these circumstances the input of a dietician is clearly most helpful but there is also a need for parent carers to refresh and update their knowledge of food groups and the contribution of each to the health of the individual, particularly once the diet becomes restricted through the diminution of the swallow reflex. Gastrostomy feeding will overcome some of these difficulties but it is important to ensure that there is adequate fibre in the feed.

A balanced diet is one which supplies all the necessary nutrients in quantities that suit the particular individual. This includes the correct calorie intake and the right proportion of different foods. A basic knowledge of nutrition and the contribution to health made by each food group would be helpful. This would include proteins, carbohydrates, fats, fibre, vitamins, minerals and liquids, particularly water.

Despite all the difficulties listed above, it should not be forgotten that in addition to a correct diet, exercise and movement are also important in that they help the child maintain a range of basic body functions which are needed for the efficient processing of food.

Section 4. Care of the Ill Child

**This section has been divided into four parts.
Each part is then further subdivided.**

- **The Carer**
- **The Child**
- **Interactions (between the carer and the child)**
- **Symptom Control**

Part one: The Carer

(1) Respecting the child as an individual

The child must be seen as a person who happens to be ill and not as an illness that happens to involve a child. Even though the disorder will result in a loss of some abilities, the child as a person remains the same: their personality and personal qualities; their interests, likes and dislikes; their strengths and weaknesses; their love for life and those about them; they do not change. It is still the same person underneath the cloak of illness.

It is important that the intellectual abilities of individuals with degenerative disorders are respected. They have not always been as they are now and any deterioration is likely to be selective and differential. Although they may have difficulties with outward communication, they are often fully aware of the communication of others and will still be using many interpretive skills, for example noting facial expression, intonation, attitude and manner, body language, posture and position in space relative to the listener.

Belief in the child reinforces their self perception of being important, respected, trusted and held in high esteem. The child will see from the actions of those about them that they are valued as individuals. In addition, if the assumption is made that the child does have an understanding of what is being said to them and an awareness of what is happening around them, then this will ensure a constant flow of sensory information which will engage and stimulate them.

*“Awareness assumed is awareness maintained”
(2004 Blackwell Publishing: D. and S. Wray)*

(2) Anticipating the Child’s Needs

A key factor in the care of very ill children is the ability to anticipate the child’s needs, often before they become evident to a casual observer. This works in tandem with good organisational skills as proactive planning is essential for effective care.

The carer needs to take every opportunity to help the child understand the apparently changing world about them. As the condition advances and their perceptions are either dulled or subjected to distortions, the individual can become very anxious and will require reassurance as to their safety.

A good carer will also have an empathetic relationship with the individual and check regularly whether or not:-

- *A change of bib, clothing or pads is needed.*
- *There is discomfort. Would a change of position help?*
- *Medication is due.*
- *A drink, food, or moistening of the mouth is required.*
- *The child needs reassurance, companionship or a cuddle.*

Part 2: The Child

(1) The Perspective of the Ill Child

It is helpful to see the world from the child's perspective of the bed or the wheelchair. For any ill or disabled child, they are at a lower level to those around them and normal social contact is based on an assumption of similar face levels. The children are slightly distanced by the wheelchair and certainly by the bed and this affects interpersonal spacing. When the child is in hospital, the situation can be made worse by bed-sides and lockers or equipment being positioned at the side of the bed.

Although the above points may seem obvious, the everyday experience of many parents and children is that it is not commonly recognised.

(2004 Blackwell Publishing: D. and S. Wray)

(2) Seeing the world from the bed

Consideration should be given to the issue of how to approach a child with a visual disability who is very ill in bed and is immobile and therefore unable to turn their head to see who has entered the room. *(See Part 4. Number 5: Impaired vision)*

It is important that the carers quietly announce their arrival, make a slow approach and go round the bed to enter the child's field of view. Initially, until the child is comfortable with the visitor, they should not lean over or dominate the bed. If physical contact is to be made, then permission should be sought before approaching and touching the patient. Wearing the same perfume aids identification but strong aromatic perfumes should be avoided if the patient is asthmatic.

The carer needs to be careful with their tone of voice as the patient is using it as a cue to assess the carer's attitude and intentions. The child may not be able to see the carer's face and smile; it is important therefore that they feel the smile through the carer's hands and voice. Any strong negative emotion, such as anger with another

person, should be left outside the care area; otherwise the patient may misinterpret the reasons and be unnecessarily frightened.

The position of the visitor's face (and also of objects to be viewed) relative to the patient's field of view is very important. It is helpful establish their near point and remember that their accommodation abilities may be impaired. The carer may have to turn their head so that the vertical axis of their face is in line with the patient's (who is lying down). They may have to also position their face closer than is usual in order to be within the focal range of the viewer. This clearly also has implications for the positioning of a television, or books which are being read and shown and even of the bed relative to doors and windows. If the patient is confined indoors, views through windows can be most refreshing and ways of achieving this should be considered.

The location of light sources, whether natural or artificial, also needs to be considered. Glare from overhead lights needs to be avoided; lights controlled by dimmers and placed behind the patient are much better. It also avoids visitors being backlight, so that their faces cannot be seen. Sunlight, although very desirable, can be very bright and may be either dazzling or damaging, particularly if the patient is unconscious but with their eyes open.

It should not be forgotten that their bed is their territory; in it they should feel secure; this security should not be destroyed; they have nowhere else to go or hide.

(3) "Slow down a little please."

Degenerative conditions, particularly as they progress into later stages, can impair some cognitive and perceptual processes and there is an increased need for the individual to be given time to absorb information and accommodate to new circumstances; thus it is helpful to give lots of preparatory warnings of some anticipated event and maintain a commentary to explain the change in circumstances.

(2004 Blackwell Publishing: D. and S. Wray)

Rapid rates of change are difficult for them as it may outpace their ability to follow what is happening:-

"Please lift and transfer me slowly."

or

"Your steady walk, pushing me down the corridor, may be far too fast for me and become a terrifying journey."

or

"I cannot to eat and drink quickly.

My swallow function may be affected and I could choke."*

*(*PLMD Ethos. Leeds LEA)*

Part 3: Interactions

(1) Communicating through the carer's hands

Some indication of the child's physical state can also be obtained by the carer placing their hand on the side of the child's chest, under the armpit: This allows a number of variables to be quite sensitively monitored, such as:-

- Depth, regularity and pace of breathing.
- The degree of effort involved in the above and an indication of its effectiveness.
- Any "sponginess" in the lungs together with any bubbling sensations.
- Any muscle spasms and stiffness in the chest wall.
- Any temperature variation from the norm with associated sweating or clamminess.

This can also be linked to visual and auditory observations of:-

- Wheezing and general respiratory noise.
- Grunting or distressed noises.
- Irregular or unusual patterns of breathing.
- Unusual postures.
- The degree and nature of eye movements and level of eye contact.
- The facial indicators referred to in Part 4: Number 7. Physiological Norms.

If this monitoring position, together with holding the child's hand, is maintained by the parent over a period, it also has the benefit of making the child feel secure and in contact with the supporting adult. It is possible, after a while, for the carer to feel "at one" with the child, even unconsciously adopting a similar rate and depth of breathing.

In addition, holding the child's hand can give reassurance and allow back and forth acknowledgement, usually by a slight movement of the fingers or a gentle increase in pressure, linked to speech, facial expression and gesture.

Some parents have also found that gently placing their hand on the area of discomfort can have a therapeutic affect. It is reassuring to the child and the gentle warmth and slight pressure often helps to reduce and sometimes relieve the pain or tenderness. Under some circumstances, just holding a cupped hand over the area (but not in contact) can produce relief.

Although the links are to some degree limited, they provide a means through which the child and carer can communicate and interact. This will help the ill child maintain a positive outlook and help to reduce any feelings of isolation. These children do communicate and are often shouting silently at the world. Those about them need to be sensitive to this and have the skills to "hear" what they are saying.

(2004 Blackwell Publishing: D. and S. Wray)

(2) Massage and physiotherapy

Many children benefit from massage and physiotherapy. A distinction might be made between physiotherapy intended to maintain limb mobility and muscle tone (which could involve games and an element of fun) and massage, which may be intended to induce relaxation and could be done in conjunction with other strategies, such as playing background mood music. Both have an additional “communication value” in that the physical contact reassures the child and makes them feel noticed and valued.

Care should be taken over the use of oils as some are not advised for children affected by epilepsy, asthma and muscle spasms. For example, nut oil can induce severe and painful muscle reactions. Body powders and baby lotion are sometimes used as options.

Foot massage can also be helpful, but measures must be taken to ensure it does not induce muscle spasms.

Part 4: Symptom Control

(1) Aphasia

Parents need to be aware that there may be a link between certain aspects of their child’s behaviour and their aphasia.

The effects on a child’s behaviour of either speech loss or a lack of speech development may not be noted immediately by the parents as the changes may be subtle, occur over a period of time and may be coincident with other changes.

Children with aphasia often exhibit higher levels of anxiety than would otherwise be the case. Their difficulty with verbal communication means that they cannot seek clarification about puzzling aspects of their surroundings or the intentions of those within it. They are therefore far more cautious and easily unsettled by unexpected change.

The parents can reduce the levels of anxiety by adopting strategies which make the child feel more secure; keeping them constantly informed and updated about their surroundings; warning them of any impending change; explaining who people are, why they are there and what they are going to do.

Clearly this raises other issues such as:-

- *The child’s degree of social contact with family, friends and peer group and the nature of the interactions in each case.*
- *Learning other communication methods (and the associated problems for children with degenerative disorders.)*
- *Language as a means of influencing their immediate environment.*

- *Language as a vehicle for personal expression and indicator of personality.*
- *Where to place the emphasis; on the sender or the recipient of the information (and the nature of the skills that are involved in each case).*
- *The need for the parents to be communication sensitive.*

(2) Controlling body and room temperatures

In the final stages of the illness the child's temperature control mechanisms may not function correctly. This could result in either:-

- Their body temperature being higher or lower than usual given that the room temperature is within normal limits.
- Their physiological responses being unable to cope with room temperatures higher or lower than normal (because of seasonal factors).

Body temperature variations require strategies to be employed either to cool or warm the child as needed. It is important that both measures are approached with caution and the process is not rushed, as differentials between peripheral and core temperatures can develop and produce erroneous readings. Also, over-heating or over-cooling can in themselves produce further problems.

This would involve the careful and monitored use of either:-

- *Fans and/or sponging with tepid water, together with appropriate medications for cooling.*
- *The careful use of hot water bottles, wheat bags or therapeutic electric blankets for warming.*

Room temperature variations require either appropriate room heating or the use of air conditioning. In the United Kingdom the usual requirement for maintaining a reasonable room temperature is to put heat into a room; in extremely hot summer weather, commonsense suggests that in order to maintain a comfortable room temperature (and avoid the child becoming distressed) excessive heat should be taken out of the room.

