Current Practice

PRACTICAL NEUROLOGY

Chronic Paediatric Neurological Disorders—Part II

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(Last week Dr. Wilson considered the approach to neurological disorders in childhood, and discussed cerebral palsy and epilepsy. The article is concluded this week.)

Progressive Neurological Disorders

Though individually uncommon, as a group these diseases come within the experience of most family doctors. Some are genetically determined, such as Tay-Sachs disease, muscular dystrophy, chronic spinal muscular atrophy, and Friedreich's ataxia, while in others the cause is not known—conditions as diverse as demyelinating diseases and tumours. Both the patients and their parents have distinctive problems.

Children.—If the child is old enough, intelligent enough, and the deterioration rapid enough for him to have insight (and the threshold for each is not so high as might be imagined) the inexorably progressive nature of certain neurological conditions causes especial anguish to the patient as well as to his parents. In some children with, for example, a brain-stem tumour, where the deterioration may be rapid though mentality is relatively preserved, the fear of dying may not be overt; but, given the opportunity, they may press very penetrating questions concerning their fate which indicate their bewildered anxiety about the course of physical events. In other less rapidly progressive disorders—for example, chronic spinal muscular atrophy or Friedreich's disease—anxieties may be less acute, but there is often more than just vague unease about the future in some children.

This is illustrated at a residential school for physically handicapped girls of superior academic ability of 11 years and over. The arrival of a newcomer is viewed with keen interest by the other girls, her physical status assessed with alarming perspicacity and related to her age and diagnosis with sleuth-like zeal by her peers, each with her special need to discover anything about the new girl's condition which might throw light on her own. Casually devastating questions about diagnosis and prognosis are usually presented not to the "expert" staff but to the relatively untrained helpers who share the girls' confidences at, for example, bath-time and bed-time. Speculation on marriage prospects and the ability to have babies take precedence (for the girls) over employment opportunities, but in the background is the anecdotal awareness that last month, or last year, or five years ago, a girl at the school died of a particular condition.

The special circumstances of having a number of intelligent patients grouped in this way intensifies their problems, but they are present in diluted form elsewhere. In a home setting it is usually the parents who are channels of information, and doctors must help them to answer their children's inquiries honestly.

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Though the time course of the more chronic disorders can be so variable that medical counsellors can honestly hide behind a confession of ignorance, this in itself may serve merely to confirm the fear of the unknown. Nor can a policy of dissimulation be advocated—the perspicacity of even preschool children will not always allow it. Precisely what is said depends not only on the nature of the condition but also on the relationship and understanding between parents and doctor, and also on the personality and insight of the doctor himself.

Parents.—Because of the difficulties of making an early diagnosis in a condition which may have an insidious onset, there may thus be a failure to recognize the existence of a problem by the doctor. An approach which on the one hand acknowledges that there is always a reason for parents' anxieties (though not necessarily expressed directly) and on the other can support parents until the lapse of time sees the spontaneous resolution of the transient and the trivial and allows the less benign conditions to declare themselves more clearly, will not forfeit the confidence of parents in situations where the help of a family medical adviser is badly needed.

Despite this tolerant and understanding approach, doctors in general, and the family doctors in particular, may yet be the object of ill-concealed hostility. This may reflect the bewilderment, the disappointment, the bitter anger, commonly seen in parents of handicapped children, especially if the condition is progressive, recognizing that their child has a condition that cannot be cured, and probably cannot even be relieved. What malignant trick has life bestowed, how inept and incompetent are these "experts" that they can't do better! It is a mistake for a doctor to personalize this antagonism, since, once understood, it is clear that it is not really personally directed.

The reaction of parents to the knowledge that their child is abnormal has been compared with "chronic mourning," and this is a helpful concept.

