Recent Developments in the Treatment of Spina Bifida in the Past Ten Years

An Honors Thesis (ID 499)

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"Like most eager couples, Steve and Intramoud Parrish had already decided upon names. David Stephen if a boy, Heidi, if a girl. Shortly before midnight on January 17, 1975, Intramoud went into labor and Steve drove his wife six miles to the hospital at Whiteman Air Force Base, near Knob Nester, Missouri. He stood close to his wife throughout the labor and delivery and at 3:30 a.m., watched David Stephen enter the world head first, face up, twenty inches long and weighing seven pounds, five ounces. The general surgeon who delivered the baby handed him to an Air Force Corpsman who began to bathe the infant. The Corpsman turned David Stephen over onto his stomach and then he stopped, curious about a silver-dollar-sized red spot on the baby's back. He called for the general surgeon. Intramoud asked her husband what was wrong. "He's got a little scratch on his back", Steve told her. A few moments later, Steve Parrish learned that the red spot on his son's back was more than just a scratch. 'Your baby has spina bifida, a very serious birth defect', the surgeon told him. Taking Steve aside in the delivery room, the surgeon began to explain..."1

Spina bifida literally means "cleft spine", a spine split in two because the vertebrae in the infant's back failed to come together during the first trimester of pregnancy.

There are three types of spina bifida. The least severe is, fortunately, the most common. Called spina bifida occulta, it involves an abnormal opening in the vertebrae, but not any damage to the spinal cord or any visible signs of deformity, so most people never even discover they have the defect. "As many as twenty-five percent of children may have such a bony defect."2

A second kind of spina bifida is meningocele, named because of the meninges, the protective covering for the spinal cord, have pushed out through the opening in the vertebrae inside a sac that protrudes from the back. The sac, or meningocele, can be as large as a small grapefruit, but since the spinal cord remains intact, the nerve pathways to the lower body are usually unaffected. After corrective surgery to reposition the meninges and remove the sac, the defect in the back is closed and the child will experience no further difficulty. However, "repair of the defect may be accompanied subsequently by the development of hydrocephalus."3

Meningomyelocele is the third type of spina bifida. With this condition, the spinal cord does not form properly and it, too, protrudes from the back.
Usually, the spinal cord protrudes from the back inside a sac.

There are no typical spina bifida children or adults. That, too, has contributed to misunderstanding about the defect. Spina bifida does not cripple its victims uniformly because it can occur almost anywhere along the spine. Generally, the higher it strikes, the more severe the handicaps. A lower opening in the spine may mean less profound damage to the spinal cord. "The usual site of the defect, the lumbosacral area, is associated with a flaccid paralysis of the lower extremities, absent sensation to the level of the lesion and loss of bowel and bladder control. Hydrocephalus commonly accompanies the defect and again, the higher the lesion, the greater the likelihood of hydrocephalus." The lucky ones wear braces that stop at the knee. The unlucky never walk. "The fortunate suffer no loss of mental functions. The unfortunate are mentally retarded, usually due to hydrocephalus. 60-75% of the children with meningocele develop hydrocephalus and because of the increased pressure on the brain, the brain cells necrose and die."  

Although doctors can now save the lives of nearly all children born with spina bifida, they still know relatively little about what causes it. There are indications, however, that genetic and environmental factors may play some causative role. "Once parents have had one child with spina bifida, the odds of a second child with the defect rise from 2/1000 to about 1/20." Some studies point a finger at industrial pollution; doctors at certain spina bifida clinics have noticed a higher percentage of children from communities near industrial parks have the defect.

The defect pays little heed to social standing. It cuts across all economic levels. Some distinctions: "it appears in girls more frequently than boys, some ethnic groups such as the Irish, Scots and British seem more likely victims; other groups, notably black, seem less likely to be affected."  

The defect is costly. It robs the child of mobility and perhaps mental ability. And, spina bifida costs money. "We used to estimate 'hard costs'-
hospitalizations, operations, braces, wheelchairs, etc— for a child living at home until age eighteen at ninety thousand dollars," says Kent Smith, Executive Director of the Spina Bifida Association of America, a non-profit group organized by parents of children with spina bifida. "That was five years ago. Now, I'd estimate those costs to be three and possibly, five, times as much." While insurance policies often cover most of these expenses, it is a rare family that does not wind up paying thousands of dollars to care for the child.

Then there are the so-called "soft costs", which strike the family budgets less noticeably, but equally hard. Countless trips to the hospital and meals away from home. A larger car, usually a station wagon or van, to facilitate use of a wheelchair. Maybe a relocation—from a two-story or split level home to a ranch, or to a different school district with a better educational or vocational program for the handicapped.

Surgery to cover the defect in the back is only the beginning of an ongoing series of operations, examinations and therapy that will probably be necessary in the coming weeks, months and years. If the child has hydrocephalus, most likely, the child will require brain surgery to implant a device called a shunt. The shunt is a thin plastic tube that runs from the lateral ventricle of the brain to the peritoneal cavity or left atrium of the heart. This shunt drains excess fluid and prevents or limits hydrocephalus. Hydrocephalus or water on the brain, can enlarge the skull and exert tremendous pressure on the brain, causing mental retardation. Often children born with spina bifida also need foot or ankle surgery. Many will require additional surgery on their spine around the age of ten to correct curvature of the spine. Because nearly all are incontinent, urologic therapy is a necessity and urologic surgery a possibility. The child's paralyzed legs will require specially fitted braces. The list goes on and on.

