Recent advances in paediatric neurosurgery

Neurosurgery has benefited from the enormous leap in diagnostic radiology provided by computed tomography and magnetic resonance imaging. Also, important technological 'tools' are now available to neurosurgeons. Against this background I will review some of the recent advances in paediatric neurosurgery.

Disorders of cerebrospinal fluid circulation

The introduction of valve regulated shunt mechanisms in 1952 was a significant advance in the management of hydrocephalus. However, the problems highlighted then of malfunction, infection, and overdrainage continue to cause concern. Altogether 80% of children will require at least one shunt revision within 10 years of insertion, some many more. Shunt revision is a procedure with morbidity and mortality. However, significant advances have been made and the goal of achieving a well functioning shunt with a low complication rate is achievable. The condition leading to hydrocephalus should now be the main determinant of outcome, as the hydrocephalus should be well controlled. With improved operative technique, obstructing lesions such as tumours are safely removed and the need for shunt avoided. The aim is to avoid the need for a shunt wherever possible.

SHUNT MALFUNCTION

With improvement in shunt hardware the previously common problems of tube fracture, disconnection, migration, or other material failures are rare. The most common cause of shunt malfunction now is related to choroid plexus occlusion of the ventricular catheter. At shunt insertion, the perforated distal 1-8 cm of the ventricular catheter is ideally placed in the frontal horn, a region where there is no choroid plexus. Performing this under direct vision with a ventriculoscope is currently under evaluation. However as the young child’s head grows and changes shape, the catheter position alters and may again come in contact with the choroid plexus.

INFECTION

Ninety per cent of shunt infections occur within six months of insertion, 70% within one month. Shunt infection in the majority of cases is a complication of shunt surgery. Attention to surgical technique, appropriate skin preparation, and asepsis are considered most important. The role of antibiotics appears to be limited to their perioperative use only. A recent report suggests that the infection rate may be reduced to less than 1%, though most series report rates of 2-5%. When shunt infection occurs, removal of the shunt, a period of external ventricular drainage, eradication of infection, and then reinserstion of the shunt are the steps required in the majority of cases.

OVERDRAINAGE

Most commercially available shunts include differential pressure values: once the valve opens flow is determined by the difference between the input and output pressure. When a child is upright a 'siphon' effect occurs due to the height of the hydrostatic column of cerebrospinal fluid between the inlet and outlet of the shunt. This 'siphon' effect results in periods of overdrainage of cerebrospinal fluid. Overdrainage predisposes to slit ventricles, slit ventricle syndrome, subdural haematoma, encysted fourth ventricle, and craniosynostosis after shunt.

A new approach aims to provide cerebrospinal fluid drainage at or below its secretion rate within a physiological intracranial pressure (ICP) range. A number of valves are now commercially available that aim to provide this but remain unproved in prospective randomised studies. It is postulated that these flow regulated valves will prevent overdrainage and reduce the associated problems. Preventing slit ventricles probably reduces the incidence of ventricular catheter occlusion as it reduces the risk of the choroid plexus coming in contact with the ventricular catheter.

ASSESSING CEREBROSPINAL FLUID DYNAMICS: THE NEED FOR SHUNT OR SHUNT REVISION?

In the majority of children with hydrocephalus the necessity for the initial shunt or the requirement for revision is clear. The clinical signs and symptoms of progressive hydrocephalus supported by ultrasound or computed tomography findings are conclusive. However in some, the clinical decision is difficult. A child with a mild degree of macrocephaly and enlarged ventricles may not have an active hydrocephalus. Intermittent symptoms of shunt malfunction not supported by ventriculomegaly on computed tomography does not always imply a properly functioning shunt.

Certainly modern radiology and ultrasound have made an enormous impact but in some children a more dynamic assessment is required of cerebrospinal fluid circulation. ICP monitoring is not new; but a more complex assessment of the state of cerebrospinal fluid circulation using computerised, consistent rate, lumbar infusion studies is possible. The resting cerebrospinal fluid pressure and resistance to cerebrospinal fluid outflow is measured and abnormal pressure waves detected.