Portable air conditioning, although regarded as unconventional at the moment, is an obvious way of ensuring the comfort of the very ill child in high, ambient temperatures by maintaining a steady and comfortable air temperature in the room.

(3) Epilepsy

Once epilepsy is diagnosed, the parent carers may be given some general guidelines about positioning, maintaining airways and medication. They may also be given a diary and asked to maintain a record of the frequency and type of episode. Parents and carers can however go beyond this quite passive role and be proactively supportive. An important principle is to allay any fears the child may have.

There can be a “halo” effect as a precursor to an attack. Some parents have found that they are able to recognise the symptoms and often avoid the episode developing by repeating reassuring and calming statements which are familiar to the child (because they have been rehearsed previously).

If the episode does develop further, it is always best to assume that child can hear what is being said, even though they appear to be unconscious. Again, the use of previously rehearsed, reassuring and explanatory statements can be most helpful.

Once the child has entered the sleep phase, some parents have found that it is helpful to stay with the child, perhaps holding their hand and quietly talking to them or reading a familiar story.

If a diary is being kept, then useful information can be gained from the data. Simple bar charts can reveal patterns relating to either the time of day, the day of the week or weeks within the month. For example, one family found that over a six month period, during term time, least episodes occurred on a Sunday but most on a Monday, the number gradually reducing as the week progressed. (See Appendices). This seemed to be related to stress levels within the family connected to work and school. Once aware of this, the family were able to initiate measures to counter the problem.

(4) Impaired vision

One of the difficulties which may face children in the later stages of a degenerative disorder is impaired vision. Increasingly poor coordination can result in double vision, problems with tracking moving objects and difficulties in focusing or refocusing quickly on a point in space. This may be superimposed on an already present visual problem which is symptomatic of the disease. This can result in blurred, unidentifiable figures or faces, suddenly appearing from nowhere within the child's field of view and then, equally quickly, disappearing without warning. For a child whose world is already limited by restricted head movement and general loss of mobility, losing the ability to scan and assess the immediate environment can be very frightening.

If the carers are aware of these difficulties then they can compensate by giving clues to their identity and their intended activities by allowing the child to use their other senses. If they all follow the same few guide-lines it will make the child's life a little more understandable and make them feel more secure and comfortable.

The Leeds Visually Impaired Service suggests that the guide-lines might include:-

- *Greeting the child by name and the carers saying who they are.*
- *Giving explanations and maintaining a commentary.*
- *Making sure their face is within the child's field of view when talking to them.*
- *Allowing the child to touch and smell a product before it is used, in order to reassure them.*
- *Consistent use of perfumes to aid identification.*

- *Using a touch, stroke or gentle word as an addition to a smile, in case it cannot be seen.*

Care also needs to be taken over the positioning and brightness of lighting. Difficulties can be caused by glare from ceiling lights or by bright sunlight coming through windows. Consideration needs to be given to the relative positions of the bed and the windows to avoid the carers being back-lit. It is better to have artificial light sources, controlled by dimmers, positioned behind the child.

(5) Less common forms of infection.

As the illnesses progress, the children may become susceptible to some of the less common forms of infection, (for example clostridium difficile). It would be helpful if the carers could be given a brief description of the nature of the infection, an explanation of why it is not normally present in a healthy population, the symptoms, their causes, the impact they have on the patient, the nature of any treatment, the ways in which the infections are communicated and strategies for avoiding infection.

(6) Pain control

There are a range of issues with regard to pain relief for children. It may well be that it is an area which would benefit from further research. This might cover:-

- The appropriateness of the medications available.
- The nature of delivery.
- The speed of acting.
- The effectiveness of the medication.
- The duration of the effect.
- The side effects.

Parents can find that pain and its control is a major issue for their ill child. It is an "invisible enemy" in that its effects are not displayed as dramatically as other symptoms and yet its impact is significant on a child already ill with other problems. Towards the end of the child's illness, as medication levels steadily increase, the whole process of judging pain levels and ensuring adequate medication can be emotionally very draining.

(2004 Blackwell Publishing: D. and S. Wray)

(7) Physiological norm

It is helpful to be aware of the child's usual temperature range and their usual pulse and breathing rates. A variation from the norm can often give an early indication of illness or impending problems even before child actually appears to be ill. It can also provide a useful datum for doctors who are unfamiliar with the child and are endeavouring to do an assessment in an emergency situation.

Other physiological signs include depth of breathing, degree of muscle stiffness, amount of eye contact, facial expression, body position, changes in the relative geometry of the lips and eyebrows and the “feel” of the skin (clammy, cool, damp, etc.)

A useful strategy, which adds the additional sense of taste and allows the parents to easily and quickly monitor variations in the child’s temperature (and also the other aspects of the child’s condition noted above), is to place their lips on child’s forehead. Clearly this does not replace conventional methods but is helpful as an additional, quick check.

(8) Throat spasms

This is a very frightening and potentially life-threatening symptom where the muscles of the throat go into spasm and close the airways making breathing, already difficult with weakened chest muscles, almost impossible. The child is in effect being asphyxiated, often turning blue.

Relief can be achieved by:-

- *Rolling the child onto their side, so that they face the parent.*
- *Holding the child’s head in both hands and using the first or second finger to gently pull the jaw forward to help clear the airways.*
- *Tilting the chin upwards to extend and open the airways.*

It is also helpful to bring the child the edge of the bed and lower the head section so that the child is tilted slightly downwards to assist in the drainage of secretions. This position will need to be maintained for the duration of the spasm which might last from five up to ten minutes.

If a reassuring commentary can be maintained throughout the episode then it will encourage the child to relax. For children with vision problems it is helpful if the parent ensures that their face is in a position where they can easily be seen by the child.

Section 5. Service Provision

This section has been divided into four parts:-

- Accessing services
- Accommodation
- Agency nurses for overnight nursing care
- Key worker
- Suitable services

Part one: Accessing services

Finding out what services are available, who provides them, who does what in each organisation, then obtaining and maintaining them is in itself an extremely difficult and time consuming exercise. Clearly the use of a dedicated Key worker alleviates this problem, but for the parent who has to act on their own, a guide as to how to set about the task would be very helpful. *(See Key worker on page 29).*

Parent carers acting on their own eventually realise that it is necessary for them to develop ways to overcome avoidance and delaying strategies. Using more than one information source is helpful in countering inaccurate, incomplete, inappropriate or misleading information. Identifying hidden agendas is always useful.

It is necessary in these circumstances to develop and use research skills and helpful to maintain a “library” of reference materials relating to services, equipment, standards, criteria, trials, surveys, reports and background information. Some materials are only available in the United States because of an apparent culture of secrecy in this country.

When services are provided, the parent acting on their own will have to liaise, coordinate, organise, monitor and (to varying degrees) manage the disparate individuals to obtain maximum benefit from their input.

Despite the expenditure of a large amount of time, effort and money on this activity, many parents feel that they still do not receive the overall quality of service that they might expect under the circumstances. This begs the question as to what would have happened if they had not been so proactive?

Part two: Accommodation

There may be an assumption that housing adaptations are overseen, free of charge, by the Local Council and Social Services. For private house owners this is often not the case as council's focus is on its own tenants living in rented council properties. Carers who are home owners are likely to face the complications of house adaptations on

their own, often starting from a position of minimal knowledge. Professionals who would normally give advice under contract are reluctant to do so privately because of the fear of retrospective litigation. It is therefore difficult for the private individual to get advice without incurring fees. A handbook would be helpful giving general guidance on how to obtain:-

- Specifications for the design of bedrooms, day areas and bathrooms for the ill or disabled.
- Information about the insulating properties of materials to be used.
- Information and specifications on heating and air-conditioning systems and their controls.
- Guidance on how to do architect's drawings for housing adaptations.
- Guidance on how to apply for planning permission.
- Guidance on how to apply for VAT exemption.

Part three: Agency nurses for overnight nursing care

Parents need to develop a range of skills in order to cope with the problems created by the use of agency nursing personnel. These include:-

- Acquiring interviewing skills so that they can properly assess the personality and abilities of the nurse who is coming to look after their very ill and vulnerable child overnight.
- Be able to plan for the preliminary visit and have ready all the necessary materials; i.e. leaflets, guidance notes on symptoms and care issues, the family support group video (which explains the disorder and the associated symptoms), home videos of the child when younger (as in the case of a child with a deteriorating condition) and other information, such as the epileptic diary or medication records
- Have the ability to explain how to deal with complex range of unusual symptoms.
- Have an understanding of the requirements of a nurse caring for a very ill child overnight in a family's home and perhaps have a basic list of dos and don'ts.
- Be assertive but polite with care professionals who are either reluctant to make a preliminary visit before they care for the child overnight (on the basis that they are skilled and all ill children are very much the same) or choose to ignore the parent's knowledge, skills and experience of caring for the child because they are the professionals and they believe that they know better.
- Be open minded, adaptable and willing to learn from other carers when they see relevant good practice being used.
- Be able to train the agency nurses in the use of the domestic variants of the "hospital" support equipment.
- Have the ability to share their home and everyday domestic life, day and night, with care staff, some of whom may initially be strangers to the family.
- Learn how to devise coping strategies to overcome the unsettling effect this has on all the family, particularly younger children.
- Acquire the skill of being assertive but polite with both the agency managers and individual nursing staff when they fail to deliver the agreed standard of service.

Initially, it might appear to be entirely beneficial to the parents to have a qualified nurse stay overnight in the home to look after the sick child. However, certain circumstances can cause it to be an additional complication which can negate the apparent benefits for the parent carers.

If the community paediatric nursing team is large enough to provide overnight nursing care for the child then this is an advantage to the parents as it removes from them the burden of ensuring that the individual coming into their home will meet the needs of the family.

If the overnight care personnel are provided by a nursing agency then this places an additional responsibility on the parent carers because, contrary to what might be expected, experience has shown that each individual needs to be screened by the parents; their personality, temperament, bedside manner, level of empathy with children, communication skills, training and recent nursing experience all need to be reviewed if they are to become a trusted member of the team caring for the child overnight, by themselves and for long periods. The parents have therefore to do what a prospective employer might do in order to check the applicant's suitability for the post and "interview" the applicant during the preliminary visit. In addition, because of the casual nature of their employment, there is likely to be a steady change of personnel, so the interviewing is on-going. This is time-consuming and is at odds with the need to devote time to caring for the ill child and the rest of the family. The agency, which has minimal contact with the parents, simplistically refers to this as "allowing the parents to see if the nurses are going to be alright for them".

