CONGENITAL LUMBOSACRAL MYELOMENINGOCELE WITH INCONTINENCE

A CONTRIBUTION TO THE UNDERSTANDING OF BLADDER PHYSIOLOGY*

EBEN ALEXANDER, JR., M.D., FRED K. GARVEY, M.D., AND WILLIAM BOYCE, M.D.

Sections of Neurosurgery and Urology, Department of Surgery, Bowman Gray School of Medicine, Winston-Salem, North Carolina

(Received for publication September 26, 1953)

Advances in the care of paraparetic and paraplegic patients during the last 20 years are well known. The mortality of patients with paraplegia has been sharply reduced and the life expectancy of the individual patient has been lengthened by the prevention of decubiti and the elaboration of practical methods for long-term rehabilitation of such persons. Of even greater importance is a better understanding of bladder physiology with a reduction in the incidence of chronic urinary tract infection, one of the more common causes of death in paraplegic patients in years past.

The value of the contributions by such pioneers as Head and Riddoch,12 Munro,20,21 and others cannot be overestimated. More recently the papers of Prather,23 Lewis,15 Freeman and Heimburger,10,11 Botterell and coworkers,1 Shelden and Bors,24 and numerous others have made possible a better understanding of the physiological functions of the normal human bladder and of the human bladder deprived of connections with the brain, or on occasions, with the spinal cord.

In brief, it has been found that in a patient with a spinal cord severance above the conus medullaris there may develop, after weeks or months, a bladder with the power to empty completely and automatically, a hypotonicity of the bladder with large residual, or a severe hypertonicity with small capacity. The weight of evidence points to the S3 anterior nerve roots as the principal nerve supply, reflex or otherwise, to the detrusor action of the bladder. The anterior S2 and S4 roots carry some fibers to the bladder but are of much less importance than the anterior roots of S3.22 Consequently, in performing anterior rhizotomy for the relief of reflex motor spasms of the legs in paralyzed patients, warning has been given to spare the S3 anterior roots.16 This warning in general has seemed to be of importance, since after bilateral anterior rhizotomy of D10 through S1, patients with severe reflex spasms of the legs with poor bladder function have usually developed improved bladder automaticity.

On the other hand, Shelden and Bors,24 and more recently Stellar,25 have
demonstrated similar improvement in bladder function in paraplegic individuals in whom they have injected 15 cc. of absolute alcohol into the lumbar subarachnoid space. Similar studies by Meirowsky, Scheibert, and Hinchey in severing the S3 and S4 anterior roots have shown improvement in bladder function in certain patients. The injection of absolute alcohol undoubtedly destroys the S3 anterior roots, previously felt to be so important in the reflex emptying of the bladder. Heimburger and Freeman have improved the capacity and emptying of the bladder of certain paraplegic patients with hypertonic bladders by procaine injection of the S3 roots bilaterally.

Patients born with varying degrees of congenital anomalies known as meningocele or myelomeningocele in the lumbosacral or lower thoracic region often present problems similar to those seen in patients rendered paraplegic by trauma, tumor or infection. There are often other congenital anomalies of the nervous system or of other parts of the body, and these, combined with the disturbed function associated with the paraplegia in early life, contribute to a high mortality in this group. The number of such patients who survive to the age of 20 is not accurately known, but is probably small. Judging from the report of Fisher, Uihlein, and Keith of 350 cases seen at the Mayo Clinic, this would be about one in ten. However, the series included patients with cranium bifidum in whom the mortality is lower, and their report did not give details as to the number of patients with such congenital defects of the lower spine who survived to the age of 20 years. Likewise, the reports of Ingraham et al. and of Cohn and Hamby do not clarify that point.