Endoscopy

Endoscopic neurosurgery is undergoing an exciting rebirth. The first endoscopic neurosurgical procedure was performed in 1910, not by a neurosurgeon but by a urologist, V L Lespinasse in Chicago. A small rigid cystoscope was used to fulgurate the choroid plexus in two infants with hydrocephalus. One child died immediately, and the other died five years later.

With burgeoning interest in endoscopy in general due to the rapid advances in optical technology, neurosurgeons have developed a renewed interest in therapeutic endoscopy. Successful endoscopic third ventriculostomy without significant morbidity in children with non-communicating hydrocephalus is now reported. Endoscopic
biopsy, the management of intraventricular tumours, dissection of intraventricular cysts, septum pellucidum fenestration, and communication of large arachnoid cysts with the normal cerebrospinal fluid pathways are all described. Modern flexible endoscopes have multiple working channels which allow a variety of instruments, including laser, to be used. The ventriculoscope may prove more useful in simplifying the process of shunt revision and eliminating the need for shunts in others. The role of choroid plexectomy remains uncertain. The major nature of the procedure, the inability to fulgurate all the choroid plexus, and the production of cerebrospinal fluid in extrachoroidal sites are all thought to limit its usefulness.

Head injury
The benefits of the road safety campaign and the requirement for safe, secure restraining of children in motor vehicles has had effects obvious to all. The slowness to promote and possibly enforce the wearing of safety helmets for cyclists is difficult to understand.

Children are particularly at risk from the low velocity injuries that safety helmets are most effective against. The secondary brain insult from a developing extradural haematoma often follows a low velocity injury that has resulted in a skull fracture.

The importance of attending to the basics of appropriate resuscitation and the early detection and prompt evacuation of intracranial haematoma cannot be overemphasised. The paediatric intensive care management of head injuries has become increasingly more complex. Strategies to control ICP are important and maintaining adequate cerebral perfusion pressure (CPP) is vital. CPP equals the mean arterial blood pressure minus the ICP. Secondary preventable ischaemic insults occur when the CPP falls below a critical value. As well as attending to control of ICP, maintaining adequate CPP with careful attention to cardiovascular dynamics and the use of ionotropic support where necessary is important. There is no place for fluid restriction.

In the central nervous system, ischaemia and subsequent reperfusion leads to the production of oxygen radicals and lipid peroxidation of neuronal cell membranes. This mechanism of neuronal injury is presently the focus of attention to improve outcome after central nervous system injury. The antioxidant 21 aminosteroids (lazaroids) were developed as compounds devoid of glucocorticoid activity but with greater antioxidant activity. They are currently being studied in severe head injury.

A better understanding of the mechanisms of raised ICP after head injury has allowed more specific targeting of treatment. Osmotic diuretics may not be the appropriate treatment for the diffuse congestive brain swelling that is more common in children, as they worsen the vascular congestion.

Neuro-oncology
The considerable improvements in operative technique and instrumentation have allowed substantial improvements in management. The biological activity of the neoplasm and the involvement of vital central nervous system structures now limit the surgeon. Surgical removal of neoplasms, when possible, is often the treatment of choice. Adjuvant therapy may be required. However, radiotherapy or chemotherapy alone or in combination are the primary treatment modalities of some central nervous system neoplasms. Surgery may be limited to biopsy alone. Radiotherapy is harmful to the developing brain. Its use is avoided below the age of 3 years. However, radiotherapy treatment protocols are changing and becoming safer and more effective.

Chemotherapy continues to have an increasing role both as adjuvant and primary treatment. It has replaced radiotherapy in those under 3 years of age. Enhanced techniques to limit chemotherapy resistance possibly combined with biologic response modifiers and monoclonal antibodies will be the treatment of the future. The challenge presented by neoplasms in those under 18 months remains formidable.

Technological advances
Neurosurgery benefited greatly during the 1980s from technological advances. Improved operating microscopes gave better magnification and illumination. They became more user friendly. Combined with lasers and ultrasonic aspiration, morbidity and mortality were reduced. Our colleagues in neuroanaesthesia have provided improved operating conditions and a more tolerant brain. Computed tomography guided and later magnetic resonance imaging guided stereotaxis allowed accurate biopsy and microsurgical procedures to deep targets within the brain. Robotics and frameless stereotaxy are being developed.

