The Worster-Drought Syndrome

A severe test of paediatric neurodisability services?

By Brian Neville

Paediatric neurodisability services are delivered by a multi-disciplinary team which has close working relationships with other paediatric medical specialities. That bland statement may conceal a lack of integrated practice able to cope with the common problems encountered in children with multiple impairments.

The Worster-Drought Syndrome (WDS)has been recognised as a type of cerebral palsy for 40 years. It is both under and late diagnosed and therefore the prevalence of this group of disorders is not agreed. The cerebral palsies themselves as a group of disorders are confusing enough, defined as motor disorders resulting from static cerebral pathology of early onset. The problematic parts of the cerebral palsies in terms of management are often those which are related to cortical grey matter damage i.e. epilepsy, cognitive impairment and psychiatric disorder. It is interesting that epidemiological studies of the cerebral palsies rarely mention the Worster-Drought Syndrome. Families in whom this diagnosis has been made met recently to inaugurate their parent support group under Contact-a-Family, and their problems in dealing with the statutory services were very clear. These problems come under several headings.

A Sense of Isolation

These families felt quite separate from their core group of disorders, that is, the cerebral palsies, because of the lack of recognition of both the syndrome and its nature.

The essential components of WDS are:

1. An upper motor neurone bulbar palsy.
2. A mild tetraplegia of pyramidal/mixed localisation.
3. An increased prevalence of epilepsy.
4. An increased rate of behaviour disorder.
5. Cognitive impairment.

Early Management Problems

The common presenting features in the first year of life are of a baby who feeds poorly, is unhappy and irritable, does not sleep well, whose development is slow and may show significant lack of social responsiveness. Frequently and understandably at this early stage, doctors cannot make any definite diagnosis. It is clear that even in children with more severe cerebral palsies, a positive diagnosis is not always made or possible until the child is rather older. Doctors should, however, examine carefully babies with feeding problems and specifically watch attempts at feeding to look for evidence of a deviant pattern of bulbar motor function, and one which is out of proportion to the other developmental problems. They should be aware that this is one way, in fact the commonest way, in which this particular condition presents.

Epilepsy

There is an increased incidence of epilepsy in this condition. This may start at any age, but sometimes can involve one of the more damaging early onset epilepsies, which may assume a high medical priority. The seizures may be of various types with an excess of attacks involving the bulbar structures. A specific syndrome of epilepsy called the opercular syndrome, defined by the bilateral peri-sylvan cerebral damage sustained usually in utero, has been defined, and has considerable overlap with WDS in which such scan findings are not commonly seen. Seizures occurring in a child with multiple neurological problems tend to be more difficult to control and doctors have to be careful not to prescribe either high drug doses or polytherapy in a way which may cause further impairment. For example, benzodiazepines, may particularly cause deterioration in bulbar functions, and barbiturates and sometimes valproate may increase irritability.

Motor Disorder

The gross motor presentation is of a mild or moderate motor delay in which walking may well be achieved between one and two years of age but progress to stable walking, running and climbing is delayed. A degree of upper limb organisational problems or 'clumsiness' are very common. Rarely, however, do the problems become recognised as a motor disorder early, despite there being pyramidal signs. None of the components of the motor disorder are usually sufficiently severe to excite specific interventions. Motor advice from an experienced paediatric

physiotherapist and occupational therapist may, however, be crucial in dealing with specific issues like the development of independent feeding.

Behaviour/Psychiatric Problems

Children with WDS commonly have a degree of attention deficit and hyperactivity disorder (ADHD). They suffer mood problems as they get older and some have features within the autistic spectrum. The recognition of these potentially treatable or manageable problems within multiple impairments is not always successful. However, it is a very great shame if a child who would respond to intensive management of ADHD is deprived of this because of the complexity of the clinical presentation.

Cognitive Impairment

The degree of cognitive impairment varies quite widely, but is not usually severe. This may present as early language delay and a global mild to moderate later impairment, sometimes with selective difficulties but without any specific patterns. Because of the additional problems of these children, they rarely seem to fit into an educational paradigm for the management of moderate learning difficulties, mostly because of their poor communication skills, behaviour problems and motor organisational difficulties.

