Overview of the Project

- In 1986, I became pregnant with my first child.
  - There was very little published information on pregnancy and hydrocephalus in 1986.
  - Since I am in the first generation of shunted women to survive to childbearing age (due to advances in imaging methods, surgical techniques, and shunt technology) there were a lot of unknowns.
    - The few articles I did find, presented conclusions based on very limited data.
    - Doctors did not have answers either.

Overview of the Project (cont.)

- I had a normal pregnancy but had a very difficult vaginal delivery due to the size and posterior position of the baby.
- Six weeks after the baby’s birth, I had a very sudden onset of shunt malfunction symptoms that lead to an emergency revision.
- My husband and I wondered if the difficult delivery and shunt malfunction were related and decided to research the subject.
The Early Years

• I continued my search for information on pregnancy in shunted women in the years immediately following my son’s birth but found very few new statistics.

• May 1994, my husband and I decided to start our own computerized database to see how other shunted mothers had fared during their pregnancies and deliveries.

• We sent computerized project updates to participants and hydrocephalus support organizations all over the United States, Canada and England

  – The first report generated by the project included the statistics of 10 pregnancies from 6 mothers

• The project has always been funded by my husband, Marty, and myself.

Mothers and Their Babies

How I found the Mothers

Participating mothers were found through:

• Hydrocephalus and spina bifida support networks
  – Articles in their newsletters (H.A.)

• Articles in disability specific publications
  – “Disability, Pregnancy & Parenthood International” in the UK

• Correspondence with practicing neurosurgeons

• Internet Web site (new address)
  – www.hydrowoman.com

Project Referenced in These Books

• Information about project published in:
  – Congential Disorders Sourcebook edited by Karen Bellenir
  – Mother to Be: A Guide to Pregnancy and Birth for Women with Disabilities, 2nd edition by Judi Rogers
The Survey Form

• Since I am not a physician and do not have access to patient records, I chose to gather data by questionnaire.
• The survey form was divided into two sections: maternal information and pregnancy information.
  – In part 1, the mothers were asked their age, medical history and type of shunt (VA, VP, LP, other).
  – In part 2, respondents were asked to evaluate shunt performance during the pregnancy, the delivery and the postpartum period.

Information Collected from the Mothers

• Several questions were asked about the mode of delivery, use of delivery aids (forceps, suction), use of anesthesia, position of the baby, and the presence of birth defects in the baby.
• The mothers were also asked to report any unusual complications.
• Each mother who returned a survey, was assigned a number to protect her privacy.
• Several of the mothers had copies of their surgical records at home for reference.

Survey Sample

Participants Are From All Over the World

• Participants come from all over the world.
  – 10 countries; 31 states
  – Many medical studies focus on participants from medical records in just one hospital or geographic location.
How Is My Study Different From Previous Studies?

- This is an ongoing study and differs from previous studies because it follows the mothers and their babies well after pregnancy and delivery.
- Subsequent pregnancies are reported to me as they occur.
- A few problems in the children of the shunted mothers, that were not obvious at birth, have been reported to me as symptoms appeared.

The Team

- Coordinator:
  - Nancy Bradley, B.S.
- Researchers:
  - Ann Marie Liakos, B.A.
  - Marty Bradley, Ph.D.
  - J. P. McAllister II, Ph.D.
- Doctors:
  - Gary Magram, M.D.
  - Steve Kinsman, M.D.
  - Cheryl Muszynski, M.D.

Goal of the Study:

Questions to Be Answered

- How does maternal shunt dependency influence the course of pregnancy and pregnancy outcomes?
- What neurosurgical complications characterize this population of patients?
- Does the underlying cause of the mother’s hydrocephalus make a difference in pregnancy outcomes?
- Is one type of shunt (valve mechanism) or shunt configuration (extracranial absorption site), less susceptible to problems?
- What complications of shunt dependency influence obstetrical management including pre-natal testing and delivery?
- What are the risks and benefits of various management options for shunt dependent pregnant women?