If the condition is genetically determined, the parents may have an intense feeling of guilt that they themselves are defective and directly responsible for the child's malady. Most of the genetically determined degenerative neurological diseases are inherited as autosomal recessive conditions, which means that both parents are heterozygous carriers, and fortunately, therefore, this sense of involvement is shared by both parents. But for sex-linked recessive conditions, such as the severe childhood form of muscular dystrophy transmitted by females, or dominantly inherited maladies such as dystrophia myotonica, when one or other parent not only transmits the condition but may also be suffering from it in some form, the sense of guilt not only has a one-sided bias but on occasions may be accompanied by the reproaches, explicit or implicit, of the unaffected parent.

Often, of course, some parents (and doctors too) have a total misapprehension about the mode and risk of inheritance of certain maladies, and may be especially confused concerning the

risk to apparently unaffected children and their offspring. In these circumstances, where there is uncertainty, parents and family doctors should avail themselves of the genetic counselling services that most teaching hospitals provide.

General Considerations

The difficulties arising over the failure to recognize the onset of a progressive disorder have been already mentioned, but this problem more often arises in the recognition of non-progressive handicaps in early life. It is worth emphasizing once more that though some mothers may be (overtly) unaware of the slow or distorted development of their offspring their close contact and intimate knowledge of their child enable them to be aware of subtle changes that may be missed by others. Such perspicacity should not be lightly discredited on the basis of a five-minute interview and a cursory examination. Even if it is possible competently to exclude any significant degree of handicap, given time and willingness, the discerning family doctor with his expertise in human relationships may recognize subtle and important causes of parental anxiety with the opportunities for help which bland reassurance only negates. I see this role increasing as the position of priest and neighbour as confidant and upholder declines in contemporary urban society.

A careful history and unhurried observation of a young child (preferably in the home) may confirm the existence of a developmental abnormality and indicate the need for a more specialized opinion, usually that of a paediatrician with a declared interest in these problems. In this way one hopes that the grosser problems will not be overlooked, and the evaluation of relatively minor variations from normal may alert the need for careful follow-up.

Intercurrent Medical Problems

Teeth and Teething.—Though the causal association between teething and convulsions has been repeatedly and justifiably challenged, nevertheless control of fits in certain children with an established seizure tendency may relapse during tooth eruption and may need a temporary increase in anticonvulsant dosage. If phenytoin is already producing gum hypertrophy the eruption of permanent dentition may be particularly troublesome, and this is a strong reason for maintaining good dental hygiene and regular dental supervision.

When extreme irritability and psychotic behaviour appear suddenly in handicapped children who cannot directly communicate their problems, simple painful conditions such as toothache and earache should always be considered as possible causes.

Salivary Dribbling.—Many retarded children and children with swallowing or postural difficulties associated with cerebral palsy may continue to dribble saliva after the usual age, and parents may need reassuring that this is not necessarily harmful and will usually resolve spontaneously. There seems to be little if any justification for the surgical extirpation of salivary glands, but atropine derivatives may be helpful in reducing salivation if it is especially troublesome and prolonged.

Immunization.—A febrile reaction which may follow immunization in any child may provoke fits in these with a pre-existing tendency, and advice must be given upon the use of antipyretics in effective dosages in vulnerable children.

Smallpox vaccination and pertussis immunization are believed to be especially apt to cause encephalopathic complications in children with pre-existing cerebral damage. Though I am unaware of any satisfactory studies of such a predilection, I nevertheless advise against such immunization in children who are suspected of having cerebral damage.

Though measles inoculation causes a mild febrile illness in a high proportion of children, and could therefore cause an exacerbation of fits or even an encephalopathy superimposed on

pre-existing damage, it seems that the risk will be no less and perhaps even greater during natural measles infections, with its known encephalopathic propensities. Since simultaneous injection of specific measles antiglobulin and measles vaccine into separate arms will effectively reduce the risk of a febrile reaction without impairing its potency, it is reasonable to offer this scheme of immunization at a mutually convenient time even in neurologically handicapped children, provided the risks are discussed with the parents, as well as the use of aspirin or paracetamol if the child becomes feverish.