The main disadvantage of the agency system is that the nurses operate as detached individuals providing casual supply. As a consequence there are no group protocols or codes of practice, no verbal exchanges of information (as in a ward hand-over), no peer group monitoring or evaluation, no training and no career structure. In addition, as they are on duty on different days, they never meet and are therefore professional strangers to one another. In effect, the parents have to assume the role of ward sister and adopt a supervisory/managerial role to oversee the situation.

For the nurse as an individual professional, responsible for the welfare of the child in their care, there are important issues of support and training that need to be addressed because they impact on the family and the ill child. There is the whole notion of having to work autonomously and independently, overnight, in the ill child's home as opposed to working on a ward with its facilities and additional staff to whom any questions can readily be referred and emotional support obtained.

Palliative care is not a "normal" nursing job as in looking after an otherwise healthy child who will recover from their illness, lead a generally full and active life and eventually be able to make a contribution to society. Care of the dying requires a change of perspective and a change of emphasis because of the eventual outcome of the illness. Caring for a terminally ill child raises many issues about the skills, experience, attitudes, values and beliefs of the staff that are involved.

A further difficulty is that because some nurses have no or only limited experience of

terminal, paediatric or domiciliary care, they have to be briefed and trained up on the job by the parents. Although this may seem surprising, it is a consequence of the limited, mix-and-match role of the agency that employs the nursing staff. For example, when caring for technology dependant children at home, many agency nurses have either not seen or are unfamiliar with the use of the domestic variants of the equipment they may have used in hospital. This includes oxygen concentrators, oxygen cylinders and their flow regulators, back-pack feed pumps and aspirators.

In addition, they are also not familiar with the maintenance of the equipment, such as changing the oxygen cylinders and their control heads, because they have previously used either piped oxygen or technicians have set up the equipment. Also, they may have to be taught the strategies for overcoming everyday problems such as jammed gastrostomy feed-line connectors, blocked feed lines or temperamental feed pumps.

Many supply staff would benefit from a handbook of dos and don'ts, which would be a quick guide to working overnight in the home of the ill child and their family, rather than individuals having to discover what is required through trial and error. This would also be helpful to parents.

Caring for a child who is dying is emotionally very draining and this raises the issue of support for the professionals who are involved. It may be an unrealistic expectation for the agency to assume that their nursing staff can cope without the support of colleagues and the opportunity to have counselling.

There is also the general issue for parents of learning to cope with having additional people in the family home (particularly overnight) and what strategies to adopt to overcome the unsettling effect this has on the whole family, particularly the younger children.

Part four: Key worker

A way of assisting the parent carers in the organisation of resources is to use a Key worker. Their support makes an enormous difference to the families with whom they work. The experience of many families, from the time of diagnosis, is that life is entirely different once they are able to benefit from the expertise and advocacy skills of a specialist key worker.

(2004 Blackwell Publishing: D. and S. Wray)

Part five: Suitable services

An unexpected problem many parents face is the absence of suitable services for their ill child. It seems as though the more ill the child becomes the less appropriate are the services that are provided. The basic difficulty seems to be a lack of awareness that a different set of "rules" apply in the case of children who have degenerative disorders.

Parent carers often find that:-

- *The service providers do not recognise the complexity of the symptoms. Frequently the decline is perceived as linear, with only one or two major, static and controllable symptoms.*
- *It does not seem to be appreciated that delays result in the service not being available when it is needed and when it is eventually provided it is inappropriate and cannot be used.*
- *The service provision is reactive. What would be better would be to have a pro-active service provision, where the diagnosis of the disorder triggers a planned response.*
- *There appears to be a commonly held belief that provision aimed at the average individual in any service category will result in a trickle-down effect to cater for those with the extreme service needs*

*“ It should be recognised that children with degenerative disorders are a group of exceptional children, in exceptional circumstances, with exceptional needs, who require exceptional resources immediately, because later is too late; their condition may have worsened or they may be dead by the time provision is made.” **

*[*NPD Family Support Group]*

Section 6. Equipment

This section has been divided into five parts:-

- **Beds**
- **Buggies, wheelchairs and ATP**
- **Child car seats**
- **Monitoring equipment**
- **Oxygen equipment**

Part 1: Beds

When reviewing specialised equipment, such as profiling beds, parents need to know how to obtain:-

- Information about manufacturers, specifications, designs, costs, suppliers and delivery times.
- Guidance from local organisations such as disability centres so that they can review the functional differences and assess the appropriateness to their own situation.
- Information about the range of ancillary equipment, such as bed-sides or air-mattresses, noting types, advantages and any potential problems in their use.
- Information about appropriate bedding, including impregnated and plastic anti-allergy mattress covers, changing and anti-slide sheets.
- Guidance on how to use to best affect the profiling characteristics of the bed for breathing difficulties, feeding, physiotherapy, avoiding reflux and assisting with drainage.

Part 2: Buggies, wheelchairs and ATP.

Parents need to be able to access information sources which allow them to check the specifications of wheelchairs, buggies and all terrain pushchairs (ATP). In this way they can obtain or be provided with a buggy or wheelchair which meets their requirements, matches the nature of their lifestyle and maintains the family's quality of life.

It may be that a family actually requires two buggies; one for domestic, school, shopping and street use and an all terrain pushchair that enables the whole family to go out walking either on day visits or longer holidays. **The fact that a child is disabled should not be a constraint on family leisure activities; the equipment should maintain or even increase opportunities, not reduce them.**

When researching wheelchairs and buggies, parents may wish to consider the following;

- **Weight.** *This is an important consideration when lifting or moving the folded equipment in or out of either public or private transport or when pushing it with an occupant over sloping ground.)*
- **Overall dimensions** *(both when in use and when folded). Some are relatively more compact when folded and will fit easily into the boot of a saloon car and still leave room for other luggage. Thought should also be given to their use in domestic premises and non adapted public buildings.*
- **Method of folding.** *Some are easier to fold (or open) than others and require one hand only. Consideration should be given to how many hands (and feet) are required for this operation, bearing in mind that the parent will also be looking after the ill child and possibly siblings as well.*
- **Posture and support.** *Will it recline for sleeping? What is the nature of the foot support, does it expose the feet to injury and will it allow the weight of the legs to be taken off the back of the thighs? Are the leg positions comfortable and how much splay is there? Can the user sit up independently, thus aiding independence? What degree of lateral support is there for head and trunk? Is the head support adjustable? What support is there for the arms and do the elbows project exposing the arms to injury in confined or busy places? Are the lateral and longitudinal profiles of the seat components correct? Are they sufficiently padded? Are the covers easily cleaned?*
- **Wheel size and types of tyres** *[solid or pneumatic]. Larger wheels give a more comfortable ride over rough ground, but there has to be a compromise because of the folded down size of the equipment. As long as they are not run too hard, pneumatic tyres give a more comfortable ride but thought should be given to the need to carry a repair kit so as to be able to repair a puncture.*
- **Fixed or jockeying front wheels.** *There are advantages and disadvantages to both arrangements depending on the circumstances of use. A system which allows the wheels to be either locked in a fixed position or free to turn is helpful.*
- **Centre of gravity.** *The point of balance needs to be near the rear wheels to allow the front of the buggy to be raised easily to access raised levels such as kerbs or low steps.*
- **Brakes.** *This covers both parking and service brakes, the latter being used when descending slopes or when wishing to reduce the speed of the buggy to bring it to a halt.*
- **Break-away straps.** *When carers with wheelchairs or buggies are negotiating slippery or uneven slopes there is an often unrecognised risk of the carer stumbling or slipping thereby losing control of the equipment with potentially disastrous consequences for the disabled occupant. This risk can be reduced by the use of a break-away strap which has one end fastened to the buggy and the other looped round the wrist of the carer. If there is a risk of the carer being towed along the ground by the wheelchair then this would suggest that the carer to wheelchair weight ratio is not correct.*
- **All weather protection.** *If the family and the ill child are to maintain normal everyday activities, then the buggy must be equipped with a means of protecting the occupant from a range of weather conditions. These might*

include an all enclosing fold-down rain cover, some means of shading the occupant from the sun and a thermal insulating layer which encloses the individual and does not just lie over the top but provides protection to the back of the body and legs and as well as feet, arms and hands. To be fully effective, the insulating layer should be windproof; if it is not then wind and waterproof nylon covers are available.

Part 3: Child car seats.

Parent carers need to be aware that child car seats, as sold, may not be appropriate for ill or disabled children. Even those that are specifically designed with this group of children in mind may not be entirely adequate.

- *Child car seats tend to have minimal padding, thicker padding usually being available only as a special order option. This means that they can quickly become very uncomfortable, particularly if a lack of mobility means that the child is not easily able to change their position. This could be further complicated by the child being susceptible to bruising and injury.*
- *The bottom part of seat can be too short and may have a raised ridge at the front which applies pressure to the back of the leg above the knee, restricting the circulation to the lower leg. This can be compounded further by the child's lower leg being unsupported. (Even healthy children may complain about "pins and needles" on long journeys.) This can be overcome by a careful choice of seat and also by supporting the child's feet so that they are not dangling in space with all the weight being taken on the back of the lower thigh.*
- *A full harness on the child's car seat is clearly preferable.*
- *It is important to ensure that the child's head is fully supported. (See next two comments).*
- *It is helpful if the angle of the child's seat can be changed slightly, allowing the child to sleep comfortably without their head falling forward, which puts undo strain on the spinal joints and neck muscles.*
- *Road surface irregularities, particularly traffic calming measures, present a particular problem. If they are in the immediate locality of the home then they may have to be negotiated frequently. This presents a hazard to children with poor muscle tone as they are vulnerable to injury from the car passing over either road "cushions" (raised rectangular areas generally the width of a car) or humps (that go all the way across the road). The pitching or rolling movement can be quite severe even at low speeds.*
- ***N.B.** There is also an important but largely overlooked issue regarding the*

emergency transfer of very ill (including terminally ill) children over these obstacles and the impact on the very fragile child of the repeated rolling and pitching movements. Also, their presence (particularly the very severe ramps right across the road) has implications for the response times of ambulances.

Part 4: Monitoring equipment

There are circumstances under which monitoring equipment can be helpful and reassuring to parents and carers, particularly when the child is very ill and their vital signs are relatively difficult to detect.

For example, the apnoea monitor warns carers of an extended break in the normal, regular rhythm of breathing and is useful overnight when the child is asleep. However it is also sensitive enough to pick-up very shallow breathing and this can be helpful for parent carers in circumstances such as gastroesophageal reflux “fainting” episodes or periods of unconsciousness, associated with epilepsy, when breathing (and pulse) can become so light as to be not readily detectable.

Part 5: Oxygen

Most lay persons are unfamiliar with the use, side effects, dosage, storage and nature of the main and ancillary equipment associated with oxygen therapy.