Two Papers Have Been Published

1st Published Paper


2nd Published Paper


One Short Article Published

• Article Published In: The European Journal of Pediatric Surgery--Supplemental Issue, December 1997

“Hydrocephalus and Pregnancy: the medical implications of maternal shunt dependency”

Study Results

2nd Published Paper

January 2000
Study From January 2000 Medical Journal Article

- Second medical journal article based on my database statistics
- Discusses 138 pregnancies from 70 shunted women
- Doubles the amount of information published in the first report

Education of the Mothers

<table>
<thead>
<tr>
<th>Categorization</th>
<th>Number of cases</th>
<th>% total respondents</th>
</tr>
</thead>
<tbody>
<tr>
<td>Did not complete high school</td>
<td>1</td>
<td>01.43%</td>
</tr>
<tr>
<td>Completed high school or equivalent</td>
<td>10</td>
<td>14.30%</td>
</tr>
<tr>
<td>Completed some college</td>
<td>18</td>
<td>25.70%</td>
</tr>
<tr>
<td>Completed an associate’s degree</td>
<td>6</td>
<td>08.75%</td>
</tr>
<tr>
<td>Completed a bachelor’s degree</td>
<td>18</td>
<td>25.70%</td>
</tr>
<tr>
<td>Completed some graduate level education</td>
<td>2</td>
<td>02.86%</td>
</tr>
<tr>
<td>Completed graduate level college degree</td>
<td>2</td>
<td>02.86%</td>
</tr>
<tr>
<td>Mother’s shunted after age of 21 years²</td>
<td>9</td>
<td>12.90%</td>
</tr>
<tr>
<td>Information not available</td>
<td>4</td>
<td>05.71%</td>
</tr>
</tbody>
</table>

- Most completed a high school degree.
- Many continued to college. One a member of MENSA.
- Data collected to demonstrate the intelligence of this population of women. (story)

Chart of Mother’s Education

- 2% did not complete high school
- 18% completed only high school
- 41% completed some college or an associate’s degree
- 32% completed bachelor’s degree
- 7% reported some graduate education or completed a graduate level degree

Initial Shunt Placement

- Initial shunting in these women took place between birth and 35 years of age for a variety of reasons.
- One mother was first shunted in 1940. She had no revisions until 1979. Until that time, she was unaware of the shunt.*
**Reasons for Shunt Initial Placement**

<table>
<thead>
<tr>
<th>Etiology—congenital</th>
<th>Etiology—acquired</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arachnoid cyst</td>
<td>CNS tumors</td>
</tr>
<tr>
<td>Pineal cyst</td>
<td>Arteriovenous malformation</td>
</tr>
<tr>
<td>Aqueductal stenosis</td>
<td>Encephalitis</td>
</tr>
<tr>
<td>Arnold Chiari malformation</td>
<td>Meningitis</td>
</tr>
<tr>
<td>Dandy Walker</td>
<td>Traumatic injury</td>
</tr>
<tr>
<td>Myelomeningocele</td>
<td>Unknown adult onset</td>
</tr>
<tr>
<td>Unknown pediatric onset</td>
<td>23</td>
</tr>
</tbody>
</table>

- Congenital hydrocephalus is hydrocephalus that began developing while the baby was in the womb.
- Hydrocephalus that is not congenital is acquired.

**Shunt Configurations**

<table>
<thead>
<tr>
<th>Shunt configuration</th>
<th># Cases (# live births)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ventriculoperitoneal (abdomen)</td>
<td>54 (84 live births)</td>
</tr>
<tr>
<td>Ventriculoatrial (heart)</td>
<td>10 (16 live births)</td>
</tr>
<tr>
<td>Ventriculopleural (lung)</td>
<td>1 (1 live birth)</td>
</tr>
<tr>
<td>Lumboperitoneal (spine)</td>
<td>1 (2 live births)</td>
</tr>
<tr>
<td>Multiple configurations¹</td>
<td>5 (5 live births)</td>
</tr>
<tr>
<td>a. Across reproductive history</td>
<td>3 (represented above)</td>
</tr>
<tr>
<td>b. Within a single pregnancy</td>
<td>2 (2 live births)</td>
</tr>
</tbody>
</table>

Total number live births² \( n = 105 \)

- Four types of shunt configurations were reported.
- Five women reported multiple shunt configurations during their pregnancy histories. All were changed from VA to VP.

**Preconception Counseling and Prenatal Testing**

- Preconception counseling and baseline CT or MRI
- Prenatal vitamins including folic acid before and during pregnancy to minimize risk of neural tube defects
- Discuss Meds/Anticonvulsants with your doctor
  - Increased risk to fetus
- Prenatal testing is an important consideration
  - 95% had ultrasounds
  - 66% had Alpha-fetoprotein (AFP) testing
    - Can detect neural tube defects
  - 28% had Amniocentesis
    - Identified a baby with Trisomy 13
    - No reported complications

**Prenatal Vitamins**

Certain drugs lessen the effectiveness of folic acid.