**Prenatal Detection of Spina Bifida**

Because spina bifida is such a traumatic illness and because the cause is
still unknown and cannot be prevented before conception, research is now being
done on detection of an abnormal fetus. The measurement of alpha-feto-protein
levels in the amniotic fluid is generally a reliable technique for the early
antenatal diagnosis of neural tube defects. "Amniotic fluid alpha-feto-
protein (AFP) levels were raised in early pregnancy in association with anen-
cephaly and "open" spina bifida. Closed lesions, including encephalocele and
hydrocephalus, were associated with normal levels as was an "open" spina
bifida at thirty-three weeks gestation."10 It is concluded that when ultra-
sound and amniocentesis are used, "most fetuses with open lesions are detected
before twenty weeks of gestation allowing selective abortion of most cases
with neurological involvement when there is a history of previously affected
fetuses."11 Closed lesions will usually be missed and maternal serum AFP
assay cannot be relied upon to detect neural tube malformations in early
pregnancy, whether open or closed. At the moment, AFP estimation seems to
offer the only practical method of prenatal detection of "open" spina bifida.

Amniotic fluid AFP before twenty-eight of gestation is a good indicator
even in a viable fetus of open, but not closed, spina bifida. In contrast,
maternal serum AFP does not seem to be specifically diagnostic of spina bifida
and is an indicator of feto-placental dysfunction.

Usually, after a defect has been diagnosed prenatally, the mother has
the option of receiving an abortion. "No false positive results have been en-
countered so far, so there have been no uncalled for abortions."12

"Amniotic fluid AFP reaches a peak about fourteen weeks of gestation,
falling to below one ug/ml by the beginning of the third trimester. It is not
entirely clear from what source maternal serum AFP is derived, but the occurrence
of maternal peak values at a time when the total amount of AFP in the fetus
is at its highest suggests a fetal origin of maternal AFP."13

Although amniotic fluid AFP assay seems reliable, amniocentesis could not
be used in more than a minority of cases. However, it might be argued that since raised maternal serum AFP levels are almost invariably associated with severely disturbed pregnancies, little hard would be done if the pregnancies were terminated even though they are not related to anencephaly or spina bifida.

But, is AFP assay enough? Some doctors say no and in order to be sure about the prenatal diagnosis, they have combined ultrasound with the assay. "The optimum time to do the testing is between sixteen and nineteen weeks. The time required to make the full examination varies from ten to thirty minutes according to the position of the fetus and the amount of fetal movement." The following case study shows the importance of combining ultrasound and AFP assay for accurate results:

"An ultrasound performed at eighteen and one half weeks showed normal sizes of the head and spine. The AFP assay done at that same time was above the upper limits of normal. This was both fetal AFP and maternal AFP. A further ultrasound one month later still did not reveal a small lesion. In view of the abnormally high AFP levels, however, termination was recommended and was carried out at twenty and one half weeks gestation age. The fetus was male, weighed 425 grams and by size was consistent with a fetus of twenty weeks gestation age. No myelomeningocele was present and dissection of the spinal column revealed no spina bifida. Dissection of the brain demonstrated no lesion and blocks of the lungs, kidneys, adrenals, sacrum and umbilical cord were normal."

It is clear that an amniotic fluid level raised very high is not an absolute guarantee that a neural tube defect or indeed any other defect is indeed present.

AFP can not define or describe the particular abnormality while ultrasound has the ability to do so and may give qualitative information to the doctor or patient which will enable them to decide whether to continue the pregnancy or opt for termination. In ultrasound, examination of the fetal head can show any ventricular dilatation and it is now clear that displaying the ventricular system by ultrasound is mandatory if the diagnosis is to be made. Ventricular dilatation would be caused by increased intracranial pressure or hydrocephalus, a complication of meningomyelocele. In an unborn child, the sutures of the skull and the bones of the skull have not hardened and grown.
together. With the increased pressure, then, the ventricles have room to expand or dilate. However, this diagnostic method would only be applicable in the child with meningomyelocele and hydrocephalus was a complication.

Ultrasound examination can also implement AFP estimation in the accurate assessment of fetal maturity. The significance of AFP levels is dependent on knowing the gestation age of the fetus. Since patients tend to overestimate the duration of pregnancy, ultrasonic dating will tend to reduce the number of abnormal AFP predictions in the amniotic fluid.

In summary, ultrasound is important because it: 16
1) confirms and defines the lesions in cases with raised AFP levels
2) possibly diagnoses at least some of the 15% of the spinal lesions that are skin covered and not amenable to diagnosis by AFP
3) if amniocentesis is performed directly under ultrasonic control, it is easier to obtain an uncontaminated specimen and estimating accurately fetal maturity, thus preventing misinterpretation of AFP values.

