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Clinical Course of Fetal Hydrocephalus: 40 Cases

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The clinical course and outcome of hydrocephalus diagnosed in utero is not well understood. To approach this problem 40 cases were reviewed of intrauterine fetal hydrocephalus diagnosed with sonography, and follow-up information was obtained regarding them. Sonograms were evaluated for cerebral dimensions, biparietal diameter, brain mantle size, ventricular ratio, amount of amniotic fluid, and associated abnormalities. Neonatal brain sonograms and computed tomographic (CT) scans were reviewed also. Clinical charts were reviewed for maternal age and parity, referral source, family history, fetal age at diagnosis and delivery, mode of delivery, physical examination and/ or autopsy findings, karyotype, amniotic alpha$_1$ fetoprotein level, cause of death, shunt placement after birth, and status of live infants. The observations indicate that the prognosis for fetal hydrocephalus is poor. Only six infants (15%) were alive after an average follow-up of 13 months. Three children were normal and the other three had neurologic abnormalities ranging from severe (paralysis and incontinence) to minimal (2–3 months delayed motor development). Thirty-four fetuses or neonates died. Nine families elected to terminate pregnancy. Ten opted for decompression at delivery for progressive hydrocephalus. Neural tube defects were present in 12 of 23 infants at delivery. Fourteen other infants had additional significant congenital abnormalities. Other abnormal sonographic findings included polyhydramnios (13 of 38), oligohydramnios or decreased fluid (nine of 38), neural tube defect (nine of 40), and other congenital abnormalities (nine of 40). These findings indicate that hydrocephalus diagnosed in utero by sonography is caused by a heterogeneous group of disorders. In general, the prognosis for normal development is poor. Individual prognoses, however, depend on the specific malformations and the interventions used.

The outcome and clinical course of fetal hydrocephalus is not well understood. Since technical advances in sonography have facilitated the diagnosis of hydrocephalus in utero [1–10], several investigators have become interested in performing in utero shunting procedures in hopes of improving the outcome [11–14]. The natural history of fetal hydrocephalus must be examined to determine whether fetal therapy is beneficial. At the present time, many fetuses are not considered for in utero shunting because of advanced age, associated severe malformations, no pressure elevation within dilated ventricles, or family refusal. In these patients, it is also important to know the natural history of fetal hydrocephalus to aid in obstetric management and provide the family an accurate prognosis; some families will elect to terminate the pregnancy.

We examined 40 fetuses diagnosed with fetal hydrocephalus that did not undergo in utero shunting. These pregnancies were either terminated or the fetuses were treated using standard obstetric therapy. We were interested in the clinical course and studied the survival, delivery, and current status and prognosis of surviving infants. We also endeavored to correlate findings at delivery with those made by sonography.

The incidence of hydrocephalus is reported as 0.5–3 per 1000 live births [16, 17]. Several reports have addressed the prognosis of neonatal hydrocephalus

[18–25]. Although the prognosis of infants diagnosed with hydrocephalus at the time of delivery who are shunted early is relatively good, our experience indicates that when hydrocephalus is diagnosed in utero the prognosis may be considerably poorer, and that of these fetuses, many will not survive long enough to receive their first neonatal shunt.

Materials and Methods

Forty fetuses diagnosed with fetal hydrocephalus were seen in our ultrasound department during a 6½ year period. Medical records, sonograms, computed tomographic (CT) scans, and autopsies were reviewed. Additional information was obtained in telephone interviews with primary physicians. Data collected included maternal age and parity, referral location, family history, fetal age at diagnosis and delivery, progression of pregnancy, mode of delivery, physical examination and/or autopsy findings, chromosomal analysis, alpha₁ fetoprotein level, age and cause of fetal death, ventricular shunt placement after birth, and status of live infants. Sonograms were evaluated for amount of amniotic fluid, biparietal diameter, brain mantle thickness, ventricular ratio, and presence of associated abnormalities. Neonatal brain sonograms and CT scans were reviewed also.

Results

Family History

The patients came from various locations: 11 were from the Denver area, 11 were from Colorado outside Denver, and 18 were from outside Colorado. The average age of the mothers was 27.1 years (range, 18–39 years). No data were available regarding parity in two mothers. The other 38 women had parities of 1 (eight mothers), 2 (17), 3 (four), or more than 3 (nine). A history of spontaneous abortion was present in nine women; three of the women had had more than one spontaneous abortion.