Any training should address the following issues:-

Oxygen Concentrators.

- How they work.
- Their size and weight.
- Positioning within the home environment (obstruction problems; power sockets; power leads and oxygen tubing runs; the need for a clean and cool air supply).
- The advantages (convenience of use, when installed and functioning correctly).
- The disadvantages (may be leased equipment and therefore old, unreliable and poorly maintained; may be contaminated with strong odours from previous user’s life-style; require regular monitoring and maintenance of filters; low flow rates and low output pressure, of consequence if bubble humidifier or long tubing runs are used; oxygen content drops as flow increases; deliver warm and therefore very dry oxygen, an irritant to asthma sufferers; constant background noise; patient at risk in the event of equipment failure or power cuts).

Oxygen Cylinders.

- Different sizes and different valves.

- Different types of flow head (variable flow type better than fixed rate of flow; can be adjusted to suit emergency situations or increased to compensate for decrease in cylinder pressure; flow rate can be easily read on graduated tube; gives better indication of cylinder running out).
- Advantages. (Higher pressure and higher rates of flow; pure oxygen as opposed to gas mixture from concentrator; gas expanding therefore cooler than compressed gas from concentrator).
- Disadvantages. (Size, shape and weight make moving them difficult in a confined space or up stairs; require collection and delivery therefore have to be handled to and from house; need to be stored away from house; need moving to and from care area as required: can be faulty, either empty when delivered or have faulty screw threads or seals so that they leak)
- Storage (space required to store several week's supply, separated into full and empty cylinder areas).
- Safety Measures (The need for caution when changing the flow heads; the advisability of informing the fire service that compressed accelerant gas is stored on the premises).

Ancillary equipment.

- Oxygen masks and nasal prongs (advantages and disadvantages of each).
- Types of oxygen mask.
- Care and maintenance of prongs and masks.

Health and safety.

- Side effects of oxygen on the patient (breathing patterns, soft tissues, asthma).
- Fire hazards:-
 1. Oxygen is an accelerant and this should be borne in mind when changing cylinders.
 2. It is also stored in pressurised containers and the fire service should be told that these are stored on the premises.

The fire service will do a free survey of the house and give safety advice as well as fit free smoke alarms; one in the bedroom where the equipment is, one on the landing and one downstairs, to give plenty of warning to facilitate evacuation. They will also familiarise themselves with the layout of the house so that they can quickly remove the sick child in the event of an emergency.

Section 7. Quality of Life

**The parts in this section are not in alphabetical order;
they have been arranged sequentially.
Some of the parts are subdivided for clarity;
this has also been done sequentially.**

- **The Environment**
- **Education**
- **Play**
- **Story Telling**
- **Music**

Part one: The Environment

The general aim is to provide a secure, happy, stable and stimulating environment in which the child can be exposed to a range of experiences, some intended to relax and others intended to excite. Overlaid on this, is the individual's physical state at any given time, which will to a large extent, control the nature of any activity.

A trained and experienced professional, with their knowledge, experience and skill can make an important contribution to maintaining the quality of a child's life and ensuring that the later periods are filled with interest, excitement and happiness. Good communication between the carer and the ill child is the key to achieving this objective.

1. The surroundings

The surroundings need to be quiet, warm and well lit, with plenty of natural daylight. It is helpful if the room is visually stimulating with some of the displays (or effects) being "active", either through auditory, visual or tactile change or by changes in orientation or location. The ceiling could well be a focus for objects and pictures; luminous planets, brightly coloured fabric shapes which move in a draught, either from an open window or a fan. Also wind chimes, moving lights on the ceiling and walls or reflective, reciprocating and rotating devices will all catch the child's attention. *(See Music Therapy on Page 43 and also Home Tuition on Page 39)*

2. Contact with other people

Contact with and knowledge of other people will positively add to the life of the child; just having a friendly and enthusiastic adult in the room will immediately catch their attention; children will see things with fresh eyes and this new experience will add an extra dimension of excitement and interest, even to previously rehearsed activities.

(See Home Tuition on page 39)

The carer can function as an “interpreter” and mediate between the child and their surroundings. A steady conversation to accompany everyday activities that the child finds difficult, for example drinking, can be worthwhile if it assists the child and offers an opportunity for praise. It will serve to make the child feel rewarded, valued and the centre of attention. This can also apply to the wider range of less routine activities that result from the carer’s input.

3. Being noticed

It is important that opportunities are created for individuals to be the centre of attention for a while. Being noticed will enhance their self image and greatly improve their feeling of well being, an important factor in their personal battle against the symptoms of a life-limiting condition. This can also be associated with positive feelings of fun, happiness, excitement and being someone special.

Although there is a general need for a quiet, structured atmosphere, there is an equal requirement for changes of pace, style, location and activity, in order to provide stimulation and excitement. Often, pain and discomfort can be forgotten for a while if there is an interesting distraction.

*“I need the opportunity to be occupied”**
(*PMLD Ethos. Leeds LEA)

Many children with deteriorating conditions enjoy watching television, possibly because it provides a vicarious experience of real life, thereby overcoming the constraints imposed upon them by their condition. A child watching a programme involving other children is given the contact and experience sadly so often denied them in normal social situations because of the caution exercised by their peer group.

4. Maintaining contact

Degenerative conditions can impair some cognitive and perceptual processes. There is an increased need for the individual to be given time to absorb information and accommodate to new circumstances. It is helpful to give lots of preparatory warnings of some anticipated event and maintain a commentary to explain the change in circumstances.
(2004 Blackwell Publishing: D. and S. Wray)

As the condition advances, there is the potential for the child to become trapped inside an uncooperative mind and body. This raises an issue of emphasis. Where should the focus be; on the child who cannot speak or the listener who may not understand? It might initially appear that children who lose their ability to speak should be taught other recognised methods of communicating. However they may have impaired cognitive and motor skills which may well preclude this.

(2004 Blackwell Publishing: D. and S. Wray)

The emphasis on the transmission of information overlooks the role and the skills of the receiver. For the perceptive carer, there are a host of cues which can give some indication of the child's physical, mental and emotional state and even of their thoughts and wishes. General indicators are posture, verbalisation and facial expression. More specific, body language indicators include gesture, positioning of head and limbs, degree of eye contact, muscle tenseness and the movement of eyes, eyebrows and lips.

In addition, holding the child's hand can give reassurance and allow back and forth acknowledgement, usually by a slight movement of the fingers or a gentle increase in pressure, linked to speech, facial expression and gesture.

Although the links are limited, it allows the child to communicate with their carers, thereby reducing feelings of isolation and helping to maintain positive attitudes. These children do communicate and are often shouting silently at the world. Those about them need to be sensitive to this and have the skills to "hear" what they are saying.

(2004 Blackwell Publishing: D. and S. Wray)

5. Language skills

Children with degenerative disorders might well start to experience difficulties with the interpretation of language, particularly if it involves abstract concepts. They seem to find it much easier working in concrete terms. For example, open ended questions involve too many stages for the child to be able to formulate a response. Closed questions, with the subject in front of them, are much easier.

(2004 Blackwell Publishing: D. and S. Wray)

Part two: Education

1. The focus

When parents are considering their child's education, they are mindful not only of academic and performance related matters, but also of care, medical and quality of life issues. For a family whose life is totally affected and controlled by the child's illness, the quality of the child's school life assumes an enormous significance.

A focus on academic issues, where care is a secondary activity, ignores the other end of the educational continuum where the emphasis is reversed. A stress on care and quality of life issues is as valid an educational approach as an academic one because it meets the child's changing needs and impacts directly on the child's ability to respond to any academic input.

(2004 Blackwell Publishing: D. and S. Wray)

- *Schools are based on an assumption of pupil progress; they are now even judged and ranked according to the rate and success of that progress. Even the curriculum is structured in terms of attainment levels. A child who is regressing as a consequence of a life-limiting illness is the antithesis of that organisational and curriculum ethos. It is not surprising that both the child and the professionals working within the school feel challenged by the situation.*
(2004 Blackwell Publishing: D. and S. Wray)
- *There are problems in trying to apply the statementing process to the situation of a child with a degenerative condition, because the documentation expands (not diminishes) and an increasing number of the objectives are not met. Being a needs/resources/objectives model, the current statementing process assumes progress and therefore requires careful and flexible application when used in its present form on a child with a deteriorating condition, who by definition, will continually fail to meet set, progress oriented objectives.*
(2004 Blackwell Publishing: D. and S. Wray)
- *For children with a life-limiting condition, education has an extra dimension. For them, it is a once only event; there is no second chance; they cannot return to it later in life. This places a responsibility on all those who make a contribution to the education of the ill child to ensure that they provide the high quality of service that these children deserve within their shortened life-span. For them, a rewarding and enjoyable educational experience will be something they do remember for the rest of their lives.*

*"Illness and disability do not necessarily disadvantage a child;
but the way we treat them could."*

(2004 Blackwell Publishing: D. and S. Wray)

2. Home tuition

The provision of education (in its widest sense) for children who are sick is essential in order to provide stimulation and improve their quality of life. Education gives them confidence and reassurance and allows them a greater understanding of their world. It is also a legal entitlement.