Bowels.—Dyschezia occurs relatively frequently in non-ambulant children for several reasons: immobility, low dietary intake, low dietary residues (if there is actual chewing difficulty), and bowel atony. It may not only cause great physical distress to the patient, but may also be accompanied by increased fit frequency in an epileptic or increased hypertonicity in a severely physically handicapped child. The inclusion of more fruit in the diet with one of the proprietary preparations increasing faecal bulk will deal with less severe constipation, and a mild stimulating aperient may be added. If, despite this, the child still becomes dyschezic, it is worth while additionally evacuating with an enema as necessary, and then using suppositories regularly, every two to three days.

Psychiatric and Social Aspects

It is implicit in what has been said already that a child's problems cannot be considered in isolation from those of his family; the repercussions on the lives of parents and other siblings may be profound and far-reaching. A recent survey² of handicapped children of school age in the Isle of Wight has highlighted both the great frequency of psychiatric disorders in children with chronic neurological problems and the frequency of psychiatric disturbance in other members of the family.

The survey did not reveal any specific association between a psychiatric disorder and particular cerebral lesions, but among epileptics there was a clear interaction between the clinical abnormality, type of fit, intellectual and educational factors, and "socio-familial" conditions. As yet the management of these difficulties is empirical and largely intuitive, but an understanding of some of the factors may enable the family doctor to be sympathetically helpful.

Drugs are of definite (if limited) value. For example, a short course of appropriately heavy night sedation may allow the re-establishment of a more socially acceptable sleep routine, while agents such as diazepam and chlorthalidoxepoxide appear to be helpful in the management of enuresis. The calming of a hyperkinetic child to permit his inclusion in a nursery group may allow his harassed mother respite and thus break what is often a vicious cycle of mutual excitement. Drugs such as diazepam, chlorthalidoxepoxide, trifluoperazine, hydroxyzine, and reserpine are mildly effective, while occasionally children respond favourably to d-amphetamine. Chlorpromazine and other promethazine derivatives are more effective but may cause extrapyramidal complications.

Families

The care lavished on a handicapped child may be at the expense of other members of the family. In a minority of cases the incessant demands of a handicapped child may accompany a morbidly intense interdependence between mother and child to the exclusion of husband and other children, who may be actively rejected. Grave marital stresses are not uncommon. Since the family doctor is much more likely than other doctors to see the family as a unit, he has the opportunity of recognizing these problems early. But unfortunately his contact with the family may be irregular and infrequent, unless he deliberately maintains contact with these families on occasions other than the acute consultations for intercurrent ailments.

It may be necessary to offer guidance about the need for long-term institutional care, and in considering the problem I reject the dogmatism of some advisers. At one extreme some practitioners urge permanent care if a mental or physical handicap is more than trivial, with utter disregard of the child's needs and the parents' feelings; while at the other there are those who exert the utmost moral pressure to reject residential care under any circumstances and wilfully ignore the pressing needs of other members of the family.

Adversity is not invariably an ennobling experience, even for the spiritually minded. For a mother who becomes socially isolated, and whose morale and physique are sapped by the intensity of her involvement with a growing and severely handicapped child, the utmost tact and understanding—as well as ingenuity—is required to arrange for residential care in a crisis. It may be almost impossible to obtain a place in a hospital or subnormality unit in a hurry, and in anticipation of such difficulties, when parents begin to acknowledge the possibility of needing such outside help at some undefined time in the future, it may be helpful to establish contact with a subnormality unit through the local authority or mental welfare officer so that a place on a waiting-list is obtained. Temporary admissions during family holidays may give much needed respite.

Since the handling of not only these arrangements but also of educational arrangements generally is statutorily defined and organized mostly by the local authorities, many parents have a vague unease, if not open suspicion and hostility, towards local authority doctors, whom they may consider more as law enforcement officers than as family friends. In so far as they are the representatives of society and society's enactments, this is understandable. A family doctor is not (as yet) regarded thus, and must be seen as an ally by the patient's family, and as someone personally concerned with the individual's needs with whom educational as well as medical problems can be discussed.

