Management of intrauterine hydrocephalus

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The aggressive treatment of hydrocephalus has been of benefit to many children. Sophisticated two-dimensional ultrasound techniques allow the diagnosis of prenatal hydrocephalus to be made with accuracy and ease. In the past, the medical decisions governing the management of hydrocephalus in utero were made by obstetricians and were directed at reducing maternal mortality and morbidity. Now, with improved diagnosis and support facilities for the newborn, neurosurgical input is being requested as more concern is expressed for the fetus.

Based on their experience with seven cases of intrauterine hydrocephalus in the past 3 years, the authors present their program for the management of this problem. If antenatal ultrasonography shows hydrocephalus without other anomalies, they recommend that the fetus be born by elective Caesarean section at the time of pulmonary maturity, and that early ventricular shunting be carried out. This plan should minimize nervous system trauma resulting from hydrocephalus and the birth process. If, however, antenatal diagnostic studies show cerebral or other major system anomalies in addition to hydrocephalus, then standard obstetrical care should be given. Antenatal ultrasonography has been found to be reliable in assessing fetal lateral ventricular size and shape, and to correlate well with the results of postnatal computerized tomography scanning.

KEY WORDS • hydrocephalus • prenatal diagnosis • ultrasonography

An aggressive attitude directed toward the treatment of infantile hydrocephalus has benefited many children. In the past, prenatal diagnosis of hydrocephalus was difficult, and labor often treacherous. Recent advances in high-resolution gray or B-mode ultrasonography have made available a safe, noninvasive, reliable method for the study of the fetus and its intrauterine environment. Using this simple technique, fetal anomalies including hydrocephalus have been detected during the second and third trimesters of pregnancy.\textsuperscript{4,9,11,13,14,16,18,19,27,32}
With this improved diagnostic ability, our obstetrical and pediatric colleagues have requested neurosurgical input into the decisions governing the management of hydrocephalus in utero in the hope of improving fetal survival. Our experience with seven recent cases forms the basis of this report.

Clinical Material and Methods

The clinical data for the seven cases in this series are summarized in Table 1. Two infants were stillborn, and the other five were delivered by Caesarean section. Ultrasonic examination was performed in all cases, using an Advanced Diagnostic Research linear real-time scanner or a Picker “B” scanner.\textsuperscript{*} Computerized cranial tomography (CT) was carried out with an EMI 1010 head scanner.\textsuperscript{†} Patients who required the placement of a shunt had a low-pressure Pudenz system inserted.\textsuperscript{‡}

Case Reports

Case 1

This woman had had a normal pregnancy and a healthy child at the age of 24 years. During her second pregnancy, the membranes ruptured at 40 weeks gestation. Obstetrical assessment was obtained because the mode of presentation could not be determined. This led to ultrasonic examination and the diagnosis

\textsuperscript{*} Linear real-time scanner manufactured by Advanced Diagnostic Research Ultrasound, 2224 South Priest Drive, Tempe, Arizona. Picker “B” scanner manufactured by Picker Corp., 595 Miner Road, Cleveland, Ohio.
\textsuperscript{†} EMI 1010 head scanner manufactured by EMI Medical Inc., 4000 Commercial Drive, Northbrook, Illinois.
\textsuperscript{‡} Low-pressure Pudenz system from Heyer-Schulte, P.O. Box 946, Goleta, California.
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TABLE 1
Summary of clinical details in seven cases of intrauterine hydrocephalus

