

Oral presentation

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Casey Memorial Lectureship :Adult consequences of spina bifida

John M Mazur*

Address: Department of Orthopaedics, Mayo School of Medicine, Nemours Childrens Clinic Jacksonville, FL, USA

Email: John M Mazur* - jmazur@nemours.org

* Corresponding author

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I am honored to be the Casey Holter Memorial lecturer. All of us in the Society miss John Holter very much! The Casey Holter Memorial Lecture was established and funded to preserve the memory of Casey, John's son who was born with hydrocephalus.

The purpose of the Casey Holter Memorial Lecture is to encourage original thinking in a way to improve the care of individuals with spina bifida and hydrocephalus. Having kept this purpose in mind, I have chosen the title "The Adult Consequences of Spina Bifida" with the intent to review the outcome studies documenting our successes, and unfortunately in some cases our failures. By looking at these outcome studies, it is hoped that we can recognize areas of potential improvement in the treatment of children so they may have more fruitful lives as adults. In this review I have included the work of neurosurgeons, neurologists, orthopaedists, psychologists, sociologists, educators as well as physical and occupational therapists.

Unfortunately no one has a great deal of experience treating adults with spina bifida. The patients who were born prior to 1950 would not have benefited from the shunts we have today for hydrocephalus. Very few adults born with spina bifida prior to 1950 are alive today, as most of those infants would have died in childhood from complications of hydrocephalus. Those individuals born between 1950 and 1970 would have benefited from having their hydrocephalus shunted, however, urological care was not as sophisticated as it is today. Urodynamic monitoring of bladder function and intermittent straight catheterization had not been developed. Obstruction and reflux lead to chronic pyelonephritis. Unfortunately many of those individuals died in their late teens or early twenties because of renal failure. This group, like those born

before 1950, is small with few survivors and little is known about them.

The children born after 1970 have benefited from both the shunts for hydrocephalus and modern urological care. This group has survived to adulthood and represents a growing population, which will present new challenges to us as caregivers. I will attempt to summarize some of the outcome studies dealing with adults who are in their second, third and fourth decades of life [2,3,8-10,13].

The major question is "how often are adults with spina bifida leading successful and productive lives?" There are several studies addressing the quality of life, level of independence and self-determination. [4,7,9,11,13,14,16,20-24,27]. Using the Objective Checklist – ACL Autonomy scale, the Days of Coping Checklist – WCC Problem Focused Coping Scale and the Denver Self Care Aging Journal, Monsen [5] showed that adolescents and young adults with spina bifida are less independent than a matched comparison sample of adolescents and adults without spina bifida. A few intervention studies (18, 19) have shown some promise in improving the independence and self care of these young adults through mentoring, coaching and peer support groups. One measure of success and independence would be the ability to seek and gain employment [3,26]. Bomalaski, Teague and Brooks looked at the employment records for individuals with spina bifida. As urologists they were looking at the long-term impact of urological management on the quality of life in individuals with spina bifida. Their study showed that only 17% of individuals with spina bifida were fully employed and 14% were part-time employed. Fourteen percent of the adults were still in training while 6% could not find a paying job and volunteered as unwaged workers. The unfortunate statistic is that 49%

were clearly unemployed and had no hopes of ever finding jobs. There are many reasons why adults with spina bifida may have difficulties in finding a job. The average IQ for an individual with spina bifida is 80, with 10% of this population having an IQ less than 70 [26]. Many have difficulties getting the level of education necessary for a highly technical job. Many are still socially incontinent and remain continuously wet with urine [3]. Being wet with urine gives them a foul odor making finding a job difficult, as they are not accepted socially. Motivation is key to any occupation. Unfortunately many of our adults with spina bifida have developed a passive co-dependency with their parents making the break from family difficult. Training opportunities may not be available to this group, depending on their community. Mobility and transportation can also be issues depending upon how independent they are with their walking or wheelchairs. Communities can vary a great deal with regards to wheelchair accessibility. Looking at all these factors that adversely affect employment, we can make some changes to improve the situation. More diligent neurosurgical monitoring of shunts can prevent shunt failures that may adversely affect cognitive function. Urologically we can strive to make them more continent. Newer advances in urology will undoubtedly improve the statistics on urinary incontinence. The antigrade colonic enema procedure has been a great advance in bowel continence. Early recognition of psychological problems can lead to early counseling to hopefully improve their motivation for independence. We can encourage our communities to provide training for our children with spina bifida. We can always work harder to help them gain mobility, either with wheelchairs or with walking. Hopefully over the next decade, we will be able to present data showing improvement in the employment statistics.