Adolescence

It is not surprising that the problems and conflicts besetting teenagers generally should also disturb the uneasy tenor of life for the handicapped adolescent. Unless he is very subnormal, the desire for independence, socially and economically, may be frustrated by limitations in physical skill and the difficulty of obtaining suitable vocational opportunities. Help from juvenile employment officers attached to employment exchanges is invaluable, and many patients are eligible for voluntary enrolment in the Ministry's register of disabled people. The Spastics Society also operates a vocational and employment advisory service.

Since adolescence also brings graduation from the supervisory care of paediatricians and child-oriented workers, this transition itself may bring a crisis to interpersonal relationships between the patient, his family, and the hospital medical adviser. Obviously such a transition does not complicate relationships with a family doctor, and this continuity can prove invaluable, especially for the epileptic.

For those teenagers requiring hospital admission for any reason, adolescent units are the obvious ideal solution, but these are very few. As an alternative, admission to an appropriately adapted paediatric unit—at least for the younger teenager—is preferable to admission to an adult unit where too many (geriatric) reminders of mortality and morbidity may heighten his fears about his own future.

Local Authority

With its staff of medical officers, mental welfare officers, nurses, social workers, and educationists, the local authority should be able to meet many of the social and educational needs of the handicapped child. If these provisions are to be fully

exploited, it is important that both family doctor and hospital-based practitioners maintain close liaison with the statutory bodies.

Some parents' educational ambitions for their children are quite unrealistic. It is usually unwise—and sometimes mischievous—to ally oneself to an educational crusade on the parents' behalf without first being fully aware of the facts of the situation—which includes a knowledge of the child's psychological assessment, parental motivation, and locally available facilities.

Voluntary Agencies

Many voluntary agencies were founded before the advent of the National Health Service, but there is no doubt of the continuing need for their help, both as an outlet for independent largess transcending fiscal and statutory obligations, and as supplementary sources of social support. Many also support research.

A list of agencies which may be helpful is given as an appendix, since many parents who feel that their family problem is unique are helped by being linked with a national organization directed to their particular needs. In some areas parents' groups exist, which serve not only as useful foci of mutual self-help but also as much-needed pressure groups when necessary. Such groups can often relieve the desolation and impotence experienced by some parents, and introduce handicapped children to a new range of social stimuli.

Conclusion

The general approach advocated here owes more to the art than the science of medical practice. The benefits are undramatic, but the need for this help was never greater, as the scientifically inspired salving of neonates and young children who might otherwise have died continues apace.

Though support for the chronically handicapped may demand skills and interest which earlier training has done little to inspire or foster, the changing pattern of medical care must encourage the community-based doctor to undertake this responsibility, utilizing the steadily improving facilities that are available, and collaborating with the various bodies whose aims are similar to his own.

Appendix : List of Voluntary Agencies

The Association for Spina Bifida and Hydrocephalus, 112 City Road, London E.C.1.

The Invalid Children's Aid Association, 126 Buckingham Palace Road, London S.W.1.

The British Epilepsy Association, 3–6 Alfred Place, London W.C.1.

The Muscular Dystrophy Group of Great Britain, 26 Borough High Street, London S.E.1.

National Association for Deaf, Blind and Rubella Children, 61 Senneleys Park Road, Birmingham 31.

The National Deaf Children's Society, 31 Gloucester Place, London W.1H 4EA.

The National Society for Autistic Children, 1a Golders Green Road, London N.W.11.

The National Society for Mentally Handicapped Children, 86 Newman Street, London W.1P 4AR.

The Spastics' Society, 12 Park Crescent, London W.1N 4EQ.

The Scottish Council for the Care of Spastics, 22 Corstorphine Road, Edinburgh 12.

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