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ROCHESTER INSTITUTE OF TECHNOLOGY

A Thesis Submitted to the Faculty of
The College of Fine and Applied Arts
in Candidacy for the Degree of
MASTER OF FINE ARTS

NO GUARANTEES

By

Barbara J. Territ

Date:

July, 1990

Advisor: Robert Wabnitz

Date: 8/22/90

Associate Advisor: Glen Hintz

Date: 9/7/90

Associate Advisor: Lawrence Williams

Date: 9/4/90

Special Assistant to the Dean for Graduate Affairs:

Philip Bornarth

Date: 9/14/90

Dean, College of Fine and Applied Arts:

Robert Johnston

Date: 9/14/90

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RD #3 Box 114B
Oxford, NY 13830

Date: August 9, 1990

ACKNOWLEDGEMENTS

Gratitude is extended to those whose help and encouragement have been invaluable to my survival and accomplishments:

The late Dr. Herbert Lourie
Dr. Franklin Vail Peale
Dr. Albert B. Kochersperger
Dr. Joseph McDonald
Dr. Gregory Liptak
Dr. Kay Francis Marshman
Dr. Ann Marie Franzen

Special acknowledgement to my extended family of friends and neighbors who look beyond the "dis" to see the ability, especially Richard.

And to Linda Ganley for her impressive typing, editing and patience.

PREFACE

What you are about to read is a medical autobiography, not merely a clinical case history. Many clinically oriented periodicals contain articles offering statistical documentation as proof that physical abnormalities can have a profound effect on the lives of those who are so challenged. Evidence has also been published concerning the effects of attitude and environment on personal health. Rarely have all of these elements been exemplified in reference to each other.

Ethical protocol in medicine today generally tends to favor an attitude of professional distance, both in practice and in literature. The patient is discussed exclusively in terms of symptoms and treatment in an attempt to assure impartiality and unbiased medical judgment. Medical technology also encourages this distance by limiting personal contact, which consequently encourages the convention of a high-volume practice. This system creates the potential for allowing a physician/patient relationship to proceed with a diminished regard for the impact that either the illness or the treatment may have on the daily life of the patient. By occasionally acquainting ourselves with the personal aspects of life and risking involvement we can enrich our own lives. The personal nature of this paper

is intended to re-introduce the reader to a more personal slice of life while providing valuable clinical information.

Professionals who are in a position to offer assistance are sometimes deified by those seeking attention. Such reverence may lead to unreasonable expectations on the part of the patient. Although physicians can offer their finest services, not one can offer an absolute guarantee of the result. An informed patient is more likely to accept a share of the responsibility in the decision making process when employing the services of a physician.

Experience has led me to believe that there are no guarantees in life. We hope for the best and prepare for the worst. Accepting responsibility for our own well-being through self-education is the best guarantee. By sharing what we have learned we empower others to honor their own responsibilities.

The medical profession is oftentimes intimidating to the uninitiated, having its own language and formal decorum. Learning the language is the first step in overcoming personal intimidation in the quest for medical knowledge. This paper is written in that technical language intentionally to introduce the reader to medical-ese. It is recommended that a good medical

dictionary and an anatomical atlas be close at hand for referral while reading this paper. The endnotes frequently contain further explanatory information to the text and should be consulted as each notation is encountered. Although the specific medical topics covered here may not have a direct application in every reader's life, this initial immersion will provide a foundation for future accessibility in self-education.

The format of the reference section is adapted from the Index Medicus, the primary reference for all medical literary research. This format was used to provide ease and consistency when locating bibliographic source references. In order to encourage further study by the reader, other sources of interest have been included in the bibliography in addition to those that have been identified in the endnotes section.

Actual medical records, information from attending physicians, family and friends has been interspersed with personal recollections to provide an over-all perspective. The "patient" as seen by the physician is not the person as a whole. A physician must consider factors beyond the immediate medical issue when any treatment is recommended. It is my hope that this paper will serve as a reminder to all who come in contact with differently abled persons that facilitating ability is the ultimate objective.

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**NO
GUARANTEES**

December, 1960. The Christmas rush had begun almost as soon as the Thanksgiving dishes had been put away. The baby was just over a year old and beginning to walk. A second child was finally on the way, delayed by a disappointing miscarriage. The expectant mother's disastrous first marriage had ended after the birth of a severely retarded son, who is now institutionalized. Her second husband works for the state conservation department. He had spent his younger days in the Civilian Conservation Corps and abroad during World War II. Now in their forties, these former childhood acquaintances are pursuing the traditional American Dream.

A new year is about to begin. There is a colonial farm in the country, complete with a Cape-style farmhouse to restore. There is a budding antique collection and thirty acres of young Christmas trees, ready to harvest. A steady stream of holiday customers knock at the door to ask directions through the woods and to borrow saws.

Constant interruptions in her usual daily routine make the expectant mother nervous. She chain-smokes while waiting by the door to collect money for Yule tree

purchases. Her uneasiness is compounded by the pregnancy. She is now entering the second trimester of pregnancy. Fearing another miscarriage, her general practitioner had given her a prescription for diethylstilbestrol when she experienced spotting shortly after conception.

Tuesday, June 27, 1961, 2:00 a.m. This is it. The soundly sleeping husband is awakened with considerable difficulty. The doctor is alerted. She is instructed to go to the hospital and the doctor says he will be in around 8:00 a.m. She shakes uncontrollably as she prepares to make the 15 mile trip to the rural county's only hospital.

She is admitted to the maternity ward at 3:30 a.m. and her husband returns home to sleep, expecting a phone call when the event occurs.

At 11:20 a.m., after a nine hour labor she is wheeled into the delivery room. With her feet high in the cold, uncomfortable stirrups of the birthing table, orders for the final push are issued by the doctor. The hushed tones of the delivery room staff and the G.P.'s flat pronouncement of "It's a girl" cause the new mother to ask if something was wrong. The G.P. holds the

screaming infant in the air, displaying a large tumor on her lower spine. He announces that there is a serious problem before sedating the mother and sending her back to her room.

She awakens in the semi-private room to the continuous chatter of her roommate, who has just given birth to healthy twins. Later that afternoon the husband wanders in to find his wife extremely upset and in tears. Although the G.P. has just left the room, she has not yet been informed of the condition of her baby. She tries to explain the problem to her husband who, not fully comprehending the magnitude of the situation, inquires about the gender of the child. "Well, I'll be darned," is his only comment.

Not until the following day is any information provided to the distraught mother. Early in the morning her G.P. returns with the county's first new pediatrician. Together the physicians confront the bewildered mother with a barrage of general technical and medical information regarding the condition of her infant. A calm and rational lay-parent would have found the presentation virtually incomprehensible. The only words the stunned mother was able to grasp were "spina bifida" and "surgery."

Specifically, their baby has a severe form of spina bifida, medically described as a lumbosacral lipomatous myelomeningocele. The Greek origins of this descriptive term illustrate the diversity and magnitude of anatomical involvement: Lipo meaning fat, myelos - marrow, meningo - membrane and kele - hernia.¹ In this case the lesion is located in a broad area in the lower region of the spine, ranging downward from the third lumbar vertebrae through the mid sacrum.

Spina bifida is a general term that describes one feature that is common to a large group of spinal malformations. In all forms of spina bifida there is an incomplete formation of the vertebral arch where the spinous process is absent. This malformation can occur at any vertebral level and may involve any number of consecutive vertebrae.² Myelomeningocele is known as one of the most complex birth defects compatible with meaningful existence, occurring as frequently as one per every thousand live births in the United States.^{3,4}

The myelomeningocele "tumor" on the baby's back is actually composed of malformed spinal cord and nerve roots protruding through incompletely formed vertebral

arches and soft tissue. These herniated neural elements are intimately entangled in a fatty tumor. This entire conglomeration is encased in a thin integument, identified possibly as an external hematomatous continuation of the pia and dura mater that normally encase the spinal cord.⁵

The causes of myelomeningocele remain undetermined and are a subject of much medical debate. Myelomeningocele is a congenital defect and occurs during the second stage of fetal development defined as the embryonic period.

A general overview of the three stages of fetal development reveals the chronology of fundamental events:

- Pre-embryonic - Fertilization to week 3.
Implantation of fertilized egg into wall of uterus and development of three primary germ cell layers.
- EMBRYONIC - Weeks 3 to 8.
Most active phase of development.
Formation of all body organs and structures.

Fetal	Weeks 8 to term.
	Enlargement and maturation of all organs and structures.

Normal embryological development of the spinal cord involves an extremely complex series of events. Many structures and organs are developing simultaneously and the development of these structures is often interrelated.⁶ Cells rapidly divide along a central axis, or primitive streak, in the embryo. Below the primitive streak the notochord forms. The newly formed tissue begins to differentiate around the notochord into many types of tissues with separate and specific functions. While the differentiation process is occurring the growing sheet of tissue, known as the neural plate, begins to thicken and fold dorsally, forming a tube. The tube will begin to mature through a process called neurulation to become the spinal cord. Neurulation commences when the edges of the folding neural plate meet and close in a zipper-like fashion from the middle in both rostral and caudal directions. At the center of the neural tube is the myocele cavity. Primary bulges will appear at the rostral end of the closing

neural tube. It is from this region that the brain will develop.

Somite formation coincides with the initiation of neural tube closure. Somites, along with cerebral spinal fluid, will provide protection to the central nervous system. Somites give rise to skeletal, muscle and integumentary systems, including the vertebral column. From the third to fifth weeks of embryonic development somites arise and form bilateral pairs along the outer neural tube. The formation of spinal ganglia, arising from the neural crest, is concurrent with somite formation. Muscle derived from a specific somite will receive its innervation from the corresponding spinal ganglia. Cells in the somite proliferate and differentiate between the third and sixth weeks, creating three distinct areas around the myocele cavity. These three areas are: the dermatome, which gives rise to the integument; myotome, which gives rise to the muscular system; sclerotome, which gives rise to the skeletal system. The cells of the sclerotome proliferate and migrate to surround the notochord, which disappears. The sclerotome cells that migrate dorsally around the neural tube will form the neural arch of the vertebrae. Chondrification of the sclerotome begins in the fifth week and ossification begins in the eighth week.

Cerebral spinal fluid provides protection to the central nervous system by its action as a fluid buffer, absorbing and distributing external forces endangering the spinal cord and brain.⁷ As soon as the rostral neuropore closes in the middle of the fourth week, the brain forms its flexures at the site of the primary bulge and the choroid plexus is formed. The majority of cerebral spinal fluid is produced by the choroid plexus in the ventricles and most csf is retained in the region of the brain. At the end of the sixth week apertures form in the fourth ventricle of the brain and the increasing pressure of the csf begins to separate out the subarachnoid space surrounding the spinal cord.⁸

The debate over the pathogenesis of spina bifida has been going on for centuries. The term "spina dorsa bifida" was coined by Professor Nicholas Tulp in Amsterdam in the year 1652. Tulp is noted for having discovered nervous tissue in the "tumor" associated with myelomeningocele during his dissections. He is probably most famous as being the central figure of Rembrandt's painting "The Anatomy Lesson."

In 1761, Giovanni Battista Morgagni proposed the "hydrops theory." Morgagni was ignorant of the presence of an open neural tube within the myelomeningocele tumor.⁹ His assumption was that at the completion of

neurulation excess fluid pressure in the myocele cavity causes the neural tube to rupture. This excess fluid originates in the hydrocephalic head of the embryo.¹⁰ The rupture, or neuroschisis, forces developing vertebrae apart and allows cerebral spinal fluid to escape, forming neuroschistic blebs along the neural tube.¹¹

Supporters of Morgagni's theory have postulated a number of scenarios based on this type of trauma to the developing nervous system in an attempt to explain the wide variety of anatomical anomalies found in spina bifida. One scenario involves the rupture and subsequent healing of the cleft. This action of spontaneous closure of the neuroschisis with variable degrees of scarring is thought to cause spina bifida with lipomas, dermoids, diastematomyelia and cord tethering. A second scenario involves the persistence of the neuroschistic bleb throughout the development of the fetus. This results in an infant born with spina bifida cystica. The degree of damage to the neural tube determines whether it presents as a meningocele or myelomeningocele. A third scenario entails rupture of the neural tube, causing a neuroschistic bleb, which subsequently ruptures and remains open. In this instance, Morgagni's followers believe that the infant presents with spina bifida aperta, also called myeloschisis.¹²

The next century brought technical advances such as the microscope into the study of spina bifida. Professor Cleland of Glasgow published a detailed description of the structure of myelomeningocele in 1883 and Freidrich Daniel von Recklinghausen's magnificently illustrated study, published in 1886, generated a new theory of pathogenesis.¹³ Both rejected Morgagni's "hydrop's theory" in favor of formation of myelomeningocele due to failure of the neural tube to close. This failure of closure theory is also divided into two major causal hypotheses. Cleland proposed that overgrowth of neuroectoderm interferes with normal neurulation. Von Recklinghausen theorized that arrest of notochordal growth prohibits closure.¹⁴

Neither the "hydrops" theory proposed by Morgagni, nor the failure of closure theory of Cleland and von Recklinghausen can explain all of the variations exhibited in myelomeningocele. The pathogenesis of all forms of spina bifida is most likely the result of an assorted combination of events that, under certain circumstances, can produce myelomeningocele.^{15, 16, 17, 18}

The doctors' overwhelming recitation in medical-ese did little to reassure the mother of the future of her infant. They impressed upon her that immediate action must be taken if they wanted the baby to survive.

In 1961 the preferred mode of treatment for infants born with myelomeningocele was aggressive. Each case was immediately evaluated on the basis of neurologic deficit, presence or absence of hydrocephalus and general health of the infant. Surgical closure of the myelomeningocele was performed as an emergency procedure for select infants at major medical centers.

The history of treatment of myelomeningocele has alternated between attempted surgical intervention and therapeutic nihilism. In Morgagni's time interest in such "monstrosities" was primarily as the study of a freak of nature rather than curative. Morgagni advised against any attempts at surgical intervention.

During the 19th century there was renewed interest in active treatment. Experimental procedures such as ligation of the sac, injection of iodine and glycerine into the sac and surgical intervention were attempted, all unsuccessfully. The insurmountable problems of

hydrocephalus, infection and paralysis defeated any initial successes of infant survival. Attempts at active treatment had generally ceased by the end of the 19th century.

The following decades of non-treatment were revealing in that the natural course of untreated spina bifida was made apparent. High rates of morbidity or severe mental and physical impairments were the fate of most born with myelomeningocele. Few new efforts at treatment were made until the late 1950's. The introduction of the Holtz-Spitzer valve and ventriculoatrial shunt to control hydrocephalus ushered in a new era in the treatment of myelomeningocele.¹⁹ Control of hydrocephalus meant that damaging pressure on the brain could be relieved, limiting or preventing mental impairment. Antibiotics could be used to reduce the risks of infection from surgery, urinary tract complications and decubitus ulcers, common to paralysis.

The doctors outlined highlights of events the parents could expect to take place if they decided to treat the infant. Surgery was at the top of the list, followed by numerous neurological, urological and orthopedic evaluations and continual follow-up

examinations. There was a possibility that the baby would never walk.

At the conclusion of their distressingly informative presentation, the physicians left the solitary parent to make her decision. She spent most of that day in her room, trying to ignore the constant chatter of her roommate, and the sobs of a mother across the hall who had just lost her baby. Nurses wheeled babies past her door on the way to visit their mothers. The unsolicited advice of one day nurse was to put the baby in an institution and forget about it. When her husband arrives later that afternoon, she attempts to explain what the doctors had told her. After her explanation the parents walk down the hall to the nursery to see their baby through the glass for the first time.

While all this explaining and decision making was going on in the maternity ward, I was lying on my stomach in the nursery being poked and prodded and testing my lungs. My hips were elevated by a folded towel to reduce the risk of tearing the onion peel-like skin covering the apex of the tumor on my lower spine. I was visited by numerous members of the medical profession, all bearing needles. Some drew blood, others pricked me with pins in an attempt to determine the boundaries of neurologic deficit. When the white coats finally left, I realized that I had not yet been formally introduced to my parents.

This was my second day in the nursery and, being the waning years of the baby boom, the nursery was crowded. My nursery-mates had been coming and going from the room at a regular pace. I hadn't left the back corner by the nurse's desk since I arrived.

Eventually my crib was moved to the nursery viewing window. A woman with auburn hair, wearing a hospital gown, and a man in a khaki work shirt and green trousers were looking at me.

The next morning a nurse walked into the nursery without the familiar mask and gown. She wheeled me out the door and down the hall. I held my head up the whole way so I wouldn't miss a detail of the trip. I was taken to the room of the woman with the auburn hair, whom I had seen at the nursery window the previous afternoon. The nurse quickly closed the door behind us and said that I couldn't stay long, as this introductory excursion was against hospital policy.

As I was being gently lifted from my crib the nurse cautioned my mother to be very careful and to keep me on my stomach. I was placed face down across my mother's lap as she sat in her hospital bed. My mother held me nervously while I squirmed to look at my new surroundings.

The conversation between the nurse and my mother was very informative. My parents' names were Leonard and Catherine, but everyone called them Slu and Kay. My mother had shoulder length auburn hair and smelled of Pacquins hand cream and cigarette smoke. I have a sister to meet. Her name is Cynthia. She is 1-1/2 years old. My father is a quiet, rugged man who, I am told, I strongly resemble.

The nurse who brought me in to meet my mother is a family acquaintance. Grace Jocelyn grew up next door to

my mother on East Hill. She seemed more concerned about us than with hospital rules. After my half hour visit with my mother, Grace returned me to the nursery.

My father came to the nursery window late that afternoon. He was wearing his State Conservation Department issue khaki and green. This time I recognized him immediately, as I do look just like him.

My father named me Barbara June.

There was no discussion between my parents about whether or not to begin treatment. They merely asked what was going to be done next.

Dr. Matt Boname was the G.P. who had delivered me. He was also a long-time family acquaintance and knew what a potentially devastating financial situation my parents were in. He immediately assisted my parents in applying for State Aid through the New York State Department of Health. My parents could stand to lose everything they had worked for in order to keep me alive.

With the decision to treat made and insurance tentatively approved, the doctors told my parents that there was nothing more they could do for me at their small, rural hospital. I would have to undergo surgery at a university hospital, 85 miles to the north.

Dr. Richard Hosbach was the new pediatrician. He was asked by Doc Boname to write up the case and contact the Pediatric Neurosurgery Department at Syracuse Memorial Hospital of Syracuse University.

I was born weighing 5 lbs., 9.5 oz. and had lost 1/2 pound in the struggle of making my entrance. A pre-surgery weight requirement of six pounds kept me in Chenango Memorial for six days before making the trip to Syracuse.

The seventh of July was warm and sunny. I was placed in a car bed that was hooked over the back of the bench seat in my father's silvery-green, 1960 Chevy Impala. Grace Jocelyn had taken the day off to accompany my parents and introduce them to what would become a familiar setting of doctors and hospitals.

I was admitted that afternoon, soon to be subjected to more examinations, white coats, needles and X-rays. This was a teaching hospital and they were learning what healthy lungs I had. My parents tried to remember names of numerous interns and doctors as they introduced themselves over my screams. Apologizing for the fuss I was making, my parents were assured that it was a good sign.

SYRACUSE MEMORIAL HOSPITAL
736 IRVING AVENUE
SYRACUSE 10, NEW YORK
ADMISSION REPORT

WILLIAM H. WAITE
Administrator

July 8, 1961

RE: Tefft, Barbara
HOSPITAL NO: 448631
BORN: 6/27/61
ADMITTED: 7/7/61

First Syracuse Memorial Hospital admission of this 11 day old girl who was admitted for surgery to her lumbar mass.

PRESENT ILLNESS: This 11 day old girl is the product of a normal, full term, spontaneous delivery. Birth weight 5 lbs. 9.5 oz. Mother is gravida IV, para III, abortus I, RH positive. Mother had been on delalutin, 2cc. IM every 2 weeks and stilbesterol orally since October 1960 because of some vaginal bleeding during the first trimester. Pregnancy and labor were otherwise unremarkable. At birth, patient was noted to have a 9 x 5 cm. trans movable mass in the lumbo sacral area and was otherwise normal. She has done well and gained weight, is now referred for treatment of this mass.

PHYSICAL EXAMINATION: Pulse 128. The baby is crying. This is an 11 day old female infant with a large lumbar mass. General physical examination is within normal limits. Neurological exam: Head: Fontanelles are open and soft. The head does not transilluminate. Eyes: Red reflexes present bilaterally. Sensation: Responds to pin prick by crying. Motor: Moves all 4 limbs well. One observer thought the patient moved the left leg very little. Reflexes: More reflexes present. Patient withdraws from painful stimulus. Biceps jerks, tricep jerks, ankle jerks, were all normal active. The knee jerk is questionable present bilaterally. There is a lumbar mass covered with normal appearing skin. The greater portion of this mass (9.5 cm.) is firm and loculated and transilluminates. Superiorly in to the right, there is an area which is softer and does not transilluminate.

LABORATORY DATA: HGB 17.8, HCT 50, WBC 13,300, 31 polys and 65 lymphs. Urinalysis: PH 5, sp. gravity (?), no albumin, sugar or acetone, 6-8 RBC. On 7/13/61, HGB was 13 gm., HCT 38. Throat and nose cultures on 7/18/61 revealed no significant findings. X-ray examination of

the lumbo sacral spine: "Examination of the lumbo sacral spine in two projections shows that there is a soft tissue mass behind the lumbo sacral junction in the soft tissues of the back. This mass measures 6 x 4 x 7 cm. through its greatest diameters. It is largely made up of water density and within it there is a linear calcium shadow which has the appearance of formed bone. There appeared to be no congenital anomalies of the spine. This most likely represents a teratoma of the soft tissues in this location."

The preliminary reports were all encouraging and I was confirmed as a candidate for surgery. My parents returned home from the first of many long trips to the University Hospital.

I was tested and observed for four more days before the surgery was finally scheduled. On July 11, Dr. Herbert Lourie, Assistant Professor of Neurological Surgery, called my parents to invite them to the event. In his strong Carolina accent he inquired, "Y'all commin' ta the op'ration?" He informed them that the surgery had been scheduled for the following morning.

On July 12, 1961, the surgery proceeded as follows:

Tefft, Barbara

7/12/61

Neuro.

Dr. Lourie

Dr. Stoops

Dr. McGinnis Jr.

: Lumbosacral teratoma

Operative Procedure:

With the child in the prone position, the area of the low back and the large mass presenting in the lumbosacral area was shaved, prepped, and draped out as a sterile field. 1/2% procaine was infiltrated across the mid portion of the mass and the child was given a whiskey nipple to suck on. This soft tissue mass was approximately the size of a medium-sized orange, attached with a very wide base over the lumbosacral area. There was normal skin covering the mass, save for a small dimple in its mid portion. The line of procaine infiltration was now incised with a sharp knife and scissors and hemostasis secured with mosquito clamps.

Beginning over the normal area on the left side, dissection was carried down to the sac of the mass. As the skin flaps were now reflected away from the mass, it was evident that there was normal-appearing intestines in the sac. The sac of the teratoma was now opened and inspected. The sac appeared to be normal peritoneum. There was a large piece of bowel measuring approximately 6 inches in length and approximately the size of a normal frankfurter in diameter. There was a normal appearing mesentery attached to the bowel and the vascular pattern in the mesentery appeared to be that of normal intestines. The bowel appeared to be small intestine rather than large intestine. There was no peristaltic activity in the isolated loop of bowel and it did contain soft material on palpitation. The peritoneal sac surrounding the bowel was now completely opened and the bowel freed up. Its mesentery was slowly divided by coagulating and incising the blood vessels and mesentery. Finally, the bowel was attached only by a small fibrous band. This fibrous band attached to a piece of bony spicule which arose in the region of the sacrum. This bone actually was formed in the muscle and fibrous tissue overlying the sacrum. The fibrous band connecting the bowel was incised and a piece of bowel removed as a specimen.

(Lumbosacral teratoma and lipomatous myelomeningocele)

We next turned our attention to a collection of soft tissue which appeared to be glandular in structure and resembled pancreas more than anything else. This soft tissue mass was dissected free of its attachments to the subcutaneous tissue and taken as another specimen. The peritoneal sac of the mesentery was now picked up again and dissected free from the under surface of the skin flap on the inferior side of the wound. The peritoneal sac was sent in as a separate specimen. There was now apparent another marble-sized mass with a stalk which went down forwards and into the epidural space. This was a firm mass. It was dissected free down into the fat of the epidural space. Its stalk was tied off and the tissue removed. On incision of this mass it appeared to be rather firm and I could not identify it as to organ type. It was sent as a separate specimen.

We were now left with one large mass in the center of the wound, which appeared to be a lipoma, however, this lipoma pulsed with heart beat and moved with respirations, so that it was evident that it connected with sub-arachnoid space or extradural space. Dissection of the lipoma was now begun and this lipoma was now freed up from the lumbosacral fascia on both sides. This brought us down to a bony defect. This defect in the dorsal elements of the vertebral column was approximately 1-1/4 inches in length and 3/4" in its widest diameter. It opened directly into the extradural space. On lifting up the lipoma it was now apparent that the dura came up into the lipoma and the dura here appeared to be very thin. One could see through the dura that there was nervous tissue in the sac. Consequently the sac was carefully opened and its contents inspected. It was apparent that the conus medullaris was buckled up into the meningocele sac and was attached to the overlying lipoma. The second stalk of the lipoma went down towards the spinal cord. The lipoma was dissected free of the meningocele sac, except for the portion which was intimately bound with the conus medullaris. The extraneural portion of the lipoma was now removed. That portion of the lipoma attached to the conus medullaris was shaved off with fine scissors as close as I could safely accomplish this. The attachment of the conus to the inferior margin of the sac was divided between ties in order that the conus medullaris might now pull back into the vertebral canal. The stalk of lipoma which ran down towards the cord was removed as best as possible, although there remained at the end of the procedure some

(Lumbosacral teratoma and lipomatous myelomeningocele)

lipoma attached in a thin layer to the conus and a second stalk which went up into the extra-arachnoidal space beyond the bony exposure. There was a free flow of spinal fluid coming down from above so that there was no question of a spinal block.

The dural sac was now carefully closed with interrupted sutures of 4-0 silk. The mass remaining was too bulky to double down into the small vertebral canal. Lumbosacral fascial flaps were elevated on both sides of the paravertebral muscles and closed over the dural closure, again using 4-0 silk sutures. We appeared to have a very adequate dural closure and good fascia over the dura. All bleeding points were now coagulated and a dry field obtained. The wound was thoroughly irrigated with copious amounts of saline.

Turning again to the skin flaps, the redundant and bluish skin was removed from both the superior and inferior skin flaps. Subcutaneous tissue flap was now closed with interrupted 4-0 silk sutures, as was the skin. The large dry, sterile dressing was applied. Prior to this Bacitracin solution was instilled into the subcutaneous tissues.

The baby appeared to withstand the procedure satisfactorily and at the close of the procedure, was able to move both lower extremities.

Dr. Lourie:amh
ED B-398 & A-395

(Lumbosacral teratoma and lipomatous myelomeningocele)

While I was enjoying my whiskey nipple at knife point, my parents waited on a hard bench in the hallway, one floor below the operating suite. For eight hours they waited and jumped every time the elevator doors slid open and another gurney rolled past their bench.

Finally, the doors opened and my parents saw me in a hip sling, being wheeled down the hall by five interns. They were laughing and joking while I screamed.

Dr. Lourie soon came to tell my parents that the surgery had gone well and he had experienced no complications during the procedure. The fact that this young doctor had operated on their daughter surprised my parents, as they had assumed the senior head of neurosurgery was to operate. Dr. Lourie appeared to be fresh out of college to my parents. He was 32 at the time. He explained the procedure to my parents and told them what he had found during the operation. He also told them what they could expect in the coming months and that he would be following the case closely.

The pathology report confirmed the surgeons' diagnosis. Letters were sent to the attending physicians at Chenango Memorial commending their initial work and bringing them up to date.

STATE UNIVERSITY OF NEW YORK

Upstate Medical Center

766 IRVING AVENUE
SYRACUSE 10, NEW YORK

PATHOLOGY DEPARTMENT

Doctor Lourie Path No. 61-4318

Old Path No. _____

Name Barbara Tefft Hospital Memorial

Ward _____ Date 7/13/61

Tissue:

History: Female. Age: 2 weeks. 9 x 5 x 7 cm. mass
midlumbar spine area, covered with skin; soft,
transilluminates. Defect in laminae.

Specimen: 1) part of mass
2) bowel appearing tissue
(PHOTOGRAPHED)
3) part of mass
4) bone
5) part of mass (mesentery)

Duration:

Clinical Diagnosis: teratomatous meningocele

Gross Description: The specimen is received in 5
parts.

The 1st part is labeled "part of
mass" and consists of a piece of
soft, lobulated tissue of a pale
brown color, and measuring 0.9 x
0.6 x 0.4 cm.

The 2nd part is labeled "bowel appearing tissue" and
consists of a hollow, tubular organ measuring 10 cm.
long, having an irregular caliber and measuring 2.3 cm.
in the least diameter and 3.5 cm. in the greatest
diameter. Both ends appear to be blind. It is curved
and on the concave surface there is a small semilunar,
thin wall with prominent blood vessels that spread out

and course along the external surface of the tubular structure. The whole specimen has a brownish-pinkish to grayish-pink color. It appears to be thin walled. Three pieces of brown lobulated tissue aggregating to 3 cm. also comes with the specimen. The specimen was received in a container with formalin.

The 3rd part is labeled "part of mass" and consists of a shaggy, fibrous, hemorrhagic, moderately soft mass measuring 5 x 1.4 x 2 cm. The point where it is greatest in diameter, the cut surface is homogenous and pinkish to hemorrhagic in color.

The 4th part is labeled "bone" and consists of a hard piece of tissue with a purplish color and measures 2 x 0.5 x 0.5 cm. Mounted entirely to be decalcified.

The 5th part is labeled "mass from mesentery" and consists of a thin membrane with a hemorrhagic surface on one side which is faintly trabeculated while the opposite side has a small amount of soft, yellow, lobulated fat attached. The specimen measures 4.5 x 2 x 0.5 cm.

DLB/k

Blocks: A - 1 - 1st part, mounted entirely
 B - 1 - 3rd part, part with greatest
diameter
 C - 1 - with least diameter
 D - 1 - 4th part, mounted entirely, decal
 E - 1 - 5th part

Addenda to description: Number two bowel contains clear gelatinous, shiny material inside. It is thin walled and trabeculated and in some areas, compartmentalized. There are at least four identified compartments. One of the compartments contains more or less white pasty material instead of gelatinous material.

Blocks: F - 1 - to include cyst with pasty
material
 G - 1 - other representative block

Diagnosis:

Teratoma with glandular, muscular, cartilaginous,
osseous, squamous and gastrointestinal tissue.
These specimens show no evidence of malignancy.

July 25, 1961

Dr. Richard Hosbach
Norwich, New York

Dear Dr. Hosbach:

I thought perhaps you would like a brief summary of the hospitalization of your patient Barbara Tefft. A more complete summary will be sent to you when this becomes available from the files at Memorial Hospital.

Your excellent letter sent to Dr. Pickett contains all the pertinent information and the history. Physical examination on admission revealed an intact infant save for the obvious soft tissue mass in the lumbo-sacro region. This mass did transilluminate but not as clearly as one might expect were it a meningocele. There were no transmitted pulsations on crying or respirations. The mass by palpation appeared to contain some areas of solid material. One bony spicule could be palpated at the inferior portion of the mass. A bony defect in the vertebral column could be palpated at the base of the mass. X-rays of the spine revealed an area of widening of the interpedicular areas over the fourth and fifth lumbar and first sacro vertebrae. Soft tissue x-rays of the mass revealed some areas of bone. The preoperative diagnosis was that of a teratoma. There was no gross neurologic deficit save for some decreased spontaneous movements in the left leg and a somewhat hypoactive anal reflex to pin prick.

At operation a most interesting lesion was found. The mass contained one loop of fully and normally developed small bowel. The bowel had a normal appearing mesentery with a normal pattern of blood supply. This loop of bowel was attached by a fiber stalk to a bony spicule arising from the sacrum. Another type of tissue present appeared to resemble that of pancreas but on microscopic study this was reported to be only fatty tissue. A third encapsulated firm mass was also reported to be only degenerated fatty material. After removal of these separate three masses the base of the sac was now seen to pulsate and to herniate up into the previous exposure. This on examination proved to be a lipomatous myelomeningocele. The lipoma external to the herniated dural sac was removed. On opening the herniated sac it was seen that the conus medullaris and cauda equina were attached to the inferior wall of the sac. The lipoma was

shaved off from the conus medullaris and the later structure freed from the wall and dropped back into the vertebral canal. The bony defect in the dorsal elements of the vertebral canal was approximately one inch long and three quarters inch wide. A water tight closure was then obtained from the dural sac. Lumbo-sacro fascia was reflected as a hinged flap enclosed over the dural closure. Good tissue was available for closure of the subcutaneous and cutaneous layers.

The child has done very well post-operatively. The head measurement on admission was 33 centimeters, it is now just 34 centimeters and the fontanel has remained flat and sunken. The neurologic status is the same as that of the preoperative evaluation.

The only thing of concern in the child now is that there is a small layer of lipomatous tissue which does involve the conus medullaris. It is hoped that this may remain quiescent but one must consider the possibility in the future that this could again grow and produce compression of the lower spinal cord. Consequently it will be necessary to follow the child closely throughout the years of growth. It is still possible that hydrocephalus could develop as it is frequently seen as an accompanying abnormality in myelomeningoceles.

Thank you very much for referring this most interesting patient to us. Dr. Pickett was kind enough to ask me to take care of the child in view of the fact that he thought preoperatively the lesion would involve the central nervous system. I should like to see the child again in approximately six weeks for follow up. If I could be of any further aid in handling this problem I hope you will feel free to call upon me.

Sincerely yours,

Herbert Lourie, M.D.
Asst. Professor of Neurological Surgery

HL:bl

For the next two weeks my parents traveled the 85 miles to Syracuse to visit me two or three times a week. My sister remained at home with our paternal grandmother while our parents visited me. My father took days off from work to make the trips.

It was the height of pruning season for my father's Christmas tree plantation. Pruning help was unaffordable and unavailable. Fortunately, Russ Mulvey, a co-worker at the Conservation Department, volunteered his help after learning of the situation. The only payment he accepted was the strawberry shortcake and beer my mother served them for refreshment.

In the early afternoon of July 21, my mother and sister were watching a history-in-the-making event on TV for the umpteenth time. The US-Soviet space race had officially begun in April of 1961. Today Walter Cronkite was at Cape Canaveral covering the 15 minute space flight of US Astronaut Gus Grissom and Liberty Bell 7. Grissom was being pulled from the water and asking for something to blow the sea water from his nose on, when the phone in the kitchen rang. My mother reached up to turn down the

volume on the large black and white Zenith that sat on top of the refrigerator before answering the phone. The caller was Dr. Lourie asking if they would like to bring their baby home tomorrow.

Ten days after surgery and nearly a month after my birth, I was released to go home for the first time. The bandages came off and the stitches were removed. Instructions were issued to my parents. There were no restrictions. A regular diet for my healthy appetite and full baths were allowed. The only requirement was for me to remain in the prone position while asleep, until my return visit six weeks hence.

world as possible. The motion of the car eventually lulled me to sleep.

I was abruptly awakened by the slam of the car door when we stopped in Oxford. My father ran into the pharmacy for a few last minute necessities while my mother stayed in the car with me. An acquaintance of my parents stopped to chat with my mother and offered her condolences upon learning of my "condition."

For the remaining three miles of the trip home I squirmed in my car bed. My mother reached over the seat back in an attempt to keep me still. We pulled into the driveway of the red and white farm house, welcomed by my grandmother who was waiting on the stone porch. She retreated into the house to awaken my napping sister as my parents began unloading the car. My mother carried me into the house through the wide entrance off the porch.

My first sight was of the hand-hewn beams of the ceiling in the keeping room. The second was of my grandmother, leading my sister by the hand as she rubbed the sleep from her eyes. The instant she saw me, my sister screamed and hid behind her grandmother.

As Dr. Lourie had requested, I returned to Syracuse for my first follow-up exam six weeks later. The exam went well. After more pinpricks and head measurements,

all was declared status quo. There was no evidence of hydrocephalus or further neurological impairment.

Frequent trips to visit Dr. Lourie were the mainstay of my first year of existence. For my parents some of the uncertainties of my physical future were clarified. The risk of mental impairment was deemed minimal because of the fortunate absence of hydrocephalus. By pinprick testing Dr. Lourie determined that innervation of the lower extremities was exceptionally good considering the severity of the lesion. There were no guarantees offered, though. Only time would reveal the maximum physical capabilities of their daughter.

My parents were encouraged to prepare for a potentially ambulatory child. Dr. Lourie expressed his concern over the external rotation of both lower extremities. He advised my parents to consult with an orthopedic specialist regarding the possible necessity of physical therapy, bracing or surgery to correct the abnormal positioning of my legs.

Herbert Lourie, M.D. November 8, 1961

Name: Tefft, Barbara Referred by: Dr. R. Hosbach

CHECK-UP
November 7, 1961

This four month old child was discharged from Memorial Hospital July 22nd following a repair of a lumbar teratoma and lipomatous meningocele. The child has done very well since returning home. There has been no problem with her eating. She has been fully alert all the time. The mother thinks that the child wets no more than did her other children. Her stools have been apparently fairly well formed.

Physical examination reveals a well appearing child. The anterior fontanel is normal in size and is soft and sunken. There is no evidence whatsoever of hydrocephalus. The head measures 16-1/2 centimeters as compared to 17-1/2 for the chest. Cranial nerves appear to be grossly intact as do the upper limbs. There is a soft tissue swelling in the lumbar area which appears to me to be slightly more prominent than at the time of discharge. This does not appear to be due to fluid but rather to an accumulation of some tissue in the subcutaneous space. The mass measures 4-1/2 centimeters in its horizontal axis and is two centimeters in its vertical axis. The mass is soft but does not pulsate nor is it fluctuant. The scar from the meningocele repair is strong and well healed. Both lower extremities appear to be externally rotated, but this may well be only a persistence of the fetal position. There is no gross deformity of the lower limbs. The child appears to feel pin prick throughout the lower extremities and movement is present in all joints including the toes. Knee jerks could be obtained at times, I could not obtain ankle jerks. There seems to be good tone in the anal sphincter although I did not obtain an anal wink reflex. Some urine was expressed when the child cries forcefully. This latter finding may indicate some faulty innervation of the vesicals' sphincters.

The child appears to be doing very well at this time. It should be noted that the lipomatous meningocele did have a stalk which did go down and become intimately associated with the conus medullaris and therefore there is always the possibility that the intramedullary portion of the lipoma could recur. I have discussed this problem with the parents. I have also advised the parents that the child should be seen by a specialist concerning the external rotation of the lower limbs.

The parents were asked to return the child in three months for another check-up.

Herbert Lourie, M.D.
Asst. Professor of Neurological

Surgery

HL:jaw
copy to: Dr. R. Hosbach
Dr. M. Boname

I was referred to the newly organized Handicapped Children's Clinic in Norwich for orthopedic evaluation. There were at that time few specialists of any sort in this rural area and no orthopedists. The clinic brought various specialists to the County Office Building once a month to provide the only medical care many handicapped children in the county received.

My first visit to the clinic resulted in a decision to begin physical therapy immediately. It was hoped that passive range-of-motion therapy would stretch and strengthen the weaker leg muscles, preventing the possibility of hip dislocation and to facilitate the eventuality of walking.

A physical therapist made a house call to instruct my mother in the finer points of muscle manipulation. She was instructed to do a five minute series of exercises every time she changed my diaper; an event which to my mother seemed to occur every five minutes.

My physical dexterity appeared to lag behind my chronological peers but I was considered healthy and alert.

Herbert Lourie, M.D. February 16, 1962

Name: Tefit, Barbara Referred by: Dr. R. Hosbach

CHECK-UP

February 13, 1962

The baby is now seven months two weeks old and has continued to do very well. She has not yet begun to crawl but does turn well. Her appetite has remained good. The diaper is dry part of the time so that it is unlikely that the child has over-flow incontinence.

Physical examination reveals a bright and alert child. The head is normal in appearance and measures 17 inches (43.2 centimeters). The fontanel is soft and sunken. The upper limbs appear strong. The abdominal muscles appear normal. There is no abnormal curve to the spine. The soft tissue mass present along the inferior margin of the transverse lumbar scar is unchanged from the prior visit. It still measures approximately 4-1/2 inches in its horizontal axis and two inches in its vertical axis. The mass is soft but is not fluctuant, it does not pulsate. There is good muscle development in the lower limbs and movement is present in all joints. It is difficult to evaluate the reflexes, the tone appears normal. There appears to be intact sensation to pin prick in the saddle area and in the lower extremities.

The baby is doing very well and is to return in six months for a check-up.

Herbert Lourie, M.D.
Asst. Professor of Neurological Surgery

HL:jaw
copy to: Dr. R. Hosbach
 Dr. M. Boname

Shortly before my first birthday, my mother was giving me a bath in the bathinet in the kitchen. As she attempted to lift my soapy, squirming body from the bath water, I slipped like a watermelon seed from her grasp and bounced across the kitchen floor. She quickly retrieved me from the cold green linoleum, anticipating evidence of any number of catastrophic injuries. To her amazement and relief I uttered not a sound and continued the squirming that had contributed to the incident.