As was mentioned before, AFP assay may present difficulties if the fetus has only a small, open spina bifida. Evidence has been produced that a careful examination of the morphology of the amniotic fluid cells, particularly those cells that adhere rapidly to glass or plastic surfaces in culture, can help in making such a diagnosis. For example, 17

"A twenty-four year old mother, with no history of affected children, had three sequential AFP levels just slightly above the upper limits of normal. There was no contamination. Nine percent of the viable cells were adherent to glass after twenty hours incubation and all these cells had an abnormal morphology. The next amniocentesis was done at twenty weeks and was normal and all the viable cells continued to show abnormal morphology. In view of the marginally raised amniotic fluid AFP concentrations, the pregnancy was allowed to go to term. The outcome was an infant with a severe lumbar myeloschisis who died after seven days."

After the Child is Born

"The main problem is the way the news is broken to the parents." 18

Usually the duty is left to a junior member of the medical staff or a nurse. This problem should be handled by the person the mother has known and trusted during her pregnancy, backed up by a pediatrician. "Questions asked are "Will he live? Will he need an operation?" 17 The person breaking the news should
be competent to answer the first question at least for the time being, the immediate future, but in respect of the second, it is best to wait until the pediatric surgeon has seen the infant. Parents should be advised, in suitable cases, that their child can compete on an equal or nearly equal basis with his normal peers; brick-building, jigsaw puzzles, card games, chess, drawing, painting and reading. Parents should also realize that calipers and crutches are not something imposed on the child by the doctor, but merely a means of gaining independence.

"One half of the children with spina bifida will go to schools for the physically handicapped and this is not really an ideal solution." If education is designed to fit a child for later life, how can one expect an employer to accept a teenager, who by implication, could not cope with normal life and competition up to that point? Similarly, workmates are unlikely to be able to accept as an equal someone who has been different during school life. If the child cannot keep up with others, special schools may postpone problems until later life.

Orthopedic Management

"The aims of orthopedic management of the child with spina bifida are to correct deformity, obtain the best possible ambulatory function, achieve the posture which allows the patient to function at his maximum capabilities and prevent or minimize the effects of sensory loss." Deformities at birth may be present due to unbalanced muscle action about joints in utero, or the effects of posture in utero or congenital maldevelopment of the skeleton. "The areas most often considered for corrective procedures are the feet, knees, hips and spine." There are several approaches to orthopedic management. First of all, there is conservative management with a delay in all operative procedures until the patient is ready to attempt ambulation. Secondly, initial attention is directed to early correction of deformities by operations on soft tissues while postponing operations on bone structures until later in infancy.
Conservative management utilizing casting and other types of corrective appliances carries the risk of injury to anesthetic skin and may not be definitive because of the continued existence of muscle imbalance about joints. "Aspects which are important in predicting the success of an orthopedic program includes the patient's general health, cardiorespiratory and urological status and cooperation and enthusiasm of the family."23

Advances have been made in the types of orthosis available for children with spina bifida. Coaster carts are used as a prebracing mobility aid. Lightweight, durable and cosmetically acceptable polypropylene inserts are used for the treatment of foot and ankle instability.

"The parapodium is a brace-like device constructed of light weight, high-strength aluminum, and consists of a foot plate, side bars, knee bar and front and back panels which fit against the patient's trunk. It is modular in construction and can be adjusted to accomodate growth. The shoes are not permanently attached to the device which can be applied and removed easily and rapidly. The parapodium facilitates standing and swiveling without crutches, thereby freeing the upper extremeties. Use of the parapodium allows many children with relatively high lesions to experience the upright position and some locomotion."24

The Verlo (vertical loading orthosis) is a simplified standing device which assists children to achieve free standing balance despite severe neuromuscular deficits. Locomotion is possible in the brace with the help of a walkerette, using either a pivot gait or a swing gait, the latter requiring greater upper extremity strength and coordination. "Until recently, pediatric bracing has utilized braces designed for an adult but reduced in size for a particular child. This approach has several disadvantages:"25

1) braces were difficult to make because of reduced size
2) difficulty in getting appropriate sizes of component parts.
3) skeletal and structural abnormalities often seen in neuromuscular handicapped children often make brace alignment difficult
4) younger patients have difficulty getting into the braces with a pelvic band or other trunk attachments
5) poor or no free standing balance
6) patient outgrows brace before it wears out with little possibility of reusing the outgrown brace for other children
7) financing standard bracing is burdensome for the family or agency involved because of frequent replacement during the growing years

Gait training with the Verlo brace is done by a regular physical therapist, either in the hospital or at a special education center, using standard protocol. Training sessions are started in the parallel bars with the patient attempting to achieve independent standing balance. The patients then learn to pull themselves along the bars, using a pivot gait. Finally, if the child's upper extremities are strong and coordinated enough, he is taught to walk in the bars using a swing through gait. After the child has accomplished these events in the parallel bars, the same series of activities is repeated outside of the bars, using a walkerette.