A positive family history for central nervous system abnormalities was present in nine families. Other congenital abnormalities were present in three siblings (one stillborn and two with tracheoesophageal fistula, absent radii, and renal hypoplasia in addition to hydrocephalus). One father had a balanced chromosomal translocation, 46 XY, t(14;2).

Maternal risk factors associated with pregnancy included hypertension/eclampsia (three), smoking (two), diabetes (one), and medications (five). The medications taken were Stelazine, Septra, Aldoril, Konsyl, Bendectin, and various antibiotics. Alpha₁ fetoprotein levels were obtained from amniocentesis in four patients; two had elevated amniotic alpha₁ fetoprotein (one of these had a neural tube defect and one did not). A neural tube defect was not present in the two patients with normal alpha₁ fetoprotein levels.

Delivery

The modes of delivery were therapeutic abortion (nine), vaginal delivery (18), and cesarean section (13). Decompression of the fetal head was performed in 10 of the vaginal deliveries. Four mothers were induced specifically for early neonatal ventricular shunt placement.

TABLE 1: **Congenital Abnormalities in Infants with Hydrocephalus**

Abnormality	No. (n = 40)
Neural tube defect	12
Dandy-Walker cyst	2
Encephalocele	1
Alobar holoprosencephaly	1
Iniencephaly	1
Posterior cranial cyst	2
Polymicrogyria	2
Cleft palate	5
Cleft lip	3
Low-set ears	3
Bilateral optic atrophy	1
Facial bone abnormalities	3
Hypoplastic left ventricle	1
Ventricular septal defect	1
Bicuspid aortic valve	1
Tracheoesophageal fistula	1
Omphalocele	1
Ventral wall defect	1
Intestinal malrotation	1
Fused kidneys	2
Abnormal spleen	2
Abnormal liver	1
Abnormal pancreas	1
Pulmonary hypoplasia	1
Absent thumb	2
Arthrogryposis	2
Short legs	1
Long fingers and toes	1
Syndactyly	1
Clubbed feet	2
Absent right arm and ribs	1
Vertebral anomalies	1
Hypospadias	1
Micropenis	1
Amniotic band	1
Total	64

Fetal Information

The average fetal age at diagnosis of hydrocephalus was 26.8 weeks (17–38 weeks). The diagnosis was made before or at 24 weeks gestation in 10 pregnancies, allowing for the option of termination. Nine chose to terminate the pregnancies. The average fetal age at delivery (excluding therapeutic abortions) was 34.6 weeks (26–41 weeks). There were four stillbirths and 27 live births. The hydrocephalic fetus was a member of a twin gestation in four cases: in each pair, the other sibling was normal at delivery.

Autopsies and physical examinations at delivery provided information regarding fetal abnormalities. Autopsies were performed on 25 infants; two fetuses were macerated and autopsies provided little information. All autopsies in our study group verified hydrocephalus. Eight types of hydrocephalus were identified at autopsy: (1) hydrocephalus of unknown etiology (eight infants), Chiari type II (six), aqueductal stenosis (four), foramen magnum stenosis (one), atresia of the third ventricle (one), alobar holoprosencephaly (one), iniencephaly (one), and Dandy-Walker cyst (one). In 36 fetuses, gender data were obtained: 18 females and 18 males. Congenital abnormalities other than hydrocephalus were present in 28 (70%) of 40 fetuses. Physical examination of the fetuses
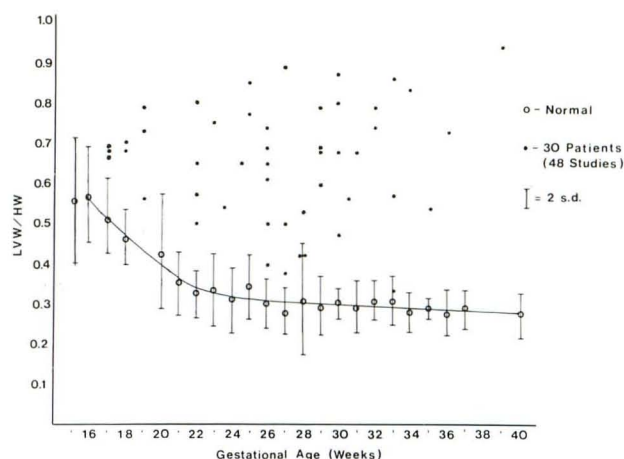
Fig. 1.—Ventricular ratios. *Open circles* represent normal ventricular ratios during gestastion (from [1]). *Closed circles* represent ventricular ratios from 48 studies in 30 fetuses with hydrocephalus. LVW = lateral ventricular width; HW = hemispheric width.