<table>
<thead>
<tr>
<th>Case No. &amp; Sex</th>
<th>Reason for Ultrasound</th>
<th>Ultrasound Diagnosis</th>
<th>Route of Delivery</th>
<th>Postnatal &amp;/or Autopsy Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>1, F</td>
<td>determine mode of presentation</td>
<td>hydrocephalus</td>
<td>Caesarean section</td>
<td>hydrocephalus, aqueductal stenosis, Arnold-Chiari malformation, myelomeningocele</td>
</tr>
<tr>
<td>2, M</td>
<td>fetal-uterine disproportion</td>
<td>hydrocephalus</td>
<td>induced vaginally</td>
<td>stillborn; hydrocephalus, tracheo-esophageal fistula</td>
</tr>
<tr>
<td>3, F</td>
<td>fetal lie determination</td>
<td>hydrocephalus, spina bifida</td>
<td>Caesarean section</td>
<td>aqueductal stenosis &amp; myelomeningocele</td>
</tr>
<tr>
<td>4, F &amp; 5, F</td>
<td>twin gestation assessment</td>
<td>both hydrocephalic</td>
<td>Caesarean section</td>
<td>aqueductal stenosis, confirmed at autopsy</td>
</tr>
<tr>
<td>6, M</td>
<td>previous birth of a hydrocephalic stillborn</td>
<td>hydrocephalus</td>
<td>Caesarean section</td>
<td>aqueductal stenosis</td>
</tr>
<tr>
<td>7, F</td>
<td>spontaneous rupture of membranes</td>
<td>hydrocephalus vs. hydroencephaly</td>
<td>vaginal</td>
<td>stillborn; hydrocephalus &amp; holoprosencephaly</td>
</tr>
</tbody>
</table>

of hydrocephalus. At this time, there was no evidence of spinal dysraphism.

Spontaneous labor ensued and a grossly hydrocephalic female child was delivered by Caesarean section. The child’s head circumference greatly exceeded the 90th percentile. The entire lower thoracic and lumbosacral spine was the site of a large myelomeningocele, 6 x 4.5 cm in size. The infant had paralysis of the lower extremities and sphincters, bilateral neurogenic clubfeet, hydrouretors, and bilateral hydronephrosis. Postnatal CT scanning revealed massive lateral and third ventricular enlargement with a relatively normal posterior fossa. Nonoperative care was instituted, and the infant died at 5 weeks of life.

Autopsy examination showed a typical Arnold-Chiari malformation with a hypoplastic cerebellum and small posterior fossa. A dorsal medullary kink was found at the level of T-1. The additional anomalies described above were confirmed.

At 28 years of age, the patient again found herself pregnant. During this pregnancy, regular ultrasound examinations and amniocentesis for alpha-fetoprotein were carried out. The pregnancy proceeded without event. A healthy normal male infant was delivered at term by Caesarean section.

Case 2

Ultrasound examination of this 31-year-old primigravida woman was carried out at 29 weeks gestation to investigate rapid maternal weight gain and clinically suspected fetal-uterine disproportion. Hydrocephalus was apparent. Follow-up ultrasonography over the subsequent weeks showed lateral ventricular enlargement outpacing the increases in biparietal diameter. No other abnormalities were detected by ultrasound or by amniotic fluid examination. There was no evidence of intrauterine infection. The patient, family, and medical consultants elected standard obstetrical care. Labor was induced and a stillborn male infant was delivered vaginally after a 400-cc transabdominal encephalocentesis had been performed.

Case 3

This 30-month-old infant girl was born at 40 weeks gestation by elective Caesarean section. The diagnosis of intrauterine hydrocephalus had been made at 37 weeks when ultrasound examination was performed to determine the fetal position (Fig. 1 left). This study also suggested spina bifida (Fig. 1 right). Progressive increases in the biparietal diameter and the size of the lateral ventricles were documented over a 3-week period. The mother had had a previous normal pregnancy and one therapeutic abortion. There was no family history of congenital nervous system abnormalities.

At birth, the infant was found to be grossly hydrocephalic. She had a large 9 x 5-cm thoracolumbar myelomeningocele and bilateral clubfeet. The only neurological function below the level of the lesion was minimal hip flexion. Postnatal cranial CT confirmed...
the diagnosis of hydrocephalus and suggested it was due to aqueductal stenosis.

