Management of Intrauterine Hydrocephalus

To THE EDITOR: The technology that allows intrauterine shunting to be performed with relative ease has raised an issue addressed indirectly by Drs. Cochran and Myles in a recent article (Cochrane DD, Myles ST: Management of intrauterine hydrocephalus, J Neurosurg 57:590–596, November, 1982). Their description of the associated central nervous system and other organ system anomalies in a significant number of infants with intrauterine hydrocephalus is a cause for concern and reflection. It is generally accepted among neurosurgeons who see a large number of infants with hydrocephalus that it is the hydrocephalus not (with rare exception) the degree of ventriculomegaly that determines outcome in the properly managed neonate. Indeed, since the cerebrospinal fluid circulation (CSF) is necessary for the development of CSF pathways, placement of an in utero shunt may convert a potentially reversible hydrocephalus into an irreversible shunt-dependent hydrocephalus. In my own experience, two infants with clearly documented intrauterine hydrocephalus (one diagnosed at 34 and 36 weeks gestation, and the other at 37 and 39 weeks gestation) had only moderate ventriculomegaly on postnatal computerized tomography scanning, with widened subarachnoid spaces. Both infants went on to develop normally without the need for shunting. Had a shunt been placed in utero, one wonders if the outcome might have been otherwise.

I think serious consideration should be given to placing a moratorium on intrauterine shunting until several issues are resolved: 1) Since little correlation has been shown between ventricular size and eventual psychomotor achievement, can one justify shunting a hydrocephalic fetus in utero rather than under more controlled conditions after birth? 2) What is the natural history of hydrocephalus in utero? What percentage of cases will resolve spontaneously, and is there an association between the time of onset of hydrocephalus and the occurrence of associated CNS abnormalities (for instance, agenesis of the corpus callosum, heterotopia, and deficient corticospinal tract), none of which are identifiable by ultrasound with present techniques? 3) A significant number of these procedures have already been done. What has been the outcome in these infants?

The age-old dictum, “primum non nocere,” holds as true today as in the days of purges and leechings and deserves careful consideration by the proponents of new technology.

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RESPONSE: Dr. Venes’ letter is timely and provocative and raises a number of issues that have been of concern to us.

Complex management problems arise when a fetus is diagnosed as having hydrocephalus in utero. At the University of Calgary, we have attempted to provide the involved families with input from experts in all relevant areas. Our interest group includes obstetricians, ultrasonographers, neonatologists, neurosurgeons, neurologists, and a geneticist. We have now had experience with fetuses diagnosed in utero as having intracerebral hemorrhage, cysts, and hydrocephalus, and in each case have individualized the treatment plans. Postnatal surgical intervention has only been recommended after consideration of the factors outlined in our paper.

Ventriculomegaly is the most easily detectable abnormality of the nervous system on ultrasound studies, although with further experience other congenital anomalies may be recognized as well. To date, we have not seen a fetus with isolated nonprogressive ventriculomegaly. In our institution, such a fetus would not be treated operatively, either in utero or after birth. Periodic evaluations of ventricular size would be carried out and, in the event of progressive ventriculomegaly, reassessment as to the cause of the intracranial hypertension would be made prior to any surgical intervention.

In fetuses such as the ones we studied that exhibited progressive ventriculo- and craniomegaly, the importance of a careful search for associated anomalies and/or infection must be emphasized. It is our opinion that these factors determine eventual neuropsychological outcome, assuming that there is adequate control of the hydrocephalus. Determination of the extent and nature of associated spinal dysraphism remains difficult with ultrasound, although with increasing experience the accuracy of the study will likely improve. Even with careful postnatal clinical and radiographic assessment, occasional associated anomalies that influence outcome will occasionally be missed. In the future, we will not recommend aggressive treatment for fetuses such as our Cases 4 and 5, in which marked asymmetric ventriculomegaly was diagnosed in utero.

We agree with Dr. Venes that more should be known of the natural history of hydrocephalus diag-
nosed in utero before treatment is started. In other areas of medicine where the natural history of a condition is unknown, treatment is rarely based on scientific data. Through collaboration, it should be possible to document the natural history of the various forms of ventriculomegaly that can be diagnosed in utero, and thus devise a more rational plan of treatment.