TABLE 2: **Relationship of Biparietal Diameter to Gestational Age Determined by Femur Length or Abdominal Circumference**

Age at Time of Study	BPD = Age	BPD > Age
10–19 weeks	11	2
20–29 weeks	25	11
30–40 weeks	9	15
Total	45	28

Note.—BPD = biparietal diameter.

TABLE 3: **Current Status of Children Surviving with Hydrocephalus**

Case No. (age in months)	Neurologic Status
1 (6)	Urinary and fecal incontinence; left leg paralysis (sensory and motor); right leg, minimal movement
2 (12)	Poor weight gain, no walking
3 (12)	Fine and gross motor development: 11–12 months; five shunt revisions
4 (13½)	Motor development: fine, 11 months; gross, 12 months
5 (18)	Motor development: 15–16 months; only crawls
6 (18)	Motor development: fine, 18 months; gross, delayed; crawls, stands with support; no speech

revealed 12 (30%) lumbar neural tube defects. Other congenital abnormalities were seen in 17 (42.5%) infants (table 1). Eleven (27.5%) fetuses had central nervous system abnormalities in addition to hydrocephalus and lumbar neural tube defects: Dandy-Walker cyst (two), encephalocele (one), alobar holoprosencephaly (one), iniencephaly (one), polymicrogyria (two), posterior cranial cyst (two), and paraventricular or intraventricular hemorrhage (two). Chromosomal analysis was attempted in four fetuses and eight infants. Cells from five did not grow; karyotypes were normal in five and abnormal in two: 47 XY + 18 and 46 XY t(14;2).

Sonographic Findings

Sonograms or reports were available for review in all patients. Serial sonograms were obtained in 27 fetuses. Hydrocephalus was diagnosed by sonography when ventricular ratios were above normal based on the charts developed by Johnson et al. [1]. The ventricular ratios are shown in figure 1. Brain mantle measurements in the parietal region were 2–20 mm. Table 2 correlates biparietal diameter with the gestational age of the fetus, which was determined by femur length or abdominal circumference. Normal variation of dates was used to determine gestational age, which was identified as 1 week at 10–19 weeks, 2 weeks at 20–29 weeks, and 3 weeks at 30–40 weeks.

The amount of amniotic fluid present on sonography was evaluated in 38 patients: normal (16), oligohydramnios (seven), decreased (two), borderline increased (one), and polyhydramnios (12). Two of the patients with oligohydramnios had premature rupture of membranes. Of the 13 patients with increased amniotic fluid, six had conditions (neural tube defect, tracheoesophageal fistula, abdominal wall defect, intestinal malrotation with ventral septal defect, and twins) other than hydrocephalus to explain the increased fluid.

Lumbosacral neural tube defects were present in 12 fetuses; nine were identified by sonography. Three were not seen on sonography at 18, 25, and 26 weeks. The report on one fetus stated that the lower spine could not be evaluated due to fetal orientation. Two of these cases were evaluated during the first 4 years of our study. The first sonograms to identify neural tube defects were obtained at 19–34 weeks gestational age.

Congenital abnormalities identified on sonography that have not already been discussed included small trunk, encephalocele, Dandy-Walker cyst, omphalocele, hepatosplenomegaly, skin edema, and increased brain density. Both neonatal brain sonography in seven and CT in five verified hydrocephalus as well as other brain abnormalities identified by sonography in utero.

Postnatal Follow-up

Six of the 40 fetuses identified with hydrocephalus were alive at the time of this report. Three of these children were premature and three were mature at delivery. The average age at follow-up was 13 months (6–18 months). Ventriculoperitoneal shunts were placed in all children at 1–90 days after birth. The current status of the six survivors is given in table 3. Three children are normal neurologically and three are abnormal.