There is little question, however, that the incidence of incontinence of urine and feces in patients with meningocele and myelomeningocele of the lumbosacral region is high. Numerous surgeons have attempted to alleviate these difficulties with control of excreta by colostomy, ureterosigmoid anastomosis, the creation of closed loops of bowel in which the urine can be stored, and numerous mechanical aids for the control of urethral emptying of the bladder. It is only fair to say that no one of these methods has succeeded in benefiting such patients with sufficient frequency to be adopted as a standard procedure. It is possible, however, that the application of knowledge gained about the reflex emptying of the bladder in paraplegic patients can be used to advantage in certain patients with congenital lesions of the lower spine. The apparent success to date of a procedure in which this was done has stimulated this report.

CASE REPORT

NCBH No. 136996. M.D., a 23-year-old single white female, was admitted on Jan. 16, 1952 for correction of urinary incontinence.

When she was born it was noted that she had a small dimple in the midline over the sacrum. She had been incontinent of urine and feces since birth. There had been weakness of the legs and deformities of the feet, the latter requiring arthrodesis. Her mental development had been normal and her physical development, except for the abnormalities already noted, was normal. There had been no difficulties
above the level of the waist, the head was not enlarged, and the neck was not stiff. She had succeeded in maintaining fairly satisfactory bowel control by remaining constipated and giving herself an enema every day or every other day.

Her urinary control, however, was a serious problem. She was wet almost constantly. In September 1951 she was admitted to another hospital where after extensive studies with cystometrograms the external urethral sphincter was tightened by an anterior colporrhaphy. This had resulted in her remaining dry for as long as 2 hours during the day but necessitated firm suprapubic pressure for emptying of the bladder. She still was not continent of urine at night.

There was no history of deformities in other members of her family.

**Examination.** Temperature was 98°, pulse rate 80, respiratory rate 16, and B.P. 120/80. The patient was well proportioned and attractive. There were no abnormalities above the level of the chest.

She walked slowly with a limp on the left side. There was a midline dimple of the skin over S2-S3, measuring 1 to 1.5 cm. in diameter and covered by thin translucent skin. When the skin was retracted downward, there was a deep indentation of the dimple, indicating an attachment cephalad to this point.

There was poor development of the legs. There was marked weakness of plantar flexion and moderate weakness of dorsiflexion of the feet, much greater on the left. The scars of operation for arthrodesis of the ankle were evident on the left side.

There was almost complete anesthesia below L4 on the left and below S1 on the right. There was complete saddle anesthesia and in only isolated spots could the patient distinguish gross painful stimuli on either leg below L5. There was relaxation of the anal sphincter.

Blood was normal. NPN was 32 mg. per cent. Urine contained 1-2 WBC per high power field; concentration was 1.014; albumin was negative.

Roentgenograms of the lumbar spine showed a widening at the spinal canal at L4 and L5, with a spina bifida at L5.

Intravenous pyelography disclosed no abnormality of the upper urinary tract. A cystometrogram showed a hypotonic type of bladder with gradual rise until the bladder reached 400 cc., at which time there was a fairly sharp rise in the column of fluid (Fig. 1). There was residual urine of between 90 and 110 cc.
Electromyograms of the bladder were made by previously described technique.\textsuperscript{2,3,6,7} An especially constructed sheath\textsuperscript{*} was passed through the urethra and two ball electrodes of silver-silver chloride were brought into contact with the bladder wall near the base. The insulated spring leads maintained one electrode in contact with either lateral bladder wall during the examination. A third channel of the sheath, connected to an irrigation flask and water manometer, permitted variation in the degree of distention of the viscus and observations of change in intravesical pressure. The electrodes were connected to the Grass electroencephalograph Model III-C and the bioelectric action potentials accompanying muscular activity of the detrusor were recorded in wave form. Spontaneous and voluntary muscular activity of the bladder was thus studied in the resting or nearly empty bladder and with varying degrees of filling. The electromotive force (E.M.F.) and duration of each electropotential shift or wave was calculated as previously described.\textsuperscript{6,7} The interpretation of these records, as based on previous experiments, would indicate that the initial A wave is unaccompanied by muscular contraction and is thought to represent spread of the excitatory process, while the ensuing B and C waves of a single electropotential shift are accompanied by muscular contraction. The magnitude of the E.M.F. of the B or C wave at any given instant is a function of the vector sum of the bioelectric action potentials of all the excited muscle cells at that particular moment and hence an indication of the mass of active muscle. The duration of a single B or C wave is considered to be a reflection of the integration of detrusor activity as successive contractions of contiguous muscle cells occur. For example, spasm of a large part of the detrusor is represented in the electromyogram by a very high E.M.F. of up to 8.0 mv. but of relatively short duration.