In our own experience, the only fetus that would have been considered for an intrauterine shunt would have been Case 6. The outcome in this case was excellent without prenatal shunting, and we do not believe that it could have been improved by placement of an intrauterine shunt. At the present time, we support Dr. Venes’ call for a moratorium on intrauterine shunting until the indications for the procedure and the clinical benefits can be more clearly defined.

We agree that neurosurgeons should take an active role in the early management of fetuses diagnosed as having hydrocephalus in utero, for neurosurgeons are in the best position to give advice on the significance of the demonstrated abnormalities and their possible treatment.

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Spontaneous Extradural Hematoma

To The Editor: Drs. Marks and Shaw report a case of spontaneous extradural hematoma (EDH) that they believe is inexplicable by current theories (that is, arteritis or progressive dural detachment by air or pus) (Marks SM, Shaw MDM: Spontaneous intracranial extradural hematoma. Case report. J Neurosurg 57:708–709, November, 1982). By citing an article of mine, they imply that I also would be unable to explain the underlying mechanism in their case. They do not seem to have read another of my publications in which I proposed that spontaneous EDH’s are secondary to cerebrospinal fluid (CSF) fistulas. This theory would explain previous cases and all of the important features of their own case, for which the following plausible sequence of events can be postulated:

1. The patient received a blow on the head or even a skull fracture, events not always subsequently mentioned to doctors. There was a bone fistula into the frontal sinus.

2. Cerebrospinal fluid leaked into the mucosa causing polyps. (This continual soaking of spongy tissues can cause ipsilateral nasal polyps.)

3. Overloading of the lymphatic drainage system occurred, hence the sinusitis.

4. The CSF dripped down into the lungs, probably during sleep, causing asthma. (Sinusitis, especially hyperplastic polyoid rather than purulent sinusitis, is strongly linked with asthma, probably via aspiration of fluid. A case of chronic asthma was cured by probing a cerebral abscess so that it drained out through the nose.)

5. Four days before the EDH, the fistula healed, and hence the headache. (The headache is often related to CSF rhinorrhea, and is relieved by decompression.)

6. Asthmatic cough or a sneeze caused elevation of the dura, tearing of a blood vessel, and EDH. Air may have passed up the still patent distal fistulous tract without opposing centrifugal CSF flow.

7. Blood seeped back along the fistulous tract, causing the hemorrhagic nasal mucosa.

8. The fistula became blocked by blood or polyps. The CSF diverted into the periorbital space, causing the edema and strabismus. (If this was secondary to sinusitis, what was the source of the high orbital pressure and how was it sustained? Orbital swelling, diplopia, and headache may be immediately relieved by copious clear watery nasal discharge.)

The above analysis presupposes that CSF rhinorrhea is very common with EDH. In less profuse forms it is probably physiological, so this should cause no problem. The difficulty with which large fistulas are diagnosed and their serendipitous discovery does not inspire confidence that smaller ones will ever be spotted. One only needs to look as far as the preceding article in the same issue of the Journal, in which the authors describe a case of CSF fistula after lumbar puncture, which was only diagnosed after 5 years of strong suspicion and intensive investigation. Posttraumatic fistulas can present as perennial allergic rhinitis and give negative glucose test results. Such tests are quite misleading. Metrizamide cisternography may fail to show the site of concurrent CSF leakage. Even so, a clear-cut history of rhinorrhea may not be volunteered. Many cases only come to light because of meningitis. Watson-Williams admitted that it never occurred to him that his patient had a CSF fistula until the postmortem examination, despite the fact that the patient suffered recurrent headache and CSF rhinorrhea for at least 26 years. In 1901, he was only the second person to diagnose a case of CSF rhinorrhea in the British literature.

There also appears in the November issue of the Journal an article on epidural abscess in which the authors summarize a case of EDH in contact with a scalp abscess along a fracture line (see Table 1 of that article). Instead of Marks and Shaw using the history of probable recent head injury as a reason for excluding such cases from their review, they should have considered the head injury an indication of the likely etiology in cases of EDH of apparently spontaneous origin in which any infection is secondary and irrelevant. Likewise, a cerebral abscess can cause purulent rhinorrhea with pus pouring out of the nose in successive pulsations.

Marks and Shaw’s failure to find a dural lesion at operation is irrelevant. A meticulous postmortem ex-