"No attempt is made to train formally children less than thirty months of age to walk in the Verlo. For these children, the brace is used only as a standing device to get them accustomed to the upright position while, at the same time, maintaining proper body alignment. The parents are encouraged to have the child stand at a table and work with activities which would develop hand-eye coordination." 26

A unique feature of the Verlo is the ability to vary the verticality of the child by adjusting the angle the upright portion of the brace makes with the baseplate. For maximum standing stability, the Verlo is adjusted so that the center of gravity of the child is over the center of the baseplate. When using the Verlo for locomotion, it is adjusted with the center of gravity of the child over the anterior portion of the baseplate. This slight anterior instability makes it easier for the child to move in the brace. It also provides a safety feature such that if the child falls, he will have a tendency to fall forward enabling him to use his hands for protection. When the child graduates from the parallel bars to the walkerette, he is trained to fall safely in the Verlo.

While the Verlo is stable, the child can still pull or push against a solid object and topple himself. If the child is to be left unattended in the
Verlo, such as working at a table, a gluteal support strap attached to the table should be used as a safety precaution. While it is possible to fit children with severe structural abnormalities in the Verlo, it is desirable to have surgical correction of the structural abnormalities if at all possible.

Body image changes experienced with the Verlo appear to be satisfactory. The children often show considerable delight and increased social responsiveness to activity when assuming their new upright position. "Investigation is in progress at the present time to determine at which developmental level training for ambulation in the Verlo is practical. Obviously, if twenty or more physical therapy sessions are required to achieve limited ambulation skills, the cost would be prohibitive in most cases."²⁷

The main disadvantage of the Verlo is the lack of provision for knee and hip flexion. Where cost is not a factor, children of school-age should probably be fitted with the standard types of bracing so that they can sit. However, the Verlo is an effective and economical way to achieve and maintain ambulation:²⁶

1) it is technically not difficult to fabricate
2) skeletal and structural abnormalities can be accommodated
3) easy to put on and remove
4) child achieves immediate free standing balance with the opportunity for progression to walkerette ambulation
5) initial cost about one fourth that of standard braces
6) when the child outgrows the Verlo, it can be used with minimal modifications for other children
"Draycott-Oswestry Splint has been used successfully in the Robert Jones and Agnes Hunt Hospital in England for about seven to eight years now. It is used for maintaining the required position of the legs after operations on children with myelomeningocele. It can be made cheaply from Standard National Service sheepskin and alleviates the pressure problems of Paris Splinting."

The splint in position shows that the Velcro bands are not in contact with the patient's skin and that those holding the legs pass through the sheepskin. The sheepskin is tailored to fit from the nipple line to feet, allowing for extensions for covering the soles of the feet to the end of the big toe. The width at the nipple line around the pelvis at greater trochanter level
should allow for one inch overlap.

In the hospital of this article, "the splint is kept continually in position, except for diaper changes and washing, until the wound is healed. Then, the splint is applied only at night."  

Incontinence is no problem because waterproof pants can be worn. The wool side of the sheepskin can easily be rubbed dry.

Pathological fracture of the femur can be effectively treated with the splint. It prevents external rotation, and shortening can be overcome by incorporating a Ventifoam extension.

**Bowel Regulation**

"Stool incontinence depends on a normal anorectal function which requires an appreciation of rectal fullness, satisfactory peristalsis and properly balanced tone of the anal rectal sphincter mechanism." The therapeutic regimens used in a spina bifida are emptying the rectum electively and regularly prior to the initiation of spontaneous defecation. This is done by setting aside regular periods for defecation, taking advantage of the gastrocolic reflex, diets and natural laxatives and foods with high fiber content. Also, the use of suppositories, enemas and oral purgatives can be a way of management.

Treatment of the neuropathic bowel can also take place with electrical stimulation of the rectum. The bladder and rectum are stimulated by the Viscero stimulator which produces a direct current stimulus. Treatment is performed through a urethral catheter and rectal tube, each of which is fitted with a silver electrode tip which is connected through a silver wire to the stimulator. The neural electrode is placed on the upper limb to avoid the possibility of burning which could occur if it were placed on anesthetic skin.

Treatment is carried out daily for between one and one and half hours and is continued for periods of between one and three months. "The current flows from the active rectal electrode through the body to the indifferent electrode on innervated skin." The electrical stimulation in no way resembles the electrical stimulation
of the sphincter muscle; it is aimed at the end organs in the bladder and rectum. As far as the rectum is concerned, "the stimulus sets up reflex contractions of the smooth muscle of the bowel wall. The stimulation has an indirect effect (regulatory) on the sphincter mechanism."33

"Seven children with myelomeningocele, neuropathic bladder and neuropathic bowel were included in the initial trials of this method of treatment. All the children had lumbo-sacral lesions, with more or less complete flaccid paralysis of the legs, but all had normal urinary tracts on radiological investigation. Within eight weeks of treatment, all seven patients had remarkable improvement in bowel function. Rectal pressure studies suggest that there has been a great improvement in the tone of the rectal musculature and some of the patients had developed a sense of rectal fullness. One seven year old boy had an inactive, paralyzed sphincter and the stool had been removed from the rectum manually. He had never had an urge to pass a stool. After treatment, he regularly had spontaneous urges to defecate and was also able to pass the stool by himself without any aid. He also achieved normal inhibition of bowel opening."34