- *The Advisory Centre for Education notes that there is a danger that home tuition may be refused by an LEA on the grounds that "a child unfit for school is unfit for education at all" (ACE June 1999). This overlooks the fact that there are many other reasons why an ill and disabled child may not be able to attend school but still benefit from home tuition, such as a susceptibility to serious chest infections, difficulties over school transport or supervision problems at school, associated with distressing, unpredictable and possibly life-threatening symptoms.*

- *Home tuition for children with degenerative disorders is not an extension of school per se but is rather a unique service provision; a specially crafted resource to meet the needs of a child and family living under difficult, stressful and peculiar circumstances.*
- *To support the situation, there needs to be an education team with a specific and dedicated role. To have maximum effect, they may have to cross orthodox professional boundaries. It follows from this that there are training implications, so that the support staff are adequately prepared for the very difficult task of working with children who are ill with a complex mix of symptoms, possibly disabled to some degree, experiencing learning difficulties and may be technology dependent. Thought will also need to be given to the organisational structure of the service, in order to give support not only to the service recipients, but also the service providers.*
- *In some respects, this is a situation already met by Paediatric District Nursing Teams who, having trained and qualified within an equipped, resourced and professionally supportive hospital setting, with colleagues usually close at hand, must to some extent, re-skill themselves in order to operate successfully in a community (and domestic) setting, still working within a team, but with a greater degree of independence, professional autonomy and direct responsibility.*
- *Every child has a basic human right to an education which allows them to access a better quality of life. The details of the provision and the strategies that are employed may be different with each individual, but the outcome should be the same.*
- *There is an assumption that education is almost exclusively about skill and knowledge acquisition as a way of preparing the individual for their place as an independent and productive member of society. Although what is taught may prepare the child for the future, a lot of it is also relevant to the here and now, (a point that is often overlooked but is quite obvious in the context of the work needing to be appropriate to the age and ability of the child). For ill children, their immediate world may be a little smaller than that of their peer group, but it is still important that they understand it and appreciate how it affects them.*
- *An issue for those providing home tuition is that although the range and nature of the activities may change and gradually become more restricted, variety is still required. Clearly the activities will be based on meeting an individual's emotional, social, physical, intellectual and spiritual needs, but there should also be an associated element of challenge, excitement and mystery, with some degree of interaction with the environment, which will vary with each child. A skilled and imaginative teacher will also be able to make use of unplanned serendipity events to add extra interest.*

- *The experience of most teachers, whatever the age of the pupil, is that children perform best in tasks that make “human sense”, or have relevance to their world and their every-day lives. To exploit fully the educational potential of the teacher’s visits, it is important to encourage the child to use as many everyday skills as possible, particularly those that are likely to be affected by the condition. This will help to maintain specific muscle and nerve functions, general coordination abilities, overall activity and awareness levels and independence. The more varied the experiences, the greater the likelihood of skill transference from one situation to another. The development of personal qualities is also important as that has both an immediate and long term use.*
- *Practical activities are always worthwhile. For example, art and craft work is a useful activity that children enjoy. It is relaxing and it allows conversation and cooperation with others. It may be two or three dimensional, done independently or with the help of the carer. The children can be involved to varying degrees, depending on their abilities and how they feel on a particular day. For children with severe motor problems, whose direct involvement may be limited, it might require the use of strategies that enable them to be represented in some way, in the project. For example, by using finger, hand or foot printing, a composite picture might be built-up and the child can then clearly see that they are “part of it” and therefore have a sense of involvement and ownership.*
- *There is a hidden value in that not only do they contribute to a finished product that they can handle, look at and show visitors, but it can also form the basis of future interaction and praise, by being displayed in the child’s room and its qualities commented on by visitors. In the longer term, the child will also be able to reflect on their involvement and when the child dies, it is an item that the parents can keep.*
- *Other activities, appropriate to the child’s gender, age and ability, might include reading and story telling, music therapy, water therapy, television programmes, trips outside, visits to a swimming pool or other similar facilities, massage, toys, games and involvement with other children and young people.*
- *Within each activity, there is scope for variations in the way the topic is presented. Also they can be combined in different ways. The essential element is the individual teacher’s style and manner of presentation; their ability to create another world to that of illness, pain and discomfort; another strand to the child’s life; something different to reflect on, look forward to and mark the days and weeks by.*
- *Contact with and knowledge of other people will positively add to the life of the child; just having a friendly and enthusiastic adult in the room will immediately catch their attention; children will see things with fresh eyes and this new experience will add an extra dimension of excitement and interest,*

even to previously rehearsed activities, giving any teacher an immediate advantage.

- *The teacher can function as an “interpreter” and mediate between the child and their surroundings. A steady conversation to accompany everyday activities that the child finds difficult, for example drinking, can be worthwhile if it assists the child and offers an opportunity for praise. It will serve to make the child feel rewarded, valued and the centre of attention. This can also apply to the wider range of less routine activities that result from the teacher’s input.*
- *As well as providing a range of interesting activities, there is also a need to provide as stimulating an environment as possible. The surroundings need to be quiet, warm and well lit, with plenty of natural daylight. The ceiling could well be a focus for objects and pictures; luminous planets, brightly coloured fabric shapes which move in a draught, either from an open window or a fan. Also wind chimes, moving lights on the ceiling and walls or reflective, reciprocating and rotating devices will all catch the child’s attention.*

Part Three: Play

Play is often a child's way of revisiting a situation to re-run the scenario in order to act-out, rehearse, become familiar with and internalise the information, strategies or skills so that they are available for later use; they are handling or processing the information in concrete terms.

Through experiencing play and organised activities in different situations and settings, they are able to acquire skills, knowledge and understandings, improve their creative abilities and develop personal qualities that have both an immediate and long term use. In addition the more varied the experience, the greater the likelihood of skill and knowledge transference from one situation to another.

A distinction might be made at this point between play and activities in the educational sense. Play might be described as a self motivated activity where the child is in control in that they make choices as to whether they do or do not play; what form the play takes, (what they play at); and when they should stop.

An activity in the educational sense is where their efforts are more directed, in that the activity (or task) is provided for them to complete, the manner and duration of their working is more controlled and they play in response to an opportunity that is provided, with predetermined outcomes in mind.

Individual creative activities and play allows the development of a child's own personal and secret world; another aspect to their life, in addition to the world of family, relatives, friends and illness.

Activities and play involving other children can at times be difficult for the individual, but when successful, it can be most rewarding. The understanding that comes from the

experience will result in the development of the personal qualities and skills that are required for successful relationships. For example, appreciating the need for a balanced interaction, the sense of unity from sharing a common goal, the challenge of cooperative working and the pleasure of a shared success will all reinforce the learning process. Clearly the positive aspects that result from the challenge of competing against each other can also be utilised.

Part Four: Story Telling

An adult reading stories to a child is provided with an opportunity to do more than just read. The story will take them into a world of imagination, where anything is possible; even the physical world about the child can also be transformed for a while by the skills and personal qualities of the adult; they can create a very close, supportive and unique world around the child.

As well as the interest and stimulation generated by the story in the book, there is the opportunity for conversation, discussion, the recounting of incidents in daily life, the telling of stories that involve the child, jokes and many other things which widen the experience of the child and give them a host of ideas and experiences to carry them through other, more tedious and perhaps painful moments.

Actors are very aware that the voice, (and the nature and style of the language used), are usually a reflection of the speaker's emotions. They use this association skilfully and to great effect. In these "closer" moments between the carer and child, the quality of the voice that is used requires some consideration, particularly if it is linked to physical contact; holding the child's hand for example.

A surprising amount of coded information can be transmitted in this way through hand and finger pressure and movement, particularly if the child has lost the ability to speak. The hand of the carer gives them another medium, through which they can communicate, in addition to gesture, facial expression, posture, body position, muscle tension and vocalisation. The value of giving an extra dimension to the interaction, through the use of variations in the voice and finger movement and pressure, should not to be under-estimated.

These human moments can also be shared by the siblings, who may be either sitting on the bed or the knees of the story teller. They can be involved in the conversation between the carer and the child or listen to the stories that are read. These shared times are often fondly remembered by all who are involved.

Part five: Music

1. Music Therapy

Music therapy is special in its use of music to encourage communication and expression by playing an instrument, singing or listening, usually in improvised music. The therapist does not *teach* the client to play an instrument; the instruments offered can all be played intuitively.

Within the therapeutic relationship there is a safe setting in which difficult or repressed feelings can be expressed and contained. Where words are inadequate (or even impossible), music can often make sense. In the restricted world of a seriously ill child, music therapy focuses on what s/he can do, gives choices and control, and raises self-esteem. [Jessie Fund web site]

For sick children, the music therapy room can be a gateway to an extra-ordinary world of rhythm and tonal colour. As well as listening to the sounds, they can feel the vibrations from the instruments. They are often delighted when their arm or hand movements, either on a keyboard or on the other specially adapted equipment in the room, can create such wonderful and exciting musical tones.

2. Natural Music

There is a place for natural music. Audio tapes and CDs are available consisting of stream noises and woodland birdsong. They are very relaxing and most effective in evoking images of summer-time family visits to the country-side.

3. Human sounds of reassurance

Many parents will have hummed or used a gentle and extended "shhh" to help settle their children. They will also have found that on occasions, it did not work. This is largely because its effectiveness largely depends on the musical nature of its delivery.

It has been suggested that humming mimics the muffled, low frequency noises that the unborn child hears in the womb and that the gentle "shh" is similar to the sound of the blood flow through the mother's or child's circulation system. For this reason, both are reassuring the young child.

There is, however, an important point with regard to the nature of both noises, in terms of their frequency and volume. If delivered in an inappropriate way, they can be unconsciously misinterpreted by the child as being threatening and an unspoken warning.

It has been suggested by researchers that there is an instinctive white-noise, alarm-response in primates which causes them to involuntarily look towards a loud and

sudden white-noise source and momentarily adopt a flight/fight state.

Some care staff, particularly when rushed and stressed, while intending to reassure and quieten a child unsettled by discomfort or anxiety, actually deliver a sharp, warning "shush" or "hiss", which indicates irritation and annoyance. The same can be true for humming. Often it can sound like a pre-occupied "Mmm", suggesting impatience. Of course, both will have the reverse effect to what is intended.

In addition, a simple repetitive series of three hummed, low-frequency notes can be very effective in settling a child when they are in pain and perhaps waiting for medication to take effect. This can again be linked to gentle but reassuring physical contact.

4. Speech and music

Research has suggested a link between speech and music. This is perhaps not surprising as music is thought to have its origins in speech and singing. Certainly music is used in some speech therapy programmes.

For a child who cannot speak and whose mobility is restricted, a useful way of maintaining contact is to play a sound pattern game. The aim is to devise a short musical phrase, consisting of a pattern of vowel noises. The other person then has to repeat the phrase exactly. It is then their turn to think of something different for the first person to repeat. Although mainly linguistic, there is a musical aspect to it as well. There is both challenge and humour in the situation.

5. Music: Background

Each person has an individual response to a given piece of music, depending on their mood, physical state and the particular circumstances at the time. Clearly some music can be inappropriate in certain settings and, in the context of background music for a care area, clearly the needs of the ill child and not the adults should be the principle deciding factor.

If the music is not the focus of listener's attention then it is important that it is not intrusive. However, the careful use of music can help introduce a little colour, interest and variety into an otherwise monotonous background.

6. Sound and Light Room

The sound and light room is invariably a great favourite with the children (and with some adults as well!). They enjoy lying on the heated water bed, listening to the relaxing music and watching the coloured patterns of the lighting effects change and merge. It induces a peaceful frame of mind and the children will often be asleep within minutes.

7. Linking vision and sound

When using a Rompa projector with background music, a much better visual effect can be achieved by using a fluid (as opposed to a fixed pattern) disc. The image that is projected onto the wall is dynamic within itself. As well as slowly rotating, the patterns are continually changing; an endless flow of coloured shapes that merge and then reform; never repeating, they seem to develop in time to the music.

Other lighting effects can be achieved by using coloured Christmas lights which flash in different sequences, moving fibre optic lights, rotating mirror balls, side-table lamps with a transparency which rotates and projects a moving picture onto the light's shade, small disco-spheres with coloured light windows and small, domestic disco-light projectors which throw moving white light shapes onto the walls and ceiling of the room.

