Medical care in severe mental handicap

The care of children with severe mental handicap—that is, with an intelligence quotient (IQ) of 50 or less—can involve most doctors. Long term medical supervision of a child with a seriously distorted pattern of development is regarded as normal practice, whereas regular medical follow up of all adults with such handicap may not be appropriate.

There is great advantage when the child's care largely involves the paediatrician in the earlier years, with referral and consultation as required with the consultant in mental handicap; reversal of these roles should occur as the child advances into later adolescence, although recruitment into the specialty of mental handicap is proving problematical. (Report of the Working Party on the Medical Care of Children with Mental Handicap, 1982.)

In a condition that cannot be cured much can be done to ameliorate the effect of the disability on the child and his family. The doctor will need to draw on the whole range of his professional skills in giving tolerant, compassionate counselling, while providing practical medical care. This medical care can be considered in various stages.

Detection

Detection of severe mental handicap may be immediately after birth, as in the case of Down's syndrome or other major abnormality, but more frequently detection is delayed while suspicions by health visitors, general practitioners, or paediatricians crystallise into fair certainty over the first months or year or two of life. It is often here that parents may feel that doctors have concealed their knowledge of the diagnosis for a period, when indeed the only reticence has been to share a suspicion that may prove to be unfounded.

Diagnosis

Cause. With a prevalence of about 3.5 per 1000, severe mental handicap must be the most common major disability in children. Although it may be possible to determine whether the origin of the disorder is prenatal, perinatal, or postnatal in up to 86% of children,¹ frequently this does not mean that a cause is established or a diagnosis made. About 30% of children with severe mental handicap have Down's syndrome, in which a diagnosis will be made without difficulty. In practice, when faced with a child with severe handicap who does not have

Down's syndrome a diagnosis may be established only in about half. It is an added burden for the parents then that the doctor is unable to offer an explanation why their child has one of the most devastating disorders that can occur. Indeed, this may reduce their confidence in him at a time when such confidence is so important for counselling on the many other issues that need to be faced. For this reason, among others, full investigation of the child is essential and is preferably done when the child is small.

In all children chromosome analysis should be undertaken and in boys a request for examination for fragile X considered. Serology for congenital infection and a metabolic assessment, including urine mucopolysaccharides in all, should be performed. Although the useful diagnostic yield from computed tomography of the brain is small, it should be considered for children who have infantile spasms, uncontrolled epilepsy, macrocephaly, or primary microcephaly. A major brain malformation may be found when cerebral palsy or facial dysmorphic features are present, even if there has been an adverse perinatal course. Parental pressure for a scan probably should not be resisted as they may wish to ensure that all possible tests have been done. 'New' diagnoses such as Rett's syndrome in girls² should be considered, and for most children referral to a geneticist is helpful for assistance in diagnosis, for sibling recurrence risk assessment, and for antenatal diagnosis.

Severity. An assessment of the severity of handicap is most important but may need to be delayed for one or two years to allow the rate of progress to be evaluated.

There is within the group of children with an IQ of 50 or less a wide range of ability. About a quarter of the children will be profoundly handicapped throughout their lives, requiring total care for all their needs, such as feeding or mobility. For up to two thirds of children it can be reasonably expected that, as adults, they will be largely independent for self care, and supervision of independent life in the community will be minimal.

When discussing with parents the future for their child's life it is useful to use the information available based on the severity to discuss a 'worst case' and 'best case' outcome for adult life. The advantage thereafter is that the doctor and all involved in management of the child can share with

parents their hope of achievement of the best outcome while feeling that a realistic picture has been painted of future life. It may be important to emphasise to parents that limited potential is compatible with a fulfilled life for the child. Developmental progress may extend into the late teens and early adult life, especially in the acquisition of mobility and social skills. Nevertheless, the child and adult's development and happiness may be threatened by the provision of a caring environment inappropriately mature for his needs or by placing expectation and demand upon him greater than he can achieve.

Associated disorders. Assessment of the presence and severity of cerebral palsy, psychotic features or other behavioural abnormalities, epilepsy, defects of vision or hearing, and cardiac and renal disorders will require collaboration with, among others, specialists in orthopaedics, ears, nose, and throat, ophthalmology, and psychiatry, some of whom thankfully have developed expertise in the assessment of these difficult patients. Joint clinics with paediatricians are ideal for this purpose but not always practicable. Plans of treatment and management will be agreed with colleagues in nursing, speech therapy, physiotherapy, occupational therapy, and psychology and based on the child's developmental level, especially in the interpretation of behavioural disorders. The doctor will need to liaise with education and social services departments to allow early and appropriate provision of nursery or school placement, financial benefits, and aids and home adaptations to alleviate difficulties in caring for the child. The doctor's and health workers' role in this may be considerable because although some specialist social workers are very helpful in this area equally many do not yet seem to accept this as part of their work with families of children with severe mental handicap.