<table>
<thead>
<tr>
<th>Patient</th>
<th>Level of spinal lesion</th>
<th>Neurological status</th>
<th>Type of bowel function</th>
<th>Type of rectal function</th>
<th>Sensation</th>
<th>Number of treatments</th>
<th>Results</th>
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<td>Boy, 7 years</td>
<td>Lumbo-sacral</td>
<td>Paraplegia</td>
<td>Chronic constipation, laxatives, stool 1 per week</td>
<td>No expression, manual evacuation</td>
<td>None</td>
<td>27</td>
<td>Complete recovery, stools 4-7 times per week, expressed by himself. Had sensations.</td>
</tr>
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</table>
| Boy, 4 years     | Lumbo-sacral           | Paraplegia          | Chronic constipation, laxatives, stools 1-3 per week | Stool expressed only occasionally | Occasional           | 15                    | Complete recovery, regular urges, 6-8 stools weekly, expressed by himself |}

Experimental Results of Electrical Bowel Stimulation
Bowel Bag

"A simple, easily available method can simultaneously protect against heat loss, drying and bacterial contamination in newly born infants with skin defects. This method applies to infants with meningomyelocele."

Typically, in the past, the child with this defect was packed in saline-soaked gauze in an effort to prevent drying. Because of this practice, the infant usually became seriously chilled and was always difficult to observe colorwise and temperature-wise because he was completely wrapped in gauze. The child frequently developed surface infections because of the open nature of the wet pack and its frequent manipulation.

The bowel bag, a commonly used device for omphaceles, gastrochisis, in the abdominal surgery suite, fits the average neonate quite well. The pre-term infant could be enclosed up to the neck.

Since the bag is impervious to water, a small amount of warm saline and a slight head-up position will keep the tissues moist for hours. There can be no evaporation; thus there is not evaporative heat loss. Although radiational heat losses occur, infrared heating devices can penetrate plastic film, allowing easy temperature control during the diagnostic and pre-op periods. "A second bag over the first could provide a dead air space to prevent convection losses. Conduction losses must be prevented by the use of insulation (a blanket) between the infant and the cold surface."

The bag is also sterile and impervious to bacteria, thus supplying an effective barrier to infection, while its transparency eliminates the practice of lifting the dressing in order to see the child.

Since the bag is small, it can be kept in the delivery room and can be applied immediately. It provides an ideal means of protecting those infants during the transportation to the surgical area.
Control of Normal Pressure Hydrocephalus

As mentioned before, hydrocephalus is a problem because with increased intracranial pressure, mental retardation can occur. Treatment of NPH is not controversial; it is clear cut and well defined if the patient has been diagnosed correctly. Repeated lumbar punctures with removal of fluids have a very slight success rate.

"The majority of the cases are treated with a ventriculovenous shunt."37 First of all, a frontal burrhole is placed just behind the anterior hairline. A catheter is inserted into the anterior horn of the right lateral ventricle of the brain.

A second incision is made in the post auricular region where a reservoir and low or medium pressure valve are placed and attached to the ventricular catheter which has been tunneled under the scalp.

A third incision is made in line with the skin crease over the upper portion of the sternocleidomastoid muscle. A radiopaque catheter is inserted through this incision into the common facial vein and threaded into the jugular vein. The cardiac end of the catheter is positioned about the level of the sixth thoracic vertebrae, with the proximal end brought through a subcutaneous tunnel to the post auricular incision where it is attached to the vein. Correct position of the cardiac or distal end of the catheter is determined by chest
X-ray.

"When the cardiac end of the catheter is threaded through the external jugular vein and the superior vena cava into the right atrium, the process is referred to as a ventriculoatrial shunt." 38

There are three other types of shunts which are used in the treatment of hydrocephalus. "They are:" 39

1) **lumboperitoneal shunt**: lumbar subarachnoid space to peritoneal cavity. Increased incidence of permanent shunt failure

2) **ventriculoperitoneal shunt**: ventricle to peritoneal cavity. Technical difficulty in threading and placement of shunt.

3) **ventriculoureteral**: ventricle to ureter. Salt loss and electrolyte imbalance.

There are common complications of all shunts—"obstruction of the ventricular catheter, thrombosis of the jugular vein, septicemia, meningitis, subdural hematoma, thrombophlebitis, pulmonary emboli and pneumonia." 40

After the operation, improvement is seen in some patients immediately. In others, the change may not be so dramatic—occurs gradually over several weeks or months. If a patient improves initially, then shows signs of deterioration, shunt failure must be considered.

Following a shunt, the most rapid improvement is usually seen in the patient's mental status. Incontinence usually clears up rapidly, also. Gait differences are usually slowest to improve and some patients are left with a residual deficit.