- **Quinine**-used to treat malaria and to relieve muscle cramps
- **Sulfasalazine**-used to treat ulceration and bleeding during the active phase of ulcerative colitis
- **Triamterene**-used to reduce fluid retention and potassium loss
- **Trimethoprim**-used to treat urinary tract infections; helps prevent recurrent urinary tract infections
- **Zinc**-This mineral has become quite popular in lozenge form to help lessen the effects and duration of the common cold.

Reference: *A Complete Guide to Prescription and Non-Prescription Drugs* by H. Winter Griffith, M.D.
Age of the Mothers at Conception

Respondents 70
Pregnancies 137
Maternal age range 18–41 years

Headaches During Pregnancy

- Nine mothers reported an increase in headaches during pregnancy
  - Eight lead to surgical revision
  - One mother reported disabling chronic headaches throughout both her pregnancies
    - Revisions following both deliveries
  - 15 mothers reported increased ICP that subsided after delivery without surgical intervention.
- Seven reported no increase
- Two reported fewer headaches

Pregnancy Related Shunt Revisions

- Five of the seven revisions during pregnancy were associated with two mothers.
- Five mothers account for 15 of the 23 post-partum revisions.
- Those who had malfunctions in the third trimester chose to wait until after delivery

Type of Delivery

- 61 of 105 deliveries were vaginal
  - 53 normal position
  - 7 posterior position
- 44 of 105 Cesarean
  - 13 performed because of shunt dependency
    - 4 mothers in shunt failure
    - In 9 cases doctors felt it was a protective measure
  - All babies in the breech position were delivered by Cesarean
Anesthesia During Delivery

- Epidural and spinal anesthesia should be very carefully administered.

<table>
<thead>
<tr>
<th>Type</th>
<th>Vaginal (% total)</th>
<th>C-section (% total)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epidural</td>
<td>24 (39.34%)</td>
<td>19 (43.18%)</td>
</tr>
<tr>
<td>General</td>
<td>N/a</td>
<td>17 (38.63%)</td>
</tr>
<tr>
<td>Spinal</td>
<td>1 (0.64%)</td>
<td>6 (13.64%)</td>
</tr>
<tr>
<td>None</td>
<td>35 (57.38%)</td>
<td>N/a</td>
</tr>
<tr>
<td>Unknown</td>
<td>2 (03.28%)</td>
<td>N/a</td>
</tr>
</tbody>
</table>

Epidural Complications*

- Total of 79 mothers had epidurals
- Epidural attempts failed in 5 mothers (6%)
  - 4 mothers reported multiple epidural attempts
  - 3 mothers reported their epidurals were only partially effective, numbing only one side
- In one case, needle slipped causing blood to mix with CSF--revision
- Another mother experienced shunt malfunction symptoms as soon as catheter was removed--revision
  - immediate headache
  - seizures and headaches for three weeks

Seizures During Delivery*

- 2 mothers reported seizures during delivery
  - In first case, doctor hit a nerve on second attempt to place epidural catheter; within minutes mother’s arms were flopping and she was unable to speak for several minutes.
  - The second mother had two petit mal seizures during delivery. Her doctor insisted she continue her vaginal delivery.

Delivery Aids

<table>
<thead>
<tr>
<th>Delivery aids</th>
<th>Cases</th>
<th>Malfunctions</th>
<th>% of live births</th>
</tr>
</thead>
<tbody>
<tr>
<td>Forceps</td>
<td>12</td>
<td>1</td>
<td>18.75% (12 of 64)</td>
</tr>
<tr>
<td>Suction</td>
<td>9</td>
<td>0</td>
<td>14.06% (9 of 64)</td>
</tr>
<tr>
<td>Pressure</td>
<td>10</td>
<td>2</td>
<td>15.62% (10 of 64)</td>
</tr>
</tbody>
</table>

- Shortened second stage of labor may be preferred
- Heavy amounts of abdominal pressure are not advised because it increases ICP in the mother.
  - 2 mothers who had heavy amounts of abdominal pressure during their vaginal deliveries had malfunctions within 4 months of delivery.

* Includes new data since 2nd paper was published
Pregnancy Outcomes

- Total of 138 pregnancies
- 103 resulted in 105 live births (2 sets of twins)
- Of the 30 miscarriages, 12 were associated with 3 mothers having both hydrocephalus and spina bifida

Miscarriages

- Although the number of miscarriages (30) may appear high (20.98%), it is important to note that at least 20% of all normal pregnancies end in miscarriage.
- Vast majority of miscarriages (27 of 30) took place in the first trimester.