The family elected nonoperative care initially, but nursing care problems at home required the placement of a ventriculoperitoneal shunt and repair of the spinal defect. The child is now 30 months of age, her hydrocephalus is controlled by her shunt, and her lower extremity paralysis is unchanged.

Cases 4 and 5

Ultrasound examination was performed at 34 weeks on this women to assess a probable twin pregnancy. She had had two previous deliveries at term. Scanning showed two fetuses, both of which were hydrocephalic. One exhibited symmetrical ventriculomegaly (Fig. 2 upper left), while its sibling showed grossly asymmetrical lateral ventricular enlargement (Fig. 2 lower left). Over the course of 3 weeks, the biparietal diameters of both infants increased, so that at 37 weeks gestation they greatly exceeded the 98th percentile.

Elective Caesarean section was undertaken to allow an atraumatic delivery and early ventricular shunting. Monozygotic female twins were delivered. Both required resuscitation at birth. Cranial CT on their 2nd day of life confirmed hydrocephalus (Fig. 2 upper and lower right) and established aqueductal stenosis as the causative lesion in both twins. Ventriculoperitoneal shunts were placed later on the same day without complications.

Over the ensuing weeks, both infants developed respiratory failure requiring intensive pulmonary support, and major motor seizures which proved refractory to medical control. The first-born infant developed shunt obstruction at 8 weeks; revision was not performed. Both children died of seizure-related pulmonary problems at 12 weeks of age.

At autopsy, aqueductal stenosis with gliosis and forking was established as the cause of their hydrocephalus, and the presence of asymmetrical ventriculomegaly in one twin was confirmed. There was no evidence of shunt infection or ventriculitis, and it appeared that their hydrocephalus had been adequately controlled. Occipital polymicrogyria was also present.

Case 6

This 32-year-old woman underwent ultrasound examination because of a previous pregnancy complicated by hydrocephalus. Two other pregnancies had terminated normally and two had ended in the first trimester with spontaneous abortions. Ultrasonography established the diagnosis of hydrocephalus and confirmed progressive ventriculomegaly over a 3-week period (Fig. 3 left). Until 39 weeks of gestation, amniocentesis showed unacceptable lecithin/sphingomyelin ratios, and therefore elective delivery was postponed. When this test suggested pulmonary maturity, Caesarean section was performed and a hydrocephalic male infant was delivered atraumatically.

On the 1st day of life, CT scanning showed lateral and third ventricular enlargement compatible with aqueductal stenosis (Fig. 3 right). A ventriculoperitoneal shunt was placed on the 3rd day of life. The child tolerated this surgery well and at 2.5 years of age is developing normally. His hydrocephalus remains controlled.
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**Case 7**

This 21-year-old primigravida woman was admitted at 31 weeks gestation following spontaneous rupture of her membranes. Other than a mild upper respiratory tract infection during the early weeks of her pregnancy, the mother had suffered no complications. Ultrasound assessment revealed a normal biparietal diameter for the age of the fetus. However, gross ventriculomegaly was also evident, to the degree that the diagnosis of hydranencephaly was entertained. Spontaneous labor began 48 hours after admission and a stillborn female fetus with semilobar holoprosencephaly and hydrocephalus was delivered.

**Discussion**

Histological and ultrastructural studies of hydrocephalus in man and in experimental animals have shown that maximal brain damage occurs when ventricular dilation is acute and severe.\(^{26,37}\) Periventricular edema, flattening and disruption of the ependymal lining, and nerve fiber destruction in the subependymal white matter and the corpus callosum constitute the acute histological changes. As edema subsides, periventricular gliosis ensues with or without reconstitution of the ependyma.