The results of this examination of the patient on two occasions prior to operation were identical. Fig. 2 illustrates the average spontaneous waves obtained from the resting bladder. These were of shorter duration and the average E.M.F. was less than that of the normal bladder (Fig. 2). Unlike the normal, which exhibits a spon-

\textsuperscript{*} Provided by Mr. Frederick J. Wallace of the American Cystoscope Makers, Inc., New York, N.Y.
taneous pattern of strikingly constant duration and E.M.F., there was a marked variation in the E.M.F. of both A and B waves, and an occasional wave was much greater than normal with a maximum of 1.52 mv. for the A and 1.90 mv. for the B waves. In no instance was there a duration of either A or B waves that approached the normal. Voluntary efforts to void resulted in electromyograms of considerably less than normal E.M.F. and duration.

Fig. 8. Artist’s drawing of operative findings. The herniation of the nucleus pulposus at L4 was considered an incidental finding.

It was concluded that these studies indicated abnormal parasympathetic innervation of the bladder with poor integration of the detrusor as a whole and occasional “spasmodic” contractions. Muscular contractions appeared to be limited to segments of the bladder that, in the absence of either normal bladder tonus or a satisfactory mass contraction of the detrusor as a whole, were incapable of emptying the viscus.

Pantopaque myelography on Jan. 21, 1952 showed an unusually wide sacral canal over the lower lumbar region with a very sharp midline defect at L4. The column of oil deviated around this on either side without obstruction. It was of interest that the patient experienced a considerable amount of pain during the insertion of the needle, but the spinal fluid was clear, free of cells and contained protein of 30 mg. per cent.
1st Operation, Jan. 23, 1952. The fibrous tract leading directly to a globular mass of firm tissue attached to the dorsal tip of the conus medullaris was excised (Fig. 3). This was a direct fibrous communication between the skin and the spinal cord. Laminectomy at L4 and L5 was carried out, the laminae of the latter being soft and cartilaginous. The ligamentum flavum beneath L5 was intact and removal of it permitted dorsal expansion of the dura mater as if it had been constricted by this tissue. When the dura mater was opened, it was apparent that the tip of the conus medullaris lay at the level of L5-S1. The canal was at least twice as large in all diameters as normal at this level. Associated with this caudal location of the conus medullaris, the L3 nerve roots passed upward at an angle of 10 to 20° in their course from the spinal cord to the intervertebral foramina. The L4 root on the left was larger than the other roots, and both L5 roots, right and left, passed downward slightly to their exit from the spinal cord to the intervertebral foramina. The sacral roots were small and were asymmetrically placed, the intervertebral foramina on the left being 2 to 3 mm. lower than those on the right.

There was a 2.0 cm. mass of scar tissue on the dorsal tip of the conus to which the fibrous tract in the skin was attached. Except for meticulous division of all fibrous connections between this and the surrounding dura mater, it was not otherwise disturbed or dissected. It resembled grossly the mass of nerve tissue often seen at the base of a myelomeningocele removed from the lumbosacral region of infants.

At the level of L4 in the midline there was evident, when the cord was displaced to the right, a very prominent bulge anterior to the cord. Because of the wide diameter of the canal and the attachments of the cord to the nerve roots, the cord was suspended dorsally and it was found that this mass was apparently not in contact with the cord. It was white, 1 cm. high, and 0.6 cm. broad at its base. The anterior dura mater was opened and the mass, which was cartilaginous, was removed. The pituitary rongeur could be passed into the intervertebral space through the opening created by removal of the protrusion, and further fragments of the intervertebral disc were thus removed. It was the impression at the time of operation that this mass, prominent as it was by myelography and at the time of direct visualization, did not actually contribute in any way to the patient’s disability. It was completely removed. Following this, the dura mater was tightly closed and the wound was closed with silk. The patient withstood the operation well.