**Urinary Control in Children with Myelodysplasia**

Difficulties have been encountered in the treatment of urinary tract infections (UTI) in children with spina bifida cystica because of organic and functional abnormalities of the renal tract and of resistance to antibiotics. "Two drugs, co-trimoxazole and cephalaxin, are effective in vitro against a wide range of gram-negative urinary pathogens, even those with multiple antibiotic resistance. Both are bacteriocidal and are readily absorbed from the gastrointestinal tract." 41
In a comparative study (14 day trial), cephalexin and co-trimoxazole were used to treat UTI in forty-seven children with spina bifida. It was found that co-trimoxazole was more effective in achieving sterility of the urine, but that neither drug was able to maintain sterility in the majority of the cases, when treatment stopped. More side effects, such as, abdominal pain and diarrhea, irritating skin rashes, were noted in the children with cephalexin. 

This study has shown that short courses are ineffective in maintaining urine sterility. Long-term treatment will be needed in the majority of these children with bacteriuria and any child whose bacteriuria recurs should be given a more prolonged course of the appropriate antibiotic.

So, what long term drug could we use? TMP-SMZ (trimethoprim-sulfamethoxazole) was continued for eight to twenty-two months in four girls with recurrent UTI, meningomyelocoeles and neurogenic bladder after the usual chemotherapeutic agents proved ineffective. Urine remained sterile in all children while on therapy. Hgb, red cell morphology and WBC count and serum folate levels remained normal. No undesirable side effects of therapy were encountered. "Long term use of TMP-SMZ was effective in maintaining sterile urine in children with repeated UTI, meningomyelocoeles and neurogenic bladders."

As an example, let me cite a case study.

"This patient (A.H.) was born with a meningomyelocoele and spina bifida involving L5 and sacrum. When assessed at age 4 2/3 years, she gave a history of having had recurrent problems for several years with UTI due to E. coli. Her urine cultures grew greater than 100,000 E. coli and she was started on ampicillin. A urine culture five days later grew a coliform resistant to ampicillin, so that she was placed on chloramphenicol for ten days. A urine culture 1 1/2 months later again grew a coliform reported sensitive to ampicillin, so that TMP-SMZ was commenced in twice daily dosages. Since TMP-SMZ has been used, all urine cultures have grown no pathogens. She has been on it continuously for twenty-one months without difficulty. A repeat excretory urogram and cystourethrogram in February, 1972, were unchanged from those when she was first assessed in December, 1970. Satisfactory renal growth has occurred over this interval. Her Hgb and WBC count have remained within the normal range as has her serum folate. Morphology of red cells is normal. Ability to concentrate urine remains normal with frequent random urinary samples showing specific gravities above 1.020.

"The ideal urologic management involves a minimum of therapeutic inter-
vention. Such conservative therapy must insure the preservation of renal function, control of urinary infection, and be age appropriate with regards to urinary continence; e.g., use of diapers in infants and timed voiding in older children. Various chemical agents can be used to increase the percentage of children who can be managed by these simple measures.  

"Clean intermittent catheterization was introduced in 1974." Results would indicate this to be an excellent method of bladder drainage. Urinary infection and functional deterioration of the kidney can be treated, controlled or prevented in most cases by this means. C.I.C. can be started in the neonatal period and continued for prescribed periods of time or indefinitely. C.I.C. is advocated in all girls and is the method of choice for boys. "A penile urethrostomy provides easy bladder access in boys when the penile urethra is very narrow or when catheterization is difficult."  

C.I.C. in infancy must be carried out by an adult, but with normal intelligence most children are able to self-catheterize from approximately six to seven years of age, sometimes even earlier. "Continence can be achieved in more children on C.I.C. by use of drugs acting on the neuromuscular system, such as oxybutinin, imipramine and ephedrine." Adjunctive surgery has also been used to increase bladder outflow resistance and bladder capacity, thereby improving continence.  

Rhizotomy (cutting of a section of a nerve root) may be indicated in some children and may change the dynamics of small spastic bladders. Functional bladder capacity can sometimes be increased by rhizotomy, making management by C.I.C. possible and obviating the need for urinary diversion.  

Asymptomatic bacilluria is frequently present with C.I.C.; it is probably of less clinical significance than bacteriuria present in catheter specimens of urine from normal children. Antibody-coated bacteria tests are usually negative, suggesting that bacilluria is confined to the lower urinary tract. This tends to be confirmed by the absence of pyelonephritis.  

"Suprapubic expression (crede) is useful in selected patients; however,
as a routine method of bladder emptying, it should be viewed with caution. Suprapubic expression is indicated only when the bladder can be easily emptied, when post-crede residual is negligible, and in the absence of vesicoureteral reflux. It is definitely dangerous in the presence of vesicoureteral reflux when high intravesical pressures are transmitted directly to the kidney during bladder expression.

No suitable external collecting device has been developed for females. In larger boys, a condom-type appliance attached to a leg bag affords urinary control if the penis is of adequate size.

A temporary new opening from the bladder may be needed in children with bladder outflow obstruction in whom C.I.C. cannot be carried out or in the presence of massive reflux with a relatively small bladder because "anti-reflux surgery is technically difficult in these patients."