Such observations suggest a sequence of events in the development of hydrocephalus.\(^{37}\) Following the obstruction of cerebrospinal fluid (CSF) outflow, CSF pressure rises and the CSF-containing spaces distend. In the ventricular system, this results in flattening of the ependyma and disruption of the intercellular junctions. Breakage of the CSF-brain barrier allows transependymal absorption of CSF into the extracellular space of the periventricular white matter, and impedes normal extracellular to CSF fluid fluxes. The resulting extracellular edema not only alters cellular architecture but may interfere with local cellular metabolism. The electron microscope has shown changes in the periventricular axons with disintegration of neurofilaments, neurotubules, and vesicles. Axonal degeneration and secondary demyelination are the end results. Proliferation of subependymal astrocytes and microglia follows thereafter. The continued proliferation of these cells has been taken as evidence of ongoing tissue damage during the late stages of the disease.\(^{30,36}\) Theoretically, optimal treatment results should follow when shunting is performed during the reversible stage of tissue damage.\(^{31}\)

Clinical studies support this concept. Satisfactory early shunting causes better neurological, intellectual, and cosmetic function in those cases where hydrocephalus is the only central nervous system insult.\(^{17,30,32}\) Prompt treatment, even in the face of extreme ventriculomegaly, may result in significant success.\(^{20,22,23}\) Delayed shunting in cases of progressive hydrocephalus, on the other hand, may not reverse already inflicted brain damage, as reflected by low intelligence quotient (IQ)\(^{29}\) or poor school performance.\(^{38}\)

In the past, the management of a pregnancy complicated by marked hydrocephalus of prenatal origin has been difficult. Not only did the diagnosis escape detection until labor arrested because of dystocia, but many cases ended in fetal and maternal disaster as the lower uterine segment ruptured. As a result, obstetrical decisions were made to minimize maternal morbidity and mortality, without concern for the fetus.\(^{15}\) Standard practice evolved to include needle or trocar drainage of the fetal head when dystocia arrested vertex presentations. As one-third of these fetuses are delivered in a breech fashion, techniques for CSF drainage through an associated myelomeningocele, surgical laminotomy, or posterior fossa puncture were described. With the recognition of massive hydrocephalus prior to delivery, these measures largely eliminated associated maternal mortality and morbidity. The fetus rarely survived. Feeney and Barry\(^{9}\) reported that 26% of their fetuses required obstetrical craniotomy for vaginal delivery, and none survived. Lorber and Zachery\(^{22}\) mentioned only one patient subjected to ventricular decompression who survived with a normal IQ.

Ultrasonic echography was introduced as a prenatal diagnostic technique by Campbell\(^{6}\) and colleagues in the early 1970's. Amniotic fluid analysis for the diagnosis of neural tube defects became available in the mid-1970's. Currently, these techniques, when combined with fetoscopy and intrauterine CT scanning in selected cases, allow reliable prenatal diagnosis of a variety of conditions.\(^{6,8,12,16}\) On the basis of this capability, prenatal treatment decisions can be made.

The anatomy of the fetal cranium is well within the reach of currently available B-mode ultrasound units. Scanning in the transverse plane can outline the lateral ventricles, the third ventricle, and the Sylvian fissures.\(^{6,7}\) Normally, the lateral ventricles become smaller in relation to the biparietal diameter as gestation proceeds. By the 17th week, the width of the lateral ventricles is less than 50% of the biparietal diameter.\(^{5}\) Increased reliability is obtained by measuring the distance between the lateral aspects of the frontal horns. At 13 and 40 weeks gestation, the bifrontal horn distance averages 1.1 cm and 2.4 cm, respectively. Confidence limits have been defined by Denkhaus and Winsberg.\(^{7}\)

During gestation, hydrocephalic ventricles increase measurably in size well before an increase in the biparietal diameter is recognized. This may allow the diagnosis of hydrocephalus to be made prior to the period of fetal viability.\(^{11}\) Later in pregnancy the diagnosis is easily made (Figs. 1, 2, and 3).