Birth Defects

<table>
<thead>
<tr>
<th>Birth defect</th>
<th>Maternal age (years)</th>
<th>Seizures*</th>
<th>Anticonvulsants*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Traumatic brain injury</td>
<td>35</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>Hydrocephalus/Neurofibra (#001)</td>
<td>35</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>Vater’s syndrome (#001)</td>
<td>24</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Central palsy (#014)</td>
<td>34</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Spina bifida/Hydrocephalus (#021)</td>
<td>29</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>Vascular/Septal heart defect (#018)</td>
<td>24</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>Hypothyroidism (#034)</td>
<td>29</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>Mosaic Turner’s syndrome (#047)</td>
<td>36</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>Hyaline membrane disease</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Persistent patent circulation (#056)</td>
<td>27</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>Symmetric parietal lobar (#061)</td>
<td>24</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>Congenital atresia (#061)</td>
<td>29</td>
<td>No</td>
<td>No</td>
</tr>
</tbody>
</table>

*Indicates if mothers had these during pregnancy

- The mother of the baby born with spina bifida and hydrocephalus did not take prenatal vitamins containing folic acid before or during her pregnancy. This mother’s hydrocephalus was acquired.

Developmental Delays in Children of Shunted Mothers

- We are still investigating any link between seizures and birth defects or developmental delays
- 4 of the 7 mothers whose children have developmental delays had seizure histories and took anticonvulsants during pregnancy
Infantile Hydrocephalus

• One child was born with spina bifida and hydrocephalus. Mother’s hydrocephalus acquired.
• 5 babies had MRI’s to check ventricles due to large head size—all normal
• One child saw neurologist to rule out hydrocephalus due to coordination problems—diagnosed with low muscle tone.
• One child experienced headaches at age 11; CT scan showed slightly enlarged ventricles. Doctors are deciding what measures to take, if any.

Unusual Complications*

• Abdominal Shunt stabbing (women w/VP shunts)
  – 52 women with VP shunts reported shunt stabbing during their pregnancies (20.47% of all women with VP shunts)
• Two women reported another unusual shunt complication. Their VP shunts disconnected, slipped into their pelvises, and wrapped around their reproductive organs. Neither woman was pregnant at the time.
  • One woman’s wayward shunt was removed using a laparoscopic technique with no permanent damage to her reproductive organs.
  • The second woman had to have a full hysterectomy as a result of the damage done by the disconnected shunt.

*Includes new data since second paper was published

Unusual Complications (cont.)

– One respondent reported trouble turning her neck in the 3rd trimester because surgeons had not left enough extra tubing.
– One mother reported that her abdomen stopped stretching in the 3rd trimester due to excessive scar tissue resulting in sharp abdominal stabbing pains.

Women with VP shunt complications affecting their reproductive organs

• Abdominal shunt complications that have made it difficult or impossible for some women to become pregnant.
  – One had a full hysterectomy due to abdominal stabbing pains by unremoved shunt catheters
  – Another is infertile due to placement of an old distal catheter in her fallopian tube.
  – A third had damage done to her fallopian tubes by scar tissue from old VP shunts.
    • Pregnancy will have to be achieved through invitrofertilization
  – In another, abdominal pain caused by unremoved catheters, was incorrectly diagnosed as ovarian cysts.
Updates Since 2nd Paper in January 2000

The Project is Ongoing and Now Has Over 150 Mothers and 300 Pregnancies

- Since January 2000, 82 new women have joined the study.
- 158 new pregnancies
  - 5 women have had subsequent pregnancies

Pregnancy Outcomes to Date*

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Count</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Live Full Term Births</td>
<td>224</td>
<td>74.3%</td>
</tr>
<tr>
<td>Vaginal Deliveries</td>
<td>144</td>
<td>64.2%</td>
</tr>
<tr>
<td>Cesarean Deliveries</td>
<td>81</td>
<td>36.1%</td>
</tr>
<tr>
<td>Live Preterm Births</td>
<td>11</td>
<td>3.7%</td>
</tr>
<tr>
<td>Twins</td>
<td>9 sets</td>
<td>6.1%</td>
</tr>
<tr>
<td>Full Term</td>
<td>3 sets</td>
<td>33.3%</td>
</tr>
<tr>
<td>Miscarried (both twins)</td>
<td>4 sets</td>
<td>44.4%</td>
</tr>
<tr>
<td>Miscarried (one twin)</td>
<td>2 sets</td>
<td>22.2%</td>
</tr>
<tr>
<td>Ectopic Pregnancies</td>
<td>3</td>
<td>1.01%</td>
</tr>
<tr>
<td>Therapeutic Abortions</td>
<td>4</td>
<td>1.35%</td>
</tr>
<tr>
<td>Still Births (cord accident)</td>
<td>1</td>
<td>0.34%</td>
</tr>
<tr>
<td>Birth Defects</td>
<td>24</td>
<td>8.1%</td>
</tr>
<tr>
<td>Infantile Hydrocephalus</td>
<td>1</td>
<td>0.35%</td>
</tr>
<tr>
<td>Babies with Anencephaly</td>
<td>2</td>
<td>0.69%</td>
</tr>
<tr>
<td>Range Newborn Weights</td>
<td>1 lb 14 oz to 10 lbs 5 oz</td>
<td></td>
</tr>
</tbody>
</table>