Getting married is another sign of independence from parents [17]. Twelve percent of the thoracic and high-lumbar level men, and 33% of the low lumbar and sacral level men were married. There was a slightly greater tendency for the women to be married. Thirty three percent of the thoracic and high-lumbar level women, and 75% of the lower lumbar and sacral level women were married. An obvious obstacle to marriage would be sexual dysfunction related to their spinal cord abnormalities and paraplegia. Despite their paraplegia, however, surveys of men with spina bifida found that 72% were able to achieve an erection with 54% being able to have an ejaculation. Thirty-one percent of the men were sexually active. The women have less of a problem in this area. Forty-nine percent of the women were found to be sexually active [6]. Looking at the population who are sexually active, 8% of the men were able to father a child and 34% of the sexually active women were able to get pregnant and deliver either vaginally or by Caesarian section [17].

Mobility is also important for independence. There are varying statistics between studies but overall there is general agreement that the low lumbar and sacral level individuals are more likely to walk than the thoracic and high lumbar level individuals [1]. Looking at adults with the low lumbar and sacral levels [5], we find 65% of this population are community ambulators while 20% can walk at home but use a wheelchair for long distances. Five percent are non-functional walkers who can walk for exercise, but walking serves no purpose or function for them. Ten percent of this group are wheelchair ambulators completely. Of the thoracic and high lumbar level patients, only 10% are able to walk in the community, 10% are domestic ambulators, 10% are nonfunctional ambulators and 70% are wheelchair ambulators [25].

For those adults with significant weakness in the legs, wheelchair ambulation is definitely more efficient and preferred by most adults with high-level spina bifida. The wheelchairs offer more speed, take less energy and are more convenient. It is much easier to get in and out of a wheelchair than to put braces on and to take them off. Walking may not be worth the extra effort.

Walking does have some advantages for those adults that have maintained the ability and motivation to walk. Walking allows you to be accessible to more places. It gives you more exercise, which could help with weight control. Walking and weight bearing may help prevent osteoporosis and prevent fractures. Adults with spina bifida who walk tend to have fewer serious pressure skin sores than those confined to a wheelchair.

Musculoskeletal pain will be one of the reasons why adult patients with spina bifida will seek medical care. Thirty-five percent of the adults surveyed complain of upper extremity pain and most of these have either elbow or wrist pain from overuse. The overuse is most likely from using crutches for support in walking. Treatments consist of modifying their activity, physical therapy, orthotics and anti-inflammatory medication.

Back pain is common and requires extensive work up. The differential diagnosis for back pain includes tethered cord, scoliosis, spondylolisthesis as well as mechanical problems related to their abnormal gait.

Pain is extremely rare in the hip and the ankle but very common in the knee. I was able to find only one case report in the world's literature where a total hip replacement was performed on a patient with spina bifida. This patient was a woman with L4-L5 level spina bifida who had right hip pain from avascular necrosis. There was no mention as to why she developed avascular necrosis. With this being the only documented case of a patient with

spina bifida and hip pain, the prevalence of painful arthritic hips must be extremely low in this population. I, therefore, believe we are correct in not being very aggressive with the treatment of hip dysplasia in children with spina bifida.

