Hydrocephalus

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Hydrocephalus is a sign of an underlying pathological process. These processes can be congenital or acquired postnatally. They range from malformations, infections and metabolic disorders to neoplasms. Unfortunately, we have made little progress in the last third of the 20th century in either our understanding of the pathophysiology or treatment of hydrocephalus. The long-term mortality and morbidity of our treatment remains disturbingly high. It is also unfortunate that there are two levels of progress, one for developed countries and one for developing countries. The alarming thing here is that the vast majority of children do now and will for the foreseeable future live in developing countries.

The future can be looked at as the next 10 years and beyond 10 years. In the shorter term, we hope for a reduction in complications of our management, earlier diagnosis and obviously more awareness by medical caregivers.

Prevention of the causes of hydrocephalus is a major area where progress can be made. If every woman of child-bearing age took a daily vitamin containing 400 µg of folate, 70% of Chiari II malformations would be prevented. Immunization against meningitis, reduction in the number and severity of head injuries in children and prevention of prematurity would be other areas where progress should be made.

Intrauterine repair of the myelomeningocele appears to reduce the incidence of hydrocephalus requiring a shunt. We will likely revisit the question of intrauterine management of fetal hydrocephalus.

In mice, there are probably a dozen known genetic defects that cause hydrocephalus. We know of an X-linked form of hydrocephalus in humans and PAX3 as a

cause of neural tube defects, and we suspect that there are multiple other genetic forms. With the completion of the human genome project, prospective parents will be able to be screened for known genes causing disease. It will also be possible to screen the embryo. This will present several ethical issues which need to be anticipated. In parents with known genetic defects, fertilization can be managed to limit the deleterious genes passed to the embryo.

Technology will offer a wide variety of both diagnostic and management tools. Micro CT and MRI with resolution down to 50 µm and below already exist. The imaging of single brain cells and gene expression is now being developed.

Only as our understanding of the pathophysiology of hydrocephalus progresses will our ability to use technology to build the 'perfect' shunt. The microchip technology is already available. Some vital questions need to be answered first.

In the more distant future, repair of the pathological defect that causes the hydrocephalus will become a reality. Using the child's own tissues is the preferred answer. Venous and skull base reconstruction will add to magnetic-guided ventriculostomy as hardware-free correction. Regeneration of the subarachnoid space or arachnoid villi, possibly using the child's own stem cells, are possibilities.

As we move beyond 2000, the possibilities are great. The fact that most children will not share in medical progress and the ethical issue needs immediate attention. Funding of the development of these areas and preservation of the pediatric neurosurgeon scientist must also be a priority.