To my mother I felt heavier than other babies at the same weight. I didn't move my legs as much on my own. The landmark events every parent waits for such as turning over and crawling were slow in coming. I began to talk before I had begun to crawl. My speech was clear and quick in developing. Crawling on the other hand was a struggle. I preferred sitting and pushing myself along, sliding on my rear. When starting out face down I used my elbow to hitch myself forward, using my legs very little. Later, I used the same elbow method to pull myself into a standing position but only after my parents had started holding me up by my hands to try walking.

Herbert Lourie, M.D. September 20, 1962

Name: Tefft, Barbara Referred by: Dr. R. Hosbach

CHECK-UP
September 4, 1962

The baby has done very well since last seen. She has grown and has begun to speak a few words. She crawls around without difficulty. She has made attempts to stand.

The baby looks bright and alert. The head is normally shaped and measures 46.8 centimeters at maximum circumference. She crawls without difficulty. The soft mass is still present at the lumbosacral junction. It measures about the same as it did last time (4-1/2 inches in its horizontal axis and 2 inches in its vertical axis). It does not pulsate when the child cries. As far as I can tell there is good sensation in the sacral area and the anal sphincter appears to have good tone. The child is doing very well.

The child is to return in six months for another check-up.

Addendum: The anterior fontanel is small, soft, and sunken.

Herbert Lourie, M.D.
Asst. Professor of Neurological Surgery

HL:jaw
copy to: Dr. R. Hosbach
 Dr. M. Boname

The anticipated "first step" was not a distinct event in my development. When I was able to pull myself into a standing position I had a difficult time balancing and would hold on to furniture for support, walking around and between things. When I fell, getting up was more than a minor inconvenience. I would roll over on my shoulder and hitch myself with my elbow to the nearest piece of furniture and start over. The weak leg muscles prevented me from standing up in the middle of a room with no support for balance. Physical therapy was continued until I was walking well.

Herbert Lourie, M.D. March 12, 1963

Name: Tefft, Barbara Referred by: Dr. R. Hosbach

CHECK-UP
March 5, 1963

Barbara has done very well since last seen. She has been walking about without difficulty. She has been talking a great deal.

Physical exam reveals a beautiful little girl who gets about the room with great dexterity. The head is normally shaped. There is no gross deficit in the cranial nerves or upper limbs. There is a slight tendency for the left foot to go into plantar and lateral deviation. However, with pinprick she seems to have good movements in all muscle groups in the lower extremities and she certainly moves her toes well. The reflexes are difficult to elicit in the lower extremities. Pin prick sensation would appear to be intact throughout. There would appear to be good anal sphincter tone. The mass in the lumbar region is the same as when seen six months ago, it still measures approximately 4-1/2 centimeters across and 2 centimeters in height.

Barbara is doing very well, I have advised Mrs. Tefft to take her back to the orthopedic clinic again so that the left foot could be evaluated. She is to return to see me in six months.

Herbert Lourie, M.D.
Asst. Professor of Neurological Surgery

HL:jaw
copy to: Dr. R. Hosbach
 Dr. M. Boname

Herbert Lourie, M.D. September 19, 1963

Name: Tefit, Barbara Referred by: Dr. R. Hosbach

CHECK-UP

September 10, 1963

Barbara has done very well since last seen. She is very active about the house and the yard. The mother notes that the child occasionally will turn the left ankle when she is walking on a rough surface, but otherwise has had no difficulties whatsoever with ambulation. She is not yet toilet trained.

Physical exam reveals a beautiful little girl with no obvious abnormalities. The lipomatous mass in the back remains the same size, measuring now some four and a half inches across and two inches in height. Perianal sensation is good. There is no atrophy in the lower extremities. There is good muscle strength in all groups in the lower limbs. There is a tendency for an equinus position of the left foot and there is slight tightening of the left heel cord. Knee jerks and ankle reflexes were obtained today.

Barbara is doing very well. I had her seen briefly by Dr. James Wray today who recommends that nothing need be done at this time for the slight increase in tone of the left hamstrings. I have advised the mother to continue with foot exercises to the child.

Barbara is to return in six months.

Herbert Lourie, M.D.
Asst. Professor of Neurological Surgery

HL:jaw
copy to: Dr. R. Hosbach
 Dr. M. Boname

After taking the first step, the next major developmental event for most children is toilet training. In my case the accomplishment of this task was delayed in part because of my comparatively slow progress in mastering the art of walking. Additional concern over possible nervous impairment to the urinary system made the occasion of my toilet training a priority event. The social stigma surrounding the inappropriate elimination of bodily wastes calls for alternative methods of toilet training when conventional methods are not physically practical.

Social concerns of controlled elimination are superceded by the physical health aspects of neurological impairment to the urinary tract. Urinary tract infections and renal failure are common problems in myelomeningocele. Faulty nervous innervation to the kidneys and bladder can cause retention of urine, hydronephrosis and pyelonephrosis. After life-threatening physiological aspects of urological management are successfully dealt with, then toilet training for social reasons can be addressed.

As long as the kidneys are filtering blood and urine is flowing through the ureters to the bladder in an unobstructed manner, major health risks are considered minimal.

The greatest problem in social management of urinary waste is impairment of nervous function in the urinary bladder. Waste produced by the kidneys is stored in the bladder for elimination at appropriate intervals.

Normal retention and elimination action of the urinary bladder involves gradual filling and synergistic emptying of the balloon-like vesicle. The bladder is encompassed by a parasympathetically innervated band of muscle, the detrusor, which relaxes as the bladder gradually fills. A ring of muscle surrounding the outlet of the bladder increases in tone. This internal sphincter is sympathetically innervated and provides continence by pinching the outlet closed.

Fullness of the bladder can be vaguely appreciated by the autonomic sensory nerves. The urge to void is most keenly sensed by somatic nerves when sudden pressure in the bladder or stretching of the detrusor muscle occurs.

Micturition occurs when the detrusor muscle contracts at the same time as the internal sphincter and perineal floor relax. This synergistic action allows the

bladder to drop, forming a funnel-like outlet. Voiding is then permitted or restricted by voluntary action of the external sphincter.

In myelomeningocele, impairment of the innervation of one or more muscle or sensory networks is common. Excess retention of urine can be caused by a spastic or dyssynergistic internal or external sphincter, an adynamic detrusor muscle or periurethral fibrosis. This condition can be detrimental to renal health causing vesico-urethral reflux, hydronephrosis, and hydroureter.

Failure to retain urine is a more socially detrimental condition. Nonretention can be caused by a hypertonic, spastic detrusor or a fibrosed, contracted detrusor. Unresponsive sphincter muscles may cause the bladder to remain small, despite a potentially normal filling capacity.²⁰

After thorough evaluation of the urinary tract is made, a number of methods can be implemented to maintain renal health and socially acceptable hygiene. Less severe nervous impairment may require only a timed voiding schedule and/or anticholinergic medication for successful control. When the impairment is more profound, more aggressive treatment must be considered. Clean intermittent catheterization is very frequently used and is less invasive than surgical intervention.

Surgical procedures such as urethrotomy, Y-V plasty, dialation or implantation of artificial sphincters are used when other methods of control are unsuccessful or unsatisfactory.²¹

Management of the bowel also has physiological as well as social aspects in dealing with myelomeningocele. Varying degrees of nervous impairment to the bowel frequently cause lack of sensation, loss of voluntary external sphincter control and/or spasticity. Chronic constipation is common when a tight anal sphincter is present.

Normal defecation, as with micturition, is a synergistic action. The pelvic floor and internal sphincter relax while abdominal and rectal muscles contract, pushing contents into the anal canal. Rectal contents touch sensitive nerve endings which send messages identifying the contents as solid, liquid or gas. Voluntary control of the external sphincter either allows the contents to pass or, if inappropriate, to contract the sphincter, pelvic floor and buttock muscles, thereby pulling rectal contents back into the abdomen.

When nervous innervation to the bowel is impaired, sensation and control is diminished. Alternative methods of training the bowel are necessary. Stool consistency may be modified by diet to avoid constipation or, more

socially detrimental, diarrhea. Fecal impaction can be avoided by establishing regularity through the use of enemas, suppositories or oral cathartics.

Personal hygiene plays an important role in the health of the integument as well as in maintaining an acceptable social quotient. Lack of sensation in the skin of the genital area can lead to unnoticed irritation and infection if not properly attended.

The extreme and unnecessary social stigma surrounding control of the elimination of bodily wastes may have a strong psychological impact, causing social withdrawal. Prompt evaluation and remediation facilitates a more normal social integration and independence.

The symptomatic difficulties that delayed my successful toilet training were urinary frequency, delayed bladder sensation, stress incontinence, incomplete voiding and constipation. These symptoms were managed by a number of strategies.

Diapers were used intermittently until I began school, along with constant reminders to use the bathroom. Fluids were restricted for outings where toilet facilities were not immediately available. Combining neurologic impairment with a pre-schooler's

reluctance to take time out from an absorbing activity just to go to the bathroom results in frequent accidents and near misses.

When constipation became a problem I was taken to the pediatrician. He prescribed a horrible imitation chocolate flavored laxative to sprinkle on my cereal, which had little effect. Recurrent bladder infections also compounded the difficulties in maintaining continence. Fortunately, the symptoms of infection such as burning and increased frequency could be vaguely sensed. Every time an infection occurred my mother admonished me for sitting on the cold, damp ground and we returned to the doctor's office. Antibiotics were administered immediately to prevent infection from migrating to the kidneys and causing more serious problems.

Continence management during my childhood was attempted mostly by trial and error methods. Such topics were cause for embarrassment and therefore not openly discussed until an emergency situation arose, such as an infection. Every physician I visited, regardless of specialty, inquired about continence and management. I most often replied that I had no problems just to avoid the topic. After we had left the doctor's office my parents would ask why *I* hadn't mentioned various

problems. No one was ever comfortable enough with the subject to bring it up or ask questions. I was treated symptomatically although as a child I was never fully evaluated or examined by a urologist.

Nevertheless, an acceptable level of continence was achieved and I was considered toilet trained at 3-1/4 years of age.

Herbert Lourie, M.D.

April 22, 1964

Name: Teritt, Barbara

Referred by: Dr. Hosbach

CHECK-UP

Barbara returns for a check-up. The mother thinks the child has been doing very well. She does on rare occasions turn the left ankle in. The baby has not yet become bladder or bowel trained, but the mother says the older sister, Cindy, was also quite late in being trained.

Physical examination reveals a beautiful little girl with no apparent deformity. Her gait is normal. The development of the lower limbs and buttocks is symmetrical and normal. All muscle groups function well. There is good perianal sensation and the anal sphincter seems to have good tone. The ankle jerks are weakly present, perhaps a slight better on the right than left. I could obtain no definite knee jerks today however. The mass over the low back is identical in size to that previously recorded.

The baby is to be brought back in six months for another check-up.

Herbert Lourie, M.D.

Assoc. Professor of Neurological Surgery

HL:pc

cc: Dr. Hosbach

Herbert Lourie, M.D.

November 9, 1964

Re: Barbara Tefft

Referred by: Dr. Hosbach

CHECK-UP

November 4, 1964

Barbara has done quite well since last seen. The mother says she is a bright and active child and speaks quite well. She has noted no particular difficulty with her gait. She is trained for bladder and bowel.

Examination reveals a very beautiful little girl in no apparent distress. Her gait is grossly normal although there is a slight tendency for turning in of the left foot. There is good tone and muscle power in the lower extremities. Anal sphincter tone appears normal. The soft mass in the mid-back is approximately the same size, measuring some 5 cm. in its vertical diameter, and 14 cm. in its transverse diameter. The tissue has the sensation of a lipoma by palpation.

The child is doing quite well and is to return in one year for another check-up.

Herbert Lourie, M.D.

Assoc. Professor of Neurological Surgery

HL:svs

cc. Dr. Boname

From the perspective of a three year old, visits with Dr. Lourie were not exactly pleasant. After a long car ride and office wait, the prospect of being systematically undressed, pricked, poked and scrutinized by multitudes of interns did not affect my temperament in a positive manner.

During one particular visit I was extremely uncooperative. Dr. Lourie, who was usually so patient and would help re-dress me, threw my shoes on the floor and left the room. When my mother tried to explain to me that he had saved my life and asked why I didn't cooperate with Dr. Lourie, I tearfully replied, "I don't like him because he has black hair!"

Subsequent trips to Syracuse were made by just my father and me, as my mother stayed home with my sister. My father would drive by the swan pond in Manlius on the way in to Syracuse and we would stop at Sno Top for ice cream on the way back. Howard Johnson's was the usual lunch stop where I would have a hot dog on a toasted New England roll, served in a crimped paper holder. I always gave my father the pickle.

I was still not sure of my liking for Dr. Lourie, but the trips were made less ominous with some amusing diversions. Dr. Lourie also began handing me lollipops after my examinations. It was not until twenty-five

years later that my father confessed to sneaking them to Dr. Lourie for him to give to me as a peace offering.

Herbert Lourie, M.D. October 20, 1965

Name: Barbara Tefft Referred by: Dr. Hosbach

CHECK-UP
October 19, 1965

Barbara was in the office and the father reports that she has done quite well. Mentally he believes that she has developed normally.

On examination she is a beautiful little girl who is so bashful that she spoke but one word "thank you" when at the end of examination I gave her a lollipop. Otherwise she was totally silent. Her gait with shoes on appears grossly normal, when she walks barefooted there is slight in turning of the left foot. The legs are normally proportioned. An ankle reflex could be obtained with the child in the prone position but not in the sitting position. The knee jerks are faintly present. There is a tendency to pes cavum on the left, and there is slight hammering of the left toes. Sensation was very difficult to evaluate for she would not respond even when pricked on the hands. The sphincters are intact by history. The mass over the back is approximately the same size although it appears smaller in that the child has continued to grow.

Barbara is doing quite well and is to return in one year for another check-up.

Herbert Lourie, M.D.
Assoc. Professor of Neurological Surgery

HL/meb

My sister had started kindergarten in September of 1965 and every weekday I watched her board the yellow bus for that mysterious place called school. I remained home with my mother to make paper chains, Santa Clauses, Valentine men and May baskets. My mother had been an art student at the Rochester Athenaeum and Mechanics Institute just prior to World War II and I enjoyed the benefits.

We lived three miles from the small village and there were no other children on our country road. Since my mother didn't drive, weekends and medical appointments were the only times I left home. My sister and I would accompany my father on the ritualistic Saturday trips to the dump, Post Office, bank and grocery store for the weekly shopping.

In December of 1965 I left home for a different reason. I had been tucked into my crib after a dinner of steak, grilled in the fireplace. Later that night I awakened with a severe internal pain that hurt more with every breath. I took my usual route climbing out of the crib, onto my parents' canopy bed and down to the floor. My father was cleaning the ashes from the fireplace when

I walked into the keeping room to announce that I wasn't feeling well. My temperature was over 102° when they called Doc Boname. The following morning I was admitted to Chenango Memorial and spent the next two days under a sheet with ice and cold cloths.

The fever and pain subsided and on the third day I was allowed to go home. No specific diagnosis was made but conservative symptomatic treatment seemed to work.

CHIEF COMPLAINT & PRESENT ILLNESS: Onset 12/16/65 at about 2 a.m. when the child awakened and complained of stomach ache. The history shows that on 12/14/65 she had an episode of nausea and vomiting which cleared rapidly and the remainder of the day and the next day she appeared to be normal. The only other symptom noted was the presence of slight cough. The child was seen in the examiner's office and following the examination it was felt advisable to admit her to the Chenango Memorial Hospital with orders for blood count, urinalysis, and surgical opinion of the abdomen.

PAST HEALTH AND ILLNESSES: This child was born in the Chenango Memorial Hospital and had an extensive birth defect which was a spina bifida. The birth date was 6/27/61 and she was discharged from this hospital directly to the Upstate Medical Center at Syracuse. At Syracuse she was admitted to the services of Dr. Herbert Lourie, Assistant Professor of Neurological Surgery. The diagnosis at Syracuse Medical Center was lumbar teratoma and lipomatous meningocele. The records show an admission date of 7/7/61 and a discharge date of 7/22/61. During this interim surgery was done for the condition noted. Subsequently the records show periodic re-examinations by the neurosurgical department of the Syracuse Upstate Medical Center and the result obtained was very excellent. The actual date of operation was 7/12/61. On Nov. 27th, 1961 she was referred to the local orthopedic clinic following observation of some degree of external rotation of the feet. Examination at the clinic and also examination and x-rays of the hips were essentially negative. The next date of examination by the present attending was 6/6/62 at which time she had some skin lesions which were presumably due to insect bites. This was entirely minor and cleared rapidly. The child's immunizations are all entirely up to date and include DPT, polio, smallpox vaccination, and measles immunization.

FAMILY HISTORY: Both parents are living and in good health. She has one sister living and well. No chronic familial diseases present.

MGB:bf
d: 12/18/65
t: 12/20/65

BARBARA J. TEFFT ADM. NO. 159
Dr. Boname 91400 12/16/65 adm.
Temp: 101.8 rectally
Pulse: 120 Resp: 32

She is very stoical and apparently bashful and answers questions with little or no spontaneity.

This child is 4-1/2 years old and appears acutely ill.

SKIN: Hot and dry, there is a marked degree of malar flush of the face.

HEAD: Scalp is entirely normal, covered with a good growth of blonde hair.

EYES: Pupils are equal and react normally. The conjunctivas are injected.

EARS: Neither membranae tympani can be visualized due to the presence of impacted cerumen bilaterally.

NOSE: Breathing is free, there appears to be no discharge but the nasal membranes are somewhat hyperemic.

MOUTH: Oral and dental hygiene is good. Tongue is slightly furred and moist.

PHARYNX: Appears very injected.

CHEST: Normal contours, normal costal and abdominal breathing observed.

LUNGS: Shows normal vesicular resonance and normal vesicular breath sounds.

HEART: The apex beat is normally situated, the sounds are of good intensity, regular and no murmurs are present.

ABDOMEN: The abdomen shows moderate distention and tympani on palpation and percussion. There is apparently generalized tenderness with possibly a point of maximum intensity located in the right lower quadrant. No definite rigidity or rebound tenderness is elicited.

BACK: The lower back shows an area of previous surgery described in the history.

EXTREMITIES: The lower extremities - reflexes are physiological and are entirely intact. No abnormalities seen as regards this area.

WORKING DIAGNOSIS:

- 1 - Probably an acute upper respiratory infection with possible gastroenteritis.
- 2 - Possible appendiceal involvement with acute infection.

MGB:bf

d: 12/18/65

t: 12/20/65

In June of 1966, my parents were notified by my sister's teachers that Cindy would need to attend summer school and undergo testing to determine the nature of her academic deficiencies. Transportation for my sister's summer school trips was arranged and I saw Cindy leave with Mrs. Stone three mornings a week.

I spent that summer "helping" a next door neighbor convert our former milkhouse into a playhouse. My parents had hired Mr. McAdams to renovate the ten by twelve foot building. When the construction was almost complete, my father removed the door to cut a heart-shaped window in the middle that could be opened and closed. My mother stained the floor and painted the walls yellow. She also made curtains for the windows and painted the heart red. A window box full of flowers was just like the one outside the kitchen window.

The summer blazed hot and dry. The spring that supplied water to the house dried up and a well had to be drilled. This proved to be a major inconvenience for my parents and great excitement for me. Being a gregarious child, I made friends with the well-drillers who remained for most of the summer. They worked their way through seemingly solid rock. After three location changes and much broken equipment, water was finally located 325 feet

down on the south side of the house. Before my drilling friends left, I presented them with balloons.

In September of 1966, I began school. I was fortunate in that I was fully ambulatory and apparently as "normal" as other five-year-olds. This was especially fortunate because there were no special schools or programs available locally that were equipped for myelomeningocele children who did suffer the common occurrences of hydrocephalus, paralysis and incontinence.

During the required pre-admission physical examination and interview with Betty Cosen, the school nurse, and my teacher, Mrs. Mott, my mother informed them of the highlights of my medical history. Since I required no special facilities I could expect to be educated along with my peers.

Kindergarten was a generally positive experience for me. I was a very social child and this was my first opportunity to interact with other children on a regular basis. As the kindergarten year progressed I was viewed by my classmates as "teacher's pet." This was primarily due to the fact that I was most often chosen as hall monitor for the daily after-nap excursions to the "basement." In actuality, Mrs. Mott had assigned my nap-time location next to the hall door in order to

facilitate my timely exit to the bathroom. By designating me as hall monitor she was assured that I would not have to wait in line for others to use the facilities. Mrs. Mott's strategy was successful in helping me avoid embarrassment, although my classmates and I did acquire a somewhat lofty and false impression of her opinion of me. At that age being "teacher's pet" placed me in a position of leadership and envy by my peers; later it would become detrimental. I learned and played with my classmates and at the end of the school year invited some of my new friends to a sixth birthday party at my house.

Herbert Lourie, M.D.

November 2, 1966

Name: Barbara Tefit

Ref. By: Dr. Hosbach

CHECKUP

November 1, 1966

Barbara was in the office for her annual checkup. She is doing quite well. She's had no problem with bowel nor bladder, the father thinks she is as dexterous and agile as is her sister.

On examination the child is to all external appearances quite normal and very pretty. The soft tissue lump on the back is about the same as when last I saw her. She seems to have good sensation in the sacral dermatoses. The ankle reflexes are not as brisk as are the knee reflexes. There is still slight pes cavum and hammer toe on the left and slight inversion of the left foot.

Barbara seems to be doing well enough. She is to return in one year for a checkup.

Herbert Lourie, M.D.
Assoc. Professor of Neurological Surgery

HL:ms

Our neighborhood was changing. We now had neighbors with children within a half mile distance of our house. I frequently wandered to various neighbors' houses to play. My favorite place to visit was the large farm just down the road. Linda and Bob Marshman's son John was three years my junior, but a great playmate as we grew older. We spent many hours in his sand pile playing with the yellow Tonka trucks and model of their barn that his grandfather had made. John would occasionally join me in our wading pool in the back yard.

As outgoing as I was, my sister was exactly the opposite. She was extremely shy, quiet and obedient, preferring solitary pastimes. Cindy rarely joined in neighborhood activities, leaving discretely and prematurely when she did. Conversely, I often disappeared from our yard without notice, to the consternation of my mother. I could be found anywhere from Marshman's alfalfa fields on the river flat to my father's Christmas tree plantation. I had also met another Barbara on my school bus who lived just a half mile north of our house. She was in my sister's class in school but only ten months older than me. Her house

became a favorite bicycle escape once I had mastered that mode of transportation.

Twice a year, every year, my family went on the typical family vacation. My father would take a week off each spring and summer when we would travel to various historic landmarks and museums along the east coast. The summer of 1967 we went to the Exposition in Montreal, Canada.

Expo '67 was memorable for me not just for the sights and atmosphere. I most recall being extremely tired from all the walking. The farther and longer I walked, the more my feet would roll laterally. This caused me to walk on the outsides of my feet and was most noticeable on the right. I frequently became so fatigued that I would just sit wherever I was. My parents would have to retrace their steps to find me. Eventually, the paternal piggy-back became my sightseeing vehicle. By the end of this vacation trip the outsides of my black patent leather Mary Jane's were completely worn, although the soles looked virtually new.

Herbert Lourie, M.D.

September 14, 1967

Name: Barbara Tefft

Referred by: Dr. Hosbach

Office Visit

Mr. Tefft had Barbara in the office on September 12, 1967 and reports that the child has been doing very well. The only thing that he has been aware of is that when Barbara seems very tired that the right foot seems to turn in a little.

On examination the child looks quite normal. I can see no obvious abnormality in the feet, certainly on specific testing there is no weakness there. There is no obvious sensory loss. The former slight pes cavus and hammer toe on the left are not as obvious as they were before, there may be minimal slight inversion of that foot, again this is barely noticeable. The soft tissue mass on the back is less obvious than before, likely due to the continuing growth of the child.

Barbara is getting along very well, I plan to see her again in one year. In the meantime she is to check in with the orthopedic clinic at her home town to see if there is anything to be done about her foot.

Herbert Lourie, M.D.

Associate Professor of Neurological Surgery

HL/ss

Routine dominated our household and interruption in the routine was conspicuous. My sister and I were never left with a babysitter. We always accompanied our parents to restaurants, antique auctions or shopping. On very infrequent occasions, my mother's sister, Grace, would pick Mom up at our house. They would not return until late in the day. Cindy and I would stay at home with Dad, enjoying a fire in the fireplace or a walk through the pines. Nothing was ever said about where my mother and aunt went on those occasions. Ultimately, I learned that they had been driving to Rome, N.Y. to visit my mother's severely retarded son, Charles, from her previous marriage. It was a difficult trip for my mother as her son barely recognized his family. My sister and I had a half-brother that we would never see.

Our neighborhood changed when Mr. McAdams, the next door neighbor who had worked on our playhouse, died. His widow auctioned the contents of the house and sold it to a young couple, Skip and Siri Rogers, who had a daughter two years younger than myself. After overcoming her initial shyness, Brenda and I became fast friends. This relationship was never accepted by my mother. As the

years passed the Rogers' family became an obsession with her. For as many times as I had asked, grounds for this feud were never formally stated and the only theory I have been able to develop is based on socio-economic differences.

Eventually, we were forbidden to have Brenda play with us in our yard or to go to her house. The rest of the neighborhood would play with us on the rope swing and trapeze which hung from the branches of a large maple in our back yard. If my mother saw Brenda playing with us she would yell "Git!" from the back porch and run outside, broom in hand.

The fatigue I had felt during the Expo '67 vacation continued to increase. I developed a permanent equinovarus deformity in both feet. In school I became fatigued climbing the three flights of stairs to my classrooms. Participating in gym class was physically tiring and always being the last one chosen for teams is not an ego-booster for anyone. I previously had had no obvious physical differences to my peers but as my orthopedic difficulties developed so did their physical prowess.^{22, 23}

Herbert Lourie, M.D.

September 18, 1968

Name: Barbara Territ

Referred by: Dr. Hosbach

OFFICE VISIT

September 17, 1968

Barbara has done well. She is doing well in school. Neurologically there is an apparent slight smallness to the calf muscles. She has a foot deformity in which she walks on the outside of the feet. The ankle jerks are present. Knee jerks are 2+. There is no sensory loss to pinprick in the perianal or posterior thigh area. The mass is much the same over the back.

Dr. Murray plans to carry out a tendon transplant to correct the foot deformity. He is to contact Mrs. Territ about this. Barbara is to see me again in one year.

Herbert Lourie, M.D.
Assoc. Professor of Neurological Surgery

/pt

September 25, 1968

Mrs. Leonard Tefft
R.D. #3
Oxford, New York

Dear Mrs. Tefft:

I have spoken with Dr. Lourie concerning the proposed surgery on Barbara's feet and he does not feel that there would be any problem from the neurosurgical point of view to such surgery. I told him I would communicate with you and if you wished, to proceed with this we should be making the appropriate applications to the Handicapped Children's Program and be working out a schedule. This surgery would consist of tendon lengthenings and transfers and she would be in the hospital approximately one week and in plaster cast for approximately five weeks.

If you have specific questions, I will be in Norwich again in October or you may write or call at any time.

Sincerely yours,

David G. Murray, M.D.

DGM:al

I continued to visit the orthopedic clinic in Norwich. Dr. David Murray evaluated my orthopedic progress and decided that surgery was necessary. Nervous innervation to some muscles in my lower legs was gradually becoming impaired. This caused an imbalance in muscle tension of the foot, resulting in equinovarus deformity. Nervous impulses to muscles serving the medial portion of the feet were apparently uninterrupted. Disruption of nervous function to the lateral foot muscles caused these muscles to become weak or flaccid. The normal tone of the medial muscles versus the weaker lateral muscles caused the lateral rolling of both feet.²⁴

On November 11, 1968, I returned to Syracuse for my second surgery. At seven years old the only time I had been away from home overnight was for hospital admissions. I had just recently graduated from my crib at home and strongly objected to the rails on the hospital bed.

Surgery was scheduled for the morning of the 12th. The night before the surgery a nurse's aide tracked me down in the play room. I was given a freezing cold bath.

Even though I protested over the temperature of the water, the aide didn't seem to care. I was brusquely scrubbed and sent back to my bed at an unjustly early hour.

My father stayed at a motel three miles from the hospital to be with me for this ordeal. The surgery went well and I was to remain at Upstate Medical Center for seven days.

Both feet were in plaster casts up to my knees and had open toes. I was supposed to remain on my back with feet elevated on a stack of pillows. This position lasted only as long as the doctors or nurses were in the room. The second they left I kicked the pillows over and sat up.

Dad came to see me every day. He commuted to work from Syracuse for part time hours and spent the rest of the time with me. He also was in the midst of Christmas tree season, spending every free minute cutting and hauling this year's harvest. One morning he had promised to be at the hospital early. Syracuse winters are notorious for snow and three feet of it had fallen during the night. My father was snowed in at his motel and the hospital staff had to suffer with me until he finally arrived. He bought a deck of cards and a tic-tac-toe

board at the hospital gift shop. Pitch and poker kept us occupied for days.

My Aunt Grace brought my mother to visit one day when I was in a miserable mood from my confinement. I said that I'd rather see my Dad and after ten minutes they left, insulted. Rarely did my mother come to visit during any of my subsequent hospitalizations.

STATE UNIVERSITY HOSPITAL

UPSTATE MEDICAL CENTER

DISCHARGE SUMMARY

<u>TEFFT, BARBARA J.</u>	<u>101 331 7 019</u>
Name	No.
<u>R.D. #3</u>	<u>Oxford, New York</u>
Address	
<u>11/11/68</u>	<u>11/17/68</u>
Admitted	Discharged

HISTORY: This patient is a 7 yr. old girl with a meningomyelocele repaired at birth and with mild sequali in both lower extremities. She has walked with increasing equinovarus deformities of both feet and comes in at this time for tendon transfer for correction.

EXAMINATION: The patient has a healed scar over the lower lumbar spine with no evidence of breakdown. She has very mild residual neurological deficit in her lower extremities with tightness of the heel cords and varus deformities of the ankles bilaterally.

LABORATORY DATA: CBC and urinalysis was within normal limits.

HOSPITAL COURSE: On 11/12/68 the patient was taken to the operating room and bilateral heel cord lengthenings with transfer of the tibialis anicus laterally and release of the tibialis posticus tendons was performed. Her post operative course was uneventful and she was discharged home 11/17/68 for office follow up.

FINAL DIAGNOSIS: Myelomeningocele, equinovarus deformities of both feet.

David G. Murray, M.D.

cc: 1 Dr. Murray
DGM/sek

I was released on November 17th. Weight bearing was not allowed for two weeks so a wheelchair was rented and I returned home. My extended absence from school brought a sunshine basket of toys and letters from my classmates. I had a tutor at home for four weeks to keep up with school work.

Finally, one inch high rubber pads were added to the bottoms of the casts, allowing me to walk without damaging the plaster. My father took me on a special trip to school to attend a class Christmas party, for which he had donated the tree.

The casts were removed in January. The surgery was considered successful although my calves were noticeably smaller and there was less voluntary movement in my left foot. After walking in heavy plaster boots for so long, my lower legs felt as if they were floating in water. When Dr. Murray swung my legs over the edge of the examining table I couldn't let them dangle - they floated back up! After he bent my knees and held my ankles down twice I was able to keep them down myself. I exited the doctor's office wearing socks and shoes following a brief session of walking practice.

Herbert Lourie, M.D.

September 22, 1969

Name: Barbara Tefft

Referred by: Dr. R. Hosbach

OFFICE VISIT

9/16/69

Barbara continues to do very well. She has no urinary problems. She is doing well in school.

On examination the lipomatous mass on the midback is approximately the same size. Her calves do appear somewhat small and the ankle jerks are absent. There is mild but definite weakness in dorsiflexion and plantar flexion of the feet. There is atrophy of the intrinsic muscles of the left foot. The glutei seems strong and contract briskly to a pinprick stimulation. She feels the vibratory sensation throughout the right lower extremity, and I think she does in the right foot. She does not feel the same tuning fork in the left foot.

Her condition seems stable and I plan to see her again in one year.

Herbert Lourie, M. D.
Professor
Neurological Surgery

/pt

The summer of 1970 was one of agonizing anticipation for me. My parents had uncharacteristically decided to add an in-ground swimming pool to our backyard. In order to reduce the cost of labor, my father did much of the work himself. One of his first jobs during his 1930's hitch in the C.C.C. had been to hand-dig twelve foot water holes for fighting forest fires deep in the woods. Having this experience under his belt gave my father the idea to tackle the excavation of the pool singlehandedly. Dad dug a 16 by 32 by 4 foot hole using a pick and shovel in the rocky soil before hiring Mr. Meade to dig the eight foot depth with his backhoe. My father was 57 years old when he added this project to his Christmas tree pruning chores after coming home from work. My mother spent *her* money to install the pool, although she would never once put more than her hand in the water.

Installation of the pool was painfully slow in the opinion of a nine year old. When the milk tanker was finally hired to fill the pool it was September. It took days to filter the milky water clear, only to put the winter cover on. The next summer cement was poured where

the slide and diving board would be erected. Cindy and I prominently left our marks in the wet cement.

As a child I insisted upon participating in numerous social activities. However, I was never able to participate without the presence of my sister. Her shyness and lack of desire to socialize with her peers contrasted sharply with my comparatively outgoing personality. Parental concern over this issue caused me to endure the shadow of my sister in virtually every activity. Requests to visit friends, attend meetings and extracurricular lessons were deferred to my sister. If she did not want to go, I was not allowed. I quickly learned the "divide and conquer" method of manipulation in order to achieve the desired response.^{25,26} Cindy could be easily cajoled to ask permission for certain activities, which was most often granted when she made the request. Mom would say "Go ask your father," Dad would say "Go ask the boss." Each would be told that the other had granted permission and off we would go.

Frequently, Cindy would "change her mind" and return home or never leave in the first place. This was the major operating mode utilized in our house. When this method failed I was reduced to the ever popular tantrum method, to which my father would usually give in just to restore "peace and quiet" to the house.

In this manner I was able to join my peers in such activities as ballet, swimming and horseback riding lessons, Girl Scout and 4-H meetings. For all of my persistence and insistence over joining these activities, my actual experiences in these groups were not always positive.

Ballet lessons were held in the Episcopal Church parish house by an instructor who commuted from Binghamton. The lessons were offered as an attempt to bring culture to our rural village. "Culture," however, did not preclude the cruelty of uninformed children. I was enrolled in ballet shortly after my tendon transfer. My left foot now toed out and fell on the arch somewhat, making for an excellent first position! I caught on quickly to the moves, although I did not possess the physical stamina to sustain the repetitive motions. It was not these problems that dampened my enjoyment of ballet lessons but the pointing and whispering of a red-headed classmate, two years my senior. She made sure that members of her clique were aware that the profile of my lower back was not like theirs. The leotards and tights we were required to wear did nothing to conceal the remaining lipoma on my spine. Ballet was the first occasion for us to become acutely body conscious and I no longer had the protection of my mother's careful

selection of clothing secreting the tumor. Fortunately, my friends either did not notice or were not as concerned about such differences and I discerned no variation in their interactions with me.

Herbert Lourie, M.D.

October 14, 1970

Name: Barbara Tefft

Referred by: Dr. R. Hosbach

OFFICE VISIT

October 13, 1970

Barbara continues to do very well. The soft tissue mass is much the same as was seen a year ago. Ankle jerks are still absent, knee jerks are equal and active. Pinprick seems intact throughout. The gastroc-soleus seems slightly weak and small bilaterally. All in all her neurologic condition seems stable at this time. She is entering her phase of growth and I plan to continue to see her, her next appointment being for about six or seven months.

Herbert Lourie, M. D.
Professor
Neurological Surgery

/blm

Herbert Lourie, M. D.

September 29, 1971

Name: Barbara Tefft

Office Visit 9/28/71

Barbara's father reports that she has been doing very well. Barbara denies urinary problems. The neurologic exam reveals small calves. Ankle jerks are absent. Right knee jerk is 1+, left is trace. Hamstrings and quadriceps and hip flexors are strong. Gastroc-soleus is mildly weak. Dorsiflexion of feet is mildly weak. Vibration is felt in all the toes. Pinprick is felt throughout the lower extremities including the buttocks. She seems to be stable. The soft tissue mass is unchanged. I will see her again in one year.

Herbert Lourie, M. D.

/pt

cc: Dr. Richard Hasbach

My mother had always selected expensive, quality clothing for my sister and me, making us the best dressed children in school. However good the intentions may have been, wearing such clothing further separated me from my peers. While most of the others were wearing jeans I was wearing clothes with labels such as "Danskin" and "Russ". The conformity that I sought at the time would have been much more attainable with a "Levis" label in our rural school district.

The experience in ballet caused me to be more self-conscious. I began wearing looser clothing and let my hair grow to my waist, giving me something to hide behind. In the locker room during gym class I was careful to change with my back to the wall into large, sloppy T-shirts. I, correctly or incorrectly, assumed that no one observed my secret.

An incident at Girl Scout camp when I was ten destroyed my early years of practiced concealment. I had begged and pleaded to attend camp with my friends from school and was again allowed only if my sister also came. The time it took to "convince" Cindy to go delayed the reservation and we were placed together in a group separate from my schoolmates. Upon arriving at camp I requested to be moved to the group that my friends were in, as there was one space available. My sister refused

to stay unless she was in the same tent with me. I conceded in order to stay at camp. We shared a tent with two girls from another town, in the tent section farthest away from the main buildings. Every activity that could be chosen by individual campers became a siamese-twin event for me, with my sister following on my heels. When it was our group's turn to use the showers I chose to wait rather than community shower with the group, saying it was too crowded. As I was approaching our tent after showers I overheard one of the girls ask Cindy why I hadn't showered with the rest of the group. She answered, "Because she has a hump on her back she doesn't want people to see." When I entered the tent they demanded to see my back and threatened to tell the rest of the camp if I didn't. I glared at my sister, spitting mad that she had revealed my carefully hidden secret. After my refusal to become the object of show and tell, they ran to the next tent and brought four other girls back, all asking questions and making threats.

Listening to their questions I realized that I couldn't answer any of them. I had never been informed of the details of my medical condition and was only vaguely familiar with the term "spina bifida." I eventually realized that Cindy had revealed my secret out

of her own ignorance rather than deliberate malice, although this realization did not temper my anger.

The remainder of my time at camp was spent ostracized from the group we had been placed with. My sister spent most of her time writing letters home saying how homesick she was. I distanced myself by spending time with my friends, regardless of assignments. Our tentmates pulled juvenile pranks in my absence, such as stealing underwear and rolling up the tent flaps by our cots during a thunderstorm, soaking our sleeping bags. The counselors were aware of this incident but did nothing to correct the situation, although we were forced to apologize en masse for an incident which hurt another counselor's feelings. An incident of which I was unaware, being with my schoolmates at the lake.

A mandatory five mile hike to the fire tower was the last group activity in which I participated at camp. We were to make campfire pizza in the woods to earn badges before returning. A thunderstorm the previous night had thoroughly soaked the woods and the remaining overcast refused to burn off. We headed out with packs full of the raw materials for preparing pizza. Everything from flour and yeast to aluminum foil was in our packs. Everything except matches.

The disconcerting fact that the counselors had overlooked the inclusion of matches was of little concern to me. As with our family vacation to Montreal, the walking took its toll. Each of our backpacks were evenly distributed to carry approximately eight pounds. Walking was difficult enough for me but the added weight was impossible for the return trip. Lacking matches or even dry wood, we each ate one orange before starting back to camp. Two hunters passed us on the rough logging road and granted the counselors' request for matches, generously including cigarettes to go with them.

I carried my pack part-way back and came to the conclusion that it was either the pack or me. Not being able to ask any of the group to carry it for me, I asked my sister. She carried it a short distance before dropping both of our packs in the road and walking on ahead of the group. I was unaware of this until I heard the others arguing about who would have to carry them back. This definitely did not increase our popularity with the group but no one approached us about the matter.

Upon returning to our tent soaking wet, sweaty and hungry, I noticed that the pain in my left knee was accompanied by swelling. I sat on my cot exhausted, ignoring the bell calling us to the mess hall for dinner. I had walked far enough that day and refused to walk

another quarter of a mile to eat. My knee continued to swell during the night and remained swollen.

Following this week-long adventure at Girl Scout Camp, I made another visit to the orthopedic clinic in Norwich. The swelling in my knee persisted. Elastic bandages were prescribed to control the effusion but any slight amount of activity aggravated the joint.

I gradually accepted the fact that the swelling was something I would just have to get used to. I continued to participate in jump rope and hopscotch games on the playground at school. I became somewhat proficient at basketball, playing on an intramural team during 9th periods at the end of the day.

The desire to participate and be accepted by a peer group was the same for me as it is with most children, my sister being the exception. In spite of the comments and actions of numerous uninformed people with whom I came in contact, I never lost the desire to participate. I joined the school band in the fifth grade, playing the clarinet, sang in the chorus and joined 4-H. I naturally gravitated toward music and artistic endeavors. Academic studies were not my forte with math being my weakest area, English my strongest.^{27, 28, 29} I was a voracious reader from a very early age. I read myself to sleep every night with the Peanuts Collection, Happy

Hollister's and Nancy Drew Mysteries. In the sixth grade my friend Karen shared a book entitled Karen with me. The book was about a girl with cerebral palsy and was written by her mother, Marie Killilea. I was impressed by the optimism and persistence Karen's parents had in raising their handicapped daughter. They founded the National United Cerebral Palsy Association to overcome the medical and social barriers to their daughter's life.