Speech and Language

The speech and language problems vary from some children who have no useful speech for many years, to those with poor articulation and disorders that may be diagnosed as dyspraxia. The term dyspraxia, however, should be reserved for selective neurological impairments, not a global disturbance of all bulbar functions. Early help with communication by simple communication systems is helpful and some children later require more sophisticated aids. However, because of additional cognitive, behavioral and motor organisational problems, such interventions may fail.

Dribbling may be a very persistent problem and if it goes on beyond four to five years of age, transposition of the submandibular ducts, or occasionally drug treatment may be considered. Such management should be combined with the advice of an recognised area of need in children with cerebral palsy and some dysphagia teams exist, though children with the WDS will of course require wider skills than just of feeding.

Where there is an upper motor neurone bulbar palsy a high rate of gastro-oesophagal reflux occurs, particularly in early years. This is because of the control of oesophagal peristalsis that normal volitional swallowing engenders. Thus the child may be having problems with oesophagal pain, acid inflammation and 'vomiting'. Oesophagal pain may often not be easy to recognise and present as a sleep disturbance.

As a result of problems with feeding, early malnutrition is a significant problem in many. Thus, artificial methods of feeding are often advocated and need management in their own right. Early nasogastric feeding is quite common. Gastrostomy feeding should be considered if a specialist dysphagia team have thoroughly investigated and advised on management. Issues involving the feeding itself, nutrition, airway protection and oesophagal reflux and the predicted natural history have to be weighed in making such decisions.

Genetic Counselling

This disorder is usually an unexplained form of cerebral palsy and as such disorders incur a relatively high recurrence risk, but one which is not very well researched. A appropriately experienced dental department because of the common secondary problems of alterations in jaw morphology and tooth eruption in this syndrome. It is important, however, that a surgical solution to one aspect of WDS is not grasped in desperation by doctors because they are unable to know what to do next. These children also have a high rate of middle ear infection and fluid accumulation secondary to the bulbar palsy. The insertion of grommets and particularly the performance of adenoidectomy should only occur after full consideration of the underlying problems and the likelihood of recurrence.

Respiratory Problems

From an early stage it may be clear that the child is choking during feeding, either on liquids or solids. However, sometimes this is not always obvious and recurrent episodes of bronchospasm, lower respiratory infection, and even brochiectasis may occur. 'Asthma' is very commonly diagnosed and is of course a common disorder, but many episodes of wheezing seem likely to be due to aspiration. Management of this aspect of the problem has to be closely linked to that of feeding.

Feeding Problems

Bulbar problems vary in their localisation, sometimes being more anterior, others having more difficulty with posterior aspects of deglutition and airway protection, but many children suffer problems in all areas, both early sucking, difficulties with chewing and problems with efficient swallowing. Fortunately this area is now a number of patterns of inheritance have been described and thus genetic counselling is required on the basis of the correct diagnosis.

Conclusions

The above series of problems are those that one might expect in children with a mild /moderate tetraplegic cerebral palsy. The crucial difference in the setting to that of severe tetraplegia is because of the children's mobility and communication needs. Close to their home families require an appropriate level of diagnostic skills, not just of the underlying neurological disorder, but of the range of specific problems that arise in WDS children. The potential for them to attend four, five, six or more specialist clinics is there, but this is normally counter-productive and would leave the families with very difficult problems in co-ordinating their child's care.

It seems reasonable to use WDS as a target condition to test the effectiveness of multi- disciplinary child development teams at secondary level to see if they have in place systems to manage such children. The founding of the parent support group for children with WDS has highlighted the need for comprehensive services which are fully integrated at secondary level and the need for easy access to specialist tertiary level services. The co-ordination between secondary and tertiary services is also the responsibility of the service, not the parents. A key worker is essential.

Educational provision is similarly very difficult because of the multiplicity of problems which, though often relatively mild in absolute terms, can cause major difficulties in using the tactics for teaching children with more focused needs.

It seems highly probable therefore, that if a child development service can manage WDS well, it can manage anything. The parent group* are keen to assist in developing such services. WDS Support Group can be contacted at**www.wdssg.org.uk** or through Contact-a-Family at:

Contact a Family,
209-211 City Road, London EC1V 1JN

Tel 020 7608 8700 Fax 020 7608 8701 Minicom 020 7608 8702
Helpline 0808 808 3555 Freephone for parents and families (10am-4pm, Mon-Fri)
e-mail: **info@cafamily.org.uk**

Worster-Drought C (1956) Congenital supra-bulbar paresis. J. Laryng. Otol. 70. 453