*Includes new data since second paper was published

Updated Miscarriage Information

- 57 out of 301 total pregnancies ended in miscarriage (18.9%)
  - 57 miscarriages from 32 moms
- 14 moms (43.7%) had multiple miscarriages
  - 3 Moms w/ Hydrocephalus and SB had 12/57 (21%) of all miscarriages
- Moms with Hydrocephalus only
  - Total of 130 moms with 263 pregnancies
    - 45 miscarriages (17.1%)
- Moms with Hydrocephalus and SB
  - Total of 10 moms with 24 pregnancies
    - 12 total miscarriages (50%)
Updated Malfunctions and Revisions

- 7 women reported intrapartum revisions
  - 10 women reported neurological symptoms during pregnancy that did not lead to surgical intervention
  - 7 women reported actual malfunctions during pregnancy that resulted in revision surgery during pregnancy
- 3 women reported symptoms during delivery
  - lost venous pulsations in both eyes (#075)
  - seizure during delivery; inability to speak (#101)
  - epidural removal that led to constant headache until surgical revision a few weeks later (#119)
- 37 women reported postpartum malfunctions that led to surgical revisions

Folic Acid and Anticonvulsants

- Researchers are beginning to explore the interaction between folic acid and anticonvulsants.
  - According to an article released by the University of Maryland School of Medicine, folic acid (B9) may decrease the effectiveness of Dilantin
  - Discuss with your doctor the use of the two together during pregnancy as well as the possibility of taking them at different times of the day to help offset the interaction.
  - Your doctor should monitor you closely for seizure activity and adjust your Dilantin levels while you take folic acid.

Anticonvulsants & Folic Acid (cont.)

- Conversely, Dilantin may reduce the levels of folic acid in the blood as well as the body’s ability to use the vitamin.
- The interaction between the two is poorly documented at this point and warrants further study.

Sources:
- University of Maryland Medicine
- Centers for Disease Control and Prevention
- Internet search - “Interaction between Folic Acid & Anticonvulsants”

Recommendations For Pregnancy and Delivery (1 of 3)

- Shunted mothers must be carefully monitored both physically and neurologically throughout pregnancy.
- Precautionary measures are vital to successful shunted pregnancies. Mothers should discuss the benefits and risks of any medications prescribed with their doctors.
- A conference should be held by all the OB on-call partners of the shunted mother (early in the pregnancy) in order to ensure each physician is aware of the shunted mother’s special medical concerns (story).
Recommendations For Pregnancy and Delivery (2 of 3)

• Baby’s position is very important. If the baby’s position is posterior or breech when labor begins, Cesarean section should be seriously considered.
• Application of abdominal pressure should be avoided (especially heavy amounts of abdominal pressure).
• Physicians should carefully consider the risks to the mother’s shunt system for any procedure involving large amounts of abdominal pressure (an example would be external breech version).
• Spinal and epidural anesthesia should be very carefully administered.

Recommendations For Pregnancy and Delivery (3 of 3)

• During Cesarean sections, great care should be exercised to thoroughly flush the peritoneal cavity in an attempt to reduce the risk of intra-abdominal infection.
  – In one case, intra-abdominal infection caused adhesion formation around the abdominal end of the catheter
  – One mother’s abdominal tubing was exposed during her Cesarean section causing a shunt infection
• Pregnant shunted women should not go past their due dates (providing the dates are correct) because the bigger the baby gets, the more intracranial pressure the mother will experience during delivery.

Summary

• The project is ongoing and now has over 150 mothers and 300 pregnancies
• Revision occurrences, unusual complications, birth defects, and other information has been documented
• Specific pregnancy management recommendations have been made including careful coordination between OB and Neurosurgeon/Neurologist
• Assuming the absence of other complicating medical conditions, women with hydrocephalus can have successful pregnancy outcomes for both the mother and child

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