Lars Leksell, a Swedish neurosurgeon, combined the accuracy of modern stereotactic equipment with a single necrotising dose of radiotherapy and coined the term 'radiosurgery'. This treatment method was proved most successful in the treatment of vascular malformations. It causes endothelial damage and progressive vascular occlusion. In the two years after treatment greater than 80% will occlude. It is limited by a vascular malformation nidus size of 3 cm in diameter or less. The combination of interventional radiotherapy, microsurgery, and radiosurgery have resulted in significant advances in the management of vascular malformations. A role for stereotactic radiosurgery is also unfolding as the prime or adjuvant therapy in some tumours.

Other important advances
Identifying medically intractable epilepsy patients who may be suitable for surgery is important. Surgery has a clear role in intractable complex partial epilepsy due to mesial temporal sclerosis, temporal heterotopic grey matter, and benign neoplasms of the temporal lobe. After proper evaluation, modern surgical techniques offer a high chance of complete abolition or significant improvement in seizure control. These children should be identified and offered treatment early to allow them the maximum chance of normal education and social interactions. There remains a small but definable role for hemispherectomy and corpus callosal section.

The procedures offered by craniofacial units continue to expand. Three dimensional computerised reconstruction allows better appreciation of the craniosynostosis and more complex craniofacial anomalies. Cosmesis remains the main indication for surgery but in a group of children correction of the more complex craniosynostotic anomalies is required to allow brain growth and avoid increase of ICP.

The incidence of congenital open spinal defects is decreasing. The later complications of dysraphic states that lead to neurological deterioration (retethering, Chiari malformation) should be remembered. Lipomyelomeningocele needs early evaluation to avoid predictable deterioration. Selective posterior rhizotomy for lower limb spasticity of cerebral palsy is commonly performed in the USA. There remains significant polarisation as to its exact role.
Body composition assessment

There is increasing recognition and awareness of the inability of classical measures of growth and development to meet the requirements of modern clinical medicine and biological research. Measurements such as weight and length provide useful but incomplete data relating to the growth or nutritional status of a child. Thus, more detailed assessment of body composition is being sought by those interested in paediatrics, nutrition, growth, and development.

There are numerous techniques that are used to assess body composition, the majority of which use a two compartment model: that is to divide the body into fat and fat free masses. Nevertheless, many such techniques are not easily applied in children because of either practical or ethical considerations. For example, underwater weighing to assess body density and hence body fat mass is clearly not applicable to young children. However, there are numerous approaches that can be used to assess body composition in the paediatric population each with their own advantages and disadvantages. The range of the technology utilised and the complexity of the various techniques is wide. Some of these techniques are described below.

Bioelectrical impedance
This technique has become extremely popular in recent years. It is based upon the fact that the electrical resistance to a flow of current of 50 kHz in the human body is related to the amount of total body water and hence fat free mass. As the technique is non-invasive, simple, precise, and relatively cheap it would seem to be an ideal body composition tool for use in paediatrics. There have been a number of validations of the technique and in contrast with anthropometric methods bioelectrical impedance seems to be much less population specific. However, problems have been reported with its use in the neonatal period but these may be associated more with the difficulty of validation in this population rather than the model itself. As the technique theoretically predicts total body water care should be taken in assuming that the same degree of precision can be applied to derived estimates of fat free mass. The conversion of body water to measures of fat free mass assumes a constant level of hydration in lean tissue. Even so the correlation between bioelectrical impedance and estimates of body cell mass via the measurement of total body potassium has been shown to be high. Current research in this area indicates that a dual frequency or multifrequency current might enable both intracellular and extracellular water to be measured. Users of bioelectrical impedance apparatus should, however, ensure that the equations built into any associated software supplied with the equipment are appropriate for children.

Assessment of total body water
It is of course possible to measure total body water rather than use prediction equations. The most straightforward approach is to use an isotope of either hydrogen or oxygen in the form of water and apply a standard dilution principle. Tritiated water ($^3$H$_2$O) is not normally used in children because of the radioactive nature of the isotope but deuter-
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