The diagnosis of spina bifida can be made when the fetal spine is scanned in the longitudinal or transverse planes. The normal spinal canal returns two parallel "tram line" echoes in the longitudinal scan and appears as a closed circle in transverse section.
Diversion of the "tram lines" or an open U-shaped configuration of the canal allows the diagnosis of spina bifida to be made\textsuperscript{6,12,27} (Fig. 1 right). This assessment is easiest between 16 and 20 weeks because the spinal curvature is minimal and the entire spine can usually be displayed on a single longitudinal scan. The assessment of fetal extremity movement and posturing may add valuable information.\textsuperscript{16}

Although the diagnostic return on ultrasound examinations is increased when specific problems are assessed, the usual indication for study is an obstetrical concern regarding gestational age, fetal lie, or twinning. Both oligohydramnios and polyhydramnios have been reported with intrauterine hydrocephalus. Abnormal lies and presentations are more common. Familial clustering of spina bifida and anencephaly is well recognized, as are cases of x-linked and autosomal recessive aqueductal stenosis. If ultrasound examinations become part of routine obstetrical care, more cases will be discovered incidentally.\textsuperscript{16}

When the diagnosis of hydrocephalus is established in utero, it is critical to document or exclude spinal dysraphism, intrauterine infection, and metabolic and genetic disorders. Amniocentesis under ultrasound will aid in this as well as provide valuable information regarding fetal maturity. Intrauterine CT has been assessed, the usual indication for study is an obstetrical concern regarding gestational age, fetal lie, or twinning. Both oligohydramnios and polyhydramnios have been reported with intrauterine hydrocephalus. Abnormal lies and presentations are more common. Familial clustering of spina bifida and anencephaly is well recognized, as are cases of x-linked and autosomal recessive aqueductal stenosis. If ultrasound examinations become part of routine obstetrical care, more cases will be discovered incidentally.\textsuperscript{16}

When the diagnosis of hydrocephalus is established in utero, it is critical to document or exclude spinal dysraphism, intrauterine infection, and metabolic and genetic disorders. Amniocentesis under ultrasound will aid in this as well as provide valuable information regarding fetal maturity. Intrauterine CT has been used by some,\textsuperscript{4} but we have not found it necessary to date.

The treatment options that we believe are available for the physician and family are summarized in Fig. 4. For the fetus diagnosed prior to the period of viability, elective abortion may be considered. The hydrocephalus will likely continue to inflict damage during the second and third trimesters of pregnancy.

**Possible Indications for Ultrasonic Examination**

<table>
<thead>
<tr>
<th>Pregnancy At Risk</th>
<th>Screening</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Polyhydramnios</td>
<td>- Gestational Age</td>
</tr>
<tr>
<td>- Previous HS of Spinal Dysraphism or Hydrocephalus</td>
<td>- Lie</td>
</tr>
<tr>
<td>- Amniotic Fluid Analysis</td>
<td>- Anomalies</td>
</tr>
</tbody>
</table>

**Possible Ultrasound Diagnoses of Hydrocephalus**

- Ventriculomegaly - Genetic
- Anomalies - Structural
- Maternal Serum Studies - Biochemical
- Intrauterine CT Scanning - Others

**Associated Anomalies**

- CNS
- Dandy-Walker
- Others

**Evidence of Intrauterine Infection**

- Traditional Obstetrical Care
- Abortion
- Vaginal Delivery with Cranial Decompression as needed
- Elective Caesarian Section and Ventriculomegaly

**Protocol for the Treatment of Prenatal Hydrocephalus**

1. **Indications for Ultrasonic Examination**:
   - Pregnancy At Risk
   - Screening

2. **Possible Ultrasound Diagnoses of Hydrocephalus**:
   - Ventriculomegaly
   - Genetic Anomalies

3. **Associated Anomalies**:
   - CNS
   - Dandy-Walker
   - Others

4. **Evidence of Intrauterine Infection**:
   - Traditional Obstetrical Care
   - Abortion
   - Vaginal Delivery with Cranial Decompression as needed
   - Elective Caesarian Section and Ventriculomegaly

5. **Follow-up Ultrasonic Examinations and Amniocentesis**

6. **Protocol for the Treatment of Prenatal Hydrocephalus**:
   - Follow-up Ultrasound Examinations and Amniocentesis
   - Elective Caesarian Section and Ventriculomegaly
   - Intrauterine CT Scanning

The nature of such damage cannot be predicted at the present time.