Supravesical (above the bladder) intestinal diversions still appear to be indicated in "patients with a very small bladder, vesicoureteral reflux and a gaping bladder outflow, or in patients in whom C.I.C. or implantation of an artificial sphincter is not feasible." A non-refluxing conduit is the diversion of choice—either a non-refluxing colon conduit or an ileocecal conduit—because long term results of refluxing ureteroileal cutaneous conduits at ten and fifteen years are discouraging. "The problems associated with refluxing intestinal conduits include pyelonephritis, calculus disease, progressive renal failure, and hypertension."

The incidence and severity of these problems increase with the passage of time. Almost all patients have problems with odor, and many have psychological problems related to the stoma or urinary diversion.

Development of an implantable urinary sphincter has introduced a new dimension in the management of neurogenic bladder dysfunction. An essential requirement for implantation of an artificial sphincter is complete bladder emptying. Patients with significant post-void or post-emptying residual are
not candidates unless they are rendered totally incontinent. "Artificial sphincter implantation, therefore, is suitable for patients with an adequate bladder capacity, an incompetent bladder outflow, and complete emptying."

The artificial sphincter is an apparently simple solution to a complex problem. However, caution and restraint should be exercised when recommending it for very young children who have to manipulate the device regularly. Growth of the child and damage to the prosthesis may necessitate revision or replacement of various components. The complication and failure rate in many centers is high and, apparently, increases with the passage of time.

"Results with the electronic pelvic floor or bladder neck stimulators continue to be disappointing and should be considered entirely experimental. Such devices should be restricted to a few designated centers where carefully controlled studies can be carried out."

New diagnostic procedures have been discovered in the past ten years. The field of urodynamics has proliferated, especially in the past five years. The studies include electromyography of the anal sphincter and the external urinary sphincter either via the perineum or the urethra, urethral pressure profiles, cystometry and uroflowmetry. Studies in children under four or five years is still technically difficult, especially in the uncooperative patient. Urodynamic evaluation is helpful during pharmacologic manipulation of bladder and sphincter function. Urodynamic evaluation is indispensable when implantation of the artificial sphincter is being considered or prior to urinary tract reconstruction.

Intravenous urography is mandatory, and cystography may be indicated prior to initial discharge of the child from the hospital as a neonate. A ten to twenty percent incidence of bladder outflow obstruction has been found and may require treatment prior to discharge from the hospital. "Such apparent outflow obstruction may be a temporary urinary retention following neurological repair of the myelomeningocele." Cystography will demonstrate
the presence or absence of vesicoureteral reflux, which if present, demands closer monitoring of the child. The child with this condition is more prone to pyelonephritis and other types of UTI infections.

Radioisotopes tracing appears to be particular useful in following children after urinary diversion. Providing that no changes in the perfusion and clearance of the radioisotopes from the kidney are detected, repeated intravenous pyelograms are avoided. "Radioisotopes studies, which are usually more expensive, will never entirely replace either intravenous urography or cystography as these latter studies yield greater anatomical detail."57

The antibody-coated bacteria test, which is not yet generally available, may prove useful in accurately localizing the source of bacilluria. This may be of particular use in evaluating the significance of bacilluria in the presence of urinary diversion. Experience to date, however, is too limited for the value or accuracy of this investigation to be definitely substantiated.

When possible, children with myelodysplasia should be treated by a multidisciplinary team. Myelodysplasia is a complicated condition affecting many organ systems so that management should be coordinated when possible by doctors and allied health professionals who have an understanding of the basic goals and objectives outside and inside their specialty. When many specialists in isolation manage problems relating only to their field, fragmented and therefore, suboptimal care is likely to result.

"Drugs may be used to improve bladder emptying or urinary control. The required sites of action of such drugs are primarily the detrusor muscle and the muscles contributing toward the bladder outflow resistance. Drugs used to decrease detrusor muscle tone or eliminate hyperreflexia include propantheline (Probanthine), imipremine (Tofranil), and oxybutynin ( Ditropan). Bethanechol (Urecholine) increases detrusor muscle tone and helps to reduce residual bladder urine volumes. Bladder outflow resistance can be increased by the use of ephedrine and/or imipremine by increasing muscle tone at the bladder neck. Bladder outflow resistance may be reduced at the bladder neck by the use of phenoxybenzamine or at the level of the external striated muscle sphincter by the use of diazepam (Valium)."58

The introduction of C.I.C. and the artificial urinary sphincter have
radically altered the management of the urinary tract in children with myelo-
dysplasia. Supravesical urinary diversion is less often needed, although there is still a place for this type of treatment. When diversion is indi-
cated, non-refluxing intestinal conduits are suggested. The routine use of suprapubic bladder expression has only limited applicability. When possible, the child should undergo urodynamic study, and the family should be made aware of the treatment modalities at the present time. Before proposing a urinary diversion or implantation of an artificial sphincter, detailed expla-
ination of the procedure and the alternatives must be given to parents and patients.

To Treat or Not to Treat?

Life or death in the nursery? Quality of life versus sanctity of life. Spina bifida presents a staggering dilemma: operate quickly and the child will probably live. But, even doctors who have operated on scores of chil-
dren born with open spine admit they cannot accurately predict how severe the inevitable physical handicaps will be. Nor can they rule out possible mental retardation. Withhold the treatment and the child will probably soon die of an infection carried through the defective spine. But, not always. "Some ten to twenty percent of those left untreated do not die, but usually de-
teriorate to little better than a vegetable existence."