The contrast of Karen's family in comparison with my own made me more aware of how other families function. A pattern of social withdrawal had gradually developed at our house. My sister and I would arrive home from school at 3:15 p.m. In the past we could usually have found our mother in the midst of a variety of household chores. Now she would just be getting dressed. Eventually she stopped doing her nails, wearing make-up and setting her hair in bobby-pin curls every night. She would stay up until nearly dawn, after the rest of us had retired. My father had long since occupied an unrestored upstairs bedroom. Our mother had taken a driver education course and obtained her license at the age of 55 but very rarely drove.

Dad continued to do all of the grocery shopping, including purchasing the weekly carton of cigarettes for my mother. He disliked her smoking as much as the rest

of us but would not verbally object. He merely refused to sit in the room where she was smoking or fanned the smoke at her with the newspaper.

We had formerly spent alternate holidays at my aunt's. My mother had never had any regular social acquaintances to our knowledge and now did not willingly associate even with relatives. The undiscussed trips with my Aunt Grace to visit Chuck at the State School in Rome also ceased.

During weekends and summers neighborhood friends could not come to play at our house because our mother was still in bed. I would grab some food and disappear hours before she arose. Permission for sleep-overs was extremely rare and guests of any sort virtually non-existent. I spent less and less time at home, instead creating a family of my own choosing within our neighborhood. The neighbor's extended farm family became part of my surrogate family, as did the forbidden clan next door. I defied my mother's orders disallowing any interaction with the Rogers'. On more than one occasion I was locked out of the house for associating with "the crud", "the thing", or "the alleycat" next door. My father either admonished me for disturbing his "peace and quiet" or ignored the issue entirely.

The intensity of my mother's dislike for our next door neighbors increased with each passing year. Shortly after the Rogers' had moved in my sister and I were taught the meaning of the word "ignore". We were instructed to pay no attention to our new neighbor as we played in our adjoining back yards. Having already befriended Brenda made this parental decree impossible for me to obey. We would play together surreptitiously at other neighbors' houses, fields and barns.

As we grew older I ceased sneaking to other neighbors' houses to play with Brenda. One day I simply rode my bike into Rogers' driveway, in full view of my mother, and entered their house through the front door. I was always welcomed as a member of the Rogers' family, frequently sharing meals and exchanging small holiday gifts. Cindy never dared defy any of our mother's orders and was further isolated from neighborhood activities, while I was ostracized at home. It appeared to me that the more independence I attempted to gain, the less I was allowed.

To my mother Cindy's introverted and obedient demeanor was preferable to my more demanding style. I was accused of stealing my sister's friends when they would call to speak to me or include me in their activities.

Cindy began assimilating and exaggerating specific characteristics that were conspicuously derived from maternal traits. Anti-social behavior, fear of heights, staying in bed late and dislike of cooking, among a number of other dislikes, became an integral part of my sister's character. Anything our mother did Cindy would do bigger.

One Sunday evening, just as we were sitting down for dinner, the phone rang. As usual, no one rushed to answer it so I picked it up. The call was for my mother. Phone calls for my mother were an uncommon occurrence and I immediately had a premonition regarding its significance. After a brief conversation of "Yes. I understand. Thank you." my mother took her ashtray and cigarettes into her room to sit in the dark.

The call had been from the State School in Rome, where Charles had been placed at the age of thirteen. My mother had just been informed that Chuck had died of pneumonia following a week long hospitalization in the school's infirmary. He had recently turned twenty-one when he died.

Dinner that night was unusually somber. My sister immediately inquired, "What did Barb do now?" when our mother left the room. Dad just said to keep quiet when I asked if Chuck had died. Cindy asked, "Who's Chuck?"

Herbert Lourie, M.D.

October 25, 1972

Patient: Barbara Tefft

Office Visit: 10/24/72

Barbara is in the sixth grade and generally doing well. She about once a month has abdominal cramping, usually during the night which seems to be related to obstipation. She claims to have good bladder control but the father says occasionally she does wet herself.

On exam she is very well developed, a pretty girl, with normal intelligence. There is failure of development of the intrinsic muscles of the feet and her calves are a bit small and the ankle jerks are absent. She seems to have some impaired pin prick sensation over the lower most sacral dermatomes distally over the lower extremities, but perianal sensation to pinprick seems good, and the buttocks seem to have normal bulk. Head is normal in shape and size. Her condition seems stable but I will follow her once a year. She is to take prune juice nightly to try to get good bowel regulation.

Herbert Lourie, M.D.

/jm

cc: Dr. Richard Hasbach

The swelling and fatigue in my left knee became chronic. Elastic bandages were used in a futile effort to control the swelling as the joint became increasingly less stable.

In February of 1973, my father picked me up at school for another visit to the Orthopedic Clinic. While walking through the parking lot on the way to the clinic, I told my father that I thought I should use crutches to prevent my knee from becoming so constantly tired. When we were called into the examining room, Dr. Kochersperger recommended a temporary non-weight bearing therapy and prescribed crutches. He wrote a note excusing me from gym class and we left to rent my first pair of crutches.

At that time no identifying name was provided to define the condition of my knee. Eventually, the progressive degeneration was labeled a neuropathic or "Charcot" joint. Historically, the condition had been medically delineated as early as 1703, although it was not until French physician Jean Martin Charcot published his detailed description in 1868 that the condition was widely recognized.

Without any appreciable external cause we may see, between one day and the next, the development of a general and often enormous tumefaction of a member, most commonly without any pain whatever, or any febrile reaction. At the end of a few days the general tumefaction disappears, but more or less considerable swelling of the joint remains, owing to the accumulation of water in the joint and sometimes also in the peri-articular bursae. On puncture being made, a transparent lemon-coloured fluid has been frequently drawn from the joint. One or two weeks after the invasion, sometimes much sooner, the existence of more or less marked cracking sounds may be noted betraying the alteration of the articular surfaces which at this period is already profound. The hydrarthrosis resolves quickly, leaving an extreme mobility of the joint. Hence consecutive luxations are frequently found, their production being largely aided by the wearing away of the heads of the bones which has taken place. I have several times observed a rapid wasting of the muscular masses of the members affected by the articular disorder...Besides the wearing down of the articular surfaces...you may notice the presence of foreign bodies, of bony stalactites...I am led to believe that they are produced in an accidental manner, and to all appearances chiefly by the more or less energetic movements to which the patient sometimes continues to subject the affected members.³⁰

The pathogenesis of a Charcot joint is also the subject of debate. Charcot believed that the arthropathy was due to spinal cord lesions causing trophic changes in the skeletal tissue of a joint. Support for this theory is observed in clinical findings of elevated temperature and blood pressure in an affected limb.³¹

More recently it has been thought that the commonly reported lack of pain sensation in an affected joint is a

preeminent factor in the development of a Charcot joint. Normal experience of pain upon injury offers protection, lack of sensation results in repeated trauma to the joint.

Charcot joints are most often diagnosed in active patients and are seen as a frequent phenomenon in those afflicted with syphilis, congenital insensitivity to pain, diabetes, leprosy and spinal cord lesions. Overuse of sensory-dulling drugs such as steroids and alcohol have also been observed in cases of Charcot joints. Another common factor in this group of afflictions is the occurrence of ulceration and infection of the integument with the potential of creating an infective osteitis in a proximate joint.

Diagnosis in the early stages of a Charcot joint may be mistaken as hypertrophic osteoarthritis. In both maladies joint effusion following even mild trauma causes distension and separation of articular surfaces. Aspiration of fluid from a Charcot joint is inevitably followed by immediate and chronic recurrence. The fluid appears clear and yellowish, usually with an elevated protein content. Blood may be present following a minor trauma to the affected joint. Fluid build up causes stretching of the articular capsule and ligaments, occasionally rupturing the capsule. Synovial thickening

also occurs. Subluxation eventually ensues when the ligaments and muscles surrounding the joint are overstretched by the swelling.

Degenerative changes in cartilage and bone are observed as debris is ground deep into the synovium and surrounding tissues.^{32,33} Evidence of frequent hemorrhage is also noted. Continued active use of the unstable joint causes further stress on articular cartilage. An active pannus of granulation tissue derived from peripheral synovium or bone marrow invades the cartilage through fissures or fractures, hastening destruction. It is thought that this pannus is the distinctive difference between osteoarthritis and neuroarthropathy.³⁴ The pannus in a Charcot joint is markedly more active and profuse than is seen in osteoarthritis.

When bone is exposed, following destruction of the adjacent cartilage, it begins proliferating and produces new bone. Reactivating endochondral bone formation, as is seen in normal growing bone, creates marginal osteophytes and sclerosis. This newly formed bone may create the numerous intraarticular loose bodies characteristically found in Charcot joints. These loose bodies may also originate from synovial villi that are

pinched off, eventually ossifying into bizarre forms.^{35,36}

The destructive atrophic mechanism of a Charcot joint is followed or accompanied by a hypertrophic repair process. Halisteristic softening of bone tissue and heightened osteoclast activity causes absorption of much intraarticular debris. At the same time, sclerotic osteophytes coalesce creating rounded, fused fragments. These fragments may adhere to articular surfaces in an attempt to restore or stabilize the disorganized joint.³⁷

Clinical examination of an advanced Charcot joint reveals a lax, swollen joint with palpable loose bodies. An audible course, grating crepitus is usually painless to the patient but unpleasant for the examiner.³⁸

Spontaneous fractures of the bones adjacent to a Charcot joint are a frequent phenomenon. These generally painless fractures can mimic symptoms of infection with elevated white count, localized redness and swelling. If not roentgen- ographically diagnosed they may remain untreated or be incorrectly managed.³⁹ The fractures may precede or follow evidence of advanced Charcot joint. This evidence suggests that the fracture initiates degenerative neuroarthropathy in some cases.⁴⁰

It was formerly believed that the fractures were manifested in the presence of a pathological weakness of

the bone tissue.⁴¹ However, histologically the bone appears normal with all stages of healing observable. The only obvious difference from normally healing fractures is that the trauma continues since the painless fractures are often not immobilized.⁴²

Localized hyperemia in the area of the fracture may cause softening and reabsorption of bone, leading to further spontaneous fractures. This softened bone becomes distorted from the stress of abnormal excess weight bearing, eventually causing avascular necrosis. Sclerotic changes in bone ends are also thought to contribute to structural weakening of bone. The combination of these destructive changes may be responsible for successive spontaneous fractures and for the collapse of joint margins and bone ends typically observed in a Charcot joint.⁴³

The attitude toward treatment of neuroarthropathic joints is often defeatist. Many physicians recommend only bracing, arthrodesis or amputation as options for treatment after the joint has reached advanced stages of destruction. Few even mention preventative therapy, although joint breakdown in the case of leprosy has been prevented or minimized by proper protection.⁴⁴

Surgical intervention by arthrodesis is often recommended to permanently stabilize the joint. Modern techniques in arthrodesis have helped to overcome former problems of non-union and rampant infection. Successful arthrodesis requires prolonged post-surgical immobilization and patient cooperation. Surgical fusion of a joint totally restricts any movement thereby saving the limb but radically restricting its normal function.⁴⁵

Avoiding trauma in a neuropathic joint by means of external support is crucial to the longevity of the limb. Bracing, awareness training and reduction of detrimental physical activity can minimize or prevent neuroarthropathic fractures and Charcot deterioration.

In a grossly deteriorated joint the blood supply may be insufficient to allow tissue to survive or heal. Infection leading to a chronic or severe sepsis is possible. Amputation becomes the ultimate necessity when the general health or life of the patient is compromised.

Total joint replacement has not been accepted as an alternative for the treatment of Charcot joints in general practice. Total knee arthroplasty has been attempted in selected experimental cases. Long term results, though promising, are not yet conclusive. Prosthetic devices designed with a posterior condyle stabilizer are inherently stable in flexion. This design

element is an advantage when attempting to correct the severe instability of a Charcot joint caused by fluid-stretched ligaments and bone absorption. Proper ligamentous balancing and bony alignment may be established by ligamentous release, adequate bone resection, bone graft augmentation or custom-augmented prosthesis. The results of total knee arthroplasty in neuropathic joints thus far suggests that proper surgical technique in combination with appropriate prosthetic devices can offer relief.⁴⁶

Any surgery attempted on a Charcot joint must be carefully considered and only performed when the joint is in its coalescent stage. Surgery performed on an actively destructive atrophic joint only enhances the degenerative mechanism and has a significantly greater chance of failure.

At first I thought it was great fun using the crutches. People carried my books in school. The second day I went to the Media Center and the librarian told me to give the crutches back to whomever they belonged. My sixth grade classmates had to try them out and hide them from me.

The novelty of the crutches was short-lived. By the end of the first week it was already wearing off. My hands and armpits were sore from the pressure, and climbing three flights of stairs to my classroom was tiresome. With the exception of my second cousin, Danny, people grew tired of carrying my books. I heard "faker" called out as I hopped through the fourth grade wing to the nurse's office. I decided not to use the crutches for a school play I was in the week after I had acquired them.

Winter had always been a favorite time of year for me. I had always loved playing in the snow, especially sledding. The quarter mile walk to our favorite sledding hill was impossible with crutches. I managed to cajole my sister into pulling me out on the toboggan but was frequently left to make my own way back as darkness fell. Often as not I left my crutches stuck standing in the snow while sliding and half walked or crawled back up the hill. My mother would watch out the back window of our

house and we would hear "Barb - use your crutches!" echo across the field. Snow and especially ice became a nuisance rather than sledding and skating fun.

Later that February, just about the time the novelty of using crutches had begun to wear thin, our usual neighborhood crowd gathered at Marshman's for hot chocolate and snacks after a day of sledding. The leftover candy hearts from Valentine's Day parties at school were passed around. Printed on the assorted pastel confections that I had drawn from the bag were the customary "I Love You"'s, "Be Mine"'s and "I'm Yours". One benign looking heart with pretty scalloped edges said "Dead Legs".

In the spring the whole neighborhood dusted off their bicycles. Since nothing had been mentioned about restricting me from my bike, I developed a crutch balancing act. The crutches rested upright on the handlebars as I peddled and I leaned on them when I stopped. Occasionally we would strap our books on the racks and ride the three hilly miles to school or summer music lessons.

That sixth grade year I had joined the Safety Patrol. The crutches gave me extra wingspread to stop students from crossing the street unsafely. The primary

incentive for all sixth graders to join Safety Patrol was the opportunity to go on the bus trip to Washington, D.C. in April. Fifty eleven year olds attended meetings and guarded their posts all year to earn the privilege of sightseeing, aggravating chaperones and spending five nights in a motel without their parents for the first time.

When I brought my permission slip home I was told that I couldn't go because of my being on crutches. My teachers tried to assure my mother that I would be fine, even suggesting I ride in the accompanying police car. She said that I wouldn't be able to keep up and that I wouldn't miss anything as they had taken us to the capitol two years before. Unable to obtain my parents' permission I remained behind, not participating in the adventures of my classmates. Naturally, they talked of little else upon returning and for several weeks thereafter. I had to accept being an observer to their inside jokes.

Being sidelined during gym class, not participating in the Safety Patrol trip, the obvious presence of the crutches and visible orthodonture that had begun the previous year, made me feel extraordinarily conspicuous. I increased my efforts at achieving public anonymity. I infrequently raised my hand in class to volunteer.

Rather than actively seeking acceptance into the all-important social cliques, I waited for the rarely offered invitation. I began to feel more comfortable around adults, who were generally more compassionate or small children who tended to be less judgmental.

Toward the end of that year I began to spend lunch hours with a favorite science teacher, rather than on the playground. Mrs. Miller once told me how much more mature she found me in comparison with other sixth graders. She commented on my positive attitude and how well I had adapted to and accepted my limitations, something my peers had yet to accomplish.

Eventually, I came to the conclusion that some of my classmates were jealous of what they viewed as the special attention or privileges I was given by the teachers. Being excused from the dreaded gym classes, being allowed to leave classes before the bell rang to avoid the crowds and to cut-in the lunch line so that I didn't have to stand so long were all punishable actions for the unprivileged majority.

The response of teachers to my increasingly more obvious physical differences were inconsistent. Some pandered unnecessarily while others went to great lengths to treat me as "normally" as anyone else, thereby emphasizing the differences. Another response was to

force me to earn the right to be viewed as normal by imposing somewhat extreme expectations on my schoolwork.

As I approached adolescence the issue of weight control was added to my list of physical challenges and peer pressures. I had always been of approximately average weight for my height and age but the added bulk of the remaining lipoma made me appear somewhat larger in circumference than I actually was. The body consciousness and comparison of adolescent peers, combined with my declining physical capabilities, hereditary and dietary factors, seemed to exaggerate the problem. Although never grossly obese, I averaged between ten to twenty pounds over my ideal weight.

I had become accomplished in the kitchen when I was quite young and always kept the cookie jar filled. Food became my security blanket. I tended to eat more when I was home than I did elsewhere. My mother's comments about my becoming "bigger than the house" did nothing to encourage better dietary habits. During stressful times at home I usually went for the cookie jar before heading out the door.

Excessive concern over obesity on my mother's part bordered on the anorexic. She was the youngest and thinnest of five sisters and their mother had died obese.

My mother ate minuscule portions for breakfast and dinner, skipped lunch, and shunned snacks and sweets. She claimed to lose her appetite in restaurants whenever overweight people were in view. My father was a meat-and-potatoes man who has always been known for his appetite. He also tended to be over his ideal weight. Cindy would be praised for her naturally petite figure while being criticized for her slouched posture, which was later diagnosed as scoliosis.

Clothes that fit had always been difficult for me to find, never mind being fashionable. The loose fitting or stretchy knit clothes that were selected for me in order to conceal the lipoma also made it easier to gain weight.

Weight control is an important issue for all physically impaired people. Excess weight creates added stress on already overtaxed joints, hastening deterioration and further limiting mobility. Obesity can lead to increased incidence of pressure sores, make the fit of orthopedic appliances difficult and decrease the life span of such aids. Obesity is commonly the cause or result of low self-esteem. Weight management requires a high level of motivation and encouragement, especially for those who are not able to participate in calorie-burning physical activity.

Participation in household activities became more difficult after the acquisition of my crutches. I was excused from standing chores such as drying dishes or vacuuming. Never being one to remain idle for long, I became possessively attached to my father's Gravely tractor. Lawn mowing became my domestic chore. The tractor was a walk-behind type with an attachable seat. Since it had no foot clutch or brake, the hand controls were ideal for me. I had been the sole operator of the tractor for several years when one afternoon I returned home to find my sister receiving her first lesson in lawn mowing. I was crushed to think that one of the few chores that had been exclusively my domain was now going to be shared. The thought that my services were not indispensable was devastating. I threw a tantrum sufficient enough to assure my authority over the tractor, which my sister never dared challenge again.

Herbert Lourie, M.D.

October 11, 1973

Patient: Barbara Tefft

Office Visit: 10/9/73

Barbara generally is doing nicely. She says she has only a rare bladder incontinence when she cannot get to the bathroom. She says she is no longer bothered with constipation. She has had some trouble recently with her left knee and is being followed by an orthopedist in Norwich.

On exam things are pretty much the same as when I saw her a year ago. She has impaired sensation in the sacral dermatomes on the left, except perianal where sensation seems quite good. She has a bilateral pes cavus and hammer toes. The feet are small. The large subcutaneous lipomatosis-like mass in the lumbar area is unchanged. Her condition seems stable. The main thing is that she remain under orthopedic follow up, and the father understands this. I will see her in one year for a check up.

Herbert Lourie, M.D.

/jm

cc: Dr. Richard Hasbach

Herbert Lourie, M.D.

June 27, 1974

Patient: Barbara Tefft
6/25/74

Office Visit:

Neurologically she has remained relatively stable. She still has considerable sacral dermatomal loss on the left, including also the 5th lumbar. The feet have remained small with atrophy of intrinsic, and pes cavus bilaterally. Ankle jerks are absent, and knee jerks are brisk. She has very good hamstrings bilaterally, but the gastrocnemius is small bilaterally. The left buttocks is small compared to the right. She is still growing somewhat in height so I will continue to follow her, at least for the next couple of years. Her bladder and bowel control have been good since last seen.

Herbert Lourie, M.D.

/jm

cc: Dr. Richard Hasbach

As usual, Dr. Lourie had inquired about and made note of the embarrassing issue of bladder and bowel function. As usual I attempted to avoid the topic by denying any problems. It was true that the constipation was no longer the particular problem that it had been in the past. During the previous year my body had developed a new pattern of relieving obstipation with attacks of colitis, occurring about every three weeks. The pain during these attacks was severe, causing cold sweat and nausea accompanied by complete intestinal evacuation over a period of several hours. These attacks usually occurred at night. They were occasionally related to emotional stress or diet, although a consistent pattern was not discernable.

Urinary tract infections had also become more frequent. I had managed in school by going to the bathroom between every class, causing my many tardinesses. When a bladder infection occurred I would have to leave class in the middle as well as before and after.

Wherever I went my first priority was to locate the facilities. I became very adept at deciphering

floorplans and plumbing layouts in large buildings. I could tell anyone where the most likely place to find a restroom would be!

Originally, Dr. Kochersperger had suggested using the crutches for six weeks. At the six week exam the time was extended for another six weeks as there had been no improvement or decrease in the swelling. Eventually, the consecutive six week sentences turned into a year. The valgus deformity increased and was attributed to unequal growth in the distal epiphysis of the femur.

Surgical correction was recommended. Staples were to be inserted straddling the epiphyseal line on the medial aspect of my left knee. Theoretically, the staples would restrict growth medially, allowing the lateral aspect to catch up. This one-sided growth would reduce the angle of valgus exhibited by the distal portion of my left leg.

The surgery was scheduled for August so that the procedure would not keep me out of school. I was admitted and sent for x-rays of my knee and wrists to determine the age and amount of growth potential of my thirteen-year-old bones.

Report of Operation:

Discharge Summary

Tefft, Barbara

Dr. Kochersperger

8/17/74

Preoperative Diagnosis:

Progressive valgus deformity - left knee

Postoperative Diagnosis: The same (myelomeningocele and aseptic necrosis of left lateral femoral condyle).

Operation: Medial Distal femoral epiphyseal stapling.

Anesthesia: Endo, Pent, Fluo, N₂O₂

This 13 year old female has a history of spina bifida and myelomeningocele (closed at birth), with a history of heel cord lengthening and tendon transfer of both feet. The patient had a large area of aseptic necrosis involving the left lateral femoral condyle, followed for several years with progressive filling in. She has been maintained on protected weight bearing. She has apparently developed unequal growth of the epiphysis, and is demonstrating progressive valgus deformity of the left knee. She was admitted for medial epiphyseal stapling. The chronologic age is 13, and the bone age is 13.

On 8/14/74, under general anesthesia, the medial aspect of the distal femur was exposed (left knee). The epiphyseal line was identified by probing with the straight Keith needle, then 3 heavy staples were inserted medially, anteromedially and posteromedially. The staples were inserted through periosteum behind the vastus medialis. X-rays in the O. R. confirmed satisfactory placement. The wound was irrigated with Betadine and then closed in layers using chromic catgut and silk for skin. A cylinder cast was applied holding the knee in maximum extension which was approximately 25° of flexion as the maximum range of motion of the knee is 25-90°. The postoperative course was uncomplicated. The patient was ambulated with protected weight bearing and crutches. The postoperative films were satisfactory.

The patient will be seen in the cast room on Friday, 8/23/74, for suture removal.

AK/bq

D: 8/17/74

T: 8/20/74

As many thirteen year old girls do at this stage, I became obsessed with horses. During my convalescence from knee surgery I read every book I could find pertaining to horses: care of, training, novels, fiction and non. After the cast was removed from my leg I coerced my sister into joining a 4-H Horse Club with me, although her interest was characteristically minimal.

Living as we did in the middle of farm country, nearly all of our neighbors had horses. After much loud insistence on my part we were finally allowed to take riding lessons. Every week we headed for the unheated indoor arena to ride while my father waited, watched and froze.

I had a passion for English hunt seat and jumping. Unfortunately, riding English requires extensive leg cues and such actions as posting-on-the-diagonal at a trot. My knee was not improved by this work and I eventually was forced to continue my riding with Western tack which was less strenuous on the knees.

Participating in a 4-H Horse Club without a horse of my own was very frustrating. Every night I looked through the classifieds under Horses/Cattle, clipping ads

to leave at my father's place at the kitchen table. After much discussion and my persistent begging we finally got our first horse.

Princess was a liver chestnut 3/4 Arab, green broke. My father replaced fence posts and strung wire around the pasture. I acquired the services of my neighborhood playmates to help clean the old, dusty hay out of the barn. Princess arrived before the fence was finished. She spent her first few days tethered to a post in the middle of the pasture. When she was finally released to run, her antics foreshadowed the problems to come. She proved not to be a good choice as a first horse, growing more obstinate by the week. Eventually, we traded her for a registered Quarter Horse named Miss Speedy Mac. Missy was a gentle former brood mare who provided many hours of pleasurable trail rides.

While the trail rides themselves were pleasant, obtaining permission to go wasn't. My mother typically forbid any contact with the family whom she despised next door. Although Mrs. Rogers was an experienced horsewoman, my mother would allow us to go riding through the woods alone before she would let us go in a group with the Rogers. I usually overcame this minor problem by riding in the mornings. Since my mother didn't get

out of bed until afternoon she had no recourse in the matter.

The more time I spent with the Rogers' the more frequently I was in trouble with my mother. One afternoon, upon returning from a swim at another neighbor's pool, where Brenda had also been invited, my mother tried to drag me into the house from the porch by my hair. She ordered me to scrub myself clean after "swimming in the cesspool with the crud." This incident took place in front of a number of people who had also been swimming with us. When I forcefully refused to comply, Cindy was sent to the workshop to get my father. He neither upheld my mother's irrational demand nor came to my defense. As usual he wanted his peace and quiet.

Another instance of parental stand-off resulted from an attempted afternoon neighborhood trail ride. I had saddled and mounted Missey by our barn while everyone else met at Rogers'. I called across the yard asking my father to open the gate for me when everyone was ready to go. He started to cross the road to oblige when my mother came out onto the porch and said, "Don't you dare." He didn't. I was not allowed out of the paddock. Meanwhile, everyone who was going on the trail ride had been sitting on their horses by the side of the road awkwardly waiting to see what the outcome of this latest

dispute would be. The sympathetic, helpless look on my father's face and his unwillingness to stand up for me made me so upset I nearly fainted. I slid off Missey's back and sat on the ground and cried.

Virtually all topics of discussion beyond that of antiques or the weather created the potential for greatly disturbing the "peace and quiet" that my father so highly valued. Issues that inevitably arise in most households that shelter adolescents carried an unmentioned, albeit well understood, taboo. The "do as I say, not as I do" philosophy was very much in use at our house. My mother chain smoked the carton of cigarettes that my father purchased for her every Saturday. Anyone else caught smoking would be threatened with a simple "I'll kill ya." The same phrase also applied to any ideas of using alcohol or drugs. Sex was a word not to be uttered other than in reference to gender. The only motherly advice my sister and I ever received was that if we ever got pregnant, don't bother to come home.

My interest in history and antiques developed partly in order to have at least one topic of conversation that could break the silence without causing an explosion. I felt that we all took second place to my mother's prize possessions. She could become emotionally attached to inanimate objects but not to people. Caressing her

possessions while cleaning and dusting was as demonstrative as she got. The only time I ever saw her kiss my father I was four years old. He handed her a pistol on the way out the door to a weekend-long conservation department convention. She pecked him on the cheek before he left.

Since nothing of any importance to adolescent development could safely be discussed at home I was fortunate enough to be able to talk to friends. My mother's "Don't tell the neighbors" demand went unheeded when it came to personal issues. Maternal threats only induced resistance and experimentation on my part. Fortunately, the one and only time I was ever caught smoking was at Rogers'. Skip and Siri returned home from a night of raccoon hunting and found a still-lit cigarette that Brenda had accidentally dumped behind the stove while getting rid of the evidence. Both of Brenda's parents were heavy smokers themselves but did not permit or encourage smoking by their daughter. They also looked upon me as a daughter and as upset as they were with us at the time they did not raise their voices. They had a long talk with us about independent decisions versus peer pressure and parental example. They wouldn't tell us not to smoke but made it clear that we could not do it in their house and that they hoped we would choose

not to smoke at all. This discussion was much more effective than any threat. Neither Brenda or I have touched a cigarette since and Ma Rogers, with some difficulty, quit smoking the day after our talk.

My actions admittedly did not contribute to any harmony at home. My headstrong conduct was constantly compared to my complacent and obedient sister. I was my own person at a very early age which was a difficult concept for my parents to manage. Their response was to try to pull the reins in tighter or to ignore my persistence.

Regardless of how my actions affected my relationship with my family, I had given myself some experiences that would have a permanent and positive effect on my attitudes and outlook on life. I refused to accept an overprotective, restrictive environment and the struggle I endured only strengthened my tenacity.

Adolescence for me was stormy, as it is for most teenagers. With the exception of our neighborhood gang I did not belong to any of the more popular cliques in school. I preferred the company of a few close friends to the soap opera-like antics of the party crowd. I was not overly interested in make-up or clothes. Most often I would be seen in worn jeans and flannel shirts in school, much to my mother's dismay.

My first independently earned income was from babysitting. I began this career in a rather incongruous manner, as I was playing in the sandpile at my neighbor's farm one morning and was asked to return after lunch to babysit. The transition from playmate to authority figure was somewhat abrupt but generally successful. Being trusted to supervise the activities in which I had so recently participated placed me firmly in the twilight zone of ambiguity that exists between childhood and adulthood. This first child care assignment at Marshman's was to lead to a number of jobs with other local families.

Babysitting was a natural occupation for me to become involved in. The maturity about which Mrs. Miller had commented was apparently obvious to other adults, with the exception of my own parents. I received commendable references and was regularly employed by parents who knew me. Occasionally, some doubt as to my physical capabilities was encountered. Once my creative adaptabilities were demonstrated parental fears evaporated. I became an often requested favorite among my little friends, as much for the opportunity to play with my crutches as for my personal companionship.

I was never spared the natural curiosity and unending stream of questions commonly asked by toddlers.

Many times their inquiries were the politely unasked questions of their parents. I always answered these questions with the best of my limited knowledge but could not begin to respond to that ever-present conundrum of curious toddlers, "Why?"

The summer before I entered high school I was allowed to stop using the crutches. I spent that summer enjoying the freedom until August. While packing the trailer for the annual family vacation I decided to throw in my crutches - just in case. My knee fatigued easily and as we would be doing a considerable amount of walking in Mystic, Connecticut I wanted to be prepared. From this point through the end of my ninth grade year I used the crutches intermittently. This appeared to give the impression to some fellow students that the crutches were used to gain attention. By the end of that school year they had become a permanent fixture.

Herbert Lourie, M. D.

July 2, 1975

Patient: Barbara Tefft

Office Visit 7/1/75

Barbara has good control of bowel and bladder. She has not yet started to menstruate but just turned 14 last week. Neurologic exam is unchanged except that the knee jerks do not seem as brisk as before. Otherwise the exam is exactly the same as recorded on June 25, 1974. I will see her again in one year, sooner if any symptoms develop. I reviewed this with the father in detail.

Herbert Lourie, M. D.

/pt

cc: Richard Hosbach, M. D.

Office Visit 10/26/76

Barbara is 15, is menstruating regularly. She says she has good control of bladder and bowel. She has swelling and fluid in the left knee, suggesting a Charcot's joint, for there is impaired sensation around that knee. The left quad is quite weak; the iliopsoas seems strong. She walks with considerable deformity in her lower extremities and a scoliosis. She uses crutches but is able to get about without them. Sensation in the saddle area seems good but there is spotty loss of sensation in the lower extremities. She cannot dorsiflex the left foot. There is some movement of the right foot. Knee jerks and ankle jerks are absent.

She and the father think there has been no change in her status in the last several years. It would appear that she is not changing significantly in her neurologic function although it is obvious that she is not as agile as she was as a young child.

I plan to see her in one year. The mass on the back is unchanged in size or appearance.

Herbert Lourie, M. D.

cc: Richard Hosbach, M. D.

During grade school I was rarely ill or absent from classes because of common childhood maladies. Absences were usually due to appointments with physicians in distant cities. Having two school nurses and several teachers in our neighborhood eliminated any notions of skipping school unnoticed.

My most extended absence from high school was for a somewhat embarrassing illness for a tenth grader, chicken pox. Those of us who had managed to avoid this infection as children would succumb as adolescents. I noticed the first spot on my left hip and initially thought it was an itchy insect bite. My sister said it looked like chicken pox. Since Cindy was speaking from personal experience, having survived an attack at age seven, my parents took me to be examined. Doc Boname confirmed my sister's diagnosis. Annabelle Smith, the high school nurse and a next door neighbor, was notified of my impending leave of absence from school. She informed my parents of school policy that does not permit chicken pox sufferers to return until the last scab is gone. Fortunately for me the lesions did not itch after the first one faded away. The only catastrophe in this ordeal was that most of the scabs were on my face and seemed to stay there forever. For three weeks in May I was not allowed in school. The weather was perfect and I spent most of that time riding

Missey. As the bus stopped to drop Cindy off after school, the other kids yelled and clucked at me out the windows. I waved and headed Missey toward Smiths' to ask Annabelle how much longer my vacation would last.

A short time after I was allowed to return to school I suffered from an acute combination of symptoms. My vision was affected initially. I couldn't see what was being written on the overhead projector in biology class. During a break I tried to go down the stairs to the nurse's office but even with the use of my crutches couldn't keep my balance. Some friends noticed my difficulty and with their help I made it to the couch in Annabelle's office. The fluorescent ceiling lights were extremely irritating to me but I couldn't keep getting up to turn them off when everyone coming in turned them on again. As I was lying on the couch my entire left side went numb, face, eyelid, tongue, arm and leg. I felt nauseous and was alternately hot and then cold. I developed a headache that was throbbing but not severe. Annabelle drove me home where I climbed into my mother's canopy bed and closed the curtains to block out the light. Eventually I vomited and afterward began to feel better. These symptoms were new to me and I had rarely been known to vomit except after surgery. By coincidence the following weekend, a long article on migraine

headaches appeared in the Sunday newspaper. The symptoms that were described matched mine almost identically.

By the time I had entered high school the progressive sensory deficit from my knees down was nearly total. This loss of sensation to pain, temperature and position contributed to the development of numerous infections from injuries of which I was unaware. I was hospitalized under threat of amputation for recurrent cellulitis in my left fifth toe during my junior year.

In January of 1979 I endured my first case of frost bite. My feet always got cold the minute I stepped outside; even electric hunting socks couldn't keep them warm. That January I attended an antique auction with my family. The auction was held indoors on a cold, wet, snowy day. I sat near the door to allow myself access for my usual frequent retreats to the restroom. This was the only advantage to the location I had chosen, as every time the door opened the snow and cold attacked me. I covered myself with the quilt that my mother had purchased and drank hot chocolate to keep warm. As I stood up to leave, my feet felt as though they were no longer attached to my body. I warmed them by the heater in the car on the way home. Later, when I removed my socks I discovered the large fluid-filled blisters on my toes. The blisters filled until they ruptured, thus

beginning a six week regimen of cutaneous debridements, soakings, dressings and antibiotics. Although I managed to avoid hospitalization I did miss a considerable amount of school.

CHENANGO MEMORIAL HOSPITAL
Norwich, New York

Discharge Summary

Tefft, Barbara 157-747 Adm. 4/18/78 Dr. Kochersperger

4/24/78

1. Cellulitis, left fifth toe.

16-year-old white female with myelomeningocele, no sensation distal to the left calf, who has had chronic recurrent infection in the left fifth toe. She is now admitted with increase draining, redness, evidence of exposed flexure tendon between the ball of the foot and toe and probably has osteomyelitis of the phalanges of the toe.

HOSPITAL COURSE:

Patient showed significant improvement, elevation, antibiotics and wet dressing. Drainage ceased. Appearance of the toe improved. She was discharged to home on oral Keflex to be followed in the office.

Cultures grew out staph coagulase positive. Strep fecalis and gram negative bacilli. There were diphtheroids. Overall sensitivity to Erythromycin, staph sensitive to Keflin.

AK/mab

d: 6/12/78

t: 6/12/78

A cold war was being waged at home and I tried to avoid being there as much as possible. I was usually considered to be the cause of any domestic difficulties although my absence from the house was not tolerated any more than my presence. It was not unusual for me to find the doors locked when I returned to the house. My father would sometimes answer my pounding and let me in but most often he refused because he "would never hear the end of it" from my mother. Usually I gained entry by throwing stones at my sister's bedroom window until I got her to come down and open the door. More than a few panes of bubble glass were broken in this manner.

Friends weren't allowed in our house to play. The former milkhouse-turned-playhouse became my summer home. As soon as the temperature moderated enough in the spring to sleep outside I took up residency in the cozy 10 x 12 foot room. From April through November I enjoyed relative freedom and invited my neighborhood friends over whenever I wanted. I slept out by the swimming pool and watched the stars, took midnight swims and had overnight guests. This arrangement was tolerated by my mother as

long as Brenda Rogers was not among my guests, and provided that no one went into the house.

I could see the lights go out when my mother went to bed, around four in the morning. Her hacking smoker's cough served as a warning that she would soon be out of bed. That cough usually wasn't heard until well after the noon whistle echoed down the valley from the village fire station. My stomach would tense when I heard that cough, in anticipation of the mood of the day.

My father retired from the conservation department when I was in the ninth grade. He would be up and out of the house before my mother arose. My sister was usually the last one up, even on Christmas.

Vacation trips and weekend outings to auctions became infrequent occurrences. Being the only ones up on weekend mornings, my father and I would take long Sunday drives together on the back roads of the Chenango River Valley as an alternative to family trips. Even trips to the various physicians I had been seeing became less frequent.

Dr. Lourie had indicated that he would like to continue following me on a yearly basis and it had always been up to my parents to call his office for an appointment. Family communication had become non-existent. Communication with the outside world also

suffered. My parents had developed an aversion to the telephone, placing few calls themselves. My mother refused to answer when the phone rang, always assuming that it was "the crud" calling from next door. The telephone became virtually my domain but I was not about to use it to make more doctor's appointments for myself. I never brought the subject of my annual visits to Dr. Lourie to the attention of my parents and they never took the initiative on their own. The last time my father drove me to Syracuse to see Dr. Lourie I was fifteen years old.

I earned my driver's license immediately after a summer of driver education classes. As usual, I had taken the class with my sister in tow and we took the road test together. I passed on my first attempt while Cindy had to try again. Obtaining my license was a major key to gaining independence, even more so for me than for my peers. I at once began going to my appointments without a parent accompanying me. I made the daily trips uptown for groceries, and could visit friends without attempting to gain maternal approval. The driver's license was also important because it allowed me to go places to which most people are able to walk.

My sister was graduated from high school on a sweltering day in June of 1978. I chose not to attend the commencement exercises. Instead I asked Brenda to join me for a swim in our pool while my parents were away. A rare treat for her and an enjoyably spiteful act for me. No family celebration acknowledged the event of Cindy's graduation.

Later that June, the occasion of my seventeenth birthday went unacknowledged by my family. I had been making my own birthday cakes for several years but that year there was nothing in the house with which to make one. Christine Cosen, a young friend from across the road, came to invite me over for dinner and had helped her mother make me a cake. When my father asked where I was going I told him Cosen's had invited me to a birthday dinner. Dad asked me whose birthday it was. My mother had never forgiven me for staying at a friend's house for a picnic supper on my tenth birthday instead of coming home. After I had returned from my dinner at Cosen's I was invited for cake at Rogers' and Marshman's. I may have been forgotten by my biological family but my extended surrogate family certainly boosted my spirits.

That fall my sister left home to attend Cazenovia College, a private, two-year women's college near Syracuse. I enjoyed my senior year of high school

without the shadow of my sister in tow. I was not known to be the world's most academically oriented student but managed to get my name on the honor roll that year.

When it came time to apply to colleges in my senior year I was called to the guidance office and told that by "virtue of being physically handicapped" I was eligible for the services of the Office of Vocational Rehabilitation. I was sent to one of that office's contracted psychologists to be tested. I was asked to put puzzles together, spell words like "cat" and "necessary", tackle mathematical calculations from 2+2 to trigonometry, and repeat in reverse increasingly long lists of numbers. After completing this test it was determined by the experts that I did not have what it takes to complete four years of college. If I were to attend a vocational or community college full tuition would be provided through their office. One counselor took me aside to privately tell me that I didn't need a college degree. He said I was pretty enough and must have plenty of boyfriends. His advice was to get married and have someone else take care of me because statistical evidence did not offer a favorable outlook for employment of the handicapped. They refused to assist in a four year program because in their opinion I would surely flunk out within the first year. This meant that when I

was accepted at Keuka College, a private, four year women's college, that my parents would have to pay the full tuition themselves. Setting personal differences within our family aside, my parents did believe in higher education and had prepared to send both of their children to college.