Thirty-four fetuses (85%) diagnosed with hydrocephalus died. The causes of death were therapeutic abortion (nine), decompression at delivery (seven), infection (five), multiple abnormalities incompatible with life (four), intrauterine fetal

demise (three), respiratory failure (two), and unknown (four). Two fetuses that underwent cranial decompression before delivery survived 25 days and 9 months, respectively; both died of infection. The life span for the other infants was 0–3.5 months. Four infants died despite placement of ventricular shunts.

Discussion

It is important to determine the clinical course of fetuses diagnosed with hydrocephalus in utero. Until recent advancements in sonographic technology occurred, few cases of fetal hydrocephalus were diagnosed. Therapeutic measures for improving fetal well-being and prognosis are now pursued while the fetus remains in utero [11–14].

It has been shown that fetal hydrocephalus may be reliably diagnosed by measuring the lateral ventricular ratio and comparing it with charts developed for normal fetuses [1, 9, 26]. The biparietal diameter was abnormal in only 28 (37%) of the 75 examinations performed in this study and, thus, is not a reliable indicator of hydrocephalus. Diagnosis of fetal hydrocephalus may be difficult during the early second trimester using the ventricular ratio charts since the lateral ventricular ratios are so large at this gestational age. Serial examinations may be required about 2 weeks apart to make a definite diagnosis of hydrocephalus. Chinn et al. [27] found that during the time period of 14–21 weeks, a normal, nonhydrocephalic brain should have choroid plexus that fills the body and atria of the lateral ventricle; specifically, the choroid plexus should lie adjacent to the medial and lateral walls in the body and atria of the lateral ventricles. They suggest that in hydrocephalus, the choroid plexus shrinks and therefore does not completely fill the body and atria of the lateral ventricle. This observation may serve as an additional criterion for diagnosis.

Of the 40 fetuses reviewed in this study of fetal hydrocephalus only six (15%) were alive at the time of this report. Three of the infants were neurologically normal; however, the follow-up time (6–18 months) was relatively short in regard to childhood development. Three infants had mild to severe neurologic abnormalities. All six living children had ventriculoperitoneal shunts placed soon after delivery (less than 3 months).

Twelve fetuses (30%) had lumbar neural tube defects associated with hydrocephalus; this combination of findings is most often identified as Chiari type II malformation. Two of these fetuses survived. Chiari type II hydrocephalus may be suggested alone by specific findings (low-grade hydrocephalus, frontal horn pointing, pseudomass) [28]. The spine should be evaluated thoroughly to identify neural tube defects in these fetuses. Congenital abnormalities other than lumbar neural tube defects were present in 42% of the fetuses. Thorough sonographic examination of the entire fetus should be performed when hydrocephalus is identified.

The quantity of amniotic fluid was not appropriate (polyhydramnios or oligohydramnios) in 57% of the pregnancies. Fetuses that do not have a normal amount of amniotic fluid should be studied for possible hydrocephalus.

In utero ventriculoamniotic shunts were not attempted in the fetuses in our study. One family was offered the option of undergoing this experimental procedure and refused. Criteria for fetuses to be considered for ventriculoamniotic shunts were gestational age of less than 30 weeks, progressive hydrocephalus, and no associated congenital abnormalities seen on sonography. Eleven fetuses were eligible for in utero shunt placement by these criteria. Three of those fetuses had a neural tube defect at delivery that was not identified by sonography; however, these examinations were performed early in this study, and a careful real-time examination of the spine in transverse and longitudinal projections is now routine.

In conclusion, the outcome of fetuses diagnosed with hydrocephalus is dismal. Only three children in our series of 40 cases are neurologically normal. Neural tube defects and other congenital abnormalities were present in about two-thirds of the fetuses. Certainly these other abnormalities account for a part of the morbidity and mortality in these fetuses/infants. The fetus should be examined thoroughly with sonography to identify these abnormalities. Sonography may show evidence of an abnormal amount of amniotic fluid (57%); these fetuses should be examined specifically for possible hydrocephalus in utero.

This study indicates that the prognosis for fetal hydrocephalus is not as favorable as for neonatal hydrocephalus. Further progress in intrauterine therapy and aggressive obstetric management (e.g., premature induction of labor with early neonatal shunt placement) may improve the morbidity and mortality of fetal hydrocephalus.

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