Degenerative arthritis of the knee, on the other hand, is turning out to be a major problem affecting the adult with spina bifida. Williams et al. [29] in 1993 studied 72 community ambulators, aged 23 to 39. Painful degenerative arthritis of the knee occurred in 24% of adults with spina bifida who walked. There are two main causes of arthritis of the knee in adults with spina bifida. The knee arthritis is related to (1) muscle weakness about the hip and (2) weak plantar-flexion of the ankle. The weak hip muscles lead to a gluteus medius lurch causing the trunk to lean laterally beyond the knee on the coronal plane. This causes a valgus force at the knee. This in turn leads to medial instability and eventual arthritis. The second cause of arthritis is the weak calf muscles. Gait analysis [12,28] has shown that the weak or nonexistent calf muscles leads to a poor push-off at the end of stance phase in the gait cycle. The patient, in order to move forward, must externally rotate the foot and tibia. The femur tends to rotate inward as the tibia rotates outward. The knee then becomes a fulcrum about which the body twists. This external rotation, valgus force leads to medial instability and eventual osteoarthritis of the knee. Three treatment plans for knee arthritis have been proposed. (1) Williams and Menelaus [29] recommend the use of a knee orthosis for prevention and treatment of the knee instability. (2) Vankoski [28] recommends forearm crutches and (3) Lim [12] suggests that a tibial osteotomy be considered to correct the external tibial deformity. Lim feels that these derotational osteotomies will delay if not prevent knee instability and later painful arthritis.

The most frequent, costly and worst problem seen in adults with spina bifida is skin breakdown. David Shurtleff [23] found that 85% of adults with spina bifida have had problems with skin sores at least one time in their lives. Some of course suffer from chronic skin ulcers. The cause of the skin sores is insensate skin and deformity, either from scoliosis, kyphosis, pelvic obliquity or contractures. Prevention and treatment must include the correction of deformity, patient education and wheelchair modifications in addition to treating the skin ulcer with wound care.

In order to improve upon the results we have seen from our adult outcome studies, we need to start the rehabilitation process while the children are still young. I will briefly summarize our rehabilitation goals and treatment plans that we set up for children beginning in the newborn nursery. Much of what we do now in Orthopaedics

is attributed to the work of Malcolm Menelous and John Sharrard.

Malcolm Menelaus was a member of this society from 1960 until his death, September 12, 2000. Malcolm gave the Casey Holter Memorial Lecture in 1976. The title of his lecture was "Orthopaedic Management of Myelomeningocele. A Plea for Realistic Goals and Minimum of Surgery." I was very close to Malcolm as he was my mentor when I did my fellowship at the Royal Children's Hospital in Melbourne, Australia. Malcolm was known throughout the world because of his interest in the care of severely disabled children, in particularly children with spina bifida. His book "The Orthopaedic Management of Spina Bifida Cystica", was published in Edinburgh in 1971 by E&S Livingstone. Now in its third edition, it will stand as a monument to Malcolm's industry and dedication to the care of his patients. Malcolm was influenced a great deal by the work of John Sharrard who was also a member of this Society and died in 2001. John was known as one of the world's outstanding orthopaedic surgeons. He was an orthopaedic consultant in Sheffield where he participated in the multispecialty clinic established for the orthopaedic management of children with myelomeningocele. John Sharrard published well over 100 papers and 30 textbook chapters, many concerned with the paralytic diseases of children. There is no question that this Society will clearly miss John Sharrard and Malcolm Menelaus.

John Sharrard and Malcolm Menelous taught us the importance of first evaluating the child's neurological status. We don't just concentrate on the spinal cord lesion created by the spinal defect but look at the total neurological picture. With the advent of the MRI we certainly recommend imaging of the brain and the entire spinal cord. We are looking for other associated anomalies such as a syringohydromelia, tethered cord and the Arnold-Chiari malformation. This slide demonstrates a syringohydromelia in a child with spina bifida. The next slide reminds us of the association of the Arnold - Chiari malformation with spina bifida. My main goal as an orthopaedic surgeon is try to determine whether or not I am dealing with a flacid paralysis or possible dealing with a spastic component to the child's neurologic picture. Once I determine this, I then try to determine the lesion level in the spinal cord. Before the spinal defect is closed, I recommend a manual muscle test to determine the spinal cord level of lesion. This manual muscle examination is repeated 5-7 days after surgery and then every 3 months. The manual muscle testing is limited and therefore inaccurate in the newborn period. We are restricted on positioning the patient after surgery as the infants need to be kept prone to protect the surgical wounds. The infant state itself makes the examination inaccurate as the infant is obviously not going to follow commands. Despite those