Clinics held in the school are of great help to such management issues, but it must be recognised that some parents will find the numbers present rather inhibiting, and additional private consultation should be offered.

Later complications. Follow up over many years will allow support to be given and also detection of conditions requiring medical management, such as deterioration of hearing in Rubella or cytomegalovirus infection; of development of ocular disease or thyroid disease in Down's syndrome; emergence of hydrocephalus in congenital toxoplasmosis or of a neoplasm in tuberous sclerosis; of deterioration of gait from peripheral neuropathy or of emergence of absence epilepsy that may reduce learning ability.

Management of acute respiratory or urinary tract infection, which may take prolonged or unusual forms in such children, requires the expertise of the paediatrician.

The restrained and judicious use of anticonvulsant drugs may be necessary, but unorthodox measures, such as the use of long term antibiotics to prevent otitis media and effective treatment of constipation, may be a more appropriate response to an increase in epilepsy than a change of anticonvulsant medication. Drugs to modify behaviour or sleep are frequently ineffective and are associated with a high incidence of side effects in such children.

It is important for the doctor to keep abreast of the voluntary societies' informal publications and to be prepared to discuss various treatments suggested in a positive way. Dietary manipulation with medical involvement is rather less likely to do harm.

Management issues

Community care for children with severe handicap is largely parent care. There is little doubt that most, and probably all, such children can live at home, whether that be with their natural parents or in a house with 'parents' provided by society in the form of foster parents, small staffed local authority or National Health Service (NHS) houses, or, less desirably, in larger group homes. It is essential that the community provides adequate resources to support such patterns of care, and the doctor will find himself involved in pressing for such support. Provision is variable over the country, but these resources must include adequate respite care sufficient to respond to emergency as well as planned needs. There has been some debate as to whether parents should be persuaded to use such facilities.² Certainly, those caring for the most disabled children and adolescents should be, and the chance of success will be greatly improved by close liaison before, during, and after placements, between parents, school, care giving staff, community mental handicap nurses, and social workers.

Mental handicap hospitals have become a major resource for respite care, and this interim arrangement must not be withdrawn until an equivalent amount of care facilities in more desirable environments can be made available. When small staffed community units, either for long term residents or short term care, are provided it is essential that staffing levels are such that the life of the child will be improved considerably on care in mental handicap hospital, although this will be costly.³ The provision of medical care to children at home will be from the general practitioner and hopefully, with increasing movement of such children into this

pattern of care, his experience in the management of their needs will increase. General practitioner training programmes should include this experience.

The paediatrician, whether community or hospital based, and the consultant in mental handicap, will be available for consultation and visits as necessary. Admission of children for long term residence in mental handicap hospitals will cease as the Department of Health and Social Security guidelines increasingly take force, and there is, in fact, movement of long stay residents into the community.⁴ Nevertheless, NHS treatment resources for children with severe mental handicap should continue to be available to support care in community, and admission to such facilities under the care of a paediatrician, as well as a consultant in mental handicap, may be necessary for periods up to weeks or months for the assessment and management of severe disruptive behaviour patterns, as well as sometimes for prolonged convalescent care of children in adolescence after physical illness or operation. For children up to the age of 16 years, some of these needs may be met in a paediatric department of a district hospital. For the adolescent of 16–19 years, the environmental and nursing needs may still be those of a child, and these will not be met in an adult ward in a district hospital.

There are better facilities available for the support of the child and adolescent at home compared with support for the severely disabled adult. The person should be allowed the opportunity of achieving the dignity of an adult life independent of his parents

and, although the parents usually will wish to continue his care, the level of burden of care placed on them should be lightened by increasing society's share of his care as he enters adult life, by providing adequate day occupational care and periods of planned residential short term care.

Progress to such facilities should be uninterrupted after school leaving for fear of losing social and other skills, which may have been acquired painfully slowly. The doctor caring for children with severe mental handicap must be involved in advocacy and planning of facilities for those under his care who are approaching adult life.

References

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