As far as deciding on a major, I knew that I excelled in Art and English and had an avid interest in biology. I was accepted as a medical laboratory technology major at Keuka initially, although I already had an idea about pursuing medical illustration. That idea was put on hold during my early college career as most parents are not particularly excited to hear the words "art major" when sending their offspring to costly private colleges. This was especially true for me as my mother had left an art program, at what is now the Rochester Institute of Technology, one semester before completing the course and my sister was doing poorly in her art program at Cazenovia.

Commencement exercises for our graduating high school class were nearly cancelled as punishment for an out of control "senior prank night" that caused considerable damage to school property. Most embarrassing of all the damage was the misspelled spray

paint graffiti painted above the main entrance to the high school building. Spelled out in four foot high letters: "Class of '79 is devine!" Restitution was eventually made but the ceremony had to be held in the gym because of unseasonable weather rather than at the traditional outdoor location on the front lawn of the school.

The morning of my commencement I slept as late as the rest of the house usually did. We arrived at school early that afternoon so my parents could get seats in the gym as there would be standing room only during the ceremony. Our school colors were red and black and I had made red velvet covers for the pads on my crutches to match our gowns. I was pleasantly surprised to be awarded an academic varsity "O" for making the honor roll. My mother was surprised to hear the complimentary comments made by my teachers as I walked across the platform with my crutches to receive my diploma. A photo was taken for the local newspaper showing off my red velvet crutch covers. After the ceremony we returned home. I changed into my work clothes and went to help a neighbor hull strawberries.

Eager to begin my own life away from home, I began to pack my belongings for college the day after graduation. In September I would be attending Keuka

College in the rural Finger Lakes region of New York State. A special significance surrounded my attendance at this particular school in that two of my maternal aunts had graduated from Keuka. My mother had been a frequent visitor as a small child while her sister Grace attended Keuka in the late 1920's.

By the time I had graduated from high school I was acutely aware that my family operated in an overtly dysfunctional manner. I had begun to question and search for clues that might offer insight into various familial behavior patterns. I wanted to find some small way to identify with the people who were my family. Consciously, I knew that their experiences and impressions would not be my own. I was also looking forward to developing my own person and being able to gain a new perspective apart from home.

The summer before leaving for Keuka I became acquainted with my assigned roommate through the mail. Chrisy Ahlberg became a great friend immediately. We roomed together for three years until we were both fortunate enough to have our own rooms senior year. We shared the same major as freshmen, chose to be lab partners and loved to canoe on Keuka Lake for recreation.

Although the campus was small, I used my bike for transportation to class, even in winter. This caused

numerous stares at first, as I pedaled across campus balancing my crutches on the handlebars. I could always be easily located since my bike was parked by the doors of the various buildings in which I attended classes.

The campus was mainly comprised of older buildings with many stairs and few elevators. Whenever possible the professors and scheduling office were very accommodating, changing the location of lecture classes to more easily accessible rooms. Only once did I experience resistance from an instructor when a room change was requested. The professor of a required English course maintained that I should not expect to be treated any differently from the rest of his students. If they had to climb the stairs to the fourth floor of Hegeman Hall, so did I. This trek became impossible for me to do on a daily basis. Fortunately, my roommate, who was also in the class, came to the rescue by turning in my papers for me and reporting what went on in class each day. I did manage to attend at least one of these classes each week. However, since the final grade depended heavily upon class participation and attendance, my overall average definitely suffered.

Getting to class became even more difficult when my bicycle was stolen from inside the dorm by two local "townies". Following a dorm guest list investigation,

two suspects were identified. One of the suspects offered to buy a replacement for the bike that was stolen, although both boys continued to deny that they were the thieves. Losing my bike also put a temporary stop to my almost daily excursions down the lake toward the bluff, which was the only form of aerobic exercise I was able to engage in.

While home on Christmas break my freshman year I accompanied my father to Cazenovia to pick up my sister. I wandered off to explore the campus while they loaded the car. On the way home I inquired about the amount of stuff that was in the car just for a two week vacation. Eventually, I was informed that Cindy had been asked not to return as her academic record was unsatisfactory. She was also experiencing social adjustment difficulties and, at the suggestion of her former psychology-major-roommate, was seeing a counselor.

My sister had developed abnormally obsessive attachments to one of her former art professors and to television star Michael Landon. Her room at home became a shrine to these people. This manifestation was ultimately regarded as merely the beginning of years of increasingly more aberrant behavior. She continued to see counselors at the local mental health clinic and to

occasionally attend the clinic's day treatment center. My father would chauffeur Cindy to these appointments at her request but would not accompany her inside.

During this already disturbing first Christmas break from college, my mother showed me some articles she had saved from recent newspapers. The articles were on the hormone diethyl- stilbestrol, known as DES. DES was thought to prevent miscarriages and was widely used from 1941 through 1971. Studies were now being released that indicated an elevated risk of rare cervical cancers in the daughters of mothers who had taken DES during pregnancy. My mother recalled taking a prescription drug during pregnancy and had experienced one miscarriage. After checking into the matter it was discovered that she had taken DES during both my sister's and my gestations.

A screening clinic had been organized at a hospital thirty-five miles from home. Our mother decided that Cindy and I should attend for testing. Although DES was also known to affect the women who had taken the hormone, my mother declined an offer to be examined.

Neither my sister nor I had had a pelvic examination prior to visiting the screening clinic. After completing the preliminary questionnaire we were taken to separate examining rooms. I was examined by a visiting physician who was assisted by a registered nurse practitioner.

During the exam the doctor asked how long I had been sexually active and what type of birth control I was using. At eighteen, I had yet to experience the pleasures of sexual intercourse and informed the physician of this fact.

My sexual experience at the time of my first pelvic exam was limited for a variety of reasons. I had spent most of my life self-consciously attempting to hide physical abnormalities, although I was fairly confident of my attractiveness. I had often received compliments on my appearance, in spite of my attempts to blend into the background. I was secure enough in my appearance to be indifferent to the nickname of "ugly" bestowed upon me by one of the boys in our neighborhood. This security had definite boundaries, however. The logistics of concealing my physical self by necessity required the building of an emotional wall that prevented anyone from getting too close. Although my desire for both physical and emotional intimacy had always been strong, I would not allow that carefully constructed wall to be disturbed.

When the doctor had completed the exam I was told to get dressed as he and his assistant left the room. Before I had finished dressing, the doctor returned to the exam room alone in order to ask me a few questions.

These questions were issued in an accusatory and patronizing manner. Based on his examination, he informed me that an "exaggeration of the labia minora indicated an extremely active sex life." Since I still refused to admit to engaging in sexual intercourse he inquired about masturbation and proceeded to define sexual terminology. After I interrupted his inquisition and lecture with a rather graphic verbal demonstration of my knowledge of such topics, he left the room slamming the door behind him. As I finished getting dressed I heard him very unprofessionally say to the assistant, just outside the door, that "No eighteen-year old who looks like that is a virgin." The PRN entered my exam room, apologizing for the physician, who I fortunately never saw again.

I was informed of the results of my exam along with my mother and sister in a conference room. A colposcopy was recommended as some ectropy had been observed in the cervix. The Pap smear was listed as class I. Their observations were consistent with findings in other DES daughters and regular follow up exams were recommended.

This was not exactly news to celebrate. The idea of frequent pelvic exams after the experience I had just had was not appealing. I left my first gynecological exam feeling that I had yet another anomaly to hide.⁴⁷

What the doctor obviously did not know, and I did not learn at the time, was that hypertrophy of the labia minora is not uncommon, especially in women born with myelomeningocele. Hypertrophy has in the past been attributed to early sexual activity or masturbation but little substance is currently placed in such etiology. Specific causes other than filarial infection with lymph blockage have not been determined. Cultural aesthetics in western society deem this condition as less desirable, in the same way that underdeveloped breasts may be considered faulty. Some cultures actually desire hypertrophy, using weights and stretching manipulation on young girls to achieve desirable enlargement.⁴⁸

In cases of myelomeningocele the condition can cause more than just cultural embarrassment. Women who rely on clean, intermittent catheterization for urinary management may have difficulty in using apparatus and with maintaining acceptable levels of perineal hygiene. Irritation of the hypertrophic labia can lead to infection and ulceration, aside from being physically uncomfortable. Deviation from societal ideals can also cause loss of confidence in personal sexuality and diminish self-esteem.⁴⁹

Surgical alteration of the genitalia in both males and females is a universally practiced procedure. Male circumcision is commonly done for religious, aesthetic and hygienic purposes. Female circumcision, in varying degrees, is routinely practiced in Islamic/Arabic countries. In Egypt today up to 75 percent of women are circumcised. This procedure can vary in degree from the removal of the labia minora to complete removal of the entire external genitalia, including labia majora and clitoris.⁵⁰

In the United States it can be difficult to find physicians who are well versed in the gynecological care of adolescents or the disabled. Even in this age of information and enlightenment many people, health care professionals included, dissociate the terms "disabled" and "sexuality". The belief that disabled people do not or should not enjoy sexual relationships is still pervasive in our society. Elective surgery to enhance sexual self-image or ability is often down-played in cases of physical disability. Plastic surgeons, rather than reluctant urologists or gynecologists, are generally called upon to perform labioplasty for women who require the procedure.⁵¹

Discussion of sexuality is often especially difficult for disabled patients. Our society is fixated

on an ideal sexual image which is reinforced by the media. Our ideal of the young, able-bodied, heterosexual married couple, perfectly matched in race and religion, is statistically in the minority of our population. The presence of a physical disability does not preclude a desire for physical intimacy. As in any able-bodied population, the sexual preferences of the physically handicapped are diverse. Heterosexual, homosexual, bisexual and asexual preferences are experienced by those with handicaps as well as those without. An entire range of sexual consequences is also possible, including sexually transmitted disease and pregnancy. A single deviation from this unrealistic ideal sexual image may create a stigma, thereby inhibiting social interaction and even access to appropriate medical care. Disabled persons who choose to participate in sexual relationships risk facing multiple social stigmas.

Access to compassionate services that provide family planning, disease control and prevention, and counseling, as well as dealing with the mechanical aspects of intercourse, is vital to the health of all sexually active people. For some disabled persons, obtaining these services may be an embarrassing task. Initiating discussion on such topics as interference of a catheter during intercourse, a penile implant as a solution for

impotence or obtaining the services of an attendant to assist in preparations for intercourse, is awkward under the best of circumstances.

The nervous impairment caused by the presence of a myelomeningocele can affect sexual functioning in a number of ways. In males, the impairment can create complex problems in potency, fertility and ego associated with desirability and performance. Many affected males are potent and fertile while others may experience a variety of problems that were unrecognized until they reached adolescence. Mechanical difficulties, including abnormal erections unrelated to sexual arousal, inability to become erect or to maintain an erection, have a detrimental effect on sexual self-image. Most males will experience orgasm and ejaculation with unaffected potency, even when the penis remains flail. Recurrent urinary tract infections caused by retrograde ejaculations or elimination apparatus can cause reduced fertility or sterility.

A female born with myelomeningocele can most often expect to function as a fully capable sexual partner. Genital sensation may be diminished or altered, although lack of sensation does not exclude orgasm. Fertility is unaffected by a myelomeningocele and successful pregnancy is possible. Not all types of contraception are suitable

for women with disabilities. Some require high levels of manual dexterity while others are chemically incompatible with other necessary medications. Internal devices may not fit because of impaired muscle tone or physical anomaly. The added weight and stress of pregnancy in combination with hormone activity creates the possibility of increased urinary tract infections, incontinence, back pain, herniation of intravertebral disks or further loss of neuromuscular function. The presence of associated kyphosis, pelvic anomalies or cloacal extrophy may complicate or prevent pregnancy.⁵²

Both genders may experience difficulty in maintaining bladder and/or bowel continence during sexual intercourse. It is possible to prevent such accidents by advance preparation. Restriction of fluids and/or emptying bladder and bowel prior to intercourse reduces the possibility of accidents. Ileostomy or catheter leakage can be prevented by careful positioning of tubing. Discussion of such possibilities with a sex partner is helpful in creating intimacy and easing embarrassment should an accident occur. Protective bed coverings can also be used as a precaution.

Sexual intercourse increases the risk of urinary tract infections, especially when a catheter is being used or in the case of urine retention. This risk can be

minimized through careful washing by both partners before and after intercourse. Emptying the bladder completely before and after intercourse is also helpful in preventing infection.⁵³

Physical intimacy in any of a myriad of forms is desired by nearly every living being. The desire to form lasting relationships and to have a family is as strong for the disabled population as it is for any other population, and personal motivation may even be higher. The risks associated with pregnancy and delivery, in combination with the financial and physical burdens of raising a family, necessarily require an almost super-human effort on the part of a disabled parent. The rewards may be just as great. When proper medical care and support is available there is no reason that those born with myelomeningocele can not become parents. The risk of genetic recurrence for an affected parent is approximately five percent. Genetic counseling and prenatal testing can identify potential problems so that informed decisions can be made.

Physician-patient relationships can be either beneficial or detrimental to the health and attitude toward care fostered in the patient. Those who have had uncomfortable experiences with the health care system often tend to avoid treatment for minor ailments.

Waiting until an emergency situation develops increases the financial burden, makes treatment more difficult and the prognosis less satisfactory.⁵⁴ Self-education on behalf of the patient can ease the burden of treatment placed on the physician by allowing the patient to play an active role in decision making. For physically disabled people, responsibility in educating the physician as to the details of their disease, unfortunately, often rests with the patient. The exponentially increasing amount of technical information that must be taught in medical schools today leaves less and less opportunity for in-depth study of exceptional anomalies. Physicians may not encounter such patients until they are in practice. Disabled patients should realize that their health care providers are human, come from varied medical backgrounds and may be uncomfortable confronting a problem for the first time. Initial visits can be made less intimidating if the patient has accepted some of the responsibility in preparing for the visit. Having copies of past medical records in hand, presenting specific medical questions and candidly answering the physician's queries will demonstrate an acceptance and appreciation of the services offered. Any level of patient awareness can also help assure proper and concerned medical treatment.

I returned to Keuka after that stressful Christmas vacation determined not to follow in familial footsteps and to remain in school. News from home came in the form of the weekly comics from the local newspaper and an occasional note. The shared pay phone in the dorm hallway was a busy place on weekends for everyone but me. During my four years at college I never once received a call from my parents. Only when I did the dialing to check up on things at home would we speak, and even then only the "safe" topics were discussed.

My sister was not one of those safe topics. Weekends and vacations that I spent at home were anything but restful. Cindy was acting out a delayed and exaggerated adolescent rebelliousness. My formerly reticent sister had now become recalcitrant. Heavy metal music, lying, vicious accusations, outbursts and tantrums replaced her formerly quiet, obedient demeanor. She insisted on moving out. My parents both disapproved of the idea, but nonetheless, Dad loaded her belongings into the car and took her to the roach infested firetrap she had rented.

My mother blamed the transformation in my sister's character on the counselors Cindy had been seeing. They had told her to be more assertive. She was asserting herself all the way to aggression. My father just did whatever it took to regain peace and quiet, including helping Cindy move to unsatisfactory living quarters.

My sophomore year at Keuka became a turning point for me, both physically and academically. My left knee had been effused and unstable for seven years. At this time I used that leg very little, bearing most of my weight on the unimpaired right leg while using crutches and riding my bike. This method worked well for several years until one spring afternoon when I began to rise from a sitting position on my bed. I felt a sudden painless "clunk" in my right knee and decided it would be wise to stay off my leg for a while. My roommate offered to bring meals back from the cafeteria and collect my assignments. By the next morning the joint was so swollen I could not bend or stand on it. I remained in my room for a week. Ice packs and elevation were used in an attempt to reduce the swelling. In order to keep up with my course work and lab assignments I had to return to classes.

The newly swollen joint also refused to extend completely. This caused considerable muscle fatigue in the quadriceps from standing with the knee flexed for extended periods of time. When the swelling had not begun to subside after a month I decided to go home and visit my orthopedist.

I was hospitalized overnight for tests. X-rays were taken and fluid was aspirated from the effused joint. An immobilizing soft cast was applied before elevating the leg in bed. Within half an hour after aspiration the swelling had returned. The x-rays showed no abnormalities and gave no hint as to the cause of the effusion. A second aspiration yielded the same result as the first. Aside from a sensation of tightness in the skin and fatigue the swollen joint was generally painless.

CHENANGO MEMORIAL HOSPITAL
Norwich, New York

Discharge Summary

Tefft, Barbara 172-601 Adm. 3/23/81 Dr. Kochersperger

SHORT STAY 3/24/81

1. Internal derangement of right knee with recurrent swelling

This 19 year old white female with myelodysplasia and a partial lesion of the lower extremities who has had a long term problem with the left knee and presented early this year with a history of recurrent swelling in the right knee and inability to extend the knee. There was no history of trauma or twisting injury. The pain is not a major component of her symptoms and there is no history of giving out but a definite inability to extend the last 15 degrees. This was not previously present. The patient had noted significant fatigue from having to stand contracting the quadriceps much of the time. She has basically been on a crutch gait because of problems of the other leg including degenerative changes in the lateral femoral condyle consistent with a neurogenic joint. She is admitted now for workup and therapy.

PAST HISTORY:

See old records.

REVIEW OF SYSTEMS:

See old records. No allergies.

PHYSICAL EXAMINATION:

19 year old white female, who is oriented and cooperative and alert. She ambulates with crutches. Blood pressure is 130/82. Pulse is 78 and respirations are 20.

Head:	Normocephalic.
Eyes:	Normal examination.
Ears, nose and throat:	Normal.
Neck:	Full supple motion. No adenopathy or thyromegaly.
Chest:	Symmetrical expansion and clear lung fields. No rales or rhonchi.
Heart:	Regular rhythm. No murmurs or thrills.
Breasts:	Not examined.
Abdomen:	No masses or tenderness.

Tefft, Barbara

Dr. Kochersperger

Orthopedic: Patient ambulates using the right leg primarily and keeps the left semiflexed. Left knee shows well healed surgical scar from previous epiphyseal stapling because of angular deformity and there is definite crepitation, clunking and gross instability in the left knee. Right knee shows a good range of motion except the final 15 degrees of extension is not demonstrated. Ligamentous stability is normal. A mild to moderate effusion is present. Patellofemoral motion is not painful. McMurray testing is negative.

HOSPITAL COURSE:

The knee was aspirated revealing yellow clear fluid of poor protein content as accessed clinically and some gross blood was obtained at the close of the aspiration. White cell count was 315 and 10 poly and 90 lymph.

SMA 12 showed normal values except for a slightly low cholesterol of 133. Latex fixation was negative. Sed. rate was 23 at 60 minutes and urinalysis was normal. CBC showed 7,100 white cell and 77 segs and 13.0 hemoglobin and 35.7 hematocrit. X-rays of the knees showed the right knee was normal to x-ray and left knee showed previous stapled fixation and fragmentation of the lateral femoral condyle, Lateral tibial plateau and lateral subluxation of the tibia. The patient has altered sensation as noted on previous admission including almost complete anesthesia of the distal leg and foot.

The patient is discharged on a program of straight leg raising 100 times b.i.d. and stretching exercises and do extension to be done for half hour each evening and Motrin 400 mg. t.i.d. and continue on a crutch gait and see me in the office in May when the semester is completed.

Exploratory arthrotomy may be indicated. I don't think arthrogram would be helpful under the circumstances.

AK/mab

d: 3/24/81

t: 3/26/81

That spring I had decided to change my major. After taking my first college drawing course with Dexter Benedict, I realized how much I missed continuing the art classes I had taken in high school. Few schools offer a degree in medical illustration so I took advantage of Keuka's student-initiated-major program. By the end of my sophomore year I had completed a number of science courses. I obtained information on required courses from R.I.T.'s undergraduate medical illustration program and designed my own program at Keuka. This meant taking major courses in art and science simultaneously, both very time consuming majors.

The latest development with my knee did not help in any way. I returned to Keuka, after my uninformative brief hospital visit at home, with more time eaten out of my schedule for leg exercises. I also began experiencing more numerous attacks of colitis and urinary frequency. Formerly, I had been getting up only once during the night. Now four or five times a night I walked down the dormitory hall, headed for the bathroom. Late nights at the lab and studio on top of the physical fatigue that I was experiencing took its toll on my grades. I was

looking forward to a summer of rest and relaxation at the end of the spring trimester.

Naturally, that summer at home turned out to be anything but relaxing. My sister had moved from the roach infested hotel to a slightly better studio apartment. She had been at this new location for three months when my father and I stopped by to pick her up one afternoon. Cindy was still in bed when we arrived. The room was piled with unpacked boxes and suitcases. Electrical cords from lamps and a clock radio were stretched taut to reach outlets from a chair in the middle of the room. The refrigerator held only rotting lettuce, bologna and sugar doughnuts. Cindy's door was unlocked and we had to climb over the tangled maze of cords to awaken her.

I told Brenda about the condition of my sister's living quarters later that day. She suggested that we offer to help Cindy unpack and straighten things up so it would be easier for her to keep house. Cindy reluctantly agreed.

Brenda and I spent nine hours cleaning the apartment while Cindy was sent out to do her laundry. She returned hours later having washed her clothes in fabric softener and rinsed them in detergent.

During our afternoon of cleaning we learned how Cindy had been spending her time when she wasn't sleeping. She continued to dabble in the art that she had formerly been studying at Cazenovia, but any level of maturity or refinement that she had previously attained was conspicuously lacking. Her work was now crude and childlike in both technique and subject matter. Brenda and I learned that Cindy had been going daily to the county mental health department's day treatment center. There she attended arts and crafts classes overseen by an art therapist with the indelible name of Marvel.

I planted myself for the summer in the former playhouse-turned-summer house, as had been my habit since junior high school days. Soon after settling my sister and myself in our respective abodes I experienced yet another orthopedic inconvenience. My left leg became swollen from just above the knee down to my ankle. I was by now used to the chronic swelling in my knees but this appeared to have a different etiology. Medical opinion of the origin of this latest problem was possibly a minor infection or a painless trauma of which I was unaware. Recommended treatment consisted of ice packs, antibiotics and elevation. I spent a considerable amount of time in my cozy playhouse patching jeans for neighbors and

playing board games with friends. Several years later, x-rays taken prior to surgery on that knee revealed a well-healed fracture of the fibula.

Shortly before I was ready to return to Keuka my sister called from her apartment. Cindy said she was scared and wanted to come home. When my father went to get her he found her huddled in a corner. He brought Cindy and a few of her things back home. She began talking of being possessed, that Satan and the Lord were fighting over her. This sudden religiosity was difficult to understand as neither my sister or I had had any religious background as children. The closest my parents had ever come to introducing us to religion was when we inexplicably received children's bibles for Christmas one year.

When it became obvious that she wasn't going back to her apartment, Dad moved the rest of Cindy's things back home. We eventually traced the source of Cindy's newly acquired religious notions to the art therapist at the day treatment center. Cindy now adamantly refused to watch television or listen to the radio and had smashed her extensive record collection. She would no longer have anything to do with art - all because Marvel had told her these things had evil, sexual connotations. Cindy told us of going to Marvel's house with other

clients of the day treatment center in the evenings, after the center had closed. Marvel was the self-proclaimed, fundamental-extremist leader of this group of mentally disabled people.

I returned to Keuka in September, not long after Cindy had moved back home. School was a welcome relief after that bizarre and stressful summer "vacation" we had just endured. The old routine of school with only the Sunday comics and a brief note from home was easier to tolerate than experiencing my sister's unexplained transformation on a daily basis. School was also more enjoyable with my new major. I developed amicable social relationships with the art faculty while spending a majority of my time working in Allen Hall arts building.

Physically, I was aware of the gradual decline in my ability to ride my bike and to walk. The fact that I could not fully extend my right knee contributed to the eversion of that foot, thereby putting more strain on my ankle. During one of my weekend visits home it was suggested by Dr. Kochersperger that a brace might help relieve the stress on my right ankle. I was fitted with a short caliper brace with a T-strap. The high top work boots that I had been wearing for support the past few

years had to have a stirrup inserted in the heel to accommodate the brace.

During these weekends and school breaks at home, I was disturbed to see the progress in my sister's mental deterioration. She was now unable to function without assistance and prompting. Cindy could not feed herself or bathe. Attempts to keep her busy with simple chores such as drying dishes only resulted in dropped and broken glass. She would march in place, roll her eyes toward the ceiling and burst out crying. It was always "Satan and the Lord are fighting over me."

My parents had spoken with counselors at the day treatment center about the "art therapist's" influence on Cindy and Marvel was eventually restricted from having any contact with my sister. Cindy became desperate to talk to Marvel. She would run out of the house in her pajamas at inappropriate hours begging to use our neighbors' phones. Because of my informing our neighbors of the turn of events at home (against my mother's wishes), they knew to keep Cindy inside and call my father. Dad would wind up chasing her up and down the road to get her in our house. Once home she would babble in "tongues", "cast spells" and issue threats saying God had told her to kill us all. At night Cindy would lie in bed rigid on her back, sweating so profusely that the

mattress would be soaked. Several times a night she would run downstairs and jump in my mother's canopy bed. Being in the room directly next to Cindy's, I could hear her get up. I had to run interference when I was home so that my mother could get some sleep. Even though my father occupied the room at the top of the stairs, he always slept through the commotion.

From my more comfortably distant location at college I would occasionally inquire about the developments on the home front. Cindy had been placed on medication by the psychiatrist associated with the day treatment center. My parents had been asked to accompany her to an office visit with the psychiatrist. The diagnosis for Cindy's symptoms was schizophrenia. My mother concluded that this was a catch-all label used to cover any type of mental disorder. In typical Freudian style, the blame for Cindy's problems fell on my mother. My father failed to acknowledge what was discussed at that meeting, although he had been physically present. Cindy's condition did stabilize somewhat after the medication was adjusted but her behavior was decidedly juvenile, undisciplined and compulsively ritualistic.

My college courses were keeping me busy but I still found time to volunteer myself as a studio assistant to the art department. While loading a kiln one morning I was approached by a professor from another department. I was asked to speak to a freshman student in an attempt to encourage her to stay in school. Although I had never met Sandy, I had seen her in a crafts class. She also walked with difficulty and sometimes used crutches. The professor told me she had spina bifida. This information intrigued me. At twenty-one, I had never met anyone else with spina bifida. That afternoon I found Sandy alone in the crafts room after class. After some small-talk I mentioned that I heard that she was leaving school and wondered why. She said she had been having physical problems and that I wouldn't understand. I told her that maybe I would since I also was born with spina bifida. We did talk for a while and I learned how embarrassed and insecure she was about her physical anomaly. Sandy had requested a private room upon being accepted at Keuka and isolated herself from her classmates. She had taken all the required tests and was receiving full financial support from the Office of Vocational Rehabilitation.

Her family was extremely protective, calling every morning and night. They hadn't been very encouraging when she decided to attempt going away to college. Sandy had decided to drop out after one trimester at Keuka. I asked what her plans were when she left and learned that she had applied for an apartment in a complex for the handicapped just a few blocks from her parents. She was looking forward to living in a place where she wasn't different from those around her and would have parents close by.

I couldn't convince her to remain at Keuka, but the conversation made me aware of how lucky I was to have Chrisy as a roommate and of how the difference in our personalities and home environment had affected our outlook on life.

Later that year I was introduced to the second physically disabled person I had ever met. Christopher Wright volunteered to teach photography to me as a favor to Gary Jurysta and Dexter Benedict, the art faculty at Keuka. Chris was a local art photographer who suffered from a rare genetic disease of the connective tissue, known as mucopolysaccharidosis IV. We all spent many hours working in Allen Hall, becoming great friends by year's end.

The summer following my junior year at Keuka provided one particularly enjoyable week. My father and I packed up the camper and the two of us spent the Fourth of July week in Cooperstown, New York at the American History Seminars. Our first night, at the outdoor orientation lecture, a little girl came across the tent and sat on my lap. I later learned that she was the daughter of Wendell and Betsy Garrett, lecturers and authors on American history and publishers of The Magazine Antiques. They apologized for their daughter, Maria's uncharacteristic boldness. I assured them I didn't mind in the least and asked Maria why she had chosen my lap out of the hundred others that were there. She said, "Because you're pretty." I met her sister Abigale and brother Nathaniel during our conversation and soon had three youngsters climbing on my lap. Betsy made a special trip back to the Otesaga Hotel each day so the children could join us for lunch at the seminar tent. Wendell Garrett was the third physically disabled person I had met that year. He had been diagnosed with polio as a child and now walked with two elegant, silver-handled canes.

My mother had worshiped Wendell's eloquently written editorials in Antiques for years. When she learned about our instant friendship, she accepted a ride to

Cooperstown with some antiquing acquaintances who were commuting to the seminars, just to meet Wendell.

At some point virtually every family develops at least one dysfunctional or problematic element in the interfamily relationship. Although my family certainly experienced more dysphoria than some, the influences of happier times in my early childhood were predominant in later years. My appreciation of art, nature, history and quality was acquired directly through parental influence. The highly social and perfectionistic dimension of my character seems to be an inverse response to the examples set at home.

Growing up in the same household did not guarantee that my sister and I had similar needs or personal experiences, in fact we developed vastly different perspectives. The values and influences that I adopted from our home environment were rejected by Cindy entirely. While I spent my days out of doors, looked forward to attending antique auctions and museums and fantasized about being a colonial settler, Cindy preferred to remain inside and developed a loathing for our anachronistic environment and materialistic sentimentality.

Respect for one another's interests and emotional needs degenerated as each of us competed for attention. Communication and affection disappeared as a result. We all began to function separately rather than as a family. We expected our needs to be fulfilled automatically without attempting to establish communication, and were most often disappointed and resentful.

When I could no longer depend on my parents for emotional support, I began to parent myself without realizing it at the time. I wanted to belong and to be accepted so I sought outside sources for fulfillment. It was easier for me to ask for personal validation from outside my family than from within the bitter circle. I was the lucky one. Neither of my parents or my sister dared to risk forming intimate friendships. Instead they withdrew from the world. What little contact my family had with the lives and activities of our neighborhood they gained through my interaction. My mother especially had retreated from any external contact. She lived as if in a fish bowl, rarely leaving the house and waiting for everything to come to her.

The last time my mother ever went into a store she was followed by two children. While in line at the checkout, one of the children tugged on her jacket and said, "Hey, lady. Are you a witch?" My mother's sallow

smoker's complexion, emaciated build and shoulder-length gray and black streaked hair, in combination with the fact that the store was decorated for the upcoming Halloween season, had made the image quite plausible. My mother's haughty rejection of the idea must have done nothing to dispel the children's impression.

The following December I observed another episode of dysfunctional behavior in my family. I had unpacked, among my clothes and art supplies, a few bottles of liquor that had been left over from a reception at school. I decided to put them in the keepingroom cupboard where my father usually kept a bottle of whiskey for his Sunday afternoon cocktail. Two weeks later, just before Christmas, I went to the cupboard to get some of the rum I had brought to make my traditional Christmas rum cake. I immediately noticed that some of the bottles were empty and the rest contained only a small portion of their original contents.

After overcoming my initial bewilderment, I recalled noticing that my father had been bringing home a two liter bottle of wine about every other night in recent weeks. Apparently my parents' custom of a nightly jigger of wine had been expanded. When I asked my father what had happened to the liquor supply he denied any knowledge of the matter, but my mother's hasty departure from the

room clued me in. When confronted, her typical and not unexpected blaming retort was, "You'd drive anybody to drink." I removed the remnants of the supply from the cupboard and told my father not to continue this enabling behavior. Since my mother virtually never left the house, he had been automatically replenishing her supplies of cigarettes and alcohol - just to keep the peace and quiet that was all important to him.

My mother's smoking habit had obviously taken its toll long ago. Her breathing was very labored, her circulation poor and her weight noticeably decreasing. I was sure she exhibited symptoms of advanced emphysema and I was not about to willingly allow alcohol dependency to compound the already unhealthy atmosphere at our house.

My senior year at Keuka was spent in the luxury of a private double room. Chrisy had obtained a room to herself just down the hall, while I kept the room we had shared for the past couple of years. I shoved the two twin beds together in the corner, creating a wonderfully decadent king size bed, unheard of in most dormitory rooms. This luxury almost caused me to miss my anticipated May graduation date for a rather unfortunate reason.

The corner where the bed was located also contained the radiator. I woke up one morning with my right foot resting on the hot pipes. I incurred full thickness burns on my foot and toe. This was especially detrimental as I used my right leg exclusively to walk with my crutches. I attempted to doctor the wounds myself in the same manner as had been previously prescribed for my bouts with frostbite. Although painless, these new wounds were more serious than I realized. They became infected, sending me to Dr. Marty Zinnaman at the college infirmary. I was ordered to stay off my foot, not to wear a shoe and was given a prescription for antibiotics. I was not able to attend

classes for weeks. Dexter and Gary brought my art projects to my dorm room. Chrisy brought the cat that I had been dissecting in anatomy class. Chris Wright, who was a friend of Dr. Zinnaman, invited me to stay with his family for a week while I was off my feet.

After two weeks of pampering it was evident that the wound was not going to heal without help. I went home to visit Dr. Kochersperger who agreed that a skin graft was necessary. I spent two weeks in the hospital to clean up the wound and clear out the infection before surgery was attempted.

positive. With cleaning up of the wound and a bit of granulation being demonstrated, she underwent mesh grafting from the right thigh to both wounds. She was covered on intravenous Kefsol for 48 hours. She has shown no febrile spikes and has no odor of the dressing. She is discharged on a nonweight bearing program for the right lower extremity and will be seen on Wednesday in the cast room.

AK/jh

d: 3/7/83

t: 3/10/83

Marty, Chris, Dexter and Gary called the hospital regularly to check up on me. Chrisy faithfully mailed me my assignments with letters of encouragement and all the campus gossip. I received fanciful crayon renderings from the Garrett children depicting my hospital ordeal, as they imagined it to be, from injury to surgery. Wendell and Betsy generously invited me to stay with them again during my next college field trip to New York City. Dr. Kochersperger was impressed with the sketches of vertebrae that I was doing from my bed, and even joined me for popcorn and to watch the final episode of M*A*S*H on TV.

After three weeks in the hospital I was allowed to go home. I couldn't walk on the newly grafted foot until it had healed so we rented a wheelchair. Since my room at home was on the second floor, I had to side up the stairs on my rear and down the hall to get in bed. The chair was useless upstairs because the doorways were too narrow to allow the chair through. I suffered a few splinters while sliding down the hall to the bathroom but the waxed plank floors got a good polish!

Chrisy came to visit me at home during spring break, which broke the monotony for me. David Marshman, from the farm down the road, came to get us the night Chrisy arrived. I was forcibly treated to a surprise wheelchair

ride down the road, in the dark, to the farm. This was the first occasion in a month that I had been outside. At the end of my half-mile ride I had to slide into the house on my rear to visit. I was beginning to notice that a majority of buildings are not very accessible. This accessibility factor was the foremost reason for my prolonged hospital stay and absence from college.

Being forced to remain at home until the skin grafts had completely healed caused my absence from an entire half-trimester of classes. If I was to graduate with my class in May I would have to successfully complete the work I had started on top of the final trimester's new work. It was assumed by some of my professors that I would be remaining at Keuka for summer session to complete the course work for my degree. This was definitely not an option as far as I was concerned. I had already begun to apply to graduate schools to pursue my interest in medical illustration.

I was specifically directing my efforts toward an acceptance at the Rochester Institute of Technology for three primary reasons. RIT's program emphasis was on the illustration and during my undergraduate study I had seen some pretty poor scientific art. The second reason was that my mother had attended RIT in the 1940's and had lived in Rochester after dropping out of the art and

design program. Attending RIT might offer another small piece of the puzzle that was my mother. My strongest reason for wanting to attend graduate school was that I needed to continue learning about my own medical condition. I had gained an understanding of normal human biological process and mechanics but wanted to know why these processes go awry and the long-term effects that are sustained by the body.

Even at the time I was applying to graduate schools I knew what the topic of my Master's Thesis would be. I was going to write and illustrate my own medical history. In order for any of my plans to fall into place, the undergraduate work at Keuka had to be completed before commencement in May.

With the help and generosity of faculty and friends, I managed to complete all of the work satisfactorily enough to be placed on the Dean's List for both trimesters.

Commencement exercises were held in the gym, just as my high school graduation had been, because of inclement weather. I once again put the red velvet covers on the pads of my crutches and sat with friends who had ridden out with my parents for the ceremony. After I had walked across the improvised stage to receive my degree, I was lifted with hugs, kisses and congratulations from the risers by Dexter and Gary.

I spent half of that summer after graduating from Keuka waiting for the mail to be delivered each day. My friend and neighbor, Barbara Fitzgerald, and I had both applied to different graduate schools and were anxiously expecting replies. In the middle of July I received the long awaited envelope. After several attempts to reach Barb by phone, obtaining only a busy signal, I gave up. As soon as I put the receiver down the phone rang. It was Barb. In unison we excitedly blurted out the words "I was accepted!"

In August I hoped to be attending the Rochester Institute of Technology's Master of Fine Arts program in Medical Illustration. A few minor details had to be taken care of before I could officially declare myself a graduate student. My parents refused to finance any further education. They believed that an undergraduate degree should be more than enough to land a high paying position and help take care of them in retirement, as they had done for their parents.

I contacted the Office of Vocational Rehabilitation and made them aware of my successful completion of a four year undergraduate degree. Since I had proved their

original assessment of my ability to complete a bachelor's degree incorrect, they allowed my request for graduate funding to be submitted. Submission of a request did not guarantee funding.

The goal of OVR is to provide services that will produce a financially viable product; in other words, a financially independent person who does not rely on government subsidies. They did not typically assist in funding education beyond a bachelor's degree. Since OVR had not assisted in my undergraduate financing they would consider a graduate education grant - *if* I could convince them of the need for such a degree in my chosen field. The Office of Vocational Rehabilitation, along with many other bureaucratic agencies, exist solely for the purpose of increasing the financial potential of their clients, with no interest in personal growth and development. I managed to provide enough evidence to substantiate the need for a graduate degree in the highly technical field of Medical Illustration, eventually receiving the services of OVR. A student loan was arranged to cover tuition with OVR providing book and art supply funding. My parents had applied for Social Security Disability for me before I had begun college. I now received the funds directly to cover rent and living expenses. I was now also eligible for SSI which carries with it the benefit

of Food Stamps. An allotment of \$15 would be mailed to me each month.

These financial resources were deceptively appealing and secure. In reality they often fell short of covering necessary expenses because of governmental restrictions on how they could be spent. I decided to apply for a campus job to cover the gaps. I learned that I was ineligible due to the sources of funding that I was currently receiving and the fact that my parents' assets were sufficient enough to contribute to my graduate education. Off-campus employment is the common alternative for many college students. In my case, a job at the local McDonald's was not an option. My physical capabilities were definitely limited and the lack of ownership of personal transportation restricted travel off-campus. It became obvious that this was not to be the beginning of my financial independence from my parents as I had hoped.

Housing was arranged on a preliminary scouting trip to RIT. I rented a one-bedroom apartment in a student oriented complex two miles from campus. A shuttle bus was to run at regular intervals providing transportation to campus. Kathy D'Abbracci, a friend from Keuka, was attending RIT as a visiting student and would room with

me part-time. I still would require a full-time roommate to meet expenses.

The remainder of the summer was spent refinishing and cleaning furniture from my parents' overflow collection in our barn. Barb Fitzgerald assisted in refinishing and my father built me a drafting table. Additional assistance was provided when a grey and black tiger kitten came to live with me. Nermal arrived at the playhouse via motorcycle from Marshman's barn. David said he brought the tiny, motherless animal for me to take care of until he was old enough to return to the barn. This pretext was very transparent, as David had known my preference in feline markings. He knew the kitten would have a permanent home. Nermal supervised the preparations for my first apartment, occasionally lending a tail to assist in the staining process.

Two weeks before I was to leave for Rochester I was informed that the apartment I had rented would not be available for occupancy in time for my arrival. I would have to arrange alternate lodging for my first week at RIT. I was rescued from this dilemma by David's Aunt Kay. Dr. Kay Francis Marshman and her housemate, Dr. Ann Marie Franzen, lived just outside Rochester. This pair of highly motivated career women generously offered to let me stay with them for the week. Ann took time from

her position as psychologist for the Rochester City School District and Kay from her duties as Dean of Special Education at Nazareth College to chauffeur me to classes. David's mother, Linda, offered to drive me to Kay and Ann's when I was ready to go.

This overwhelming support from the neighborhood was greatly appreciated, especially considering the comparative lack of attention or support I was receiving at home. My sister and her erratic behavior tended to be the main preoccupation at our house for the past few years.

After orientation, the campus Health Center was on the top of my list of departments to become familiar with at RIT, although not exclusively for medical reasons. Dr. Marty Zinnaman, who had seen me through the long trauma of my radiator-burned foot at Keuka, was now practicing preventive medicine with several other physicians at RIT. Marty was pleasantly surprised to see me and I was very glad to have a physician who was familiar with my history close by.

After a dose of Ann Marie's persuasive encouragement, directed at the complex management on my behalf, the apartment was finally ready. My father rented a trailer and brought the furnishings that had been restored during the summer. Kay sacrificed part of

her precious weekend to help me set up housekeeping in my first apartment. My friend Kathy was also an invaluable help, driving me to classes and helping with chores such as laundry. Being on the third floor, getting laundry to the lower level was a huge task for me. Grocery shopping was another exhausting and time consuming task in which Kathy efficiently assisted.