It has been proposed that infants who have any one of the following should not be given active treatment, but should be given normal nursing care, to-
gether with symptomatic treatment to avoid pain, discomfort or fits. These criteria are:

1) gross paralysis of the legs (paralysis below third lumbar segmental level with at most hip flexors, adductors and quadriceps being active)
2) thoracolumbar or thoracolumbosacral lesions related to vertebral levels
3) kyphosis or scoliosis
4) grossly enlarged head, with maximal circumference of two centimeters or more above the ninetieth percentile related to birth weight
5) intracerebral birth injury
6) other gross congenital defects—cyanotic heart disease, ectopia of the bladder, mongolism

Further, no active treatment is advised for those children who after closure develop meningitis, or ventriculitis and who already have a serious neurological handicap and hydrocephalus, or later, if any life threatening episode occurs in a child who is severely handicapped by gross mental and neurological defects.

Nevertheless, such policy does not solve the problems and occasionally, leads to disaster. If the object of the selective non-treatment is the early painless death of the infant, then one must do nothing to prolong life. This means no antibiotic therapy for infections, no intensive care, no oxygen or tube feedings and infants should be fed on demand and no more. Active euthanasia is not only illegal, but also could be an extremely dangerous weapon in the hands of the wrong individual.

The natural worry is that in spite of scrupulous adherence to the criteria, some children might live long and with more handicaps than if they had been treated. However, experience in several large hospitals indicate that only a very small minority of such untreated infants would live very long. If they do survive for about six months and appear likely to survive indefinitely, such infants must be taken back into the fold and all their problems treated as if they had been treated from birth. Occasional "hard" cases should not sway the doctor to do what he considers best for the patient and the patient's family.

Like every medical system in the world, Britain's government-run National Health Service faces a chronic dilemma—whether to use new, life-saving techniques for every patient, regardless of the quality of life that can be saved and the cost.

A decade ago, for example, British doctors applied new techniques to
save babies born with spina bifida. The operations were extremely successful, but the surviving infants were deformed and needed continual attention. Thus, scientific innovation contributed to fresh medical and social problems. Today, in contrast, most physicians in London have abandoned the imperative to save every life. Infants with irremediable abnormalities are left untreated, to die, usually within nine months or so after birth.

"A doctor must always ask himself whether he has the right to inflict suffering on another human being and his family." In other words, the doctors determined, consideration must be given to the burden imposed on others by merely keeping an infant alive, since an incurably handicapped child can destroy a family. The National Health Service in Britain operates with a fixed budget and without any open-ended commitment to offer the maximum feasible treatment at all. Doctors recognize, consequently, that doing too much for one patient may deprive another.

"Given the rising cost as well as the scope of technology, a policy of striving to prolong life could cheat others of the opportunity to improve their chances for an active existence." 62

So, is euthanasia the new and only "treatment" for spina bifida? It is a fact that no one in this country has ever been prosecuted for withholding treatment for a child with spina bifida. However, this hardly makes the practice right. It may only mean that local prosecutors are unaware or uninterested in facing such a volatile issue.

Many legal experts feel prosecutions are justified. In the opinion of John Robertson, professor of Law at the University of Wisconsin, "withdrawing care would appear to be a serious infringement of a basic right—the right to life." He concludes that parents, physicians and even hospital staff members who permit the involuntary euthanasia of a child may be guilty of crimes ranging from homicide and manslaughter to conspiracy, child abuse and neglect.
Four years ago, a Superior Court in Maine answered the very serious question when it ruled on the case of a newborn with birth defects other than spina bifida. "The issue before the court is not the prospective quality of life to be preserved, but the medical feasibility of the proposed treatment compared with the almost certain risk of death should treatment be withheld." That court also cast aside family considerations and emphasized the right to life of the child. This case supplies an answer, but not necessarily the answer for the lawyers, doctors and especially, the parents involved.

This thesis attempts to point out some of the new and upcoming treatments or lack of treatment with regard to spina bifida. Is more research into the treatments needed or is not saving the child the right method of care? I cannot even begin to attempt to answer that question, however, I do feel that the research that is done about the disease known as spina bifida should be geared toward finding the cause and then prevention.
Patients Get New Hope

DETROIT (AP) — An artificial urinary sphincter which allows some children afflicted with spina bifida to control their bladders has been implanted successfully at Detroit's Children's Hospital.

Dr. R. Lawrence Kroovand, a urologist, performed the surgery twice on boys aged 6 and 11. He plans implants on three girls later.

He says the surgery is still rare.

The device is commonly called the Scott sphincter after Dr. F. Brantley Scott of Baylor University School of Medicine, who developed it several years ago.

Spina bifida is the second most common birth defect next to congenital heart disease, Kroovand said. It originates in the embryo during the first month of pregnancy if cells which later form the spinal cord do not develop.

The spinal cord then remains a flat plate of nerve cells which the bony vertebrae of the spine are unable to enclose.
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