limitations, we try to determine the presence or absence of muscle function with a simple grading system. We record an "X plus" if the muscle appears strong and an "X minus" if the muscle contracts but appears weak. A zero is recorded if the muscle does not function at all. If they have no muscle control of the lower extremities, we classify the child as a thoracic level patient. Lumbar one level (L1) would have hip flexion. Lumbar two (L2) level would have hip adduction and flexion. All of these patients (thoracic and upper lumbar) would have the same functional capabilities and therefore we would put all of these patients into one group. This thoracic and upper lumbar group would have no sensation in the lower extremities, and are prone to develop skin sores. The patients with the Lumbar 3 level of lesion have some quadriceps function but the strength is weak and may not support weight bearing without long braces. The patients with the Lumbar 4 level have strong quadriceps and hamstrings. The patients with the Lumbar 5 level lesion have all of the functioning muscles as those with the Lumbar 4 level with the addition of strong dorsiflexors of the ankle. The patients with Sacral level lesions have all the functioning muscles as do the patients with the Lumbar 5 level of lesion with the addition of the strong plantarflexors.

Once we determine the level of lesion and come up with reasonable goals and expectation for the infant we begin parental education as soon as they can handle the emotional impact of having a child with spina bifida. Many times the parental education is delayed because of the grief that sometimes is initially overwhelming to the parents. Physical and occupational therapy is started immediately to begin range of motion and stretching exercises to prevent joint contractures. The parents are warned that motor development is normally delayed and they can not compare their infant to a child without spina bifida.

Most infants with myelomeningocele learn to belly crawl as their first means of mobility. Infants with strong voluntary hip flexion and knee flexion may creep on all-fours (assume the all-four creep). Children with low lumbar and sacral level lesions learn to walk by two years of age with or without bracing at the ankles. Children with high-lumbar level of paralysis often require crutches and bracing up to the hip. Children with thoracic and high lumbar paralysis may eventually stand upright and walk but need support of the hips, knees and ankles. This support must be provided by extensive bracing and mobility devices such as a parapodium, reciprocal gait orthosis (RGO) or hip-knee-ankle-foot orthosis(HKAFO) used in combination with crutches or a walker. As a result, children with the thoracic and upper lumbar levels are unlikely to walk before the age of three or four.

The sacral level patients should have no problems walking and they require little intervention. Orthotically they may require an AFO if the plantarflexors are weak, a supramalleolar AFO if there is a flexible ankle deformity, or a foot orthosis (insert) for some minor foot deformity such as a cavus or flat foot. Many times they require no bracing at all.

The low lumbar (L4 and L5) level patients should be able to walk but usually function better wearing ankle foot orthoses (AFO). Several studies have been performed assessing the ability to walk with various orthotic devices. In children with low level lesions, AFO's are helpful. Velocity of walking is improved with better mechanics of walking using an AFO. The AFO is used if the quadriceps strength is grade 3 or better. The AFO prevents foot drop and allows foot clearance during gait. We would anticipate 75% of our children with low lumbar levels to maintain the ability to walk as adults. Patients with L-3 spinal cord level of paralysis need long braces or wheelchairs as their quadriceps are usually weak even if they show some innervation.

For the high lumbar and thoracic level individuals, HKAFO's or RGO's are needed to maintain hip and knee extension. The velocity of walking seems to be faster with HKAFO's than RGO's if a swing to gait is used. If a reciprocating gait is used, the RGO seems to be superior. Ambulation with either the HKAFO or the RGO offers some physiological and psychological benefits. These benefits include improved bone density, better bladder emptying and improved self-esteem. These benefits, however, have not been proven with random trials. A recommendation for the future would be to do a randomized controlled study looking at the benefits, costs and functional outcomes of walking versus using a wheelchair. We need to look at outcomes such as getting around in the home, school, or community. Elements of such a study should also include: bone density, incidence of decubitus ulcers, upper extremity function, activities of daily living, incontinence, employment, marriage and participation in recreational activities like sports.