For all of Kathy's help, I still needed a full-time roommate to cover the financial aspects. After placing posters around campus, another roommate was acquired. What I did not learn until after she had moved in was that this former Catholic had just undergone a religious transformation. She now declared herself saved as a born-again Christian and her mission was to save the rest of the world, including me. I began to notice striking similarities to my sister's behavior in this new roommate. Being curious, I decided to discuss this religious transformation with my roommate one evening. I was initially concerned that I would be watching my roommate's mental stability deteriorate in the same way that Cindy's had and I was not looking forward to living with the same situation both at home as well as at school.

During our conversation and the weeks that followed, I gradually began to realize that my roommate was not mentally ill, merely brainwashed. Her piety was hypocritical. The materialistic orientation of her religious sect was overtly incongruous with the biblical quotations which she quoted to everyone with whom she

came in contact. Although we shared the same apartment, and even the same bedroom, we did not speak for the second half of the year. I was too concerned with other things to expend any more energy trying to understand this roommate or to bother to find another. The strained atmosphere at the apartment frequently discouraged me from returning any earlier than necessary. If I missed the last shuttle bus to the apartment complex I would just stay in the studio to work, catching a few cat naps during the night.

The apartment lease contained a "no pets" clause. Because I had foreseen being too busy for a pet, I had left the kitten to which I had become foster mother during the previous summer at my parents' house. This was not as simple an arrangement as it appears. My mother had adamantly refused to allow Nermal in the house from the moment he arrived. But Nermal had been aptly named by Christine Cosen, one of my adopted neighborhood "daughters". He was the namesake of a character in the Jim Davis comic strip Garfield, after Garfield's nephew, Nermal - The World's Cutest Kitten. Nermal managed to charm his way into the house unlike any of the number of pets that had preceded him.

By the time I came home for my first break from graduate school, Nermal had taken charge of the house. He slept on my mother's bed and any other precious antique of his choice. (My father still had to sit on a lawn chair to watch T.V. in the newly renovated "living" room.) When Nermal wanted to go out or come in, activities which usually followed one another in rapid succession, his requests were catered to immediately. A pathetic meow in front of the refrigerator was answered promptly with a bowl of milk and a pat on the head. Endearing names, the likes of which had never been heard in our house before, were now in common use. My mother called the cat "sweetheart", "honey" and "dear", among other affectionately syrupy names. As with all emotional matters, Dad's practiced indifference prevailed in his relationship with Nerm. Privately, he also enjoyed the company of this charismatic tabby in sharing his nightly bowl of ice cream and inviting the cat to accompany him out to the workshop. Although I envied all the attention that was lavished upon this cat, at least I knew I had helped provide an outlet for the repressed affection that existed in my family.

When I returned to my Rochester apartment I was not pleased to find that a poodle-mix puppy had been added to the roster of occupants. A note attached to the

cardboard box in which the dog was confined announced "This is Pookey. Isn't he cute?" I broke the rule of silence that had prevailed in our apartment to inform my roommate that dogs were in violation of the terms of our lease and that it had to go. Keeping a puppy under the circumstances also bordered on cruelty as no one was actually at the apartment for much more than to sleep. She sweetly recited some biblical quotation and said that no one would even know he was there. That the dog would remain unnoticed was an extremely unlikely assumption. The minute the door closed, a high-pitched whine followed by incessant barking filled the building. I decided that it would be better to let the management in on the problem from the beginning rather than risk future reproach. I went to the rental office and requested an eviction notice. After explaining the situation they gladly obliged, sending a forty-eight hour "comply or quit" notice to our apartment. Two weeks of howling puppy finally came to an end but the tension in our apartment noticeably increased.

Shortly before spring break, the residents of our apartment complex were notified that the shuttle bus service provided by RIT would be discontinued due to inadequate patronage. This announcement caused no major

hardship for most residents, considering that a two mile walk or bike ride is generally not unpleasant. For me, the news was devastating. Kathy, who had been invaluable the past two academic quarters, would not be returning as she had completed her degree credits. Without the shuttle or Kathy I would have no transportation to campus. I had given up my bike soon after arriving at RIT, being unable to pedal up the steep ramps on campus.

I spent spring break at home trying to convince various people of my dire need for transportation, including my parents and Vocational Rehabilitation. To my parents, a car was a luxury children did not deserve. The OVR could offer funding for transportation services, such as a taxi, but limits on such funds would have transported me to classes only three of the six days a week that I needed to attend. With only one week left of break, I told my father that I needed help, either with buying a car or with bringing my things home from Rochester and subletting my apartment. I had to submit written evidence that the shuttle had been discontinued in order to prove to my parents that I needed transportation. When I finally convinced them of my need and made them aware of the limitations of my options, my father agreed to take me car hunting. We managed to find a suitable used vehicle on the last day of break and with

great relief I was able to continue my graduate school education.

The roommate situation as it had existed during the past year was completely unacceptable. To avoid risking such an occurrence for the following year I began my search for a roommate early. I placed a listing in the off-campus housing directory for "Female Roommates Wanted". The first caller who responded to my ad was in no way female. He introduced himself as Brian, saying he had inadvertently noticed my ad and liked my sense of humor. He wondered if I had ever considered sharing an apartment with a male roommate, adding (as an element of security) the fact that he was currently engaged. The thought actually hadn't occurred to me, but I agreed to meet him the following afternoon.

Immediately after I had arranged to meet this caller I began to imagine what his reaction would be when I walked in with my crutches. I tended to see just about everyone I met as a potential new friend and had become aware that how I handled an initial introduction usually set the atmosphere for any type of new relationship.

The next day I left my ceramics class splattered with clay and drove to the campus apartment address I had been given. The person who answered the door could

easily have stepped out of the pages of *Gentleman's Quarterly*. He was impeccably groomed, tall and lean with unmistakably Scandinavian blonde hair and warm blue eyes. I said I was looking for Brian and was informed that I had found him.

We spent the next couple of hours discussing past roommates, current roommates, graduate school and solved the problems of the world in general. If Brian had any reservations about my physical difficulties he did not make them known. He did ask me a number of questions that I had formerly considered unanswerable. I was now able to respond using my newly acquired knowledge without feeling embarrassed or inferior. Since Brian and I hit it off so well right from the start, we decided that the following weekend we would begin looking for an apartment to share. Before I left that evening, Brian admitted that he had not yet mentioned sharing an apartment with a member of the opposite gender to his fiancée and was anticipating a bit of resistance to the idea. He asked if I would mind his emphasizing that I was "handicapped" and maybe even "kind of unattractive". I said I didn't mind in the least and if it would further his cause of reassurance he could even add a wart or two.

As far as informing other interested parties was concerned, I had to tell my parents about my new

apartmentmate. I decided to wait until after we had signed the lease, knowing that a legal document involving money might temper some objections from the home front.

Brian and I met on campus for movies and spent time together looking at apartments for the coming year. By the time we signed the lease we had become good friends. I called home the night the lease was signed to give my parents the news. As usual, Dad answered the phone and received the announcement first. I told him I had an apartment for the fall and had found a roommate. He sounded relieved that the financial burden we had experienced in the absence of a roommate the previous fall would not be repeated. "Good," he said simply. I said, "My roommate's name is Brian." "Oh. Maybe you better talk to your Maw about that," came the reply. He handed the phone to my mother and I repeated the news. Her instant and emphatic reply of, "Oh, No! You can't do that!" almost made me burst out laughing. I used similar tactics to Brian's negative press that he was providing his fiancée, informing my mother that he was engaged and omitted any details regarding physical appearance. I told my parents we had already paid a security deposit and signed a lease on a two bedroom apartment off campus.

After the news had been absorbed and partially accepted, my mother advised me not to tell my Aunt Grace

(who later told me not to tell my uncle or certain neighbors or...). I found this advice very amusing. For years I had been fully aware that my immediate family was at least two generations removed from my sister's and mine. My parents had grown up during the "Great Depression" and were raised by parents with a strong Victorian influence. During my mother's childhood, mentioning Darwin's Theory of Evolution at the dinner table was a blasphemous enough offense to be banished to the garret without supper for a week. Coming from me, the news of this living arrangement should not have been a tremendous shock to my parents. If anyone in our family was expected to deviate from tradition, I would be the one.

At the end of the semester my father helped move my things into the new apartment. Brian would be staying for part of the summer and Dad helped move some of his things in too. The "born-again" roommate with whom I had suffered through the past school year disappeared, owing forty dollars on the last phone bill. She had apparently decided that the most worthy candidate to receive her particular brand of "Christian Charity" was herself.

Cindy had accompanied my father on this trip to Rochester. She did not speak unless spoken to and her shyness and immaturity gave the impression that she was

the younger sibling. Brian later asked me how old she was and was shocked to learn that she was my senior by a year-and-a-half. He said he was embarrassed because he had been speaking to her as if she were twelve rather than twenty-three. I assured him that his assumption was not uncommon and had occurred many times from the time we were children. When he asked why she behaved as she did, I told him she had been diagnosed as schizophrenic. All of a sudden I realized that I had no real idea of what the term meant. I was as ignorant and intolerant of Cindy's disability as she was of mine.

Brian's questions about my sister inspired me to begin a summer's worth of information gathering. I had begun to collect my own medical records and other research on spina bifida in anticipation of writing and illustrating my master's thesis. Now I began to simultaneously research my sister's disease. The small amount of material available at the local libraries at home was outdated at best. I read the basics on mental illness, Freud, Jung, etc. Although I could see where some of the theories originated, I could not believe that bad parenting or toilet training were the absolute cause of my sister's symptoms. If the parents were to blame why didn't I suffer the same symptoms as my sister? We were close in age and had been raised in the same house, yet each of us were completely different.

In August, Brian and I officially set up housekeeping and returned to classes. The new class of first year Med. Ill. grad students consisted of two people, one of whom left after receiving an affirmative reply to his inquiry, "Do you have to do much work in here?" The remaining student, Carol, was a Cornell graduate of Bolivian heritage who quickly became one of my closest allies and friends. I could count on Brian and Carol for any type of assistance I might need.

By now I was carrying a demanding course load, majoring in Medical Illustration with a double minor of computer graphics and ceramics. Most of my major time was spent researching information and rendering illustrations for my thesis exhibition. The frequent trips to the library and book store for art supplies were becoming more tiring. I stopped going to the store for groceries because the nearly quarter-mile walk just to get milk from Wegmans' most distant corner was far too exhausting. Brian took over the vacuuming, shopping and did the laundry in the basement laundry room. In the winter I would often find that he had brushed all the snow from my car before we left for classes. Carol

challenged herself to carry both of our combined collections of books and art equipment from the parking lot every day, in one trip - without a wheelbarrow.

By November of that year I knew that something had to be done about my exhausting mobility problem. I was becoming uncharacteristically dependent on others for the necessary tasks of everyday life. I was also beginning to believe the accusations of laziness that had been directed at me in childhood by certain relatives and some former teachers. I found myself allowing people to exercise all of their caretaking impulses for my benefit and requested assistance when I really didn't need it. This forced inactivity was also contributing to weight gain. Instead of walking with my crutches to shop for food or go to the cafeteria, I would either ask Carol to bring something back to the studio or make my way via elevator downstairs to the vending machines. A hot meal usually consisted of popcorn made in the studio. I was now medically astute enough to realize that spontaneous improvement in my physical condition would not occur.

I asked one of my med. ill. instructors to recommend a good orthopedist. Mr. Wabnitz had worked on illustrations for several local physicians and would know who to consult. He recommended Dr. Carl Harris at Genesee Hospital in Rochester. I was able to arrange an

appointment immediately when I mentioned Mr. Wabnitz' name.

My car had proven to be anything but the "luxury" my parents had first considered it to be. The handicapped parking permit I had obtained allowed me access to places that I previously would have avoided. Now the car would also provide access to medical attention that would otherwise have been very difficult to arrange.

Dr. Harris, as every other physician I had ever visited, held a medical convention in my examining room. He invited his colleagues in to observe my problems and comment on how amazingly well I was doing. After everyone had been consulted it was decided that Dr. Franklin Peale would follow my case. Dr. Peale was an orthopedic surgeon associated with the University of Rochester's Pediatric Birth Defects Center. He had been instrumental in the development of the Rochester Parapodium brace, used primarily to help spina bifida children maintain an upright orientation and independent mobility.

All of my options were discussed during this initial meeting. Amputation, arthrodesis, orthosis, wheelchair exclusively or orthosis and wheelchair, were all mentioned. My previous experience at such consultations had led me to believe that a wheelchair was the only or

best solution. One physician even suggested my moving to Arizona because the level terrain is more accessible. (Recent exploration of the Grand Canyon State has proven to me that Arizona is neither flat nor completely accessible). Dr. Peale was concerned that I understood the social as well as physical implications that accompanied each of the options they discussed with me.

Most radical of all the proposed options was the suggestion of above the knee bilateral amputation. X-rays of the knee joints demonstrated grossly disorganized joints bilaterally. Varus-valgus deformity was obviously present but the limbs were at this point still somewhat functional. More importantly to me, they were my legs and I happened to like them attached in their original location.

Arthrodesis to fuse both knees in a permanent semi-flexed position was offered as a less extreme option but was also not entirely desirable. This would mean neither leg could be bent nor completely straightened. Sitting, driving or riding in automobiles, climbing stairs, dressing and even walking would become very awkward activities at best. The only purpose served by arthrodesis would be saving the limbs. Mobility would not be greatly enhanced.

Bracing bilaterally with knee-ankle-foot-orthosis would offer stability, reduce stress and slow further degeneration of the knee joints. Mobility would be improved through the reduction of muscle fatigue. Drawbacks to the proposal of KAFO's were identified as financial and medical issues. The braces would have to fit precisely in order to prevent the formation of pressure sores, which could lead to serious infection. The fittings could be time consuming and the fabrication expensive. Continued expense would be incurred for adjustments, repairs and, when necessary, refabrication.

Exclusive use of a wheelchair would limit accessibility and independent mobility. Few buildings or outdoor environments are entirely accessible to wheelchairs and those that claim to be have often been designed by well-intentioned but non-disabled people. Accessible housing is a necessity and difficult to find. Wheelchairs also come with an attached stigma. Public perception of wheelchair users often tends to associate mental deficiency with physical disability. The world also seems very different when always viewed from a level that is physically beneath the perspective of others. The advantage of body-language through position, posture and stance is limited for wheelchair users. Eye contact requires that a standing person physically look down upon

someone in a wheelchair. This positioning alone can create a patronizing atmosphere in social situations, and it is most always the responsibility of the person in the chair to change detrimental preconceptions.

Exclusive use of a wheelchair by a previously ambulatory person also disallows the use of certain muscle groups formerly used in upright posture and walking. Atrophy from disuse can cause circulatory problems in unused limbs and weight control becomes an even more important issue in a sedentary lifestyle. Physical therapy to maintain muscle tone, burn calories and assure overall fitness may be necessary.

Dr. Peale felt that a wheelchair would be advantageous in extending the longevity of my knees as continued use of the unstable joints would likely hasten the degenerative process that was occurring. He also expressed his opinion that the human form is intended to walk upright and that social interaction is strongly oriented in that direction. Self-perception must play an important role in what ever decision I made. I was encouraged by this fatherly and concerned physician to choose what was best for me.

I knew that I was definitely not ready to sacrifice all ambulatory abilities and opted for the KAFO's. Dr. Peale was glad to learn of my decision but encouraged me

to at least think about using a wheelchair for long distances. At the time, my impression of both braces and wheelchairs was not favorable. Until I was enlightened by Dr. Peale, I had visions of cumbersome steel and leather braces with heavy orthopedic shoes and the hospital-variety fifty pound Everest and Jennings type wheelchair. Dr. Peale showed me the new breed of wheelchairs which was like comparing an Edsel with a Porsche. Braces were now made with high-tech plastics rather than leather and steel. Some of my misconceptions and reluctance about this new mode of transportation were relieved. Dr. Peale also mentioned that arthroscopic surgery to remove the loose bodies in my knees could be performed once the braces had been adjusted. Before he would guarantee that braces could be made for my benefit, Dr. Peale informed me that I would have to be seen by an orthotist at the Birth Defects Center at Strong Memorial Hospital.

Carl Harris, M.D., P.C.
Genesee Hospital
220 Alexander Street
Rochester, New York

Nov. 7, 1984 This girl is 23 years old and has myelodysplasia. She's got an extensive medical history that accompanied her when I saw her first on 11-7. She's been treated by Dr. Kochersperger in Norwich, NY most recently. She's had some foot surgery done by Dave Murray. She's presently a medical illustration student at R.I.T. and Bob Wabnitz recommended that she come here. She developed a Charcot knee on the L at age 12 or 13 and in the last several years, has developed a similar problem on the R. She has been ambulatory up to this point but it's getting increasingly more difficult for her. She's also been able to ride a bicycle in the past and ride her horses.

I had Frank Peale take a look at her and I think he's going to follow up with her at the Birth Defects Clinic and the suggestions that were raised as we discussed things is some type of bracing vs. amputation. It appears that she has reasonably good sensation on both sides to at least the knee. The R side is probably below that and the musculature around the hip and the knee is surprisingly good.
CH/n

An appointment was arranged and I drove to the hospital where I had studied human gross anatomy and dissected cadavers the previous year. I learned that the BDC was located in an office shared by the Obstetrics and Pediatric Neurosurgery departments. The waiting room was teeming with mothers and toddlers. At twenty-three I was the oldest new patient they had ever seen. When the nurse called my name, she said my child would be examined alone first. I had to inform her that I was the patient.

I was examined by several doctors at the BDC who all remarked at how wonderfully I was doing physically. Dr. Peale introduced me to Gerry Tindall, from Rochester Orthopedic Laboratories, who would oversee the fabrication of my braces. I recognized Mr. Tindall as the man who several years earlier had taken measurements for the short brace that I was now wearing on my right leg. He thought I would benefit from more extensive bracing and that they could construct them to fit. I would have to go to their lab across the city to have plaster casts of my legs made before they could begin. Several appointments for fittings would also have to be made.

Driving the ten miles through city traffic and the "twelve-corners" intersection proved to be time consuming. Even more time consuming were the fitting

sessions. The braces would have to fit precisely to avoid causing pressure sores or causing further internal damage. I would often be at the lab for more than three hours at a time.

When they brought the newly fabricated orthosis into the examining room for my first fitting, I was not exactly overjoyed with their appearance. The braces would go most of the way up my thighs and were made of a milky-white polypropylene plastic with aluminum uprights on either side of my leg. The plastic had holes for ventilation, giving it the appearance of an anemic baby swiss cheese. They were hinged at the knee with drop-locks that would lock the joints in extension while standing. I immediately decided that I would only use them when absolutely necessary. The look on my face must have told Gerry what I was silently thinking. He tried to assure me that I'd like them so much I wouldn't even want to take them off to sleep. I had my doubts.

I took them back to the apartment and put them in the closet. It took me a week to adjust to the idea before I could bring myself to put them on. I was afraid that I wouldn't be able to drive as I had been, without any alterations to my car. I would put the braces on after I returned from classes, only wearing them around the apartment. Brian offered to drive if I needed a ride

but I was too self-conscious to wear them in public just yet. I also found out that my shoes would not fit over the plastic foot mold and that I would have to find new shoes that would fit. Brian took me shopping and, after many unsuccessful stops, I found a pair of suede hiking boots that would fit and that I could afford.

Another problem concerning the bulk of the braces was that I could barely pull my jeans over them. I had worn little else since high school so my entire wardrobe consisted of denim, flannel and sweatshirts. I couldn't afford to buy all new clothes to accommodate the braces so tight jeans would have to do.

The wheelchair also had to be fitted for proper posture. Measurements were taken by a supplier who came to my apartment. I learned that I had a choice of colors and attachments to accessorize my new chair. The chair would weigh under twenty-five pounds, light enough for me to manage alone. I also learned that the chair would cost more than my first car and was fortunately covered by my father's medical insurance as I was still considered his dependent.

I was missing a considerable amount of studio time during the day for the brace fittings and doctor's appointments. To make up the time I would stay at the studio to work on illustration assignments, sometimes

until three o'clock in the morning. One night, when I returned to the apartment, Brian had hung large signs with arrows and words of encouragement from the ceiling pointing the way to my room. "Sleep is just a few steps away!" they announced. Having a friend at home certainly made life easier.

After wearing the braces only inside my apartment for a couple of weeks I finally made the decision to try a brief trip outside. Carol had come over one Saturday to work on a project and needed to make a purchase at a local hardware store. I offered to drive. In order to get used to the feel of driving with my braces on, we made several trips around the apartment complex parking lot before venturing out onto the main highway. The experience was not half as nerve wracking as I had anticipated. The only adjustment I had to make was in how hard I stepped on the brake, as it seemed that I now had more direct power in my leg.

The trip to Chase-Pitkin was pleasantly uneventful. Previously, this store had been one that I avoided because Chase-Pitkin's size was comparable to its sister supermarket, Wegmans. Carol and I wandered around the store for nearly an hour. I was amazed at how long I could stand without being overcome with fatigue. I also noticed that I was considerably taller. I had never

thought of myself as short, but before bracing my legs would not fully extend or support my weight. I was now able to look Carol in the eye without glancing up.

Collecting my medical records proved to be a major project in itself. In order to write my own case history I would have to obtain the information from all of the physicians who had treated me throughout my life. Some of these doctors I hadn't seen in years, including the neurosurgeon who had performed the initial surgery on my spine, Dr. Lourie.

When I called his office in Syracuse to inquire about getting the records, I was immediately put through to Dr. Lourie himself. His Carolina accent was as strong as ever. He was delighted to hear from me and inquired about how I was doing. When I responded to his question of, "Ha ole are yew now?", he said that I made him feel like an old man and he could remember my surgery of 23 years ago like it was yesterday. I told him about my proposed thesis project and received an enthusiastic affirmative reply to my request. He offered to assist in any way he could and said he would like to have me make an appointment so he could examine me again. My case was unusual enough to have made a lasting impression on Dr. Lourie and I received copies of my records in the mail the following day.

All but one of the physicians I contacted were as enthusiastic as Dr. Lourie and provided all the information they could. I even obtained an immense collection of x-rays from the hospital where I was born, as they had saved every one.

The medical records were gathered piecemeal, leaving numerous gaps in my physical puzzle until information from yet another source arrived. Frequently, pages would be missing and eventually I found it easier to collect the information in person. I traveled the Binghamton, Syracuse, Rochester triangle of more than 350 miles several times in search of missing pieces.

The folders of records were gradually acquired and I began to compile a single chronologically organized file. As I attempted to skim through the reports of operations and discharge summaries, I became aware of my limited medical vocabulary. I was by now familiar with labels and descriptions of normal anatomy but would have to dramatically increase my medical vocabulary in order to comprehend and illustrate my own case history. My medical dictionary became severely dog-eared during this learning process.

The braces provided an amazing amount of freedom. I could now stand to do simple chores like washing dishes.

I even began to take a few short steps around our tiny kitchen without using my crutches; I only needed to hang onto the counters for balance. Being able to wash dishes may not seem like a big deal, but for someone who likes to cook, expeditious clean-up is a necessity. From the time that my right knee had joined my left in Charcot deterioration during my undergraduate years, I had been supporting myself with my crutches, bearing most of my weight on my hands and armpits to stand. My hands would go numb while standing at the kitchen counters washing dishes or preparing food. When I reached for something from an overhead cupboard one crutch would fall as I leaned on the other to stretch. Bending to pick up objects from the floor was difficult with crutches supporting all of my weight. I would have to place them in a splayed position in order to bend enough to reach the floor, much as a giraffe must do to take a drink from a waterhole. I had to hope that the crutches were firmly planted and would not slip out from under me.

When I realized that while wearing my braces I could take a few steps without my crutches, I wondered if I would be able to improve my balance enough to walk across a room. While Brian was studying in his room, I practiced walking the five feet between my drafting table and the daybed that served as a couch in the living room.

The carpeted floor proved to be a hindrance by throwing me off balance and the linoleum floor in the kitchen was too slippery for the smooth plastic foot plates of the braces. After several attempts I was able to walk between my desk and the daybed without bobbling tremendously. At the time I was elated, thinking that I would be able to walk around the house or on other level surfaces without crutches. I figured that I would probably never be able to carry anything substantial as it would throw me off balance but just getting from here to there unaided would be wonderful! I practiced walking across the room when I needed a break from the anatomical illustration of a human heart that I was working on.

Later that evening, when I had gained enough balance to attempt walking the entire length of the living room, I became very excited and daring. I wanted to share my newfound ability. I walked down the long hallway and into Brian's room on the pretext of borrowing a pair of scissors. Preoccupied, he handed me the scissors without looking up from his work. I said thank-you and started back down the hallway. Half way back to the living room I heard a loud "Hey!" as Brian ran after me. "You're not using your crutches!" was his surprised observation. I told him I had been practicing that evening and showed him how well I was managing. As had become our custom

for minor breaks and celebrations, he took me to a nearby "Friendly's" for a late night ice cream sundae.

I adjusted to wearing the braces full time and began to experience frequent mechanical difficulty. My activity level was now much higher than anticipated by the technicians who had fabricated the braces. I was therefore placing an excess amount of stress on the metal components that had been used. My semi-flexed knee stance also contributed to force exerted on the drop-locks at the knee, causing the locks to give.

Adjustments were made gradually to decrease the valgus and flexion contractures. Initially I was told that they would adjust the braces over an extended period of time, as the orthodontist had done to straighten my teeth. Each time I returned to the lab for repairs they were able to adjust the braces to a more straightened position.

Every time an adjustment was made to straighten my knees or relieve pressure areas I had to also make a separate visit to Dr. Peale, who was supervising the process. I developed several sores on my feet during this adjustment period which also demanded medical attention. The more my knees were straightened, the more noticeable the loose bodies within the joints became. Surgery to remove these aberrant pieces of bone was

scheduled for late February but was delayed by yet another prolonged infection.

Franklin V. Peale, M.D., P.C.
Genesee Hospital
220 Alexander Street
Rochester, New York

Feb. 8, 1985

Her braces have markedly helped the comfort of her knees. She is able to take a few steps around in her apartment without crutches, feeling stable. She's having trouble keeping her knee locks locked. They tend to pop out. The L knee does not yet fully extend because of a tight biceps tendon which should be better after it's released. R knee extends almost completely now. Both knees are in only mild valgus. The braces look as though there might be a little less valgus possible in the knees but this will depend upon the knee itself. The R brace was not removed.

L foot has a large superficial blister abrasion 2nd degree at least over the end of the great toe and the smaller but full thickness one over the heel cord which is granulating in. The one on the toe is angry, shows some evidence of cellulitis in the toe itself and an area of full thickness necrotic about 1 x 1/2 cm. just beneath the tip of the nail. She thinks she did it walking around the apartment at night without her brace on at bedtime.

MANAGEMENT: Cultured, detergent soak, Betadine dressing and re-check in 10 days. Ampicillin P.O. 500 q6h xs 3 days, then 250 q6h. FVP/n

Feb. 12, 1985

I spoke with her on the phone 2-12 re: her culture and the possibility that we would not be able to do surgery now. I also suggested to her that perhaps surgery was not absolutely indicated since she is much more comfortable now that she has good bracing, than she was before. The one aspect of her situation that does really need some relatively immediate surgical care is the valgus deformity of the left knee with a very tight biceps femoris tendon which does need to be released. It certainly should not be done in the face of any intercurrent infection in her foot, but it is of some urgency even tho her knees are now much more comfortable, properly braced and protected from the rapid valgus deterioration of the lateral compartments that has taken place over the last 3 or 4 yrs. FVP/n

Feb. 19, 1985

The surgery scheduled for next week for exploration of her knees has had to be cancelled because of continued skin loss over her L great toe. The area of eschar and clear necrosis has enlarged to about 12 x 10 mm., but the surrounding area looks pink, reasonably healthy granulation, and is beginning to epithelialize from the margin. The erythema is diminished from last week. X-ray of the toe today shows no evidence of osteo-myelitis.

I've dressed her with Xeroform and she will continue to do this herself. She has a small blister in the flexion crease of her great toe and another small blister over the pulp of her small toe which she thinks may be related to walking without braces in the apartment sometimes and trying to protect the big toe from pressure. She'll go to the Lab and get her braces finally adjusted to remove all residual valgus if possible from her knees, and re-check here in 2 weeks. FVP:ek

Mar. 5, 1985

Big toe is coming along. It's now clean, epithelializing from the margins, and the eschar will soon separate. I debrided the margins of it slightly. I think 6 or 8 weeks to coverage.

L knee - Any time she gets up from sitting she has trouble getting the knee extended and the brace locked because of tension in the biceps femoris, but the valgus has been pretty well adjusted out of the brace now and except for the natural external rotation of the tibia she seems satisfactory. The R knee is improving steadily. She is getting more and more extension all the time, and she goes back to the shop and gets them to set the knee out a little straighter all the time. She gets a little pain occasionally from a loose body that gets caught. We'll have to get these out, but for now she's doing fairly well. She ambulates on a crutch, bears full weight on the R lower extremity. The L is shorter and she sort of swings through and puts the toe down slightly. She does like to walk around in small spaces without her crutches, but it's critical that she use them to protect the knees and preserve their longevity as best she can. Not sure yet about when she could have surgery. FVP:ek

Mar. 26, 1985

Her toe wound is now nicely granulating. The eschar has fallen off. The center is a little yellow but it's coming along satisfactorily. She says it hasn't closed in anymore for the last 10 days, but there is early epithelialization visible at the margin of the wound which does not appear inflamed. There is nothing to do here except wait for it to close before planning any other consideration. 4 wks. with continued Xeroform dressings and Betadine washes intermittently in between until then.

I have x-rayed her pelvis which has not been done in many years. Her left hip lies in marked external rotation and shows laterally on an AP view. The R hip is in neutral position. Both show excellent normal hip joints with good joint space, normal femoral heads, good acetabular coverage, etc. FVP/n

Apr. 23, 1985

Her R great toe is just about completely healed now. She is getting along satisfactorily. Wants to re-schedule the exploration of her knees for 6/17, which is reasonable. Carry on with her braces, take excellent care of her skin, and re-check preoperatively. FVP:ek

May 20, 1985

Stopped by to check her braces preparatory to new fabrication because her legs have changed so much since they were applied that they no longer can be made to fit her. The R can come out of the degree of external rotation that it is in. The L generally could fit better to stabilize her knees better. The hinges are broken, the locks don't work, and her general alignment of her knees has improved sufficiently that I think this is worthwhile even though they are only 5 months old. Prescription given.

When her L brace broke recently, she was out of it for several days. When she got it back, her L knee began to swell and did so for 2 or 3 days to the extent that she couldn't wear the brace. It is now beginning to go down, but it is significantly swollen in the joint. It is not hot, nor unstable, but simply has a marked effusion again.

NOTE TO EDNA: It's important to check with the OR. We will definitely need TV when this is done, and would like to use the VCR from the Orthopaedic Department to tape the procedure. In addition, one of the medical illustration students from R.I.T. would like to observe. FVP:ek

Between trips to the Orthopedic lab and Dr. Peale's office I still had classes to attend and had a required showing of thesis-related art to prepare. Time constraints would make the written portion of the thesis impossible. I would have to forego the research requirement until after I had completed the required coursework.

Transportation in all of the varied forms I was now using proved to be time consuming and costly, including my car. For some reason my grey Chevy Monza seemed to be a target for accidents, mostly of the anonymous bump-and-run parking lot variety. On several occasions when I was about to leave RIT for my apartment I found new dents, gouges and cracked parking lights. Twice, while stopped for red lights at intersections, the car was hit causing "minor" damage in excess of \$500.00. Fortunately, the known incidents were covered by the offending driver's insurance but still more precious time was consumed for accident reports, estimates and repairs. For all the inconvenience that accompanied car ownership, the independence it provided was worth the trouble.

Arthroscopic surgery on my knees was delayed until after the thesis show in order to allow my foot time to heal. Even with all the late nights at the studio, I was still putting finishing touches on my watercolors with an airbrush minutes before the opening reception. Carol frantically cleaned glass and put my artwork into the frames while I finished. The last illustrations were hung as the gallery gates were opened.

I quickly returned to the apartment to change my clothes for the opening reception. When I was ready to put my shoes on I realized that the only shoes I owned that could be worn over the braces were my brown suede hiking boots. Fashion statements were not to be the highlight of this show as far as I was concerned.

When I returned to the reception for this master's group show, I was asked to respond to a number of questions about my display. I had presented my work as an educational exhibit on spina bifida. A friend from the ceramics studio had volunteered to construct a light box so that I could include x-rays as well as a normal human skeletal vertebral column in the display for comparison. I was surprised at the interest in my exhibit and the number of people who confessed to having relatives or acquaintances with the condition. More than a few people asked for further information and for copies

of the written thesis after completion. I collected names and addresses so I could honor these requests. I even received one request from the parents of an adolescent with spina bifida to call their child to offer encouragement. I was also honored that my former instructor of photography at Keuka, Chris Wright, had brought his family from Penn Yan to attend the reception.

After the thesis show work had been accomplished the remaining coursework had to be completed and attendance at a commencement ceremony was required. Because of extended study courses and the limited time in which I had to devote to classwork, I planned to remain at RIT for summer quarter. Brian would be returning to Toronto and our lease had expired, so I was to share an upstairs apartment with Carol and her roommate, Geraldine, for the summer.

Because participation in a commencement ceremony at RIT is required in order to eventually receive a degree, I accompanied my classmates in this formality. For the third time in my life I attended commencement exercises in a gymnasium. This was to me the ultimate irony. For me to receive any honor in such a temple of physical ability I likened to revenge.

After commencement my father came out once again to move my things, this time to my summer nest at Carol and Geraldine's. My surgery was rescheduled for the break in between the spring and summer academic quarters. I was admitted for bilateral arthroscopy and debridement of the knees.

THE GENESEE HOSPITAL
HISTORY - PHYSICAL EXAMINATION - PROGRESS

ADMISSION NOTE

UNIT NO.

Barbara Tefft

Barbara Tefft has mid-lumbar meningomyelocele. She has been ambulatory all her life, and her activities have in early times included both bicycle riding and horseback riding. In 1973, she underwent medial epiphyseal stapling of her left distal femur to correct a progressive valgus deformity of the knee with degenerative changes in the lateral femoral condyle of a Charcot nature. She first came to attention here in November of 1984 because of progressive changes in both knees with marked swelling, valgus deformity and diminishing ability to ambulate in an AFO, which was her only bracing to that time.

Her physical examination demonstrated markedly swollen knees bilaterally with palpable osteochondral fragments in each knee, quadriceps and hamstring power of a good nature bilaterally, knee flexion contractures associated with valgus deformity bilaterally, and insensitive flail feet.

Initial management consisted of fabrication of bilateral KAFO's which have succeeded in returning her right knee to a full range of motion and stabilizing her quite well against valgus deformity when ambulating. The left knee still carries a 20 or 30° flexion contracture. The left lower extremity appears to be approximately 2" shorter than the right to clinical examination, which seems a large discrepancy to have resulted from a unilateral stapling of the knee at the age of 12.

She enters at this time for arthroscopic evaluation of both knees with the intent to remove osteochondral fragments, and make some determination as to what further corrective action may be warranted to protect her knees from further deterioration.

Her urine has been asymptomatic for many years. She voids. Has occasional stress incontinence. At the time of this dictation, urine culture has been obtained. We have no results. 5 to 7 days of hospitalization is anticipated.

Signed _____
for Franklin V. Peale, M.D., P.C.

FVP/n
MF 47B

The
Genesee
Hospital

OPERATIVE REPORT

PATIENT NAME	DATE OF OPERATION	DATE DICTATED
Barbara Tefft	6/17/85	6/17/85

DATE TRANSCRIBED
6/18/85

PRE-OPERATIVE DIAGNOSIS

Bilateral Charcot knee second to meningomyelocele.

POST-OPERATIVE DIAGNOSIS

Same.

OPERATION

Bilateral arthroscopy; debridement of knees through open arthrotomy.

SURGEON

Dr. Peale Dr. Black

OPERATIVE PROCEDURES AND PATHOLOGICAL FINDING

Anesthesia: general.

PROCEDURE

Under general anesthesia, the right knee was arthroscoped through a lateral infrapatellar approach. Suprapatellar pouch was unremarkable. The patellofemoral joint revealed grade II chondring, mostly at the crest of the patella. Joint surfaces were reasonably maintained. Medial meniscus was visible. Weight bearing portions of medial femoral condyle and tibial plateau seemed reasonable. There was considerable chondring and no evidence of anterior cruciate ligament in the intercondylar notch. The lateral tibial plateau was grossly destroyed. There was a marked drop off of about a centimeter into the lateral joint space from the intercondylar notch and articular cartilage was no longer present. There was a cobblestone, reddish surface of bone and several loose bodies toward the margin of the joint which could be readily moved about by palpating with a finger externally. No lateral meniscus was visualized. At this point the tourniquet was inflated. Arthroscopy was discontinued. A lateral arthrotomy was made through the ilio-tibial tract. The step surface of the lateral femoral condyle was quite smooth to finger palpation. There were numerous synovial enchondromata along the lateral margin of the joint which were excised sharply. The origin of some was not clear. The origin

SIGNATURE OF OPERATOR _____

The
Genesee
Hospital

OPERATIVE REPORT

PATIENT NAME DATE OF OPERATION DATE DICTATED
Barbara Tefft

DATE TRANSCRIBED

PRE-OPERATIVE DIAGNOSIS

POST-OPERATIVE DIAGNOSIS

OPERATION

SURGEON

OPERATIVE PROCEDURES AND PATHOLOGICAL FINDING

TEFFT - CONT.

of others was pretty clearly the very lateral cortical edge of tibia. The posterior half of the ilio-tibial tract was divided completely to prevent further flexion forces from it. The anterior third was left intact. After extensive saline irrigation, the wound was closed with 0 Vicryl interrupted sutures in capsule and ilio-tibial tract, 3-0 plain in subcutaneous tissues and steri strips in the skin. #13 French, round Jackson-Pratt drain was placed in the joint prior to closure through the superolateral drain site. Tourniquet time was 40 minutes.

The left knee was then scoped similarly. There was much more degenerative charding in the synovium and articular surfaces of the left knee than the right. The lateral joint space could not be well visualized at all and when it was, it was apparent that the changes were very similar to those on the right with loss of the articular surface of the tibia laterally. A number of large, marginal osteochondral bodies, movable on palpation, but apparently attached to two synoviums were present. Visualization was difficult but it was eventually proven that the medial meniscus was intact. The articular surfaces were not severely damaged. There was a lot of charding in the intercondylar space but I think that we could see the anterior cruciate ligament. We did not

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Hospital

OPERATIVE REPORT

PATIENT NAME DATE OF OPERATION DATE DICTATED
Barbara Tefft

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PRE-OPERATIVE DIAGNOSIS

POST-OPERATIVE DIAGNOSIS

OPERATION

SURGEON

OPERATIVE PROCEDURES AND PATHOLOGICAL FINDING

TEFFT - CONT.

probe the joint in this case but, when it was apparent that there was a large amount of tissue to be excised laterally, arthroscopy was discontinued and another lateral arthrotomy performed under tourniquet. A number of bodies, all that could be palpated, were excised. There was some very fine, synovial osteochondromatosis lateral to the superior pole of the patella. Some of this was excised. Closure was similar. Tourniquet time was 1 hr. 7 min. Dressing was the same. It should be mentioned that the lateral joint space on the left side is unusual in that the tibial surface is convexed, domed, and the femoral surface is concave. Both surfaces are fairly smooth and intact but bony contact is not made until the knee drops into considerable valgus. It should also be mentioned that the posterior aspect of the ilio-tibial tract and the biceps femoris were both divided on the left knee, prior to closure.

It is not clear whether this procedure will make a major difference to the function of her knee. We do now know clearly the state of the joint.

sj

SIGNATURE OF OPERATOR _____

Dr. Peale had video taped the surgery and early the following morning collected me for a premier showing in the doctor's conference room. He thoroughly explained what I was seeing at every angle. During the five days I was hospitalized Geraldine and her fiancée, Mike, came to visit every day, sneaking in a beer on one occasion. I had hung one of my recent general illustration projects next to my bed to remind me of one of the things I looked forward to doing when I recovered. The painting of recently deceased author Richard Brautigan drew a steady stream of hospital staff at all hours of the day and night. Dr. Peale brought several of his colleagues from the operating suite, this time to see my painting rather than me. My former painting instructor from Keuka, Gary Jurysta, was up from New York City and made a special effort to visit me in the hospital.

When my father arrived to take me home to recuperate until summer classes began, he informed me that Nermal, the world's cutest kitten (now cat), was also in the hospital. He had been bumped by a car on the dangerous curve in front of our house and was at the vet's for observation. Late that afternoon we stopped at the vet's on the way home to discharge Nermal from the hospital. When we pulled in the driveway at home my mother and

sister's first question as they came out to the car was, "How is Nerm?"