Pathological Diagnosis. Fibrocartilage.

2nd Operation. After satisfactory healing of the wound had taken place, the patient was returned to the operating room on Jan. 30, 1952 and under general anesthesia the wound was re-opened. A #18 Foley catheter was inserted into the bladder and attached to a water cystometer apparatus. The nerve roots were again identified, using as identification the 4th lumbar interspace at which the protrusion of cartilage had been removed and the 2nd sacral interspace at which a silver clip had been placed at the 1st operation and confirmed by a roentgenogram taken before the 2nd operation.

The level of the cystometer was adjusted to that of the partially filled bladder. Stimulation with 8 volts of faradic current resulted in movement of the hips and legs when the L3 and L4 roots were stimulated. Stimulation of the 5th lumbar and 1st and 2nd sacral roots resulted in no muscular movement and no change in the level of the cystometer.

Stimulation of the 3rd sacral root on the left resulted in a rise of fluid level in the
cystometer of 11 cm. On the right side the rise was approximately 5 cm. These Results were obtained on repeated stimulations of equal voltage.

Stimulation of the S4 roots and of the filum terminale gave no response.

The 3rd sacral roots were then carefully isolated bilaterally and crushed with a straight hemostat. Anterior and posterior roots were not separated because of their small size. All remaining roots were left intact. The wound was closed in layers with silk.

Postoperative Course. An indwelling catheter was left in place. On one occasion when the catheter was removed, a residual of 1200 cc. developed and the patient was not able to empty her bladder at that time. She was discharged from the hospital with the catheter in place.

Feb. 13, 1952. The patient was able to void 200 cc. of urine with a residual of 60 cc. Her urine contained a few pus cells.

Feb. 18, 1952. The patient voided 300 cc. with complete emptying of the bladder.

Feb. 28, 1952. The bladder held 400 cc. of urine; no infection, and no residual. She found that she could hold her urine 8 hours without voiding and was not wet at night.

Aug. 7, 1952. Patient maintained her bladder control 4 to 5 hours at a time without emptying her bladder. She then emptied her bladder completely without residual. She had had no pain. She was not incontinent at night.

Sept. 26, 1952. The cystometrogram was repeated (Fig. 4). The bladder showed the typical hypotonic chart. There was no infection of the urinary tract. Electromyograms of the bladder at this time indicated only an occasional wave occurring about once in 3 minutes in the resting bladder with very low E.M.F. and of short duration (Fig. 2). There was complete absence of any bioelectric potential with voluntary efforts to void.

DISCUSSION

The exact mechanism by which the function of this patient's bladder has been so strikingly improved by crushing of the 3rd sacral roots is not at once evident. Correlation of the cystometric and electromyographic studies suggests that she had marked hypotonia of the detrusor with contractile hyperactivity of segmental character. These hyperactive segmental contractions were completely abolished by block of the 3rd sacral roots. It is conceivable that elimination of these segmental "spasms" may permit an integrated contraction of the detrusor as a whole. The "trigger" mechanism for such an emptying contraction may arise from the intrinsic innervation
of the detrusor, either as a result of physiologic filling of the bladder ("stretch reflex"), or a sudden increase in intravesical pressure secondary to voluntary increase in intra-abdominal pressure applied as bladder filling approaches the critical capacity.

The electromyograms in this patient following sacral nerve crush are in complete accord with the recent report of Corey and Vest, who have demonstrated that the typical "A–B" wave complex in a variety of experimental animals can be elicited only by stimulation of the parasympathetic nerves or by administration of acetylcholine.