*However, it is **essential** that the rate of change of the light patterns is very slow and that the lighting effects do not dominate the room, otherwise they can become very disturbing and disorientating, particularly for patients who are experiencing visual distortions and associated balance problems.*

The ceiling can also be a focus for objects which can add interest to the room and catch the child's attention while they are listening to the music. These might include brightly coloured fabric shapes, unusual mobiles and small wind chimes which move or rotate slowly in the draught from an open window or a fan.

Section 8: Siblings

Many of the difficulties encountered by the parents will also have an impact on the children. Like them, the siblings also suffer a restriction of freedom, gradually "lose" a member of their family with whom they still live, have to accept additional responsibilities, do not have a normal, (as perceived by their friends) home and life-style and have to deal with the death of a family member.

For younger children particularly, they are at a formative age and there are future consequences to their current experiences. The way in which they are guided through the unfolding events will shape their reactions to future circumstances; opportunities taken or missed now will return later to influence the child's adult world.

(2004 Blackwell Publishing: D. and S. Wray)

Although there are many issues concerned with being the sibling of an ill child, the more significant ones are **Isolation, Quality Time** and **Bereavement**.

- **Isolation.**

There is the potential for siblings to feel isolated. This may be brought about by either a reduced level of contact with their peer group and wider family members

or by the lack of someone to talk to who has a full understanding of the issues that are troubling them. It may be that the child's experiences are unique in the area where they live.

(2004 Blackwell Publishing: D. and S. Wray)

- **Quality Time**

Quality (or dedicated) time is a pre-identified period when the child knows they can look forward to the undivided attention of one or both of the parents. It requires careful planning and is clearly difficult in the unpredictable circumstances of the illness, but many parents find that it is most rewarding and highly valued by the siblings. Input from other adults can supplement these positive, security periods.

(2004 Blackwell Publishing: D. and S. Wray)

- **Bereavement.**

Preparing the sibling for the death of the ill child, giving them an understanding of the concept of death, making them aware of its implications and preparing them for its consequences is a difficult exercise. Each child will have their own base level of knowledge and from this will need to be built up a progressive understanding of the issues, to the point of explaining what will eventually happen to their ill brother or sister. *(See Section 8 on Bereavement)*

If the sibling can also be sensitively involved in the funeral arrangements and the service, then that will contribute to the healing process.

(2004 Blackwell Publishing: D. and S. Wray)

Owing to the enormous care demands placed on parents by the need to look after the very ill child, siblings often have to be far more independent and resourceful than would otherwise be the case.

Many parent carers find that by default they are in danger of becoming "bad" parents to the siblings. This is on the basis that poor and inadequate parenting is characterised by the adults in charge of the children, to varying degrees:-

- *Being in-attentive, unresponsive and seemingly indifferent to the needs and demands of the child.*
- *Being short tempered, irritable, lacking in humour and constantly tired or unwell.*
- *Not being prepared to devote quality-time to the child when they can have the opportunity to be close to them, interact in positive and differing ways and communicate meaningfully, effectively and appropriately.*
- *Continually using deferral phrases and avoidance or blocking strategies such as.... In a moment... Not now... Can't you see I'm busy...Look, I'm dealing with this at the moment... I'm just going upstairs... I must get this done now... Don't do that now, do it later... Don't get anymore toys out... etc".*
- *Not providing treats, pleasant surprises, visits, trips or holidays or doing things that reflect affection, love, care, interest, concern, sympathy and understanding. From a child's perspective, it is actions (and not just words) that count.*

- *Not engaging in meaningful, purposeful and at times, entertaining conversation which recognises the contribution of the child, gives positive reinforcement, reflects the values of the group and helps to create a unique family ethos and identity by having a bonding and unifying effect.*
- *Not providing adequate explanations for what lies behind certain actions or unfortunate circumstances which may impinge adversely on the life of the child and reduce their quality of life.*

There is clearly a need for the caring parent to compensate for the unintentional disadvantaging of the siblings; there is a dilemma that by being a good parent-carer and meeting all the care demands of the sick child they might then, by default, become a poor parent to the siblings.

While it is readily recognised by support organisations that poor parenting brought about by social or intellectual inadequacy requires additional input to support a family, what does not seem to be generally acknowledged is that reduced parenting resulting from coping with extreme care demands can also adversely affect the family and siblings and have long term consequences.

If the family is not provided with adequate and appropriate support during the period of the illness, difficulties can develop, which have the potential to adversely affect their future perceptions of each other and the nature of their relationships.

This emphasises the need for the support to be both holistic, (viewing the family as one unit with internal interactions and dynamics) and individualistic, (providing differentiated and adaptable support for each family member, whether parent, or sibling).

Section 9: Bereavement

All family members will require help to prepare for the death of the ill child.

For the parents, it may be readying themselves for the child's final moments; rehearsing mentally what should be done and what might be said. As there is a large element of the unknown about the situation, it may also include attempting to fill in some of the blanks in their knowledge, perhaps visiting the cool room in the hospice where the child will stay once they have died or speaking to experienced staff about how the situation might best be approached. Clearly any of this is difficult to do while the child is still alive and the degree of preparation will depend on the individual.

Preparing younger siblings for the death of the ill child, giving them an understanding of the concept of death, making them aware of its implications and preparing them for its consequences is a difficult exercise. Each child will have their own base level of knowledge and from this will need to be built up a progressive understanding to the point of explaining what will eventually happen to their ill brother or sister.

Depending on the age of the child, they are likely to go through several phases in their understanding of the concepts and it will be necessary to provide the information in stages, giving them chance to absorb the new ideas.

Young children often have difficulty in understanding the finality of death so it is helpful to encourage them to take the opportunity, while there was still time, to say and do all the things that they feel need to be said and done.

Deciding whether or not the sibling should be with their brother or sister at the moment of death will depend very much on the circumstances and the individuals involved. It does not have to be a frightening experience however and if the death is peaceful then it is likely to be accepted by the prepared sibling as another natural stage in the whole series of events, both sad and joyful that has marked the life of the ill child.

Equally, if the sibling can be sensitively involved in the funeral arrangements and the service itself then that will all contribute to the healing process.

Following the death of the ill child, it is likely that all members of the family will require some degree of counselling. This may be individual (as in Cruise or the Castle Project) or may be family based (as in the family follow-up sessions organised by the hospice which cared for the child). The bereavement sessions are often of great benefit, allowing the parents and the siblings to move on from the trauma associated with the child's illness and death and prepare for a changed future.

Preparation

The family, both as a group and as individuals, will need to prepare, in their own way, for the death of the ill child. It is very likely that everyone will need some degree of guidance and support from experienced palliative care staff in the period immediately before the death.

When discussing forthcoming events, it can help to think of the events happening to a third party so that the focus of the discussion is away from the child. The detachment can then allow difficult and distressing issues to be discussed.

Unless the family has been through the situation before, they may be uncertain as to how best to support their dying child and cope with the impending events. It is most helpful if they are given some general guidelines so that they can ensure that all that needs to be done is done. This will also allow the parents and siblings to quietly prepare themselves mentally and emotionally for the forthcoming situation.

The guidelines might include:-

- *What supportive things to say.*
- *What supportive things to do to ensure the child will feel close to the family, but is comfortable and settled.*

- *What things the family (both as a group and individually) might need to do to help them cope with the immediate and longer term situation.*

Other issues to be addressed might include;

- *The cessation of the caring role*
- *The return home (if the child died in a hospice)*
- *Family readjustments*
- *The gap in the family left by the death of the child and change of status for the siblings*
- *The impact of bereavement on the siblings*
- *Dealing with the now redundant housing adaptations made for the ill child and adapting the family home to the changed situation*
- *Returning or disposing of equipment [and perhaps personal belongings]*
- *Visiting the grave site and attending to the grave*
- *Addressing financial issues*
- *Returning to work*
- *Post traumatic stress disorder*
- *Future reminders of the child's absence such as anniversaries or important events which mark out the developing lives of the dead child's peers.*

The last few days

During the final few days, it is helpful to everyone if the family assumes that the very ill child is aware of their presence and can hear and feel their contact with him. They should try to:-

- *Reassure the child (who may detect their anxiety).*
- *Make the child feel loved and valued.*
- *Emphasise the child's contribution to the family.*
- *Perhaps stress personal qualities. (Often these children are inspirational).*
- *Possibly reflect on all the happy times the family has had together.*

There is also the issue of saying goodbye to the child (though great care needs to be taken when doing this as the child might misinterpret the situation and become upset, thinking that it is the family that is leaving and not them). Plenty of reassurance is necessary, with perhaps some comments about:-

- *Not being afraid.*
- *The family being with him.*
- *Everyone is staying and they are not going to leave him.*
- *His being surrounded by their love and therefore he can relax and be settled and quiet.*

When the child has died.

After the child has died, there are the immediate issues of washing, dressing and positioning. Many physical problems often so evident in life will recede so that the child looks peaceful and at rest.

If the child is to stay at the hospice until the funeral there are the preparations that need to be made for moving the child to the cool bedroom. A collection of personal items can be taken with the child into the room, so that they are in a more personal setting. This will also help those who visit the child to say their last goodbyes.

Making the funeral arrangements.

Many parents find that, contrary to what they expected, when their child dies, the duty of care pressures are not immediately removed; they merely change. There is still the feeling that there are jobs to be done and despite the exhaustion, it is they that must do it for the sake of their child.

The Children's Hospice can help greatly in this respect by supporting the parents and guiding them through this difficult time; reviewing and discussing options, establishing outcomes, providing resources and facilitating action in order to achieve the family's wishes.

It is helpful if the whole family is involved in the arrangements, particularly siblings, as these experiences will eventually become part of the healing process. Gaps in their knowledge are unhelpful and it is important that they feel involved and able to make a contribution. This is why adequate preparation before-hand is so important. They should be able to say goodbye to their brother or sister, just as they would have done had they still been alive.

They can be:-

- Involved in the arrangements with the funeral director over the style of both the coffin and the headstone and make a contribution to the nature and content of the memorial inscription. They may also wish to visit the proposed grave site, so that they can mentally prepare for the burial.*
- Involved in the arrangements with the vicar (or religious leader), over the service, burial and, perhaps later, the memorial service. They may be able to help with the design and content of the order of service, propose ideas for hymns, readings and music and, if possible, be involved in the service itself, even if it is limited to carrying a prized personal possession of their brother or sister in the procession down the aisle and placing it on the coffin so that it is there during the service.*

After the burial

After the burial, so that there is not a sudden cessation of care, which can heighten the feeling of loss, the family (or individual members) can focus on keeping the grave site tidy and attractive with flowers (or even helping with maintaining the church grounds generally). This provides a way of preserving memories and showing that the deceased child is not forgotten

In the longer term, there is the issue for all members of the family of how to deal with questions (and the associated explanations) which occur in either work or social situations about gaps in service, changes of occupation, family size, or a lack of resilience in certain circumstances.