While I convalesced at poolside, Dr. Peale called long distance to make sure I was behaving myself. I stayed off my knees and used my wheelchair as much as I could, considering that there were no ramps, inaccessibly narrow bathroom doorways and an upstairs bedroom. The playhouse now proved to be a much more accessible living space than the house. The former milkhouse had a ground level doorway and was only fifteen feet from the three steps leading to the back door next to the bathroom. Crawling up the three steps was preferable to crawling the flight of stairs to my room. Pushing my wheelchair over the uneven lawn to the house was moderately difficult but the real inconvenience came if I wanted to go inside the house. I had to get out of the chair, lift it up onto the stone porch, push it across to the door, lift it over the five-inch threshold while holding the screen door open and get back up into the chair from the floor once inside. When I asked my parents to build a short ramp alongside the porch, my mother rejected the idea because a ramp was not an historically accurate architectural structure for her house. More than a year later, my father constructed a wood ramp in his shop and

began installing it one morning before my mother got out of bed.

The denim jeans I had been wearing over my braces were by now in shreds because of the unavoidable metal protrusions at the knees. Rather than investing in new jeans that would soon follow suit, I decided to sew myself some dresses. This was a radical change of style for me since I hadn't even owned a dress since I was in junior high school. Having my legs encased in plastic also made me extremely hot during warm summer weather. I wanted something cool and summery to wear so I made several sundresses. I found a pair of shoes that were more suitable than my suede hiking boots and got matching leg warmers to cover the braces below the dress hemline.

I returned to Rochester for my post-op exam with Dr. Peale completely transformed with new clothes to complement my newly renovated knees.

Franklin V. Peale, M.D., P.C.
Genesee Hospital
220 Alexander Street
Rochester, New York

Barbara Tefft

Jun. 27, 1985

She feels very good about the surgery on both knees, each of which feels better. She said there is "no nail" in the R knee which was there before. She feels she can extend both knees better than she could before. The R has a mild effusion but she said yesterday it had almost none at all, to the extent that she's having a little patellar pressure from her anterior shell with less swelling present. She became aware of a ridge on the medial femoral condyle. It's an osteochondral ridge at the margin of the articular surface. This too is more noticeable when the swelling is down.

The left knee had quite a marked effusion as it did preoperatively. It is the more severely involved. Her leg length films which demonstrate a 3/4" differential L shorter, still shows a couple of round masses in the central portion of the knee which must be posteriorly in the popliteal space. We could not reach them surgically and I think it would be more than a reasonable procedure to try and go after them. For the time being, she is going to get her right knee brace fixed and padded or flared in some way so that it doesn't produce patellar pressure, use the braces full time and re-check here in August. FVP/n

Aug. 26, 1985

No complaint. She had broken one of the screws in her lateral side of her R orthosis and had to have that replaced but she has no symptoms in her knees, has been getting around using her wheelchair very little. On exam both knees lack 5° to 10° full extension. The R has a mild, the left a moderate effusion. The R tends to lie in slight valgus but can be corrected down to about 6° of valgus. Her brace is set at about 13 and prescription sent to the Lab. to try and modify the brace to less valgus. As I worked with the R knee into full extension, there was a definite clunk as a piece of firm fibrous feeling tissue moved in the knee joint. This happened twice. Neither she nor I could localize it.

The L knee with the greater effusion has a better alignment. It collapses into valgus as does the right, but can be brought completely straight, and her brace is set nice and straight.

I've encouraged her again to be good to her knees, to use the wheelchair when she can for distance activities, as their longevity is precarious.
Return prn. FVP/n

I divided my time during summer quarter between both of my minors, ceramics and computer graphics. These subjects greatly contrasted and complimented each other, providing me with an enjoyably challenging course load for the summer. The season was made even more enjoyable when Carol and I took an extended weekend camping trip to the Adirondacks and visited my Aunt Hazel and Uncle Walter.

When I was not in class, I spent the majority of my time in the medical library at Strong Memorial Hospital gathering information to write my thesis. During my research I noticed that information pertaining to those born with birth defects had a noticeable age termination point. There was virtually no information about myelomeningocele patients past the age of twenty-one. I began to wonder what happened physically, emotionally, socially and financially to this lost population, a demographic sector which I had joined only a few years ago.

At the end of the summer my father came to Rochester to move my things back home. I continued my research and

contacted physicians using the playhouse as my base. I also began to send out resumes in my search for employment. The playhouse worked well as long as the weather stayed warm. By the end of October, however, I was forced to move back into the house. The saying "You can never go home again" began to ring true. The house was much too small to contain the mentally, emotionally and physically disparate group that attempted to exist under its roof. I escaped for extended periods to stay with friends in Rochester where I was closer to the reference materials and medical care I needed.

During this time I learned the details of my physical condition. Issues that had been of unspoken concern for most of my life now were explained. I now knew why my physicians asked certain questions and how important my answers were. I learned how the damage to my spinal cord affected my lower limbs and organs. This newfound knowledge changed the way I looked at and cared for my body.

Being comprised of fat cells, the lipoma that remained entangled in the cauda equina of my spinal cord would be metabolized much as any other body fat. If I gained weight more fat could accumulate at the site of the lipoma, increasing its size and pressure exerted on the cord. If I lost weight, size and pressure could be

reduced. Increased pressure on the cord, however slight, may cause further neurologic symptoms and damage. As with any excess fat accumulation, absorption of carcinogenic toxins or mutation can create the potential for malignancy. Urologic symptoms could be treated by any number of methods including drugs, corrective surgery or prosthetic implants. Because colon, breast and skin cancers have been diagnosed in both of my parents' immediate relatives, and because of the fact that my mother was given DES while pregnant, I became aware of the importance of self-examination and routine medical evaluations.

I visited Dr. Lourie at his office in Syracuse as he had requested when I contacted him about obtaining my medical records. I also made an appointment for urological evaluation in Syracuse with Dr. Michael Kendrick, who was experienced with implantation of the artificial sphincter and other corrective urologic surgeries and treatments. Dr. Kendrick's tests yielded a not uncommon diagnosis for a myelomeningocele patient - classic spastic bladder. Surgery would not be necessary as the problem could be controlled pharmacologically. He prescribed an anticholinergic called Ditropan at a dose of 5 mg. to be taken four times each day. The drug acts

as a muscle relaxant on the over-stimulated smooth muscle of the bladder, increasing vessel capacity and delaying the urge-to-void sensation. All drugs are metabolized and distributed throughout the entire body; therefore, the muscle relaxant property of Ditropan affected more than just my bladder. The 20 mg./day prescription relaxed me into at least a two hour nap after each dose! Although the drug definitely had a positive affect as far as my bladder was concerned, my hours of productivity were greatly reduced. After gradually reducing the dosage, I found the optimal amount for me to be 2.5 mg. taken only at bed time. This meant I had to cut the little blue pills in half but that was a minor inconvenience for the relief it provided. The Ditropan relaxed my bladder enough to allow me to sleep through the night for the first time. It also helped the insomnia I had developed from being unable to relax aching muscles and having to get up several times a night. I could now sit through a movie without having to leave the theater, attend lectures and not have to conspicuously leave in the middle and go to new places without having to find the restrooms in advance. A small degree of spontaneity was added to my life.

KENDRICK & TEMES, M.D., P.C.
ST. JOSEPH'S HOSPITAL PHYSICIAN'S OFFICE BUILDING
101 UNION AVENUE, SUITE 705
SYRACUSE, N. Y. 13203

MICHAEL J. KENDRICK, M.D.
SANFORD P. TEMES, M.D.

TELEPHONE: 479-6626
479-6627

Referring physician:

Age: 24

Patient's name: BARBARA TEFFT

Date: 11-6-85

REASON FOR REQUEST: (Complaints & findings, including pertinent diagnoses, neurological exam and lab data)

- myelomeningocele
- HTN

REPORT OF CONSULTATION

FLOW RATE: PEAK _____ cc/sec MEAN _____ cc/sec TIME _____ sec
NSG TOTAL VOLUME VOIDED _____ cc

CYSTOMETRY POSITION supine POST VOID RESIDUAL 0 cc
CO₂ ✓ FIRST SENSATION 80 cc CAPACITY 90 cc
WATER _____ PEAK PRESSURE 100 + cm H₂O DETRUSOR CON-
TRACTION? yes ABLE TO SUPPRESS? yes
POSITIONAL CHANGE? NT TEMPERATURE
SENSATION? NT

SPHINCTER EMG: SITE rectal POSITION supine VOLUNTARY CONTROL _____
UNINHIBITED SPHINCTER ACTIVITY? none
BULBOCAVERNOSUS REFLEX NT
POSITIONAL CHANGE? NT
DETRUSOR-SPHINCTER DYSSYNERGY? NO

URETHRAL PRESSURE PROFILE POSITION supine FUNCTIONAL LENGTH 6 cm
MAX. CLOSURE PRESSURE 55 cm H₂O
CO₂ _____ ANATOMICAL LENGTH 1.2 cm
RESPONSE TO ALPHA BLOCKER NT
WATER _____ ABNORMAL AREAS OF HIGH PRESSURE? none

PHARMACOLOGICAL TESTING:

ADDITIONAL DATA:

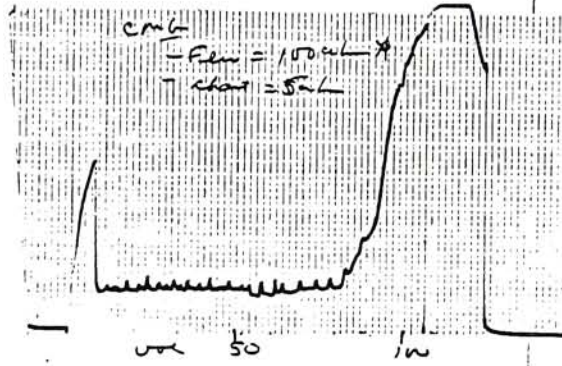
VP - myelin

INTERPRETATION:

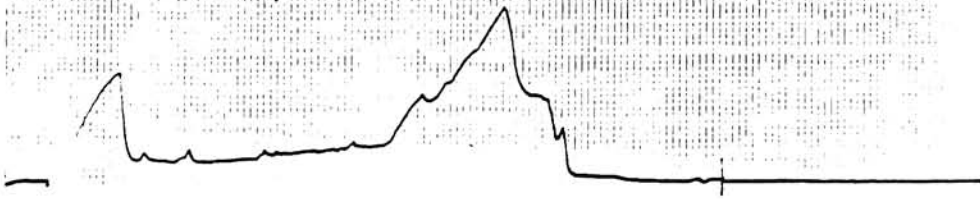
- classical spastic bladder

BARBARA TEFFT 1-6-85

PMIS



TP
Flw = 100 ul *
clat = 25 ul
pullin = 12.5 ul





**St. Joseph's
Hospital Health Center**

301 PROSPECT AVE., SYRACUSE, NY 13203

E. W. Carsky, M.D.
F. C. Brownell, M.D.
G. A. Shaheen, M.D.
F. Azimi, M.D.
A. B. Foster, M.D.
J. P. Marangola, M.D.
D. M. Haswell, M.D.

DEPARTMENT OF RADIOLOGY

PATIENT NAME		AGE
LEFT, BARBARA		24
DATE	OP	
11/6/05		
X-RAY NO.		
24-10-97		
PHYSICIAN		
Dr. Kendrick		

IVP:

The preliminary film of the abdomen shows marked abnormality in the lower half of the lumbar spine. There is widening of interpediculate distances. There is anacolous formation of the posterior arches or perhaps surgical laminectomies. The patient is known to have had a myelomeningocele. Certainly this can be explained by the changes I see on the radiographs.

After infusion of contrast, the kidneys are visualized promptly. They are of normal size, shape and position. The pelvicaliceal systems are well seen and normal. The ureters follow a normal course into the bladder, which is unremarkable.

IMPRESSION: The intravenous pyelogram is normal.

mq 11/06/05

Alan B. Foster, M.D.

Dr. Lourie performed a thorough neurological evaluation, and this time I answered all of his questions without the embarrassment I had experienced as a child.

* * *

LOURIE, STEWART AND SHENDE, M.D.'S, P.C.
Suite 504, 725 Irving Avenue
Syracuse, New York 13210

ADULT and PEDIATRIC NEUROLOGICAL SURGERY

315) 470-7961

HERBERT LOURIE, M.D.
DONALD H. STEWART, Jr. M.D.
MICHAEL C. SHENDE, M.D.

Herbert Lourie, M.D.
12/2/85

Office Consult

Patient: Barbara Tefft

This is a now 24-yr. old girl on whom shortly after birth I performed a repair of a lipomatous myelomeningocele but the pathology was not a straightforward MM for there was also a teratoma or at least a partial twin associated with the mass. The mass contained adult bowel and glandular tissue as well as skeletal and muscle tissue. Her condition was stable with a partial cauda equina syndrome until 4 yrs. ago when she had increasing numbness below the knees and later ascending numbness up above her umbilicus in an area that was not previously numb. Two yrs. ago she noted her hands going intermittently numb. She thought it was due to heavy use of crutches for when she got long-leg braces last year and had to use less weight onto the palms and axillae, she thought the hand numbness subsided. However, being

an artist, she knows her hands still do not feel just right. She feels as tho she has a thin glove on such that she is not actually getting primary contact with the object. Never has she had pain in her neck, back or legs. She thought that once her genital sensation was better than it is now.

Dr. Kendrick put her on Ditropan about a month ago for a spastic bladder, which has helped somewhat. She also suffers from colitis but does not soil herself.

On exam, upper limbs seem grossly normal for there is full strength and mobility. Reflexes 2+; no pathologic finger signs. No Horner's syndrome. Object recognition is acutely intact in the hands as is vibration. Grips are powerful. Superficial abdominal reflexes are present in all 4 quadrants and there is no Beevor's sign. Pinprick is markedly diminished over the entire lower limbs, worse below the knees where it is all but absent. She feels sensation in the upper lumbar dermatomes but sensation is not normal until one gets into the T-9-8 dermatome junctional area. She has some strength in the iliopsoas and quadriceps bilaterally and can weakly dorsiflex some of the right, none of the left toes. No response to plantar stimulation. Right ankle jerk trace; left absent.

My concern is whether she has a tethered cord syndrome. I recommended that she come into the hospital for myelography, that we get plain films and CT scan today as an outpatient. She is reluctant to consider further surgery. I will try to arrange for a second opinion and she has promised to stay in phone contact and we shall talk it over once I can arrange for a second opinion.

Herbert Lourie, M.D.

A second opinion was arranged with the Chief of Pediatric Neurosurgery at Strong Memorial Hospital in Rochester, Dr. Joseph McDonald. Again I waited among the expectant mothers and small children where I had visited Dr. Peale at the Birth Defects Center.



THE UNIVERSITY OF ROCHESTER
MEDICAL CENTER

601 ELMWOOD AVENUE, BOX 661
ROCHESTER, NEW YORK 14642
AREA CODE 716-275-5255

SCHOOL OF MEDICINE AND DENTISTRY · SCHOOL OF NURSING
STRONG MEMORIAL HOSPITAL

Joseph V. McDonald, M.D.
Professor & Chairman
Division of Neurological Surgery

December 6, 1985

NEUROSURGICAL CONSULTATION

RE: Barbara J. Tefft

Shortly after birth, this lady had a lesion over her lumbosacral area explored. Intestine and pancreas, as well as lipoma tissues, were found and excised.

She has always been paraparetic. However, up until she was about 11, she could get along pretty well walking. Then, she began to have more difficulty. Her downhill course since then has been very gradual. For the last seven or eight years, her left foot has been completely paralyzed. She can feel things most of the time below her knees, although she has suffered severe burns on the right foot and has not felt them.

She recently had a urologic examination which showed that she had a small, spastic bladder. She didn't think her bladder status had changed significantly for many years.

She has a bowel movement about every two weeks, usually accompanied by a lot of abdominal cramping. She used to have many autonomic discharge symptoms when her colon became distended and she had a bowel movement. She would sweat profusely, have cramps and pain. Those things are not as prominent now.

She has poorer sensation in the left leg than in the right one.

She has Charcot knee joints. Dr. Peale had operated on her left knee to remove numerous cartilaginous fragments which prevented knee extension. She could extend her knees now. Her right one hurt. The left one had no sensation.

Neurosurgical Consultation
December 6, 1985
Page 2

RE: Barbara J. Tefft

She has buttock numbness and perineal numbness. She thought that the buttock numbness had increased during the past four or five years. She had also had some difficulty with sensation in her hands and couldn't feel hot things with her fingertips at times.

She denied headache or pains in the neck or scapular regions.

Cranial nerves were normal. Range of motion in the cervical spine was excellent. There was no spasm or tenderness or deformity there or in the thoracic or upper lumbar region so far as I could see. The knee joints were distended with fluid, especially the left one. She wore long leg braces. There was atrophy of the muscles below the level of the knees. When she dorsiflexed the right foot, it everted. She couldn't move her left foot. She could not extend the right ankle. She did perceive vibration and touch in her legs below the knees. She perceived touch in the perineal area but couldn't distinguish pin prick from a dull object there or over her buttocks out to about S1.

There was a soft mass over the lower lumbar and sacral region. It wasn't tender. It didn't distend when she strained.

No atrophy or fasciculation or sensory loss was found in the upper extremities. Intercostal motion was normal. Abdominal muscles had good tone and strength down to the lowest part of the abdomen.

She had had a teratoma excised and the tissues were reported as showing intestine, pancreas, and lipoma. It is quite possible that some of the teratoma persisted and has grown. Tethering of the cord is very likely. The slow progression of her symptoms over the past seven years suggests growth of a mass rather than simple cord tethering since she stopped growing in her mid teens.

However, it is known that adults can get into trouble with cord tethering so that is certainly a possibility here.

Neurosurgical Consultation
December 6, 1985
Page 3

RE: Barbara J. Tefft

She might have an associated lesion somewhat higher such as diastematomyelia, but I think if that were true, there would be some obvious deformity, either visible or palpable over her spine.

I think that she ought to have an MR scan to demonstrate the entire length of the spinal cord. Both intramedullary and extramedullary lesions such as a lipoma, teratoma or tethering would very likely be demonstrated on that study.

It is possible that supplementary studies such as localized CT or myelography with cervical injection might be needed. I doubt it. After that, a decision could be made as to the need for appropriate surgery.

Joseph V. McDonald, M. D.

JVM:sm

xc: Herbert Lourie, M. D.

Dr. McDonald and Dr. Lourie agreed that an image study would be helpful in determining my neurological status. I was scheduled for a scan using the new technology of Magnetic Resonance Imaging. The MRI uses magnetic energy rather than radiation to capture a detailed image. I was slid into the MRI machine's tube while lying flat on my back. This had never been a comfortable position for me as the tumor on my lower spine was like having a pillow in the small of my back. Being strapped to the table and told not to move for the two-and-a-half hours it took to image my entire spine was not pleasant. I had never been claustrophobic but having the white roof of the tube only two inches from my face for such a long time made me panicky. I broke out in alternately hot then cold sweats and felt as though I couldn't breathe. Knowing that if I moved the entire series would have to be done again, I took deep breaths and clenched my fists.

Before the imaging series was begun I was warned that I would hear a "tapping" sound while the image was being taken. In reality the "tapping" was more like a jackhammer! Positive and negative charges are aimed at each other inside the machine in regular, rapid pulses. This creates the noise, much in the way thunder is heard when opposing charges create lightning. I came out of

this sophisticated piece of machinery stiff, deaf and phobic, but at least they got what was needed and I didn't have to go through it again.



MRI Imaging of Syracuse

Suite 125
550 Harrison Center
550 Harrison Street
Syracuse, N.Y. 13202
(315) 476-7711

January 16, 1986

Dr. Herbert Lourie
725 Irving Ave. Suite 504
Syracuse, New York 13210

RE: BARBARA TEFFT
00272

Dear Dr. Lourie:

Your patient, Barbara Tefft, had a magnetic imaging study of the entire spine performed on the Picker 0.5 Tesla superconductive magnetic resonance imaging unit. Sagittal images were obtained utilizing both T1 and T2 imaging sequences.

The examination demonstrates a normal cervical and thoracic spinal cord. The termination of the cord however is low at the mid T3 level where there is a large posterior mass which extends from L3 to the upper S1 level into the soft tissues posteriorly. There is posterior dysraphism with lack of elements at L3, L4 and L5. The mass itself measures approximately 5 by 4 centimeters. The intensity on both T1 and T2 images is predominately that of fat. It is however inhomogeneous with less intense regions which may be related to fat necrosis, prior hemorrhage, contained neural elements or even calcifications. If calcifications, this could be dermoid instead of simple lipoma. Overlying the mass but separated from it by a well defined tissue plane is prominence of the fatty tissues over the back throughout the lower lumbar and sacral regions. The spinal canal itself is capacious through the thoracic spine but particularly through the lumbar and sacral spine inkeeping with the myelodysplasia.

IMPRESSION: There is tethering of the cord posteriorly down to the mid L3 level where there is a predominantly fatty mass as described. Central areas of lower intensity suggesting dermoid or a teratoma as the etiology of this mass. There are no abnormalities of cervical or more superior thoracic cord identified. There is no evidence for a diastematomyelia.

Thank you for the courtesy of this referral.

Sincerely,

George Petro, M.D.

LOURIE, STEWART AND SHENDE, M.D.'s, P.C.
SUITE 304, 725 IRVING AVENUE
SYRACUSE, NEW YORK 13210

ADULT and PEDIATRIC NEUROLOGICAL SURGERY

(315) 470-7961

HERBERT LOURIE, M.D.
DONALD H. STEWART, Jr., M.D.
MICHAEL C. SHENDE, M.D.

Herbert Lourie, M. D.

Office Visit 3/11/86

Patient: Barbara Tefft

Barbara was here both for a consultation on her thesis and for medical follow-up. Her condition is unchanged. I reviewed with her the symptoms in her hands and they are not typical of carpal tunnel or ulnar neuropathy.

On exam, hands appear normal to motor and sensory testing. We had a long discussion on pros and cons of spine surgery. Her personal condition is such that she cannot make a decision if she wants the operation and, if so, when. She is to stay in contact with me.

Herbert Lourie, M. D.

ml

After all the neurological examination results were explained to me and after I had read volumes of medical textbooks and journals, I decided to try a more conservative approach to relieving the symptoms I had been experiencing. Medical opinion was in general agreement that some sort of pressure and/or traction on my spinal cord was the cause. Since I admittedly had gained weight during college and graduate school, I wanted to see if losing weight would relieve pressure from the cord by reducing the tumor.

My biggest obstacles to losing weight were lack of aerobic activity and living at my parents' house. I started a regular exercise routine of sit-ups, leg lifts, etc. for an hour every morning. Unfortunately, the plank floor boards in the 175-year-old house would creak and groan with every sit-up. Since my mother did not get up until after noon, the complaining floor boards would elicit even more complaints from my mother. I was also compelled to partake of the meat and potatoes meals that were served. Living at my parents' house meant living as my parents' child under the same conditions I had experienced in childhood.

At twenty-five years of age, I knew it was time to get out. The question was, how? I had had several job interviews but no offers as of yet. The most common reason offered in the rejection letters was that the employer was looking for someone with "professional experience". I was still considered a "dependent" under my father's medical insurance as long as I lived at the same address. Once I left I would no longer be covered. The only income I had was approximately \$325 each month from Social Security Disability.

After contacting several agencies and organizations, I learned that I was eligible for subsidized housing under the Department of Housing and Urban Development's Section 8 Rental Assistance Program. The Chenango Housing Improvement Program had an apartment complex consisting of sixteen apartments in the Village of Oxford. These apartments were specifically targeted for financially eligible elderly and handicapped tenants. The problem was that all the units were occupied and there was a waiting list. Tenants were usually long term and vacancies occurred when a tenant either had to move to a nursing home or died. Thus began my initiation to life on public assistance. I checked the obituaries regularly to see if any "vacancies" had occurred.

When my mother learned of my intention to move to a place of my own she expressed her objections in her characteristically adamant manner. She would rather have me live at home, forever in disagreement, than to have me live my own life. Her objections appeared even more irrational considering the fact that I would be living merely three miles away.

Fortunately, I didn't have to wait for an obituary announcement before being notified of a vacancy. An eviction left a specially designed handicapped unit available. It was a one bedroom apartment with an open kitchen/dining/living room. The bathroom was wheelchair accessible.

I moved in the spring with no help or encouragement from my parents. Brenda and her husband David took a load of furniture in their truck and I managed to get Cindy to help me with some smaller things. I spent what little money I had saved on a waterbed, which proved to be a great investment as far as my unconventional body was concerned. For the first time I was able to sleep on my back. The mattress conformed and relieved the pressures that a traditional mattress caused. I was no longer awakened by tingling limbs or pressure on the pillow-like lipoma on my lower back.

Cindy continued to attend the County Mental Health Clinic's Day Treatment Center during the week. Her behavior had moderated since she had been placed on medication. She was taking an antipsychotic drug with the trade name Navane. Antipsychotic medications, as with many other types of medications, can cause side effects. In my sister's case the Navane caused her eyes to roll upward. To counteract this side effect Cindy was given an anticholinergic, Cogentin.

I was only able to learn second hand, through my parents, about the treatments prescribed for my sister. My father just dropped Cindy off at the day treatment center and my mother refused to become actively involved in her therapy. My parents were told that Cindy would probably need to continue taking the medication for the rest of her life. To my mother medicine was something you took to get well and, once recovered, stopped taking. She frequently expressed her opinion to us that Cindy ought to get off the medication. When I asked what the pills were for and how they worked, neither of my parents knew.

I decided it was time someone in the family knew what was wrong with my sister. I learned the date of her next appointment for a prescription review with her psychiatrist and drove two-and-a-half hours from Rochester to Norwich to accompany Cindy to her appointment.

My father dropped Cindy off as usual, only this time I was waiting in the parking lot. I explained to her that we needed to know more about what was causing her problems if we were expected to be able to help. Her reply to me was, "I knew you were here to ruin everything. You're all against me."

I was told by the staff at the Mental Health Clinic that I could not accompany Cindy into the psychiatrist's office or discuss her case with the doctor without her permission because of patient confidentiality. I convinced her to agree to another meeting, this time with her counselor from the day treatment center and myself included. For the first time since my sister had been diagnosed with schizophrenia five years earlier, did I begin to learn the fundamental aspects of what the term encompassed. Cindy's counselor recommended a book by Dr. E. Fuller Torrey, entitled Surviving Schizophrenia, A Family Manual. This book described my sister's behavior

and explained what modern medicine had recently learned about her disease.

Schizophrenia is a biological disease of the brain. The limbic system and its connections are primarily affected. Schizophrenia often runs in families and the brain damage may occur in prenatal life. Diagnosis can often be aided by a CT or MRI scan which may demonstrate enlargement of the ventricles of the brain, atrophy or necrosis of brain tissue and abnormal symmetry of the brain. Research has shown that the brains of those with schizophrenia are pathologically dissimilar to those without. Because of the indescribably complex biochemical processes carried out in the brain it should not be difficult to accept the possibility of malfunction.

Schizophrenia is not multiple-personality disorder. Persons suffering from schizophrenia may experience thinking disorders, delusions and auditory or other hallucinations. The limbic system, located at the deep base of the brain, performs complex selective, integrative and unifying functions. All incoming stimuli to the brain must pass through the limbic system where it is normally organized into a functional reality. Abnormalities in this area of the brain are known to cause profound changes in emotion, inappropriate

behavior, distortions of perception, illusions, feelings of depersonalization, paranoia and catatonia.

Schizophrenia is an autobiologically induced disease, just as epilepsy and multiple sclerosis are known to be. Similarly, the symptoms of these diseases can often be controlled by prescription drugs, but as of yet there is no cure.

The prognosis for each case of schizophrenia can frequently be predicted through the evaluation of a number of factors. The length or suddenness of symptom onset is an important indicator. A sudden, acute onset tends to indicate a more favorable chance for complete recovery, whereas a prolonged, gradual onset followed by acute symptoms indicates a poor prognosis for recovery. Social adjustment prior to the onset of acute illness also may provide clues to outcome. Those who have been described as withdrawn, delinquent or strange children are more likely to fall into the poor outcome category. The presence of subtle "negative" symptoms such as flattening of emotions, poverty of thoughts, apathy, ritualistic obsessive compulsive behavior and social withdrawal are also indicative of a poor prognosis. Most cases of schizophrenia are diagnosed between the ages of 15 and 30, with a more favorable outcome observed in the older segment of this suffering population.⁵⁵

During the consultation with Cindy, her counselor and psychiatrist I learned from my sister how she felt and from her doctor the course of treatment and prognosis. The medication Cindy was taking was an attempt to counteract the chemical imbalance in her brain that caused her symptoms. She would need external discipline and guidance in decision making. Cindy's prognosis for a complete recovery was not favorable based on descriptions of her behavior from childhood, gradual onset of symptoms and the presence of "negative" symptoms.

The aspect of heredity was also a consideration but difficult to ascertain because of the reluctance of my family to discuss any "skeletons in the closet". My mother vaguely recalled her sister Ruth's "nervous breakdown" suffered at the age of sixteen, but diagnosis and treatment of a psychiatric illness in the 1920's was generally Freudian rather than scientific. Ruth apparently made a functional recovery, worked as a department store clerk and was very religious until her death during heart surgery at the age of 40.

When my mother asked what was discussed at our conference I handed her Dr. Torrey's book and told both of my parents to read it cover to cover. My father refused to read more than the first chapter because the

book was in no way entertaining. My mother was surprised to learn that "bad parenting" was no longer considered to be the cause of my sister's problems and that so many other parents had experienced a similar situation. She now encouraged Cindy to take her medication and participate in the day treatment program.

The compassion, discipline and understanding that my sister desperately needed had not existed in our family in recent memory. Although we now knew more about my sister's disease, the ingrained patterns of family relationships were virtually impossible to change. I had begun to fill my unmet emotional needs outside my family circle as a child; my sister attempted to do the same as an adult.

Cindy became infatuated with a fellow client at the day treatment center who was several years her junior. Mike had the long-haired, earringed, rock-n-roll look Cindy seemed to find appealing. She followed him like a puppy obeying his every command. When Mike told her to move in with him, she obeyed. Cindy announced her plans to my parents, who both voiced their objections. Being of legal age and possessing a new-found assertiveness she made it clear that nothing could stop her. As usual, my father gave in and helped her move her things to Mike's

tiny, run-down studio apartment. My father's peace and quiet was temporarily restored once again.

After reading Dr. Torrey's book I located a support group for friends and families of the mentally ill in Syracuse. Sharing and hearing how having a mentally ill person in the family affected other members, and learning how to use the mental health system for help, provided both frustration and insight about my sister's future. I joined the National Alliance for the Mentally Ill and the National Organization on Disability. I began to write numerous letters to government officials on the need for vast improvement in our nation's health care system.

Since I had moved out of my parents' house I had been indoctrinated to the systems of Social Security, Medicare, Medicaid, Food Stamps and public housing. The laws which govern eligibility, access to services and even how subsidies can be spent, are so complex and restrictive that the recipients of these so-called "benefits" are virtually forced to remain in poverty or lose financial support. There is also a tremendous discrepancy in the financial quality of life between those who acquire a disability and those who begin life with a disability. The origin of a physical disability frequently determines liability and the source of financial support. A victim of an automobile accident who suffers spinal cord injury, and is symptomatically very much like a person who was born with a myelomeningocele, will most likely receive insurance compensation for medical expenses, living expenses and lost wages as well as being considered for government funding. Those who can afford the best legal assistance will most likely receive the maximum financial settlement to provide a comfortable lifestyle.

Severe birth defects often rule out the possibility of employment for physical reasons. Even when suitable employment is physically possible, attitudinal and economic barriers may prevent financial independence. Unless parents are able to take some form of legal action and prove external liability, as in the case of thalidomide affected children, those who are born with a disability must rely on families and government funding exclusively.

Social Security Disability Insurance provides funding with the monthly amount based on the work record of the recipient or, if age twenty-one or under, the record of the parent if that parent is retired. If a physical or mental disability develops after the age of twenty-one or before twenty-one and the parent is not yet retired, Social Security Insurance pays a monthly amount based on the recipient's income and assets or the family's income and assets.

I was eligible for SSDI by virtue of having a disability from birth and a retired parent. When I moved out of my parents' house I also became eligible for SSI. The "needs test" for SSI counts SSDI as income and counts bank accounts and personal property, such as automobiles, as assets, thereby making the contribution of SSI minimal to non-existent. Cindy, after my parents provided

substantial documentation to prove that she suffered from a severe mental disability, was eligible for SSI exclusively.

Eligibility for SSDI automatically qualified me for Medicare insurance. Medicare pays medical suppliers and physicians a portion of the billed fee directly. Medicare is most commonly associated with Social Security Retirement benefits for the elderly. The physician or supplier must agree to accept this amount as payment and/or bill the patient or a supplemental insurer for the remainder. SSI eligibility made me eligible to apply for Medicaid which is also a needs based program. Medicaid will pay a portion of the portion not paid by Medicare when separately billed by the provider. Food Stamps are another benefit of SSI that are based on need. Mathematical formulas are used to determine the amounts paid to recipients. Any changes in income, living situation, marriage, assets or gifts are supposed to be reported immediately. This includes reporting gifts of food, clothing or cash and inheritances, so that payments to the recipient can be reduced accordingly. Determinations of overpayment can affect the amount of checks in the future. Employment will also affect amount and eligibility in SSI and associated programs. Of course, there are voluminous forms to fill out during the

eligibility process and even more after eligibility has been determined.

Virtually everyone who is determined to be eligible for SSDI or SSI will require a substantial amount of medical or psychiatric care, myself and my sister included. I quickly found Medicaid to be a questionable benefit at best. Many physicians refuse to accept Medicaid patients because of the length of time it takes to receive a payment that is often less than half of the usual fee charged. After paying office personnel to fill out the complicated, time consuming paperwork, profitability is greatly reduced. Medicaid will only cover the most basic or generic in prescribed medical necessities. I was very glad I had obtained my wheelchair while I was still covered under my father's private insurance. Under Medicaid I would most likely be using an awkward, heavy hospital type chair which would definitely limit my independent mobility. Dr. Peale assured me that although it was not his policy to accept Medicaid patients, I could always count on him for my orthopedic needs, and as a friend.

The complexity of the laws governing Social Security and associated programs creates the potential for tremendous error in administration and payment of benefits. Keeping track of payments, learning of

entitlement for various programs and managing daily finances becomes time consuming and confusing, especially for someone suffering from a mental illness.

Because virtually all of the government programs have some type of financial needs test, recipients are discouraged from saving money or owning property such as a house or automobile. Preparing for a future without government financial assistance becomes a challenging game of strategy which requires perseverance and a certain degree of intellectual acumen. So-called "incentive" programs such as "Plan for Achieving Self-Support", designed to encourage independent financial lives, require extremely high motivation and ability on the part of the recipient. Striving for an ordinary life "just like everyone else" is not financially satisfactory. Severe physical or mental impairment restricts the amount of energy and time that can be devoted to an income producing job. Someone who is able to work a steady part-time job runs the risk of earning an income in excess of the limits set for eligibility in a government program, while not earning enough to be entirely self-supporting. Those who manage to replace subsidized income with earned income often barely meet ordinary living expenses, while risking their health to survive financially. The opportunity to save for things

many people take for granted, such as a home or car, is virtually non-existent.

Ironically, in many instances, a specially equipped automobile or accessibly designed housing would allow more time and energy to be put toward earning self-supporting income. Someone who must use a wheelchair, living in a privately managed second floor apartment because of affordability, must be extremely motivated to be willing to crawl up and down stairs to attend a minimum wage job that provides no medical or other benefits. Accessible public housing is rarely available and also has restrictive income eligibility limits.

Another irony is the consequence of receiving a substantial gift or inheritance from a well meaning friend or relative. The intention to relieve some of the burdens of a physically or mentally handicapped person may actually cause more financial harm than good. I was alerted to this fact while researching estate planning in an attempt to inspire my parents to have their own wills drawn.

From the time I entered high school I was very much aware that my parents were a generation older than the parents of most of my peers. Their age and my mother's threats of disinheritance (aimed at discouraging my association with the neighboring Rogers family) prompted me to wonder what would happen to the house that had been so laboriously restored. It did not take long to learn that in cases of intestacy the state has sole power in the distribution of an estate. Everything would be sold and divided equally between the closest surviving relatives, after the state took its share in fees and taxes.

For both my sister and I this action would jeopardize our eligibility for the government programs we depended upon. Our inheritance would not be enough to invest for interest income that would cover our combined living and medical expenses. The principal of our inheritance would have to be spent down until we were impoverished enough to reapply for the benefits we were now receiving. Considering the cost of medical care, the inheritance would last less than one year.

My mother's primary concern appeared to be making sure that none of the Rogers family ever stepped foot on the property while the estate was preserved intact, down to the arrangement of the china in the corner cupboard. My concern in the matter was in not losing my accessible apartment, medical care, or the option to keep the house I grew up in, and possibly my only opportunity for a place of my own in the future. For a number of reasons, my parents stubbornly avoided the issue of estate planning and mention of the topic only produced arguments and excuses. Since this was an issue that could impact my life in a detrimental manner, I decided to research all the options thoroughly and present them to my parents in hopes of providing an incentive for action.

It took several months to learn the effects of inheritance and property ownership on Social Security. I also researched the option of living and posthumous trusts and life insurances. Early in my research I learned that anyone who anticipates giving any gifts or inheritances to a person likely to depend on Social Security as their primary source of income must become familiar with trusts. Outright gifts will be considered as assets and will be used to cover basic living or medical expenses until funds are exhausted. If the intent of the donor is to provide for activities or

personal extras that are very much restricted by the receipt of government subsidies, a trust must be designed using specific terminology that forbids any payment of interest or principal for expenses that would otherwise be covered by government benefits. At the present time such trusts are legal and if the language provides an uninterpretable content the law will uphold the terms of the trust. Laws are constantly in a state of evolution, however, and there are no guarantees that in the future these laws will not change.

After providing my parents with volumes on estate planning and explaining the consequences that intestacy would have on the surviving spouse and offspring and how the estate would be dispersed, I hoped to inspire my parents to act. All of my work appeared to be in vain. I was thanked for my efforts and the information was added to my father's mountainous pile of as yet unread magazines, stacked next to his lawn chair in the back room.

Just after Thanksgiving my mother came down with a cold. The usual viral infection that everyone picks up on occasion can be miserable; in my mother's case it was nearly fatal. Although she had somehow managed to quit smoking "cold-turkey" a few years earlier, the damage had already been done. When she could no longer get out of bed I insisted that my father take her to a doctor. She refused to go anywhere until she had taken a bath and washed her hair, a process which took several hours and much moaning with her extremely labored breathing. They finally left for the hospital emergency room at 7:30 on a Sunday night. After waiting two hours to see a physician and have chest X-rays taken, she was told that it was just a cold. Drink fluids and get some rest was the advice she received.

My mother's condition continued to deteriorate over the next couple of days. I was infinitely dissatisfied with the diagnosis made at the emergency room so I contacted the local American Medical Association to locate a pulmonary specialist. When I called Dr. Richard Baron's office in Binghamton and explained the situation, an appointment was arranged for early the following

morning. I had to tell my mother that the hospital had referred her case in order to elicit her cooperation. She was still the parent and it was not my place to take such action as far as she was concerned.

I obtained her chest X-ray and report from the emergency room visit and accompanied my parents on the forty mile ride to Dr. Baron's office. My mother was shown to an examining room while the doctor reviewed her X-rays. I was called in to answer Dr. Baron's questions. He was appalled that the X-rays had been taken so recently and that treatment was not offered immediately upon examination. The X-rays showed my mother's lungs as solid black shapes. For the first time since she had given birth to me twenty-five years earlier, my mother was seeing a doctor for a personal examination. She was admitted to the hospital at once for stress tests, medication and oxygen therapy. At the time she was admitted my mother weighed eighty-seven pounds.

Every Christmas Eve the Marshman farm hosted its annual barn party during evening milking, complete with drinks and hors d'oeuvres. As was the tradition, guests would bring a homemade specialty to share. With my mother in the hospital and my sister and I both having left the house, I decided to rekindle the Christmas

spirit at home with my father. Since Dad and I were the only ones in our family that ever attended the barn party, I acquired my father's assistance in making a scale replica of Marshman's barn (complete with five silos and one hundred cows) entirely of gingerbread. By the time it was assembled this edible barn barely fit on the half sheet of foil-covered plywood my father had cut to contain the structure.

My father and I worked on baking and icing this project between trips to visit my mother in the hospital. The day after my mother was admitted we took my sister down to visit. Cindy's first question upon entering the hospital room was, "You're not going to die, are you?"

Final touches were put on the gingerbread barn Christmas Eve Day, just before my mother was released from the hospital. She would be on medication and oxygen for the rest of her life. After my mother had inspected the festive structure, it was delivered to the barn to await the hungry team of demolition specialists. It became my responsibility to initiate the destruction of this masterpiece confection in order to free the red and green cows trapped within. Once the milkhouse roof was destroyed, silos, walls, doors and cows disappeared rapidly.

During one of my many organizational meetings, I was approached by a visiting committee member from another chapter of the National Organization on Disability. I was given information on a fellowship program in the state capitol to encourage women in government. An offer of recommendation was extended if I would be interested in applying. The Fellowship for Women in Government would offer a stipend for 18 months in Albany working with a government representative. The opportunity to learn how to institute change from within the system was very appealing. I had learned from personal experience and from having served on several committees just how inadequate our political-legal system could be. I filled out the application for the fellowship, answered the essay questions and obtained glowing references from former professors, physicians and even politicians I had met in passing at various organizational functions.