Reference should be made to a similar case reported in 1944 by Meredith. His Case 2 had difficulty with his legs and bladder. At operation the tip of the conus medullaris seemed to be bound down by scar tissue at the lumbosacral level and freeing of the adhesions resulted in "recession cephalward of the entire cord." Postoperatively there was improvement in bladder function, apparent loss of anesthesia in the legs and increase in motor power of the legs.

In our case, the conus did not move upward when freed of adhesions and in fact the cord was well relaxed. The finding of a sharp rupture of the nucleus pulposus at the L4 interspace was felt not to have contributed to the production of cord signs or symptoms, although this possibility cannot be denied. It was our impression that crushing the S3 nerve roots effected the change in bladder function. There has been no change in the legs since operation.

Although this approach to bladder incontinence has been used in patients rendered paraplegic from other causes, it has not been reported as useful in cases of the defect termed myelomeningocele. This new technique may offer hope of bladder control in these patients with myelomeningocele who survive and are otherwise relatively intact. The value of the anterior colporrhaphy performed several months previously is difficult to assess. Possibly it has contributed to the success in bladder control following the crushing of the S3 nerve roots. The fact that it required several weeks for this individual to develop bladder control after crushing of the S3 roots would indicate that conclusions concerning alteration of bladder function by procaine block of various sacral roots might not be valid.

This particular method has apparently not been previously applied to a patient with a congenital defect of the spine and it is entirely probable that only a relatively small number of patients with such congenital defects would be amenable to such therapy. However, this approach to the problem in patients with myelomeningocele and urinary incontinence may in the future result in great improvement of bladder function in these individuals. It is most distressing in certain young people who are otherwise well and able to get about fairly adequately to see the social ostracism that results from constant urinary incontinence.
MYELOMENINGOCELE WITH INCONTINENCE

SUMMARY

1. An adult patient with urinary incontinence from a small myelo-
meningocele has been reported.

2. Operative findings included the tip of the conus medullaris at the
lumbosacral level and a midline ruptured nucleus pulposus at L4, the latter
apparently causing no encroachment on the cord.

3. Bladder continence has been gained since operation, apparently as a
result of crushing of the S3 nerve roots, the only apparent efferent nerve
supply of the bladder. It would appear that blockage of the abnormal para-
sympathetic outflow permitted a more effective coordination and mass
contraction of the detrusor in response to physiologic filling of the viscus.

REFERENCES


2. BOYCE, W. H. Bladder electromyography: a new approach to the diagnosis of urinary bladder


1948, 5: 316-320.


6. COREY, E. L., BOYCE, W. H., and FRENCH, C. R. Electropotential and pressure variations in the


8. COREY, E. L., and VEST, S. A. Electromyography of smooth muscle in the human bladder and


10. FREEMAN, L. W., and HEIMBURGER, R. F. The surgical relief of spasticity in paraplegic patients.

11. FREEMAN, L. W., and HEIMBURGER, R. F. The surgical relief of spasticity in paraplegic patients.
II. Peripheral nerve section, posterior rhizotomy, and other procedures. J. Neurosurg., 1948, 5:
556-561.

12. HEAD, H., and RIDDOCH, G. The automatic bladder, excessive sweating and some other reflex con-

13. HEIMBURGER, R. F., FREEMAN, L. W., and WILDE, N. J. Sacral nerve innervation of the human

Spina bifida and cranium bifidum. Papers reprinted from the New England Journal of Medicine
with the addition of a comprehensive bibliography. Cambridge, Mass.: Harvard University Printing

15. LEWIS, L. G. Treatment of bladder dysfunction after neurologic trauma. J. Urol., 1945, 55:
284-295.


17. MICHOWSKY, A. M., SCHEIBERT, C. D., and HINCHRY, T. R. Studies on the sacral reflex arc in
paraplegia. I. Response of the bladder to surgical elimination of sacral nerve impulses by rhizotomy.

18. MICHOWSKY, A. M., SCHEIBERT, C. D., and HINCHRY, T. R. Studies on the sacral reflex arc in