Our experience with hydrocephalic fetuses diagnosed after the period of viability, in which massive or asymmetrical ventriculomegaly is recognized or in which intrauterine infection has occurred, has been discouraging. In these cases, the mortality is related not to the hydrocephalus but to the associated anomalies and the complications of premature delivery (Cases 4, 5, and 7). The benefit to this group of patients of an atraumatic delivery\textsuperscript{9} does not outweigh the risks of Caesarean section\textsuperscript{1,5} in our opinion, and we therefore feel that standard obstetrical care should be instituted in these cases.

Patients with or without associated anomalies, going into labor spontaneously prior to the period of pulmonary maturity as indicated by amniotic fluid lecithin/sphingomyelin ratios or dates, should be allowed to deliver vaginally with small-needle ventricular decompression as required.\textsuperscript{4,13}

In cases of fetal death, it is critical to obtain full pathological examination in order to assess the effects of the hydrocephalus on the developing brain and to establish the etiology. The genetic implications for the involved family can then be appreciated.\textsuperscript{10}

In the few cases where the fetus has symmetrical ventriculomegaly and no other detectable anomalies and there is no intrauterine infection, the family should be offered elective Caesarean section. This is carried out when pulmonary maturity is reached and prior to any rapid increases in ventricular size as indicated by at least weekly ultrasound examinations. In cases where sequential ultrasonography shows rapidly increasing hydrocephalus and amniocentesis does not confirm pulmonary maturity, steroid administration prior to delivery should be considered.\textsuperscript{34} This will decrease the risk of respiratory distress and associated hypoxic events which may predispose to subependymal germinal layer hemorrhages. The objective is to minimize the additional risk of neurological damage associated with vaginal delivery\textsuperscript{25,26} and to allow earlier definitive ventricular shunting.

Since repeated ventricular taps are not the treatment of choice for progressive hydrocephalus after birth,\textsuperscript{20,25,28} it seems unlikely that repeated transdominal ventricular taps should be the treatment for hydrocephalus in utero.\textsuperscript{3} Until a satisfactory shunt is available for placement in utero,\textsuperscript{24} we believe that this select group of patients is best served by early definitive treatment (atraumatic delivery and traditional shunting). Encephalocentesis should be reserved as an aid for vaginal delivery when indicated.

The problems resulting from our current ability to diagnose hydrocephalus in utero are due to the lack of definitive treatment options for the fetus. The proposed suggestions can only serve as guidelines. The value of genetic counselling for the parents in Cases 4, 5, and 6 is obvious.

Ultrasound has provided physicians with a safe,
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reliable, noninvasive method for the assessment of the fetus and its environment. A multifaceted diagnostic approach in prenatal hydrocephalus is required, and treatment guidelines, as above, will require the test of time. Through early diagnosis and atraumatic delivery and shunting, it is our hope that the disability borne by these children can be minimized.

Addendum

Since the preparation of this manuscript, there have been further developments in the in utero treatment of hydrocephalus, both in experimental animals and in humans (Clewell WH: Personal communications, 1981, 1982).

We have encountered six additional cases of hydrocephalus diagnosed in utero, and in all cases additional anomalies have been identified by ultrasound. These anomalies have been the critical factor in determining the choice of treatment, and aggressive therapy has not been instituted in any of these cases.

Acknowledgments

We are grateful to Dr. Davis Elliott, Department of Radiology, Foothills Provincial General Hospital, Calgary, Alberta, for his invaluable assistance.

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Manuscript received May 4, 1981.
Accepted in final form May 28, 1982.

This paper was presented in part to the 15th Canadian Congress of Neurological Sciences, June 18–21, 1980, in Ottawa, Canada.

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