To my surprise I was selected to participate in the interview stage, before final selection of the fellows could be determined. My father accompanied me on the trip to Albany for the interview, after which we toured

Rockefeller Plaza and the New York State Museum before driving the three hours back to Oxford.

A few months after the interview I received a phone call from the chairperson of the fellowship selection committee. I was congratulated for my acceptance as an alternate fellow, should one of the fellowship positions become vacant. Of course it had been a difficult decision as all of the candidates who had been selected to be interviewed were highly qualified. All of the candidates, except for myself, were political science or law students. I should be honored that as an illustrator my selection as an alternate had been based solely on my experience as an advocate and committee member. I was asked to please reapply for the following year and it was suggested that I take a few "poli-sci" classes at my local S.U.N.Y.

This response was entirely opposite from the replies I had been receiving from prospective employers after job interviews. As far as my career as an illustrator was concerned I was suitably educated but underexperienced. I went to our humble local library and checked out Catch 22, which I had not read since junior high, to rediscover the humor in life's discouraging situations.

Continuing my daily exercise routine and moving out of my parents' house contributed to my weight loss of fifteen pounds. I noticed a definite improvement in my neurological symptoms, although the numbness in my hands did not disappear entirely. After several telephone conversations with Dr. Lourie, we decided to schedule an appointment early in March for a follow-up neurological evaluation and consultation on my thesis. Discussion about possible dates for surgery would have to wait until I returned from a long-planned trip to Hilton Head, South Carolina with Chrisy and Michelle, my former Keuka roommate and a Keuka friend.

A few days before our anticipated drive to South Carolina, I began the usual vacation preparations of laundry, ironing and packing. I turned on the television news for company while I ironed. The lead story nearly caused me to drop the iron in disbelief. I immediately went to the local drugstore for a newspaper to verify the story in print.

SYRACUSE
Herald American

FRIDAY, MARCH 6, 1957

THE POST-STANDARD

THE POST-STANDARD
 CORTLAND EDITION

DEWITT DOCTOR SLAIN AT HOME

By Jeff Stage,
 John S. Tonello
 and John Doherty
 Staff Writers

A prominent Syracuse surgeon was shot dead and his wife was wounded Thursday by a uniformed man who came calling at the front door of their DeWitt home.

Dr. Herbert Lourie, 57, of 104 Litchfield Drive, died at 8:34 p.m. in the emergency room at University Hospital, where he was taken after the 7:45 p.m. shooting.

His wife, Betty, 58, was reported in stable condition today at Crouse Irving Memorial Hospital, where surgeons removed a .38-caliber bullet from her right forearm.

DeWitt police said the Louries were shot with a handgun wielded by a man who came to the front door in the upper-class neighborhood, rang the doorbell and asked to see Dr. Lourie.

Police were not sure whether Mrs. Lourie was shot intentionally or was struck by a stray bullet.

More than 50 investigators, 45 of them on loan to DeWitt police, spent the night interviewing witnesses, neighbors, friends and colleagues of Dr. Lourie.

"We have not identified an individual as being a suspect," DeWitt Police Capt. Bruce Wahl said this

morning. "We've been talking to a couple of people."

Wahl said investigators also interviewed Mrs. Lourie but declined to discuss information she provided. Also interviewed was a neighbor, Dr. Armand Cincotta, who ran to the Lourie home seconds after the shooting and provided first aid to the victims.

Wahl said both victims were conscious and talked to Cincotta. He also declined to reveal what was said to Cincotta.

The assailant was described as a 5-foot 7-inch white man with a medium build and wearing a uniform.

DeWitt police said the suspect was in his 40s, wore a thin mustache and drove a small vehicle, which was parked in the Lourie driveway as the shooting occurred.

Early reports indicated that the assailant may have been wearing the postal carrier's uniform and may have driven away in a Postal Service Jeep, but Wahl said that has not been "100 percent confirmed."

Wahl said investigators also examined Dr. Lourie's patient records and other documents that may lead police to the assailant.



Dr. Lourie Mrs. Lourie



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"At this point it appears we may be dealing with someone who was disgruntled in some way," said DeWitt Director of Police Larry Lynch. "There appears to be no motive of robbery or any other kind of street crime motive."

A guard was posted outside the door of Mrs. Lourie's hospital room, and Dr. Lourie's partners, Dr. Michael Shende of Turnwood Drive and Dr. Donald H. Stewart of Marville Road, Fayetteville, were given police protection, Lynch said.

Lynch said Lourie's partners also were questioned to try and determine whether they had any clues as to who the killer was.

"There's a guard because we don't know what we are up against," Lynch said. "We don't know where this guy will go next. We don't know if someone had something against just one doctor or the office."

Security also was tightened at Crouse Irving Memorial Hospital, said spokeswoman Melanie Rich.

Lynch said investigators were trying to determine whether any threats were made prior to the shooting.

"Doctors get complaints all the time," Lynch said. "We're playing this as safe as we can."

Syracuse and state police, along with the Onondaga County Sheriff's Department and district attorney's office were assisting in the investigation.

Dr. Lourie, a neurosurgeon, practiced at Crouse Irving Memorial Hospital and University and Veterans Administrations hospitals in Syracuse. He was professor of neurosur-

gery at the SUNY Health Sciences Center, attached to University Hospital, and neurosurgeon for the Syracuse University football team.

Lourie would have celebrated his 58th birthday today. He was one of a handful of neurosurgeons in Syracuse.

As a member of the board of directors of the Medical Liability Mutual Insurance Co., the state's largest malpractice insurance carrier, he was heavily involved in the heat of the debate over medical malpractice insurance.

Gov. Mario Cuomo sought his advice on the issue.

Lourie served more than six years on Syracuse Stage's board of trustees, more than half of that as chairman. His wife, Betty, has been a long-time member of the Syracuse Stage Guild.

Emergency workers from Eastern Ambulance brought Lourie to the State University Hospital emergency room at 8:15 p.m. Police said he was conscious when he was taken from his home, about five miles from the University Hill hospital.

Lourie was a former chief of neurosurgery and president of the medical staff at Crouse Irving Memorial Hospital. The hospital's director of communications, Melanie Rich, said he had been shot three or four times in the abdomen and chest. Rich said Mrs. Lourie was told of her husband's death between 9:30 and 10 p.m., before undergoing emergency surgery on her wounded arm.

Lynch said Mrs. Lourie had answered the door around 7:45 p.m. and greeted a visitor who asked to see Dr. Lourie. When the doctor came to the door, the visitor opened fire, hitting the doctor three times in the chest

and abdomen.

"It was a downward diagonal pattern from the heart," Lynch said.

Mrs. Lourie was hit by one bullet, police said, but they did not know whether the shot was intended for her or was a stray shot intended for her husband.

Police were alerted by a neighbor who reported the shooting as a possible suicide, said Investigator Larry Rice.

The staffs at both hospitals were shaken. "It was very, incredibly sad," said Rich. "He was well-liked by everyone. It was quite devastating to those in the hospital that knew him."

"I don't think we even knew who it was until we were half-way through. People don't look the same in that situation," said State University Hospital Nursing Supervisor Theresa Laporta, who was in the emergency room where doctors tried to save Lourie's life.

The Louries had lived on Litchfield Drive for more than a decade, neighbors said.

The couple has two sons, Gary, 30, of Durham, N.C., and Gerald, 32, of Denver, Colo., and two daughters, Karen, 35, of Philadelphia, Pa., and Suzanne, 28, of San Francisco, Calif., Rich said.

Litchfield Drive is in a neighborhood three-quarters of a mile north of Tecumseh Country Club and one-sixth of a mile south of the Syracuse city line. Of the nine homes listed on Litchfield Drive in the city directory, four are owned by doctors, two by company presidents and one by a corporate vice president.

Staff writer Mike Gormley contributed to this story.

Lourie murder 'beyond belief' to neighbors

By John Tonnello and Mike Gormley Staff Writers

The clogged snowblower lurched and then in an instant it flung a crowbar into the head of Al Robinson.

Robinson was rushed by helicopter to Syracuse, the crowbar protruding a foot from his head.

In surgery at University Hospital, Dr. Herbert Lourie struggled mightily to save Robinson's life. It was the kind of life-and-death work Lourie was called to do often.

Robinson's injury was too severe, and he died Jan. 14 at the age of 40. But Robinson's family remembers the effort of Lourie.

"He was very, very good, a very honest man in dealing with my daughter-in-law and

myself," said Eleanor Robinson of Constableville, Robinson's mother.

She said she had only short dealings with the doctor, but the news of his killing struck her as "a bad shock," she said.

Around Syracuse hospitals, among neighbors, in the theaters Lourie helped to support, the shock was similar.

"He was a lovely man and a good friend," said James Clark, Syracuse Stage's managing director. "We'll all feel his loss. It's a shock. He had a wonderful style. He was friendly and funny."

"He was a marvelous person with a kind of Southern gentility with a smile and a twinkle in his eye," said Frank Funk, president of Syracuse Stage's Board of Trustees. "And he was a very soft-spoken person. But underneath he was a brilliant neurosurgeon who was also doing quite a bit for his community."

Lourie was a member of many prestigious national medical societies and was a dedicated teacher to neurosurgery residents and medical students.

Lourie was born March 6, 1929, and would have celebrated his 58th birthday today.

He was a native of South Carolina. He completed college in less than two years, and at

the age of 19, graduated magna cum laude from the University of South Carolina.

He earned his medical degree from Duke University School of Medicine in June 1952 and did his internship at there. Before coming to Syracuse in 1960, he completed two residencies at Barnes Hospital in St. Louis, Mo.

Dr. Luciano Modesti, a neurosurgeon at University Hospital, knew Lourie for several years.

"It's an irreparable loss to the community, not only as a physician, but as a man," Modesti said. "I had the greatest respect for him as a friend and a colleague. I am very distraught."

"He was a very smart, very kind man," said Dr. Charles Hodge, a neurosurgeon at University Hospital. "It's certainly a tragedy for the medical community in the city. He was very, very well liked by everyone who knew him. His patients loved him. This is beyond belief."

Lourie was active in fund-raising campaigns which led to the renovation of the old Regent Theater and construction of the John D. Archbold Theatre, 820 E. Genesee St.

'He's the only reason I'm talking right now'

By Geoff Hobson Staff Writer

Jim Rooney's mind, left for dead on the Carrier Dome football field until Herb Lourie brought it back, couldn't stop churning Thursday night.

How could the man whose precision saved Rooney's life three years ago have his own life snuffed out in a matter of mindless seconds?

"What can you say about a guy who saved your life?" asked Rooney as he tried to make sense of the slaying of Lourie, a Syracuse neurosurgeon.

"He's the only reason I'm talking right now," said Rooney, 23, now a step away from the Pentagon as budget analyst for the U.S. Defense Logistics Agency and living in Arlington, Va.

"In fact, I even think he fixed more than what was wrong in my head. I think he made things click more smoothly."

On April 12, 1984, Rooney, then a Syracuse University inside linebacker, collapsed after making a tackle during a spring practice.

Within minutes, SU trainer Don Lowe contacted Lourie, the team's neurosurgeon, and soon Lourie began saving Rooney's life, scraping blood off his brain in a 5½-hour operation at Crouse Irving Memorial Hospital.

"I called (Lourie) at his office, told him the symptoms and he said he'd meet us at the emergency room," Lowe said. "He told me what he thought it was, and he was right on. If he wasn't, Jimmy wouldn't be here now."

Rooney and Lowe were just a few of the members of the SU athletic community who reacted with first shock and then grief Thursday night. Tim Neal, Lowe's assistant, got off the phone with Lourie 4½ hours before he died.

"Just an everyday chit-chat," said Neal, who used to watch his 4-year-old son, Brooks, grab the M&M's Lourie would sneak to him from their adjacent seats at SU basketball games.

"He wanted to talk about spring (football) practice coming up, and he said he had this

Saturday night free so he was also going to stop by the lacrosse game. Then I said, 'I got to run,' and he said, 'I'll talk to you soon' and that was it."

Lourie also wanted to know if he could get tickets for college basketball's Final Four in New Orleans next month. He planned to be in Louisiana then, and besides, tickets seemed to be his only payment when he worked on athletes.

"As far as I know," Neal said, "there was a guy who always put his neck on the line to make tough decisions every day, and all he got out of it was our thanks and a few tickets."

Lourie earned a Domeful of tickets during a busy fall last year. He examined starting strong safety Cooper Gardiner and reserve offensive lineman Ken Jenkins and advised that because of head injuries it would be a good idea if they never played again.

And he observed inside linebacker Tim Pidgeon, the team's leading tackler, during a scary two-week episode in which he suffered what turned out to be migraine headaches.

"He called the house once and was very reassuring," said Joe Pidgeon, Tim's father. "It was after a story in a Syracuse paper came out telling how headaches were killing football players. But he told us that Timmy wasn't going to drop dead on the field."

But it's the Rooney case they still talk about. How Rooney made a hit on a third-down. How he collapsed while the punt team was coming on the field. How he was unconscious and unresponsive and had an irregular heartbeat when they shoved him into the ambulance.

"When I finally had all my mental capacities a few days later, he told me about the whole shebang in common terms," said Rooney, who suffered a hematoma — blood on the brain.

"It wasn't a run-of-the-mill deal. The blood seeped from my arteries into an opening in the skull and he had to go in and wipe the blood off the brain to relieve the pressure. If he didn't know what he was doing, I would have died very quickly."

And Rooney remembers Lourie's soft Southern accent telling him that, no, he couldn't play the game ever again: "He knew sports. That was part of his understanding. He knew how badly I wanted to play."

About eight to 10 months later, Rooney and his parents were eating dinner at The Concord on Erie Boulevard when Lourie saw them and came over to chat.

"That's what I'll always remember about him," Rooney said. "How many people come over from across a crowded dining room? That's how caring he was."

That was the first and only time John Rooney, just as stunned as his son as he spoke from Mahanoy City, Pa., Thursday night, met the man who saved Jim's life. He recalled how strange it was that this talented doctor was thinking of getting out of the business because of the malpractice situation.

"The doctor who worked on Jimmy's been shot and killed," John told his wife, Eleanor, as he heard the news over the phone. "My God, He'll be in our prayers tonight, that's for sure."

Even the people in the sports corner of Lourie's life were aware of his preoccupation with the malpractice question. SU lacrosse coach Roy Simmons Jr., who claims Lourie "put me back together" after he injured discs in his back, remembers seeing a chart in Lourie's office that reflected the financial ramifications facing a neurosurgeon.

"I know he felt this area would lose doctors if they felt they could get a better rate some place else," Simmons said.

But you wouldn't catch Simmons filing a suit. Not when his nurses would come to his bed, check his chart and smile at him. "You've got no problem at all. You've got Dr. Lourie."

Lowe frequently brought in Lourie to operate on SU's sports medicine staff with lectures. Lowe, a chew-chewing straight shooter from the Midwest, appreciated Lourie's gentle common sense.

"He'd tell you what you as trainers needed to know on the field with head and neck injuries," Lowe said. "He didn't go into what he as a neurosurgeon had to do. We have people come and speak and then we have other people come and speak. When he spoke, we talked about it two and three days after."

But Lourie will be with Rooney a lot longer than that.

"What more good can you say about a guy who's responsible for saving your life?" Rooney could only ask again.



ROONEY

Shooting kills neurosurgeon, injures wife

DEWITT (AP) — Police officials say they have a strong suspect in the shooting death Thursday night of a prominent Syracuse neurosurgeon and hope to make an arrest today.

Police said a gunman opened fire Thursday night on the front doorstep of Dr. Herbert Lourie's home in a wealthy neighborhood of the Syracuse suburb of DeWitt, killing him and wounding his wife.

Police said the doctor was shot three times, at least once in the chest, and his wife, Betty, was shot once in the right forearm by a man who came to their door about 7:40 p.m. The killer then left an envelope addressed to the doctor and fled.

Lourie, who would have turned 58 today, was taken to University Hospital and pronounced dead on arrival at 8:30 p.m. His wife was in good condition at Crouse Irving Memorial Hospital early today. Doctors operated late Thursday to repair two damaged bones in her right forearm.

DeWitt Police Captain Bruce Wahl described the suspect as a white male, about 5-foot-6 or 5-foot-7 with a medium build. Initial police reports said the suspect was wearing a postal uniform, but police said later they were not sure what kind of uniform the gunman wore.

Investigators were reviewing the files of Lourie's patients with CIM President James Mahar and Lourie's secretary early today.

Lourie, chief of neurosurgery at

Crouse Irving, was a highly respected surgeon who was considered one of the best in his field.

DeWitt Police Chief Larry Lynch said a man wearing what appeared to be a uniform rang the Louries' doorbell. When Betty Lourie answered the door, the man asked to speak to "the doctor," Lynch said.

As the doctor approached the door, the man pulled out a handgun and opened fire, Lynch said. Betty Lourie then walked back to the door and was shot in the arm, the chief said.

Police sources said the killer left an envelope addressed to the doctor on the doorstep, then fled in a "box-type" vehicle. The envelope, which had two stamps on it, was inspected by the Onondaga County Sheriff's Department bomb squad about midnight and then opened, the sources said. The envelope contained miscellaneous papers, they said, but would provide no further details. The papers will be examined by evidence technicians.

Investigator Larry Rice said there did not appear to be any scuffle or words exchanged between the doctor and the gunman. Police said shell casings found at the doorstep indicated the murder weapon is an automatic handgun.

Dr. Armand Cincotta, a neighbor, said he rushed to the Louries' home after the shooting and gave both of them first aid. He said both of the victims were conscious and could talk.

"I just tried to get him through his discomfort," Cincotta said. "He was the one severely hurt. I tried to console her."

Cincotta said Lourie was extremely well-liked by neighbors and respected by his colleagues. "This is very, very tragic," he said.

Cincotta declined to say what Lourie or his wife were able to tell him.

As sheriff's deputies and DeWitt police videotaped and photographed both the driveway and front lawn of the home, neighbors gathered in the chilly night air and mourned the loss of a friend.

Dr. Rao Bhaskar Davuluri, an anesthesiologist at CIM, said he worked with Lourie on an operation Thursday morning and described him as a fine neurosurgeon who was good-natured and at ease when he last saw him.

Lourie, a native of South Carolina, was a graduate of Duke University, according to neighbor John Maroney.

The posh neighborhood, located south of the city, is populated mostly by professionals, including at least five doctors, neighbors said.

The Louries resided there for more than 10 years, according to neighbors. They have four children — Karen, 35, of Philadelphia; Gerald, 32, of Denver; Gary, 30, of Durham, N.C.; and Suzanne, 26, of San Francisco.

Dr. Lourie's violent demise came as a complete shock. After having decided to undergo surgery with Dr. Lourie and depending on his consultations for my thesis, I was lost. The trip to Dr. Lourie's southern homeland seemed almost a pilgrimage. Television network news reported updates on the investigation and search for the murderer, as did local Carolina radio and television stations. I was reminded of Dr. Lourie each day of the trip. The gruesome conclusion of the story was not revealed until after we had returned.

Businessman linked to neurosurgeon

SYRACUSE (AP) — A Syracuse businessman who killed his wife and then shot himself to death this weekend has been linked to the slaying last Thursday of a prominent neurosurgeon, authorities said.

Armando Di Nolfi, 58, was described by police as a disgruntled patient of Dr. Herbert Lourie, who was gunned down last week at the front door of his DeWitt home.

Investigators have yet to say officially that Di Nolfi killed Dr. Lourie, but several police sources say they have no doubt.

State police were expected to release a statement today.

Police found a .38 caliber pistol at Di Nolfi's home that they say was used in both the Lourie slaying and the murder-suicide at the Di Nolfi residence on Saturday, said State Police Capt. Joseph Loszynski, who has been heading the probe by 30 state police investigators.

Di Nolfi, the owner of the Onondaga Uniform Co., reportedly was angry about the after-effects of back

surgery performed by the doctor.

A friend of Di Nolfi who sold the uniform company to him in 1962 told the Syracuse Herald-Journal that Di Nolfi harbored a grudge against Dr. Lourie for a back operation in 1985.

The bodies of Di Nolfi, 58, and his wife Pasquelina, 48, were found in their home by a daughter at about 11:30 p.m. Sunday, said Robert Burns, spokesman for the Onondaga County Sheriff's Department.

Both were shot once in the head with a handgun, Burns said.

"It appears as though Mr. Di Nolfi may have shot his wife while she was sleeping and then turned the weapon upon himself," Burns said. "The wife was found in an upstairs bedroom and Mr. Di Nolfi was found in an easy chair in a downstairs family room."

Di Nolfi left no suicide note, Burns said.

Sheriff's Department investigators said they believe the couple had been dead for about 24 hours before their

bodies were discovered.

"They had last been seen Saturday afternoon and were discovered by a daughter when the daughter had been trying to contact them throughout the day but had received no response," Burns said.

Both bodies have been turned over to the county Medical Examiner's office. Autopsies were to be performed today.

Dr. Lourie, 57, was gunned down Thursday evening and died in the emergency room at University Hospital about an hour later.

Investigators said a man wearing a uniform — perhaps that of a postal worker — rang the front doorbell, asked for the doctor, handed over an envelope and began firing.

Di Nolfi's company supplies uniforms to letter carriers, local police agencies, ambulance attendants, firefighters and bus drivers.

Dr. Lourie's wife Betty, 58, was shot once in her right forearm by what investigators believed to have

been a stray bullet that was intended for her husband.

Di Nolfi was a meek man with "some malice in his heart" toward Lourie, a friend and former employer told the Syracuse Herald-Journal.

The friend revealed Di Nolfi had been unable to walk or stand up until two months ago following surgery performed on his back by Lourie in November 1985.

Frank Ballard described the man he had known since Di Nolfi moved to the United States from Montreal to work at Ballard Uniform Co. in the 1950s. Ballard said the company told Di Nolfi in 1962.

"He was a very meek individual, not capable of what happened," Ballard said. "Up until two months ago, he couldn't walk or stand up."

"I guess it's typical of anyone if something doesn't work out," Ballard added. "He did have some malice in his heart. He did express it on an occasion or two."

When I returned from the Hilton Head trip I learned of my sister's latest plans to prove that she was an independent adult. She was getting married to Mike and nothing anyone said could stop her. None of the potential in-laws were in favor of this union for many well-founded reasons. Ever since Cindy had moved in with Mike there had been problems. She had left their apartment several times to move back in with our parents, citing everything from sexual incompatibility (bordering on abuse) to his excess consumption of alcohol, as reasons to leave. Each time she would succumb to Mike's repentant pleading and return to live with him. Cindy insisted that my parents pay for an elaborate, fancy wedding, which they refused to do.

In the case of my sister and Mike, two mentally ill people trying to deal with the issues of marriage when they had not yet been able to cope as individuals, was a recipe for disaster. To complicate matters, the fact that each of them depended on SSI as their only income was a major consideration when discussing marriage. As two unrelated individuals living in the same household they would each receive a monthly allowance. As a

married couple that amount would be reduced to almost two thirds of what they received before marriage. Under Mike's financial management they had begged to borrow a substantial amount of money from both sets of parents, presumably to pay the rent and to buy food. Cindy had been cashing her checks and handing all of her money to Mike which he used to buy lobster, beer, and stereo equipment.

Cindy continued to remind everyone who opposed the marriage that she was a legal adult and could do as she pleased. She was getting married. My parents eventually were coerced into contributing the money for her wedding dress, which was a floor-length lace and ruffles prom dress with tiny pink hearts all over the skirt. The wedding was held at the American Legion in Oxford. I was asked to be the maid of honor and my father was asked to give the bride away. My mother used her convenient excuse of being tethered to an oxygen tank to avoid attending the ceremony.

Cindy's future mother-in-law assisted in the bridal toilette, dressed her hair and applied the make-up. When she was finished Cindy was led to the mirror. Her astonished remark as she viewed her reflection was, "I look beautiful!" This was the first time I had ever heard my sister make a positive statement about her own

appearance. She spent the entire day gloating over her success at having acquired a mate, while I felt like we were all watching a couple of children play grown-up. Cindy sipped her milk from the nuptial champagne glass during a congratulatory toast offered by Skippy, the best man. After the toast she leaned toward me to ask, "So when are *you* going to get married?"

Another question that had no answer. Even with my limited experience, I had encountered nearly every type of matchmaking and pick-up line imaginable. I had been on blind dates, obligatory family acquaintance dates, bar dates, pity dates and even a few sincere dates. I had had crushes and had been the subject of several undesired crushes from kindergarten through graduate school, which included all of the silly, manipulative mating rituals we initiate and endure. The problem was that I allowed no one to break through that impenetrable wall that I had spent a lifetime building. By the time I was twenty-five, my sexual experience consisted of occasional encounters with a very dear, long-time friend who was able to overlook the physical abnormalities that contributed to my self-consciousness. My sister's inquiry made me more aware of why I had not yet allowed a relationship to pass a certain point in its natural

development. I would either have to learn to accept myself as I was or change what I could, physically as well as emotionally.

Dr. Lourie's untimely demise had cancelled the tentative plans for surgery to remove the lipoma on my lower spine. Reaching the decision to have surgery had been a difficult process. Being able to trust Dr. Lourie's abilities and knowing of his personal interest in my case had confirmed my decision. If I still wanted to have the surgery I would have to find another surgeon I could trust.

I contacted Dr. Lourie's colleague, Dr. Joseph McDonald, at Strong Memorial Hospital in Rochester, to see if he would be interested in performing the operation. Dr. McDonald reviewed the evaluation he had done as a second opinion for Dr. Lourie and asked me to come in for a re-examination. I travelled to Rochester for my appointment with Dr. McDonald and stayed with my friends, Drs. Marshman and Franzen. Dr. McDonald agreed with the decision to remove the lipoma, and since my losing weight had actually lessened the neurological symptoms, decided against the more risky aspect of untethering the spinal cord. Surgery was scheduled for the end of July.



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ROCHESTER, NEW YORK 14642
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SCHOOL OF MEDICINE AND DENTISTRY • SCHOOL OF NURSING
STRONG MEMORIAL HOSPITAL

Division of Neurological Surgery
Joseph V. McDonald, M.D. (275-5255)
Shige Okawara, M.D. (275-5709)
Curtis N. Nelson, M.D. (275-5993)

June 12, 1987

OFFICE NOTE

RE: Barbara Tefft

Barbara Tefft was seen again on June 11. Her lipomatous lumbar mass is uncomfortable when she sits or drives her car. Hand and arm function is better. I don't think there is any indication for untethering of the cord now, but the lipomatous mass could be removed superficially down to the spinal level. Films will be reviewed and then she will be admitted, will be seen by plastic surgery and then have the removal of the tumor.



Joseph V. McDonald, M. D.

JVM: sm

As soon as I returned to Kay and Ann's from my appointment with Dr. McDonald, I received a phone call from my mother. I immediately knew something was wrong as this was the first time I had ever been on the receiving end of a phone call from home. My father was in the hospital with a strangulated inguinal hernia. He had acquired the hernia shortly after an appendectomy, from carrying heavy water tanks while fighting a forest fire in the Catskills during the 1950's. Although it had bothered him intermittently in the past, he refused to have the simple corrective procedure until it became a life threatening problem.

He had driven himself to the hospital that noon, after spending the morning carrying in firewood and taking care of the horse. He sat doubled over, insisting that he felt better, until my mother convinced him to see a doctor. He was admitted immediately but the surgery would have to wait until he had stabilized. I told my mother over the phone that I would be home in a few hours, left a note for my hosts and headed for Oxford.

I arrived at the hospital around 11 p.m. and stopped in to check on my father. He was lying on his side in a

fetal position pretending he wasn't in pain. Not at all the big, strong man I knew as my father.

When I arrived at home my mother told me how she had to feed the horse even though I had said I would do it when I arrived. She could barely lift the ten pound bucket and had to come back inside for oxygen twice before she could walk the sixty feet to the barn.

Dad spent just over a week in the hospital and was warned against any strenuous activity for several months after surgery. My parents were very stubborn about hiring help with yard work, firewood, and other strenuous chores but eventually saw the necessity for assistance. I found their reluctance in this matter very frustrating. People in our neighborhood had always helped each other and trading favors was a common neighborly courtesy for everyone, except my parents. If the possibility of becoming indebted for any reason existed, that possibility alone was reason enough to do without.

Accepting and requesting assistance had always been a fact of life for me. From the time I first acquired my crutches I had learned to accept that I needed help with simple tasks such as carrying books or picking dropped articles up from the floor. Most people who offer to help or are asked for assistance seem to derive at least some small pleasure from the act and I have never been

made to feel indebted even when I could offer no more than a thank you. The friends I have made from simply accepting offers of assistance have greatly enriched my life. These friends have enabled me to participate in activities that otherwise would have been unavailable to me and meeting new people through them provides a continuing source of mutual pleasure.

For me, incidents of aggressively offered assistance or risks of indebtedness have been relatively few. On occasion, I have had things forcibly removed from my hands after declining an offer of help or, when obviously struggling with an unwieldy load, have been stared at and ignored. Children have been told by parents not to "bother the lady with the crutches" and to get out of my way when they offer to help. If I had allowed myself to be offended by the actions of those who, for the most part, were trying to be helpful, I would probably never venture out in public again.

After my father had recovered from his surgery and begun his convalescence at home, I returned to stay at Kay and Ann's while more adjustments and repairs were made on my braces. My psychologist friend always lent an ear and support when I needed it and both offered encouragement in my academic and professional efforts.

One Sunday, I decided on a spur-of-the-moment trip to Penn Yan while Kay and Ann were attending to the details of their recently accepted purchase offer on a house.

I had not been to visit my Keuka College friends in the past two years, although I had kept in touch by mail. My first stop was at my former part-time RIT roommate's house. Kathy D'Abbracci, now Mrs. Gernold, was sunning herself in the backyard when I arrived. Without opening her eyes or turning over to see who was approaching from behind she said, "Hi, Barb! Where have you been, stranger?!" Kathy's uncanny ESP warned her that I might show up.

After visiting with Kathy I decided to drive to the opposite side of the lake to see if one of my former art instructors was at home. When I turned into the Benedict's driveway, the number of cars indicated an event. Dexter's wife, Faith, was hosting her annual spring pottery open house in the studio. I walked in to find virtually everyone I had known at Keuka in attendance and received an enthusiastic welcome. The only person I did not see was photographer, Chris Wright. I had been to see Chris while he was in the hospital for surgery but no one at the open house had seen him recently, except Richard Sprentall, a carpenter who had been working on all three of my former art instructors'

houses. Richard offered to drive me out to the bluff to visit Chris after the open house.

I had met Richard briefly several times before while he was working on Wright's house and visiting Chris in the hospital. On the long ride to Wright's I learned that Richard was about to start on an addition at Benedict's and had been working on Jurysta's cottage. What I didn't learn until later was what an important part of my life Richard would become. After my surprise visit to Wrights, Richard took me to Lloyds, my favorite pub, for pizza and wings. We talked until ten o'clock that night, when he took me to Benedicts to pick up my car. The affectionate hug he gave me when we said good night was the beginning of a lasting relationship.

I returned to Penn Yan to spend a glorious Fourth of July week with Kathy's family at a cottage on the lake and at Richard's rustic cabin in the woods. I realized just how special this man was when I drove up to the cabin as he was putting the finishing touches on a handrail along the stairs just for me.

As we got to know each other, Richard was never afraid to ask questions or to touch me. He was concerned and interested to know about my medical condition but more interested in getting to know me as a person. Little by little my carefully constructed wall began to

crumble. It was difficult to tell him so early in our relationship that I was having a long awaited surgery at the end of July.

Kay and Ann had offered to have me stay at their new house to recuperate after the surgery and escorted me to the hospital to check in. I was admitted two days before the surgery as a show-and-tell example for medical school interns. Richard helped me escape for ice cream and a late night visit to Kay and Ann's on the first night of my hospital stay. After being introduced to Kay and Ann and seeing their moving preparations, Richard offered to help with their relocation.

The morning of my surgery, Richard arrived at 6 a.m. He brought my stuffed Muppet, Kermit, dressed in a surgical mask and gown that Richard had made himself. Kermit accompanied me to the O.R. tucked under my arm. The surgery was originally estimated to be around two hours in length. When I had not returned to my room four hours later, Richard became more than a little concerned. The nurses finally convinced him to leave the hospital for a little while and assured him I would be there when he returned. When he came back from visiting his mother outside of Rochester, I was enjoying a pizza that the floor nurses had ordered to share. Richard was very relieved.

The surgery had taken more than four hours but was successful. I noticed immediately the lack of bulk as I lay flat on my back. I had to remain in the hospital for two weeks with only a brick wall outside the very green decor of the neurosurgery ward. Fortunately, a steady stream of cards and visitors, including Kay's brother's family from Oxford, provided diversion. Richard drove the hour-and-a-half trip from Penn Yan every day after working on Benedicts addition.

SMH 557

THE UNIVERSITY OF ROCHESTER
STRONG MEMORIAL HOSPITAL

OPERATIVE NOTE

116-59-89

LOCATION	NAME, ADDRESS	AGE	P SP DIV	UNIT NO.
			ANES	General
DATE	7-24-87			
NAME	TEFT, Barbara J.	SURGEON	Joseph V. McDonald, M.D. Webster H. Pilcher, M.D.	

PREOPERATIVE DIAGNOSIS: Lumbar lipomatous mass, 26 years status post removal of myelomeningocele and associated teratomatous tissue in lumbosacral region.

POSTOPERATIVE DIAGNOSIS: Large lipomatous mass located in the subcutaneous tissue plane in the lumbosacral region.

OPERATION: Resection of lipomatous mass of lumbar region.
Excision of lipomatous mass of lumbosacral region.

OPERATION: The patient was taken to the Operating Room and general endotracheal anesthesia was obtained. The patient received preoperative intravenous prophylactic antibiotic therapy. The patient was placed in the prone position on supportive pillows, exposing the lumbosacral region. This area was shaved, prepped and draped in sterile fashion, draping a wide area, so that a generous operative field was available. Using a skin knife, a large curvilinear incision was made superior to the preexisting myelomeningocele scar and the skin incision was carried to the depths of the subcutaneous fat plane. Using pickups with teeth and skin hooks, the skin was gently retracted and using a scalpel blade and occasionally a Cavitron, the subcutaneous tissue plane was entered, with removal of large volumes of fat from this area. The skin was undermined superiorly and inferiorly and the subcutaneous fat deposit was removed, generously, laterally to the lateral border of the iliac crest and superiorly and inferiorly with significant undermining of the skin flap above and below. Hemostasis was intermittently obtained with the Bovie cautery and bipolar coagulation, and the resection of fat was limited to satisfy cosmetic requirements, primarily. Therefore, the spinal canal and the vertebral elements were not visualized or entered. Once a substantial amount of fat was removed laterally, superiorly and inferiorly so that the skin when replaced lay in an anatomically normal position, a sterile marking pen was used to delineate the area of redundant skin along with the preexisting scar, which was to be removed. This redundant skin was resected in a curvilinear fashion, along the superior margin of the wound and at this juncture, the wound was copiously irrigated with Bacitracin solution, hemostasis was vigorously obtained, a subcutaneous fascial absorbable suture layer was placed to take tension off of the wound and the skin edges were apposed with staples.

Postoperatively, a pressure dressing was placed on the wound which included a number of cotton fluffs, and an abdominal binder, and the patient was then extubated and taken to the Recovery Room in satisfactory condition. Estimated blood loss was approximately 500 ccs. The patient was maintained on prophylactic antibiotic therapy postoperatively.

WHIP:14
DD: 9/28/87
DT: 9/30 87
#478

Joseph V. McDonald, M.D.

SURGICAL PATHOLOGY REPORT
THE UNIVERSITY OF ROCHESTER - STRONG MEMORIAL HOSPITAL - ANATOMIC PATHOLOGY LABS
601 ELMWOOD AVENUE
ROCHESTER, NY 14642

NAME McEETL, Barbara 2-2162-87 AGE 66 SEX F UNIT 165-52-89 DATE 7-24-87

DIAGNOSIS:

DR. McDonald

DIV. 5-36

DX. BY JH

CONF. BY TAE

Mature adipose tissue, clinically lipoma.

GROSS: The specimen is received in two containers.

The first specimen is received unfixed, labeled "lumbar lipoma", and consists of 100 cc of yellow adipose tissue. RS, labeled "IA" and "IB".

The second specimen is received unfixed in a cavitron container, labeled "lipomatous lumbar mass", and consists of 500-600 cc of brown fluid in which numerous particles of yellow adipose tissue are suspended. Gross only.

MICRO: Microscopic description omitted.



Thomas A. Eskin, M.D.

SMH-54 7/86

SLIDES EXAMINED

PHYSICIAN'S COPY



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SCHOOL OF MEDICINE AND DENTISTRY · SCHOOL OF NURSING
STRONG MEMORIAL HOSPITAL

Joseph V. McDonald, M.D.
Professor & Chairman
Division of Neurological Surgery

August 5, 1987

DISCHARGE SUMMARY

RE: Barbara Tefft

Barbara Tefft was discharged from the hospital on August 4. She had been admitted for removal of a large lipomatous mass which lay in her lumbar area and was part of a lipomenigocele, present since birth.

The lesion was excised through an elliptical incision.

Postoperatively, the wound healed well, but there was an accumulation of fluid within the very large dead space created by removal of the fat.

On two occasions during the first postoperative week, this was aspirated and 200 cc of fluid were obtained each time.

The fluid collection decreased in volume and a third aspiration yielded only 60 cc. That was on August 2. Her skin sutures were removed on that day. Her wound had healed well. Subcutaneous fluid volume remained small.

she was discharged after having had a revision of her long leg braces and she was up walking in them at the time she went home.

Procedure was excision of lipomenigocele and diagnosis is lipomenigocele.



Joseph V. McDonald, M. D.

JVM:sm

Having the lipoma removed was one of the best decisions I have ever made. I could now buy clothes two sizes smaller than before the surgery and without having to attempt to disguise an abnormal contour. Shopping for a bathing suit was no longer the same dreaded frustration it had been the past 26 years. I could now sit straight in chairs and while driving, and for the first time in my life I could lie comfortably in a supine position on virtually any surface.

The medical events of the past few years, including surgeries, KAFO's, wheelchair, Ditropan to improve bladder control, and self education all contributed to a much greater self-esteem and confidence. My body was still by no means perfect, but knowing that I had taken measures to assure my most optimum functional capabilities gave me a greater sense of control over my life. Feeling better about myself made it much easier to interact with people who were less tolerant or understanding of my differences.

Both my parents and Richard's mother were vaguely aware of our relationship almost from the beginning. I was not introduced to his mother until some time after the surgery. Meeting "the family" can be awkward under the best of circumstances and we were anticipating some

resistance, mostly regarding age discrepancy, Richard being fifteen years my senior. Amazingly, the issue of age was a minor consideration for our relatives and friends.

My introduction to Richard's mother was very polite and civil. We went to her house for dinner after which we were invited to accompany his mother on an errand. I decided to remain at the house rather than ride along, my sixth sense realizing that a mother/son chat was brewing. The inevitable proof that we are always our parents' children, regardless of age, was made apparent to Richard during this errand. His mother's concerns were directed more toward the potential for similarly affected offspring and that Richard knew what he was getting himself into. Richard's attempts at reassurance did little to alleviate her concerns. I knew that this initial meeting was a bit overwhelming for her and that my physical problems would gradually become less of a concern as she got to know me. Within a year we progressed from the formalities of Mrs. Sprentall, to Dorothy, to greeting cards signed "Love, Mom". I am now accepted by his mother as a very positive asset in Richard's life.

My introduction to Richard's mother was less traumatic for me than Richard's introduction to my

parents, for reasons which had nothing to do with Richard. During the introductory supper at my parents' house, Richard was virtually ignored as my parents and I got into a typical loud family discussion. Richard had cleared the table and started washing the dishes before we settled into civility again. A very unceremonious, but typical, welcome to the family!

So much had happened during the past few years that it was difficult to absorb it all. Learning about myself and my sister, seeking help, attending college and graduate school, caring for ill, elderly parents, surgeries, learning the Social Security maze and fighting to maintain benefits, job interview, committee work, the occasional freelance art jobs, and even working in Marshman's milkhouse were the highlights of the past five years. Falling in love and being loved in return was the highlight of my life.

I decided to take a breather from the frustrating activities I had been involved in and spent a winter on Keuka Lake with Richard. All of these new events and adjustments contributed to a very disconcerting period of anxiety attacks which I managed to overcome with the help of my psychologist friend, Ann Marie. I commuted to RIT for a graphic design class in order to improve my portfolio and keep my hand in the arts. Job hunting had proven to be a fruitless effort as far as any offers of full-time employment were concerned. I had received numerous compliments on my work, had nearly always been called back for second interviews and received offers for

freelance work. Reasons for my not being hired as a full-time employee ranged from my not having prior "professional experience" to "you'd be bored here." The complicated process of reporting income to Social Security made the benefit of earned income seem more like a drawback. It began to appear that the Social Security system was an all or nothing proposition.

My sister's diminished financial status following her marriage was a good warning for me. I realized just how much of a financial liability I would be to a spouse. Unless we suddenly became extremely wealthy, marriage was out of the question. I also began to suspect that this liability was a factor in my lack of success in becoming employed.