As the children with thoracic and high lumbar lesions approach adolescence, most rely increasingly on wheelchairs for mobility. This occurs for several reasons. As the children get taller and gain weight, mobility becomes more difficult. A lot of energy becomes necessary to walk and mobility using a wheelchair becomes more efficient and faster than walking. Because most children with myelomeningocele will not become effective community ambulators, the supplemental or primary use of the wheelchair should be considered in children. Wheelchairs offer the advantage of speed, efficiency and are now socially more accepted. Motorized wheelchairs can be

used as early as 2 or 3 years of age if the child is mentally normal with normal hand function. In addition to the severity of muscle weakness, impaired hand functioning, decreased IQ and motivation may affect the ability to walk. Walking may not be a realistic goal and wheelchair training should be started early for those children with a combination of a high level lesion, spinal deformity, joint contracture, and cognitive impairments.

It has been controversial as to whether or not we should immediately prescribe wheelchairs for these patients or make an attempt on trying to teach them to walk with braces. Our outcome studies show that even if these patients can learn to walk as children, they will definitely choose a wheelchair in adulthood. In Australia where walking was strongly encouraged, an outcome study there showed only 10% of the high level adult patients were able to remain community ambulators, only 30% retained the ability to walk but used a wheelchair in the community for long distances, 15% became domestic ambulators walking only in the home and 45% used a wheelchair everywhere. Walking is not the most important aspect of life to these individuals. Self care may be the most important priority for our children as they go into their teenage and adulthood years. Communication skills is the second priority and ambulation would be the third priority. Included in self care is the need to be continent of stool and urine. Our urologists work diligently to evaluate bladder function with urodynamic studies early in life and work hard to keep these children dry. I mentioned earlier in this talk, staying continent of urine and stool is crucial for social integration. Success in adulthood as in getting a job, having friends and getting married is a function of staying dry from urine and staying clean.

Orthopaedically for the children with high level paralysis, we primarily concentrate on preventing contractures so they can better fit into their braces or wheelchairs. We try to encourage the use of the wheelchair early in childhood. We do have the occasional parent who insists that their child with spina bifida be given the opportunity to walk. In these cases we do prescribe standing devices like this parapodium. The parapodium is particularly useful if the sitting balance is poor and there is a spinal deformity. For the older children and young adults we can use this type of standing device called a swivel walker. Propulsion in the swivel walker is initiated with a lateral and somewhat forward weight shift using the abdominal, arm and upper trunk muscles to rotate forward advancing the brace. This allows them to be at eye level with their peers, possibly stand at a table or a blackboard at school. Standing could have some psychological benefits. In some isolated cases with highly motivated parents and children, we do prescribe the reciprocating gait orthosis (RGO). Forward propulsion with RGO is initiated with a lateral weight shift

and trunk extension causing hip flexion on the non-weight bearing side. We would prescribe an RGO around age 4. A prerequisite is that they have good head and trunk control. Most of our children who walk with these devices will still need a wheelchair for long distances. There may be some benefit from a walking program. The adults who walked as children seem to be more independent in their activities of daily living than the adults who never walked. The adults who walked as children are more independent in their transfers to and from a chair. I would not recommend this for everybody and would only offer it to people who actually ask for it and show the motivation. If the motivation is not there, the patients, parents and caregivers end up wasting a lot of time and money in a process that is doomed from the beginning.

To switch gears, I would like to discuss some Orthopaedic management issues.

The clubfoot is the most common orthopaedic deformity seen in children with spina bifida. We begin treatment of the clubfoot with serial casting to try to stretch out the deformity. We do know this could possibly lead to skin sores because of the lack of sensation, however, we have not had any significant problems with pressure sores in the infants as long as we change the cast frequently. We consider surgical intervention if the casting is unsuccessful. Surgery is delayed until age 2 or 3 years of age when the children begin to show signs of walking. We usually make a fairly long incision on the back of the foot. We release all the ligaments and tendons. We then pin the foot in the corrected position. A cast is used for 2 to 3 months post operatively. Afterwards, the children are given AFOs to maintain the correction. The rear entry floor reaction AFOs with solid ankles are the most appropriate short leg brace for children with spina bifida. This type of brace helps with proprioception as well as helps support the ankle. As the children walk, the brace vibrates with each stride and the vibration can be felt near the knee where they have sensation. By having the plastic touch the skin on the proximal tibial area, the vibrations are felt from the foot through the brace to the knee. This will substitute for their lack of proprioception at the ankle. Hopefully, this will prevent ankle problems such as a Charcot joint.