Facing the future

The family (as a group but also as individual members within the group) has to face the issues associated with readjusting to the changed situation and if possible devise ways round the problems they face. These include:-

- *The change in focus and the necessary readjustment of family dynamics.*
- *Finding new focal activities which preferably involve all the family.*
- *Examine behaviours within daily routines and reassembling fragmented activities; for example, re-establishing unhurried, family meal times at the table).*

Many parents and children find that a creative activity is very therapeutic. It allows them, either consciously or unconsciously, to express their thoughts and feelings about recent events and identify with positive aspects of their past and present life. It may take a variety of forms and vary in its complexity.

For many, simply putting their thoughts on to paper allows them to be recorded, commented on and then filed for later reference (if necessary). Making sense of a difficult and traumatic experience is always very helpful. It also means that the events of the past do not have to be carried in the memory of everyday events; they are safely held elsewhere and can be returned to when the need arises. Writing allows the person to move on and not be worried about forgetting the experience.

In addition, if through writing about their own experiences, the parents are able to assist other families then that will also give a sense of purpose to what has happened: "My child's experiences can help many other boys and girls"

Section 10. Conclusion

Clearly the huge care demands of looking after children with life-limiting conditions makes everyday life very difficult for carers and their families; there is much to be done and a lot to be learned. It is hoped that this paper, "A Summary of the Skills, Knowledge and Understanding needed to look after a Child with a Genetic, Degenerative Disorder" will help all those who are involved in the care children with life-limiting or degenerative disorders and perhaps allow them to be more effective in their support by giving them a greater understanding of the issues that are involved.

Section 11. Text References

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Section 12. Useful Reference Books

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Complete Book of Mother and Baby Care
Dorling Kindersley Ltd. (1990)

Family Doctor. Home Advisor.
Dorling Kindersley Ltd. (1986)

Family Guide to Prescription Drugs.
Diamond Books (1995)

Family Health
Dorling Kindersley Ltd. (1987)

First Aid and Family Health.
Marshall Cavendish Ltd (1992)

Human Body
Dorling Kindersley Ltd. (2001)

The British Medical Association Complete Family Health.
The British Medical Association Complete Family Health.
Dorling Kindersley Ltd. (1990)

The British Medical Association Guide to Medicines and Drugs.
Dorling Kindersley Ltd. (1991)

The Children's Medical Handbook
Dorling Kindersley Ltd. (1991)

The Mothercare Guide to Child Health.
Prentice Hall Press. (1989)

The NHS Direct Healthcare Guide
NHS Direct and DPP 2000 Ltd

Appendices

(1) At-a-glance summaries: Examples 1, 2, 3.

Most medical records are presented in the form of written reports. These take time to read and unless read in completeness do not give an overall view.

This section contains examples of “at a glance” summaries which are in the form of charts or graphs. These proved to be of great value when briefing care staff or attending clinics.

The raw data was either draw from the carer’s everyday experience (*Appendix 1*) or was based on carer’s notes recorded in the Epileptic Record Book provided by the hospital.

They are reproduced here in their original form.

(2) Observations on Good Practice: Examples 4, 5, 6.

We were asked by the Primary Care Trust if we could give examples of any good practice we had experienced from service providers during the period when we were caring for Andrew. The three examples shown were our response.

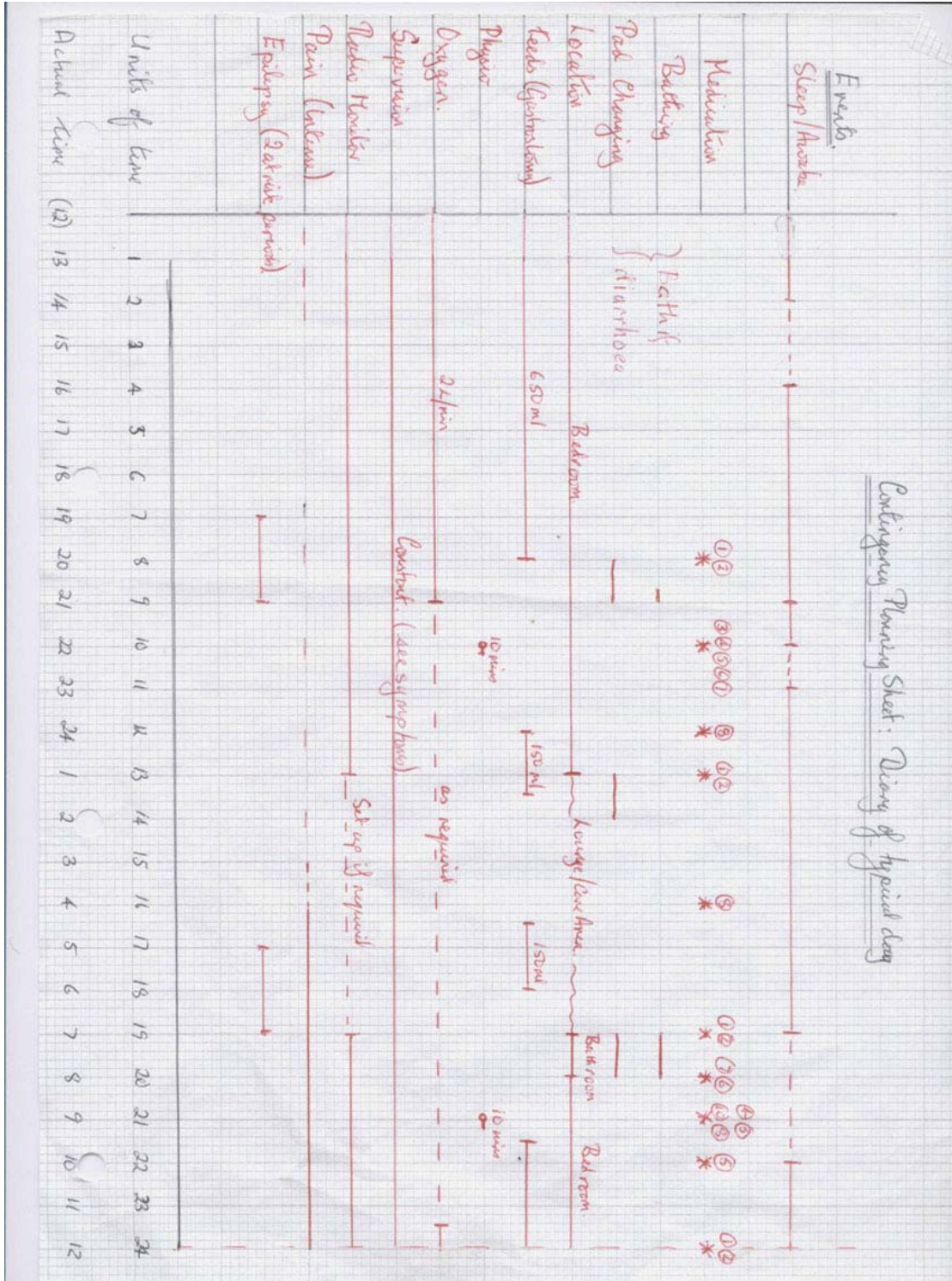
(3) Friends, Neighbours, Members of the Community and Relatives who made a difference.

The help we received from the wider family and people within the community.

Appendix 1

Contingency Planning Sheet

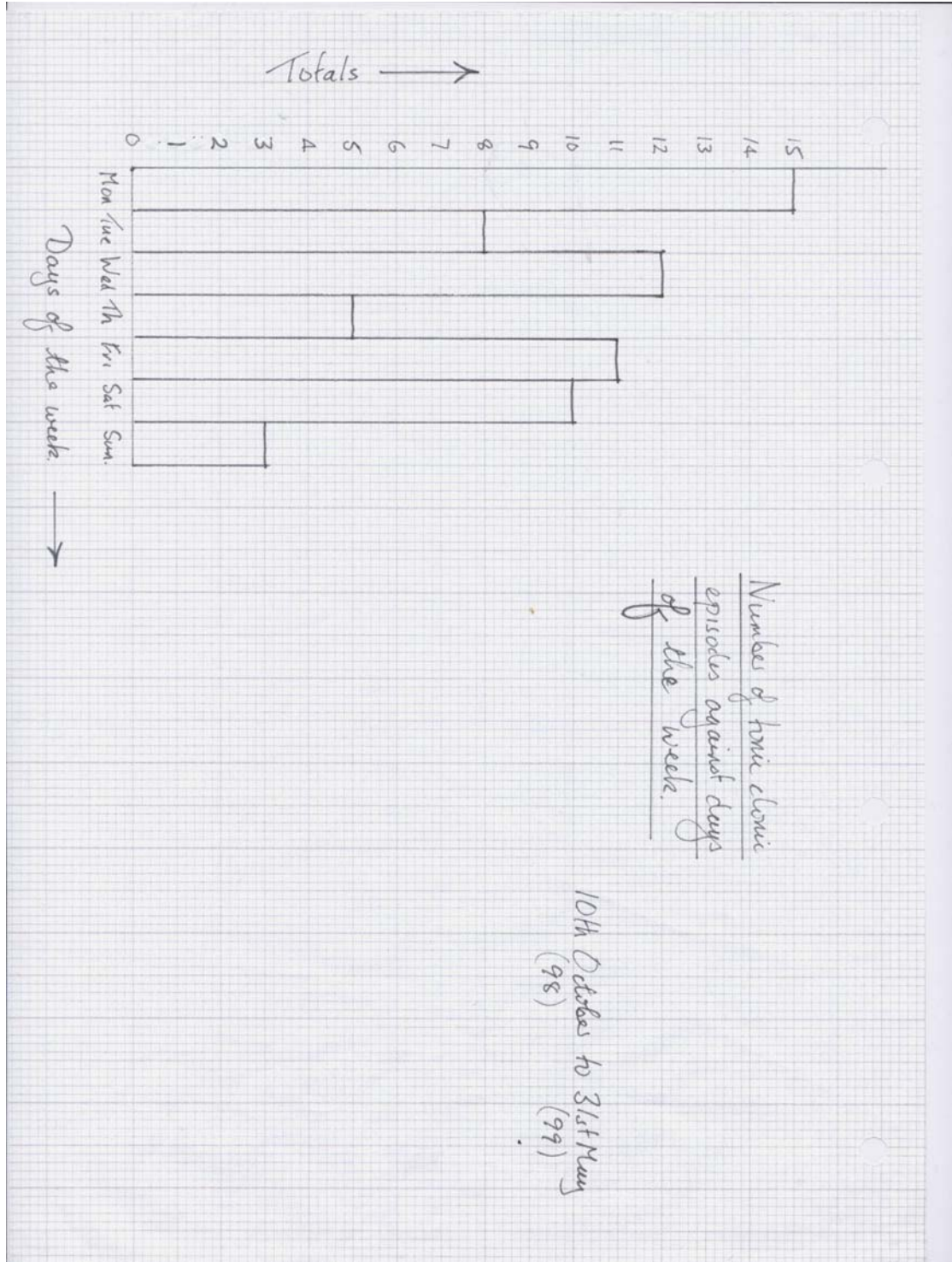
An example of a hand-drawn chart, produced for new carers, showing care patterns over a 24 hour period at a particular stage of the progressive illness