We left Keuka Lake to spend Christmas at my parents' house. My sister had moved back home a few days earlier, after the police had admitted her husband to a state psychiatric hospital. Cindy was adamant that she would never return to live with him and wanted a divorce, less than eight months after their wedding. Mike was released on New Year's Eve and, as usual, he ordered Cindy to return with him to their apartment. She left the house without even the courtesy of a good-bye.

In the spring, Richard accepted a construction job in Oxford. He rented an apartment in the house that my father had been raised in, not far from my apartment. While looking through the "help wanted" column of the local newspaper, I was surprised to find that the Oxford High School was looking for a long-term substitute to teach art. First thing in the morning, I was speaking to the principal about substitute teaching for my former art teacher while she was out on maternity leave. I was hired immediately.

The excitement of being hired for my first job, however temporary, was tarnished by government bureaucracy. I responsibly notified Social Security and the administrators of my government subsidized accessible apartment. After I had made these agencies aware of my pending increase in income, I received the distressing replies. My temporary employment would render me ineligible for the food stamps and SSI allowance I was now receiving. My income would also be in excess of the allowed monthly ceiling for tenants in the apartment complex where I was now living. I was told that I would have to move out of my accessible quarters.

After much discussion with a variety of agencies, I was allowed to remain in my apartment, provided that I paid the estimated full market rent during the months that I was employed. This meant that the rent would be more than triple the amount I had been paying.

Several unsuccessful attempts to notify the Social Security office by phone of my recent employment yielded no acknowledgement. Because I was teaching during the day, I could not drive the 35 miles to Binghamton to spend a day waiting in line just to make sure they knew of my change in income. I mailed a number of letters and sent copies of my paychecks every two weeks. I had expected the monthly SSI checks to cease or be substantially reduced but the direct deposit amount on my bank statements did not change. Nearly two years after this employment ended, I was notified by Social Security that I had been overpaid during that time and that I had the option of repaying the amount in full or of having my checks substantially reduced until the overpayment was corrected. Fortunately, I had kept copies of every letter and paycheck sent to the Social Security office as proof that they had been notified as required. Eventually, I received a letter from Social Security admitting to their oversight and I was not forced to repay.

I found teaching art to high school students challenging and rewarding. I was responsible for planning the curriculum, teaching and grading six different types of classes. Because of another infected pressure sore on my left foot, I spent most of my time teaching as a tripod, using crutches and one brace. The classroom preparation, clean-up and discipline was exhausting, especially using only one leg. Before school was out for the summer we hosted an art show and reception for Karen and Pat Mills and their new daughter, Lorraine Emma.

While readjusting to being unemployed again, I spent some time figuring out exactly how much I had gained financially from my employment. After Social Security, taxes, my increased rent and elimination of other benefits were taken into consideration I calculated that I had earned approximately \$7.30 more per day than I received on Social Security. If this had been a permanent position I also would have had to leave my accessible apartment for less satisfactory accommodations. If Richard were to move into this apartment the rent would have become his responsibility as the amount was adjusted based on income.

I have yet to discover a working example of the highly touted government incentive programs designed to

make independence more easily attainable. The maze of paperwork, inflexibility and insensitivity of the ever-changing bureaucracy that oversees our "public welfare" programs only serves to discourage even the most sincere attempts to gain independence. The majority of handicapped people who do manage to find an end to the maze may only find employment as advocates and counselors to others with handicaps. Rather than being able to truly overcome the limitations of a disability many spend their lives concentrating on that disability as a means of livelihood. Disabled individuals are as varied in abilities and interests as non-disabled but only those with an intense determination have much hope for even moderate success in our competitive, money conscious society. Achieving the goal of being "ordinary" may actually be a hard-fought dream for some.

After contributing so much time to advocacy oriented committee work, I eventually realized that I really did not want to spend the rest of my life focusing on my limitations. As noble as a career in advocacy may be, I would rather put my interests and acquired skills to work in the field in which I was trained.

Richard and I decided to relocate to a more active urban area and settled on Ithaca when Richard got a job

in a commercial cabinet shop. He began to understand my reluctance in leaving my accessible apartment in Oxford when he attempted to find suitable and affordable housing in the city. After a long and frustrating search, a small apartment on the first floor of a house was rented through Ithaca Neighborhood Housing Services. INHS is a non-profit organization dedicated to rehabilitating run-down neighborhoods and providing affordable housing. The apartment was not designated as accessible since there were seven steps up to the main entrance, and the doorways were barely wide enough to accommodate a wheelchair. The management was willing and open to minor modifications, such as removing an interior door to make the apartment more accessible. After residing in the apartment for a short time I was asked to join the INHS Tenant Review Committee, which oversees the tenant application process and provides grievance services for tenants. The following year I was named Chairperson for this committee.

My hopes for better access to freelance work or even full-time employment in my field was initially based on the proximity to the research being conducted at Cornell University and the small city's strong ties to major metropolitan markets. I've learned that strong potential requires even stronger resolve and patience, like an actor pursuing that big Hollywood break.

AFTERWARD

I am a product of modern technology. The fact that I exist at all is the result of this technology. The potential for birth defects can now be diagnosed even before conception and surgery can be performed in utero. The fierce debates over the ethics of abortion, euthanasia, in vitro fertilization, surrogate parenting, animal research and countless related issues will continue to be fueled by the advances made in medical technology. Are we prepared to accept responsibility for the inevitable mistakes of nature and those that will occur while attempting to perfect technology?

Every year the lives of an increasing number of infants born with severe defects are saved by the heroic measures of physicians armed with knowledge and access to state-of-the-art equipment. Immediately, the financial burden of the care necessary to maintain these lives becomes apparent. Who is to pay the cost? Family resources are bled dry until they are financially eligible to receive limited government assistance from programs that taxpayers accuse of mismanagement and overspending. The already strained institution of the family often dissolves under the added financial and

emotional stress of raising a handicapped child. Who suffers the most in this situation?

These children are raised in a society that places emphasis on financial success as a measure of human worth, while dreams of participating in the activities of this society are often impossible for them to attain. Judgments of a person's ability to contribute to society are based on physical appearance, mental capacity, financial background and career ambitions. These judgments begin from the moment of birth with remarks such as "What beautiful dimples!", and who hasn't asked a toddler what they want to be when they grow up? The condolences offered to my mother by the first person to learn of my defect are indicative of the impressions and expectations our society has for the disabled.

The nature of a disability also shapes public perception and reaction. The "victim" of a car accident who is in a wheelchair appears to be more easily accepted by our society than someone who has had to use a chair since birth. Sympathy is expressed for what the victim has lost, pity is the expression for something deemed never to be. The inquisitive onlooker who is brazen enough to ask "What happened to you?" generally expects to hear a simple answer. "I was hit by a car" is much more comprehensible than "I was born with spina bifida."

Although both are simple, direct statements, everyone instinctively understands the damage that can be inflicted by a car. Few have even heard of spina bifida. For some reason, the results of an act of man seems to draw people together and inspire our society to take action. Repulsion and distance are, unfortunately, common reactions to a defect of nature.

The technical ability that allows us to determine the presence of a defect before birth carries with it an ethical dilemma of the options available. Should the parents of a severely deformed fetus be forced to deliver a child they are not financially or emotionally prepared to raise? Should selective abortion based on the presence of a defect be an option? Following the birth of a defective child, should life sustaining treatment be performed or should the child be allowed to die of natural causes? Are the parents, physicians, judicial system or the public allowed to decide the child's fate? Recent judicial setbacks in upholding the right of a woman to choose abortion will undoubtedly lead to an increase in the number of deformed infants delivered in this country. The case of Baby Jane Doe, a spina bifida baby, whose parents chose not to have surgery performed on their infant daughter, were forced into a public court

battle to defend their informed decision. Is there only one right answer to any of these questions?

All children are expected to grow into adults who will contribute to their society. Children are nurtured, encouraged and educated according to their abilities and the hopes of their parents and the society in which they are raised. Our society invests heavily in the education of its children in hopes of gaining a return on this investment. Is this return only to be measured in financial terms? Do we not provide an education to those who are considered a financial risk or a burden, or do we invest with the goal of achieving personal enrichment?

Once the decision to treat an infant has been made, a tendency to become short-sighted develops. Many organizations exist to aid in the development, education and medical needs of a handicapped child. What is forgotten is that this child will, hopefully, someday become an adult. The goals and needs of a disabled adult are very different from those of children. Desires for independence, of feeling needed, of establishing intimate and sexual relationships and to have a family must be taken into consideration when raising a handicapped child. When family is no longer able or available to care for a disabled adult child, where will that person

live? Who will be responsible for this person's personal and financial needs?

Those of us who are fortunate enough not to need such intensive support may become more handicapped by societal attitudes than from our physical limitations. Employment of the handicapped is often restricted by perceived financial burdens and fear of the unknown. Lowered expectations of the capacity for work also keep many of us, who do have the potential for other than menial tasks, out of the market.

Statistics indicate the presence of a substantial population of physically disabled adults in this country. Yet, how many members of this population do you come in contact with at work? How many people with obvious physical disabilities do you socialize with or encounter on a daily basis? If those who work in the field of health care are discounted, the numbers are few. Where are these adults? Are they leading full and productive lives with every emotional and material satisfaction available to the able-bodied majority? Did you ever wonder?

EPILOGUE

A neat and tidy conclusion is rarely possible in life. Since moving to Ithaca I have continued to seek full-time employment in my field. The process of writing and researching this paper has proven to be cathartic as well as a great source of information and, best of all, new friends. Involvement with horses, gardening, antiques and art adds color and interest to my life. Richard adds the warmth.

Medical aspects that are of constant concern include the prolonged process of fabricating new braces, nursing pressure sores, keeping a wheelchair in working order and anticipating the need for further surgery. The lack of physicians who will accept Medicare and Medicaid patients means that I am always searching for quality health care. Twice, I have experienced the loss of services of excellent physicians who have had to give up private practices. One of these physicians who formerly accepted Medicaid patients was advised by his accountant to "get a real job."

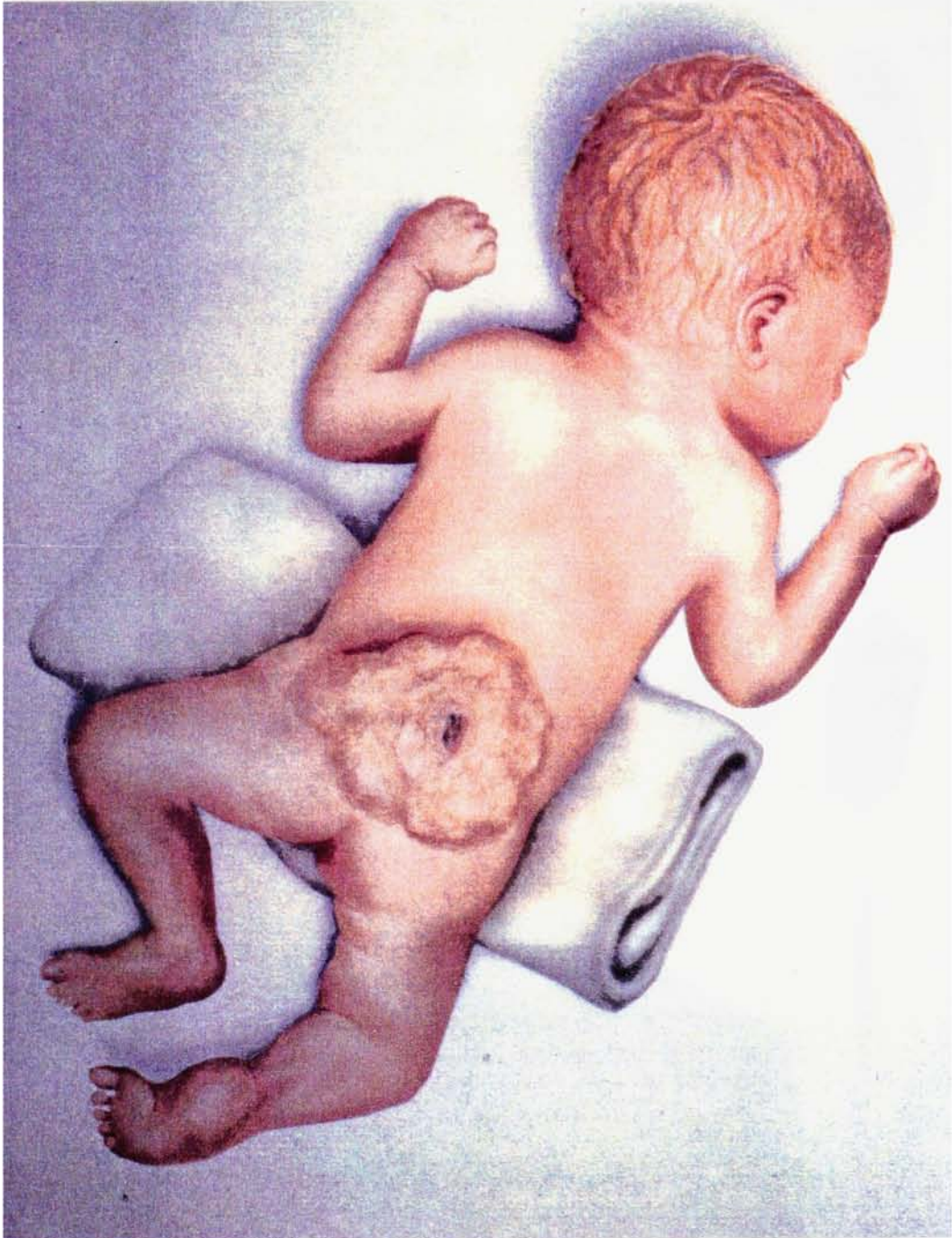
I have continued my involvement in a limited capacity with advocacy groups for the mentally and physically handicapped. After several regressions, my sister finally obtained a divorce and was accepted into a

halfway house that provides a supervised living skills program for the mentally ill.

My in-depth research into estate planning, in combination with the unpredictable status of my sister, finally convinced my parents to sign wills. Although there are no guarantees as to what the future holds, we can take some comfort in knowing that we have done all that we can.

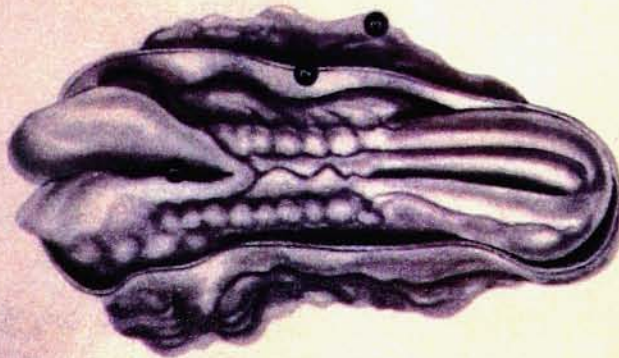
ILLUSTRATIONS

Dorsal View of Infant Exhibiting Myelomeningocele



Theoretical Embryological Development of Myelomeningocele

normal formation of neural tube

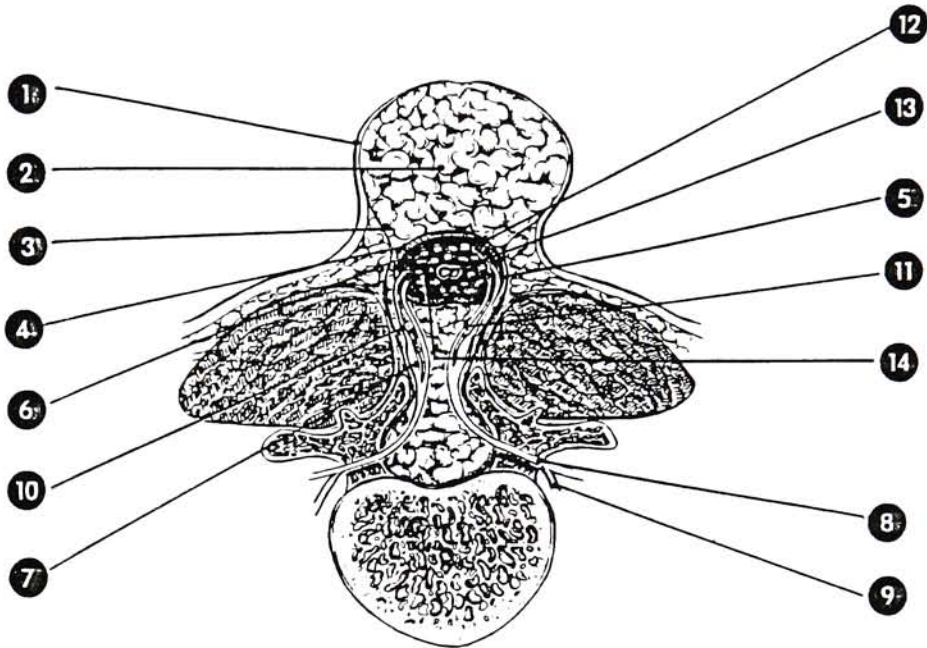


dorsal aspect of 22 day human embryo

rupture of tube causing abnormal reclosure



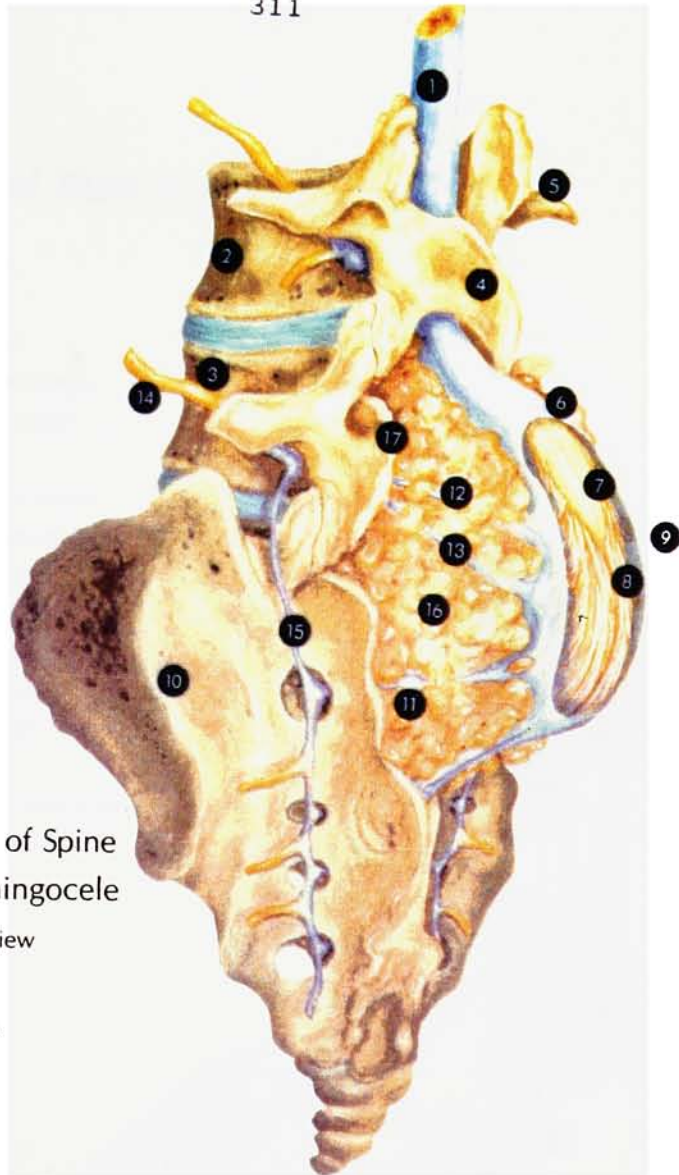
- 1 neural plate groove tube
- 2 neural crest dorsal root ganglion
- 3 somite
- 4 ectoderm
- 5 notochord
- 6 amniotic sac
- 7 yolk sac



Myelomeningocele

Cross Section of Fifth Lumbar Vertebrae

1. skin
2. lipoma of epidural space
3. periosteal layer of dura
4. meningeal layer of dura
5. arachnoid
6. subarachnoid space
7. dorsal root ganglion
8. dorsal root
9. ventral root
10. dorsal ramus
11. ventral ramus
12. pia matter
13. filum terminale
14. cauda equina



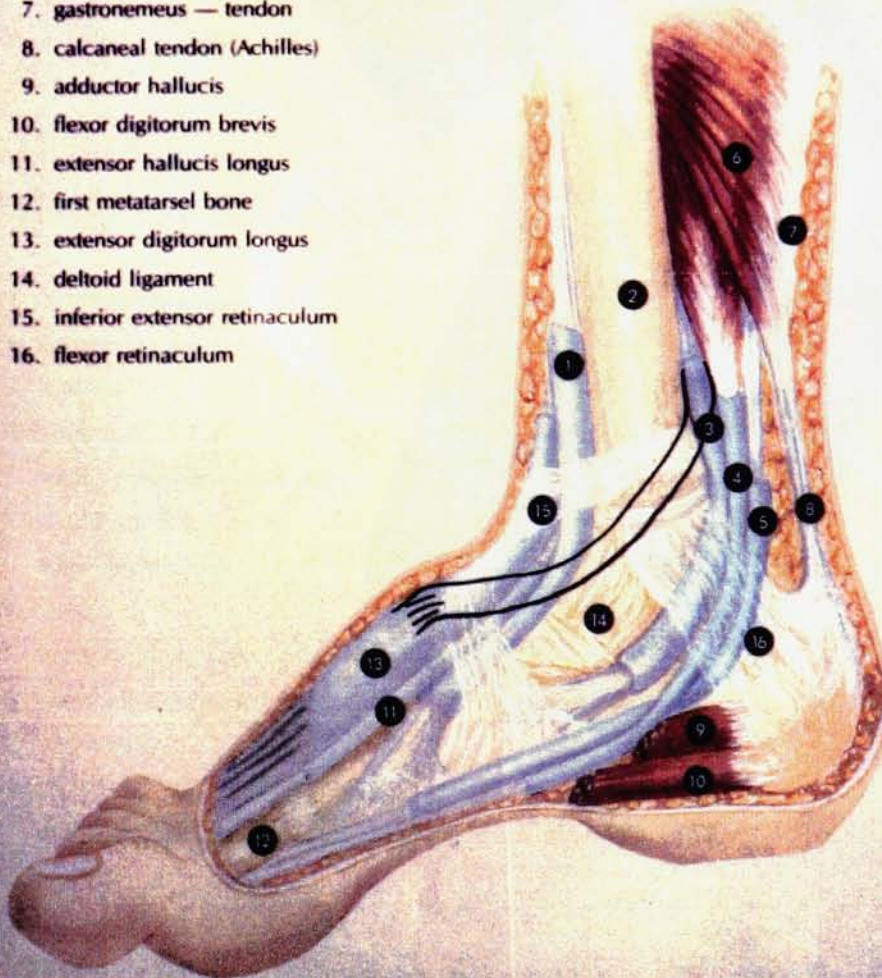
Lumbosacral Region of Spine Exhibiting Myelomeningocele

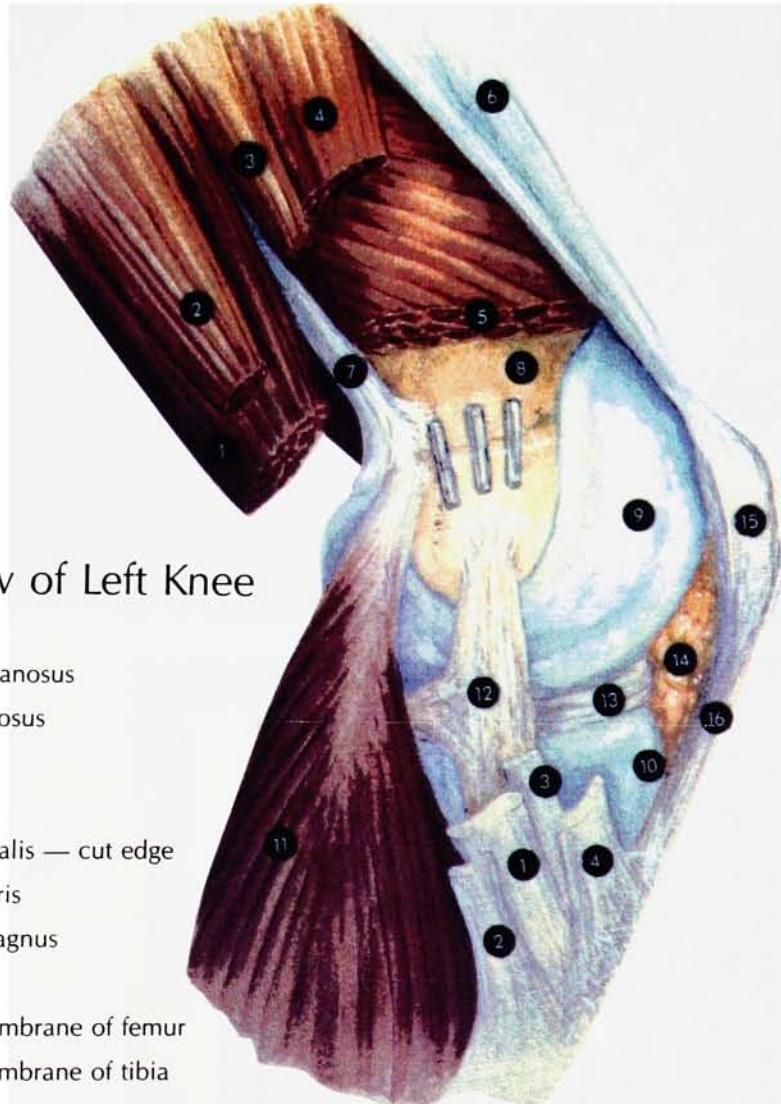
left posterolateral view

1. spinal cord
2. fourth lumbar vertebrae
3. fifth lumbar vertebrae
4. spinous process
5. transverse process
6. dura — cut edge
7. conus medularis
8. filum terminale
9. cauda equina
10. sacral vertebrae
11. dorsal root ganglion
12. ventral ramus
13. dorsal ramus
14. dorsal root of spinal nerve
15. ventral root
16. lipoma of epidural space
17. bony defect — absent spinous process of 5th lumbar and 1st — 3rd sacral vertebrae

Medial View of Right Foot

1. tibialis anterior
2. tibia
3. tibialis posterior
4. flexor digitorum longus
5. flexor hallucis longus
6. soleus
7. gastronemeus — tendon
8. calcaneal tendon (Achilles)
9. adductor hallucis
10. flexor digitorum brevis
11. extensor hallucis longus
12. first metatarsel bone
13. extensor digitorum longus
14. deltoid ligament
15. inferior extensor retinaculum
16. flexor retinaculum



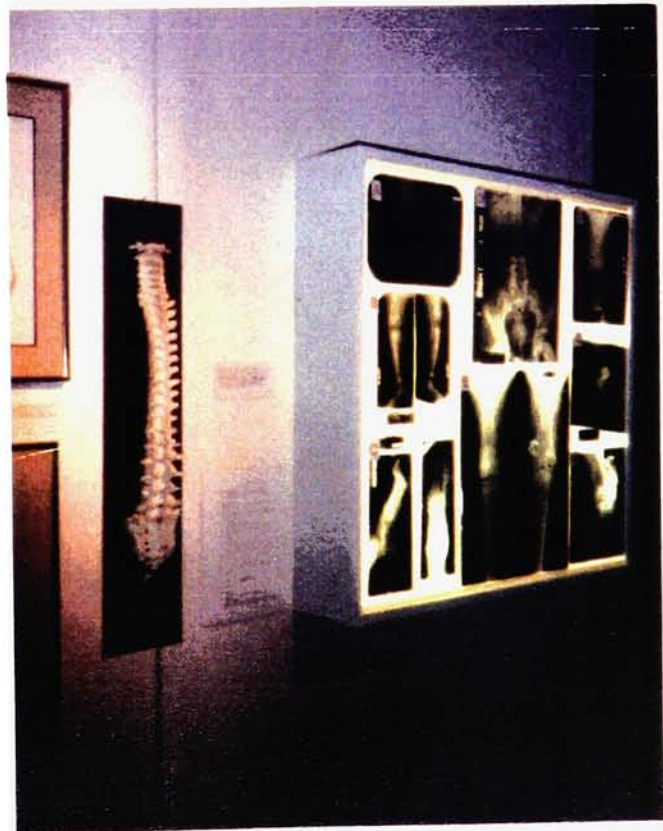


Medial View of Left Knee

1. semimembranosus
2. semitendinosus
3. gracilis
4. sartorius
5. vastus medialis — cut edge
6. rectus femoris
7. adductor magnus
8. femur
9. synovial membrane of femur
10. synovial membrane of tibia
11. gastrocnemius
12. tibial collateral ligament
13. medial meniscus
14. infrapatellar fat pad
15. patella
16. patellar ligament



Thesis
Exhibition, RIT
April, 1985



ENDNOTES

1. Dorlands Illustrated Medical Dictionary, 26th ed., p. 859.
2. Freeman, p. 3. The following is a glossary of terms defining the wide variety of spinal malformations. These terms are frequently discussed in medical literature in association with the term "spina bifida."

Glossary of Terms

1.	Diastematomyelia	Splitting of spinal cord, often associated with bony or cartilaginous spur tethering cord.
2.	Diplomyelia	Reduplication of part of the spinal cord, often associated with diastematomyelia.
3.	Dysraphism	Incomplete closure of neural tube; may be of variable severity from spina bifida occulta to myeloschisis.
4.	Lipomeningocele	Overgrowth of fatty tissue which involves the meninges and/or spinal cord and may be associated with tethering of cord.
5.	Meningocele	Spina bifida cystica without neural elements in the cyst; operationally used for such defects without demonstrable neurological deficit.

6. Meningomyelocele Spina bifida cystica with neural elements in the cystic cavity; operationally any cystic lesion of the spinal coverings with evidence of neurological dysfunction.
7. Myelocele Same as myelocystocele.
8. Myelocystocele Cavitated spinal cord lying in abnormal meningeal sac, often used synonymously with spina bifida aperta or myeloschisis.
9. Myelodysplasia Abnormalities of the spinal cord or roots of varying type, including diplomyelia, myeloschisis, hydromyelia, syringomyelia, and diastematomyelia.
10. Myelomeningocele Synonymous with meningomyelocele.
11. Myeloschisis Split cord, exposed to the surface without cystic coverings, identical to spina bifida aperta.
12. Rachischisis Splitting of the spinal column; should mean same as spina bifida, but is often used to mean nonfusion of neural folds in myeloschisis.

13. Spina bifida Nonfusion of the dorsal arches of the spine of variable degree; the skin, subcutaneous tissue, spinal cord, and contents of spinal canal may or may not be abnormal.
14. Spina bifida aperta Spina bifida with open cord on surface, possibly covered with thin membranes, no cystic covering; identical to myeloschisis and myelocele.
15. Spina bifida cystica Spina bifida associated with a cystic lesion on the back; the cystic lesion consists of dura, meninges, and normal or abnormal skin and may or may not include neural elements; this term includes meningoceles and meningomyeloceles.
16. Spina bifida manifesta Spina bifida associated with external manifestation; abnormalities ranging from hemangioma and skin lesions to an open or covered cord are included.

4. The Infinite Voyage, p. 5. In 1986 the skeletal remains of a civilization that existed approximately 7,210 years ago was discovered in southeast Florida. One hundred seventy individual skeletons were uncovered, among them an adolescent with spina bifida.

This child was totally impaired, unable to contribute to the community. Yet from skeletal evidence it is clear that his people were determined to keep him alive. He suffered from an opening in the spine, a defect known as spina bifida. The condition would lead to a terrible loss of bone in one leg. Through daily care, his people would arrest further deterioration for at least two years before the child would finally die. Archaeologists have found remarkable evidence of compassion in a society 7,000 years old.

5. Lemire, et. al., p. 373.
6. Lonsten, John E. "Spine Embryology." In Winter, p. 1.
7. Netter, Frank. Ciba Collection, Vol. 1, Nervous System. p. 31.
8. Freeman, Chapter 3, p. 1.
9. Gardner, Clinical Neurosurgery. p. 57.
10. Osaka, p. 720.
11. Gardner, Clinical Neurosurgery. p. 57.
12. Freeman, pp. 3, 10-11.
13. Stark, pp. 1, 2.
14. Stark, pp. 1, 2.
15. Renwick, pp. 321-324.
16. Angevine, pp. 95-113.

17. Lemire, et. al., pp. 280-288.
18. Emanuel and Sever, pp. 325-332.
19. Stark, p. 2.
20. Shurtleff, Current Problems in Pediatrics. pp. 55-58
21. Shurtleff, Current Problems in Pediatrics. p. 62.
22. Liptak, et. al., p. 6. "Children with myelomeningocele often...have restricted physical development and decreased ability to engage in many of the activities that other children their age can do. These factors make them different than their peers and may lead to loss of self esteem."
23. Pearson, et. al., pp. 27-30.
24. Freeman, p. 26. Innervation of the lower limbs originates in the lumbosacral region of the spine. The lesion on my spine was located in this area. The following chart indicates muscle groups used for ambulation that have the potential for impairment because of interruption in nervous system function.

Segmental Innervation of the Lower Limbs

L ₁	L ₂	L ₃	L ₄	L ₅	S ₁	S ₂	S ₃
Ileopsoas							
	Sartorius						
		Adductors					
		Quadriceps					
			Tibia ant.				
			Tibia post.				
				Gluteus med.			
				Hamstrings med.			
				Toe extensors			
				Peroneals			
				Gastroc. and soleus			
				Hamstrings lat.			
				Gluteus max.			
				Toe flexors			
					Foot intrinsic		

25. Liptak, et. al., p. 6. Children with myelomeningocele may feel different from their peers due to restricted development and frequent hospitalizations. They "may develop manipulative and demanding behavior or may demonstrate regression and withdrawal as a response to stress."
26. Mattson, pp. 801-811.
27. Tew and Lawrence, p. 122. Some studies of the academic achievements of spina bifida patients indicate a significant deficiency in the mathematical capabilities of these patients, as compared to control studies. "The attainments of the [spina bifida] index cases were significantly poorer than their controls, with mathematical ability especially weak."
28. Liptak, et. al., p. 4.

Most children with myelomeningocele have overall IQ scores in the normal range. For example, Mapstone reported that the mean overall IQ for children with myelomeningocele was 104. Although children with hydrocephalus scored slightly lower than those without, the lowest scores were seen in those who had had ventriculitis, hypoxia, or uncontrolled hydrocephalus. Most children with myelomeningocele have selective cognitive disabilities and score better on verbal than on performance scales. Specific cognitive testing often reveals deficiencies of selective visual attention, visual-spatial perception, tactile perception and auditory concentration. Common manifestations seen as a result of these problems include short attention span, distractibility and difficulty with subjects requiring visual-motor integration such as arithmetic....

29. Results of standardized tests administered to me at an early age seem to substantiate the evidence of mathematical deficiency exhibited by spina bifida children. I also found subjects with abstract concepts difficult to understand, such as chemistry or physics. Although such subjects were difficult they were not impossible for me if proper instruction was offered.

During my years of studying art I found myself struggling with subjects requiring an emphasis on geometric perspective, such as landscapes. Subjects with a more narrow focus and fine detail were noticeably less difficult for me to render accurately.

The following documents were accessed through the freedom of information act and released to me by Oxford Academy and Central Schools. These scholastic records contain I.Q., achievement and aptitude scores of tests administered through grade 9.

Last Name		First Name		Middle Name		Form Number of Progress		Exempted		Identified	
Birth Date: Month 6, Day 27, Year 1961 Residence: Norwich						CHECK AUTHORITY OF BIRTH CERTIFICATION <input type="checkbox"/> Birth Certificate <input type="checkbox"/> Father's Signature <input type="checkbox"/> Passport <input type="checkbox"/> Mother's Signature <input type="checkbox"/> Bible <input type="checkbox"/> Church Record <input type="checkbox"/> Club <input type="checkbox"/> Hospital Record Licenses Issued in the State: _____ Class: _____ Certificate: _____					
PARENTAL RELATIONS AND GUARDIANS											
NAME		NATIONALITY		RELATIONSHIP		LIVINGS					
Leonard Tefft				Father		yes					
Catherine Tefft				Mother		yes					
FATHER'S OCCUPATION		PLACE EMPLOYED		PHONE		RESIDENCE		PHONE		DATE	
						R.D.#3, Oxford, N.Y.					
OTHER TESTS:											
DATE		NAME OF TEST		FORM		SCORE		S-ILE RANK		OTHER	
5/16/67		Palmer-Hingham		B		41		7-2		5-10 117	
CE _____ Retested 12/3/68 until 1/6/69											

Standard Test Record - Grades K-6 (Continued)

Aptitude and Intelligence Tests

Date	Grade	CA	Form	Test	Total DIQ	Grade %ile	Raw Score
				NEW YORK STATE TEST GRADE 3			
10/69	3		A	Reading Math		90 70	49 40
				OTIS-LENNON TEST			
11/69	3	8-6	J		118	87	60
				LODGE THORNDIKE			
5/72	5	10-11	A	Verbal: DIQ 117 Non-Verbal: DIQ 112		92 84	Raw Score _____ Total - 115
				NEW YORK STATE TEST GRADE 6			
10/72	6			Math - 40 Reading - 80			

3174 7 - - MLAT - local % tile 71

Name Tefft, Barbara

OXFORD ACADEMY & CENTRAL SCHOOL

Standard Test Record - Grades K-6

Achievement Tests

Date	Grade	Form	Test Results																													
05/70	3	W	SAT - PRI 2																													
			WD HNG IPAR HNGSC SS (I) SPELL W SY SK LANG AR COMPAR CONC																													
			4.4644	5.3887	6.094	4.0684	5.4724	6.4928	3.0324	4.9787																						
4/71	4	W	SAT - INT 1																													
			WD HNG IPAR HNG SPELL W SY SK LANG AR COMPAR CONGR APP LOGIC SY SCIENCE																													
			7.5949	6.1891	4.6843	7.2928	8.8989	4.9505	5.5704	4.4884	5.6704	9.1889																				
TEFFT BARBARA			IOWA TESTS OF BASIC SKILLS										GRADE 4 LEVEL										IOWA 3 DATE TESTED 03/72									
			80	66	69	76	69	71	71	46	45	44	45	59	59	59	64															
METROPOLITAN ACHIEVEMENT TESTS			LEVEL INT										IOWA 3 DATE TESTED 12/72										GRADE 4									
			99	99	99	99	99	99	99	99	99	99	99	99	99	99	99	99	99	99	99											
METROPOLITAN ACHIEVEMENT TESTS			LEVEL ADV										IOWA 3 DATE TESTED 10/73										GRADE 7									
			99	99	99	99	99	99	99	99	99	99	99	99	99	99	99	99	99	99	99											
METROPOLITAN ACHIEVEMENT TESTS			LEVEL ADV										IOWA 3 DATE TESTED 0/74										GRADE 8									
			99	99	99	99	99	99	99	99	99	99	99	99	99	99	99	99	99	99	99											
DIFFERENTIAL APTITUDE TESTS			TEFFT BARB J										119791										755 L 08 F									
			34	20	54	32	29	52	37	72	38																					
			90	55	80	60	10	97	90	65	85																					

Name Tefft, Barbara

OXFORD ACADEMY & CENTRAL SCHOOL

Standard Test Record - Grades 7-12

Intelligence & Aptitude Tests

Date	Grade	Form	Test Results																			

Achievement Tests

10/75	9		METROPOLITAN ACHIEVEMENT TESTS																			
			LEVEL ADV																			
			IOWA 3 DATE TESTED 10/75																			
			GRADE 9																			
			99	99	99	99	99	99	99	99	99	99	99	99	99	99	99	99	99	99	99	
N.Y.S. Basic Competency Test			Reading-39 - 98%										Math-41 - 82%									
10/75	9																					

30. Rodnan, p. 1095.
31. Bruckner and Howell, p. 48.
32. King, E.S.J., Archives of Pathology. pp. 693-671.
33. Horwitz, pp. 579-588.
34. Bruckner and Howell, p. 50.
35. Bruckner and Howell, p. 51.
36. Katz, et. al., p. 969.
37. Kidd, p. 599.
38. Bruckner and Howell, p. 52.
39. Townsend, p. 255.
40. Johnson, J.T.H., p. 10.
41. Johnson, J.T.H., p. 1.
42. Bruckner and Howell, p. 50.
43. Bruckner and Howell, p. 51.
44. Johnson, J.T.H., p. 3.
45. Bruckner and Howell, p. 66.
46. Soudry, pp. 199-204.
47. The Task Force on Concerns of Physically Disabled Women. Toward Intimacy: Family Planning and Sexuality Concerns of Physically Disabled Women. p. 9. Exposure to and support from other disabled people can be beneficial.

Many congenitally disabled women have grown up exclusively with nondisabled people, with parents who were trying to help them be "Normal". This is done with the best of intentions, but can make loneliness and sense of being physically different overpowering. It can be reassuring to know other people with similar - or different - disabilities.

48. Radman, p. 79 S.
49. Kato, et. al. p. 296.
50. Hodgkinson, p. 414.
51. Radman, p. 78 S.
52. Shurtleff, p. 63.
53. The Task Force on Concerns of Physically Disabled Women. Toward Intimacy: Family Planning and Sexuality Concerns of Physically Disabled Women. p. 44.
54. The Task Force on Concerns of Physically Disabled Women. Toward Intimacy: Family Planning and Sexuality Concerns of Physically Disabled Women, pp 22-24.
55. Torrey, pp. 129-169.

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