The children who have excessive dorsiflexion of the ankle or calcaneus deformity can have significant problems. Their feet become deformed because the anterior tibial muscle is strong and has no muscle working against it. This leads to a prominent calcaneus and a cavus deformity as seen in this x-ray. This foot deformity is particularly problematic by the fact that there may be no sensation over the heel. The deformity can lead to a severe heel sore especially in children with no sensation on the bottoms of

their feet. Prevention is key. We recommend releasing the anterior tibial muscle or transferring it to the calcaneus in early childhood. Afterwards these children benefit from wearing the floor reaction AFO as previously described.

The sacral level patients have very few orthopaedic problems. Clawing of the toes can occur from intrinsic foot muscle weakness. The clawed toes can be treated surgically by releasing the long toe extensors and transplanting them into the lateral cuneiform bone of the foot. The big toe is corrected by transferring the long toe extensor into the first metatarsal and fusing or tenodesing the interphalangeal joint. The tendon transfer to the little toes is called the Hibbs procedure and the operation for the great toe is the Jones procedure. The postoperative results are usually quite good.

Hip surgery has been an area of controversy over the years. We have in the past recommended surgical management of the dislocated and subluxed hips. There are essentially four situations as described by this two dimensional Punnett square. We can have a high level patient requiring high braces and crutches with bilateral hip dislocations. In this situation we would never reduce the hip. In the low level patient with one hip dislocated, we would always recommend reducing the hips. In the case where you have a high level patient and a unilateral hip dislocation, the hip is reduced only if the other leg is good or there are contractures that need released. If there are bilateral dislocated hips in a low level patient, we would treat them again only if contractures need to be released. The treatment includes varus osteotomies with internal fixation and tendon transfers to help balance the muscles around the hip joint.

As we have gained more and more experience with children with spina bifida, we have come to realize that surgery around the foot is the most beneficial. Knee surgery may play a bigger role in the future. Hip surgery is rarely indicated except in the low level patients who can walk without support. Of course, spinal surgery preventing scoliosis is necessary in both the high and low level individuals.

In summary, I have presented the results from some of the available outcome studies dealing with adult patients with spina bifida. I have presented a summary of the Orthopaedic management of these patients. It is hoped that our treatment plans will improve the quality of life of our patients. The adults with spina bifida will present new challenges to the medical community. We must be there to help these adults with the quality of life issues such as employment, recreation and social integration. The babies born today with spina bifida will survive. We must try to improve their quality of life as caregivers and scientists involved with the research and treatment of individ-

uals with spina bifida. In adults, the most common problems will be skin sores, back pain and knee arthritis. We will be prescribing braces for walking or wheelchairs for mobility. We will be performing surgery for correction of deformity and contractures.

Before concluding, I would like to pay tribute to John William Holter who established for this Society, the Casey Holter Lecture and the Casey Holter Essay Prize. John was an engineer and successful businessman. In 1955 his son Casey was born with hydrocephalus. John Holter devoted his professional skills to the development of shunting systems for the treatment of hydrocephalus. Working with Dr. Spitz, the first one-way valve was inserted at the Children's Hospital of Philadelphia in March of 1956. British surgeon George MacNab visited Dr. Holter, brought his device to England and started to use it at the Hospital for Sick Children, Great Ormond Street. This eventually led to its worldwide use. In 1963 John Holter was made an honorary member in the Society for Research into Hydrocephalus and Spina Bifida, becoming its first nonmedical member. He was a vigorous participant at the society's annual meetings, which he attended regularly. In 1976, Sheffield University conferred an Honorary Doctor of Science degree for him. In 1998 he was invested as an Honorary Commander of the Order of the British Empire, an honor of which he was immensely proud. John Holter was a dear friend to all of us in the Society. He remained active in the Society attending its meetings until his death in December of 2003.

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