Appendix 2

Chart showing relative frequency of epileptic episodes over a week

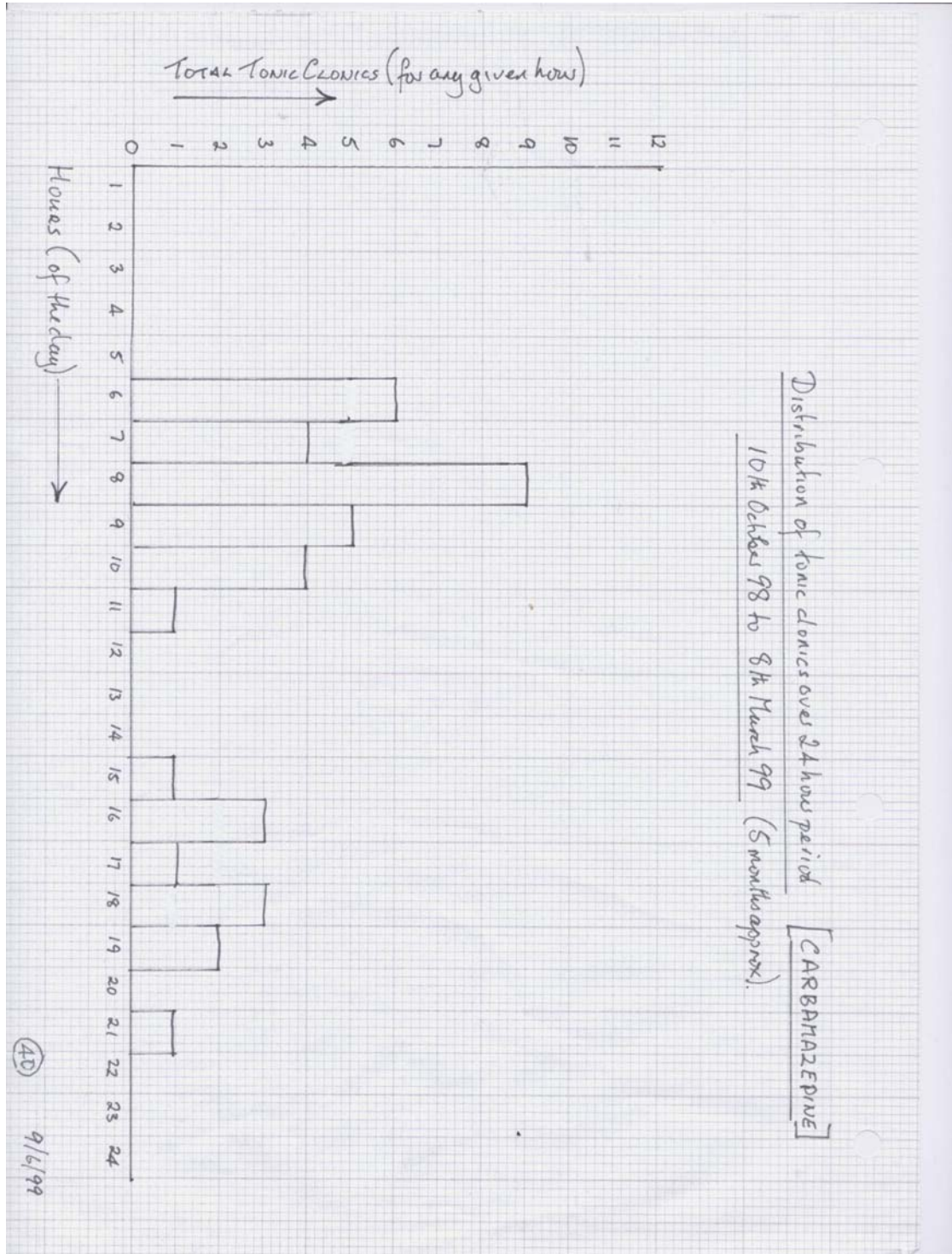
A copy of a hand-drawn Bar Chart showing relative number of epileptic episodes for each day of the week over a period from 10th October 1998 to 31st May 1999.



Appendix 3

Chart showing relative frequency of epileptic episodes over 24 hours

Copy of hand-drawn Bar Chart showing distribution of epileptic episodes over a 24 hour period from 10th October 1998 to 8th March 1999.



Appendix 4

Professionals who made a difference.

We found that the professionals who made a difference to Andrew's quality of care exhibited certain characteristics. These were as follows:-

- A true professional was a person who was empathetic and well organised. They found their work intrinsically rewarding and were therefore motivated to provide a high quality service. They made a positive and even memorable contribution to the lives of the patient, the siblings and the parent-carers.
- They were individuals who were prepared "to go the extra mile" and do things which were beyond their basic job description. This may, for example, have been arriving early or staying beyond their allotted duty period to help the patient and the carers cope with a particularly difficult situation.
- With regard to the qualities they displayed, we found they had initiative, were usually well qualified and were experienced and skilful in working with children. They were also reliable, trustworthy and consistent, recognising that when on duty, patient care should come before personal convenience.
- In order to work successfully within the domestic setting, they recognised the need to be present but not conspicuous. Being polite, patient, resourceful, conscientious, creative, supportive, friendly and helpful facilitated this. The fact that they were, in addition, modest, gentle, caring, interested in and clearly concerned about the patient was reflected in their bedside manner and the way they treated the parent carers. They were careful to keep to a minimum any disturbance to the patient and avoid distress and pain.
- Generally, their positive and optimistic outlook, together with a quiet humour, had a positive effect on the moral of the patient and the carers.
- They were inspirational, in that they made others aware of just how much could be done and even did what at first sight seemed to be impossible.
- They were teachers, either by example or by intent, skilfully passing on their knowledge to those with whom they worked. They valued the patient and the carers, recognising, encouraging and developing the carer's knowledge, skills and personal qualities.
- Often the people who made the greatest contribution were those who did not realise how skilful they were.

Appendix 5

Good Practice Guide-lines.

- It is important to see the patient as a child with an illness or disability; not an ill or disabled child.
- The parents may not actually be demanding and fussy; they may just be concerned and anxious.
- Before caring for the child at home, it is essential for the professional to make a prior visit, in order to better understand the needs of the patient and family.
- The professional should be prepared to admit to a lack of knowledge but also be prepared to research the topic so that they are equipped to help others in future.
- In the home, they should work as part of a team where the parents are equal partners, so that each person can make their own unique contribution to the group's efforts.
- They should always endeavour to find a solution to a problem and respond to messages and requests promptly, in order to provide a high quality of service.
- They should be open minded and be prepared to learn from others, continually updating their skills and knowledge. They should be experts at learning as well as experts at knowing.
- There is a need for them to be reflective and review their own practice, having the confidence to change if that will improve the level of service they offer.

Appendix 6

Examples of good practice

- A GP who, of his own volition, would call-in when passing on his rounds, to check on the welfare of the patient and see if there was anything further he could do to support the very ill child and the family. This was greatly appreciated by all members of the family.
- A surgeon who made a home visit in his own time and provided vital guidance as to how to deal with spasms of the patient's throat muscles, which inhibited the child's breathing.
- A consultant who slid a business card across the desk and said, "Here is my home telephone number; ring me if you have a problem."
- A GP who said, "If you need to contact me, my home telephone number is in the 'phone book."
- A support worker who, in a time of crisis, was prepared with minimal notice to stay overnight to ensure the well being and safety of an ill sibling and the exhausted father.
- A voluntary, sibling-support worker who, through her conscientious efforts, helped a troubled sibling through a difficult period of her life.
- Care staff who found the time and energy to listen sympathetically to tired and distressed parents who needed the opportunity to talk through the many insuperable problems associated with the care of a child with a degenerative disorder.
- A skilful professional, who upon seeing injustice, did not just accept the situation, but through her efforts, resolved the problems.
- A paediatric nursing team who ensured the prompt delivery of services and equipment. (If they said it would be provided, it was, within days.)
- Nurses, doctors and care personnel, who by their personal qualities, dedication and skill won the admiration of both their colleagues and the families with whom they worked.
- A counsellor from a voluntary bereavement organisation who skilfully led a carer through a maze of problems to a situation of self resolution. ("You already know the answers; I've just helped you to find them!")
- Staff who rang work when they were off duty to check on the progress of a patient.

- Ambulance crews, who respected the parent's views, listened carefully to the information that they were given and went to great lengths to make sure the emergency transfer of a very ill child was done with as little disturbance to the patient as possible.
- An A&E Department that always dealt immediately with an ill child, recognising that the child's condition could deteriorate rapidly.
- An A&E nurse who spotted the early symptoms of meningitis.

Appendix 7

Friends, Neighbours, Members of the Community and Relatives who made a difference.

- A friend who avoided just saying “Let me know if there’s anything you need” but would use her mobile telephone to contact us and say “I’m at the supermarket *now*. What do you need?”
- A friend, who on more than one occasion drove several miles to the city centre late at night to obtain medications prescribed by deputising doctors.
- Retired neighbours who were compassionate and understanding and always responded positively to sudden and unexpected requests for help, including driving the parents to chemists, hospitals or the child’s hospice.
- Neighbours who, without prompting, on several occasions prepared a full evening meal and brought it to the house already served onto plates.
- A senior member of a local church group who was prompted by his own, similar experiences to visit the family and offer practical support.
- Members of the local church who, without prompting, drove the fifteen miles to the Hospice to offer practical support and help with the funeral arrangements and the services.
- A member of a nearby village church who without question agreed to sing at both the funeral and the memorial service
- A former colleague who brought a group of fellow musicians to play at the memorial service.
- Relatives who were prepared:-
 - To do research on our behalf on the internet using recognised and reputable sites.
 - To collect by car (at short notice) urgent medical supplies or replacement support equipment.
 - To take a sibling on surprise day trips and also look after her for a week in the summer break so that she had a holiday that summer.
 - Facilitate the repair of the family car while it had broken down during an emergency visit to the Children’s Hospice.