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Discussion

DR. JAMES A. O'NEILL, JR. (Nashville, Tennessee): Dr. Hendren is clearly the world expert on this subject of the persistent cloaca and its many variants. There are two entities that he dissected, if you get down to the basics. One is the cloaca and its multiple variations. Second is cloacal exstrophy. He stressed that there are principles of staging, while at the same time bringing in some new concepts, basically posterior sagittal anorectoplasty approaches in order to do almost complete primary repairs.

Now, it is very difficult to hear a ten-minute presentation on a subject that would take months to study. When you look at the manuscript, carefully, there is an enormous amount to be gained.

The evaluation of these anomalies is critical. And of course he uses endoscopy and a number of other things in order to make an evaluation of what needs to be reconstructed, in what order, what kind of staging, and the like. The principles are really the basics of both internal and external reconstruction. And the key here, whether he presented it strongly or not, the key is the urinary tract. Because that is the thing in the end which will kill these patients. You can do a pull-through for the imperforate anus and whether the patient is continent or not is one issue. But long-term, intelligent, well-done reconstruction of the urinary tract is key. And I think that is where Dr. Hendren and his results are clearly outstanding.

I have a couple of questions for you, Dr. Hendren. First of all, in the cloacal exstrophy group, about 50% of those patients have spinal dysraphism. A hundred percent will have tethered cord, as you pointed out. And the question comes up, really, whether any of those patients should have a pull-through. Now, you have indicated that perhaps half of them can have a pull-through. Should we use things such as MRIs to evaluate whether there are deficiencies in pelvic musculature in order to make a more considered decision about that issue? Secondly, what do you think is the ideal approach to vaginal reconstruction? Now, if you have sufficient material to pull through or to pull down the vagina from a posterior approach, well, that works pretty well. But in many of these instances it will be necessary to use some other tissue which is available. What do you think is the ideal?

DR. W. HARDY HENDREN (Boston, Massachusetts): Thank you, Dr. O'Neill. Those are two incisive questions. The first, regarding which cloacal exstrophy patients may be candidates for pull-through? They are the ones who have a good perineal muscle complex which contacts on electrical stimulation. Those babies with a rounded bottom and no gluteal cleft or contractile muscle should in most cases remain with a colostomy. Severe orthopedic disability with confinement to a wheel chair is another state which makes pull-through not feasible. Pull-through was reversed in three cases because loose stools were uncontrollable because colon length was very short. Each family is warned, therefore, that colostomy reversal may be needed in pull-through does not work out well. I have not relied on the pelvic MRI studies to make this decision.

Regarding vaginoplasty, if a vagina is present I try to free it and do a pull-through. If it is too short, the gap can be bridged by using perineal flaps or splicing in a segment of bowel to lengthen the vagina. The cardinal point is to not waste colon to make a vagina in cloacal exstrophy patients. They need every possible bit of colon mucosa for water absorption to give them a solid stool.

DR. BARRY O'DONNELL (Dublin, Ireland): I feel I should first recognize Dr. Jonathan Rhoads. It is 50 years since Dr. Rhoads did the first abdomino-perineal pull-through of a high imperforate anus. That was a milestone. This is another.

It is difficult to discuss Dr. Hendren's paper without descending into the lower levels of hagiography and hero worship. I have assisted him on two cloacal extrophies on his five operative visits to Ireland. The first one lasted 16 hours; the second 24-½ hours. I have had the somewhat unnerving experience of booking an operating room for two days for a single operation. His system requires Cromwellian self-control and nobody has influenced my operating technique in the last 25 years more than Dr. Hendren.

Michelangelo was once joshed by an assistant who said, "Why are you spending so much time over that? It is only a detail." Michelangelo said, "Details make perfection and perfection is no detail." Again I have been reassured that no objection will be taken to my mentioning Michelangelo in the same paragraph.

Now two questions and a request. Did you use the Mitrofanoff principle of using a pedicled appendix with an abdominal stoma for access to the bladder? I have the impression that it is more popular in Europe than here. The second concerns the issue of the tethered cord. On MR all these cords look abnormal but do the patients benefit from intervention? Most of this group do not depend on voluntary control for continence. Most neurosurgeons will say, "It needs to be untethered." To which I say, "Don't ask your barber if you need a haircut." How do you select the group that will benefit?

Dr. Hendren, these are dangerous waters and silted harbours. We need charts. We need an atlas of the variables that you have found. Let us hope that your first paper here for the new millennium will be on this atlas. Thank you.

DR. W. HARDY HENDREN (Boston, Massachusetts): Thank you for your comments, Mr. O'Donnell. It is always risky to ask Mr. O'Donnell to discuss a paper because you never know what is coming! But it is always with good humor.

The first question is about the use of the Mitrofanoff procedure. Paul Mitrofanoff is a very clever pediatric surgeon from Roen, France, who described using the appendix as a catheterizable conduit to empty the bladder. One end of the appendix is brought to the surface of the abdomen and the other is implanted into the bladder or into an internal bowel reservoir for urine. Leakage is prevented by tunneling the end of the appendix into the bladder just as one would reimplant a ureter. Alternatively a catheterizable conduit can be made from tapered small bowel, a segment of stomach, or a ureter. The Mitrofanoff principle is a good one. I use it liberally when we cannot construct a continent bladder outlet.

Regarding the tethered spinal cord seen in one-third of "regular cloacas" and all but one of the cloacal exstrophy patients, I rely on the judgment of Dr. Michael Scott, chief of Neurosurgery at Children's, to make that decision. Unlike your barber, he does not always operate. Dr. Scott bases the decision on the degree of tethering, the presence of neurological changes, and the coexistence of lipomatous or hamartomatous tissue with the filum terminale. Urodynamic studies, including sphincter EMG data can help make the decision. Untethering does not reverse already established nerve weakness. However, it can prevent increasing deficit which can be caused by greater stretching of the cord as the child grows.

DR. JUDAH M. FOLKMAN (Boston, Massachusetts): I would like to add a note of admiration. I have had the wonderful opportunity over the past 16 years to see up close a true master surgeon at work on the most difficult problem in all pediatric surgery.

These operations, as Dr. O'Donnell said, are intricate and arduous and take a long time. These operations have the potential for an array of frightening, terrible complications, because there are so many anastomoses that depend upon each other. But if you talk to our senior and chief residents who help on the postoperative care of Dr. Hendren's patients, they say that there is nothing to do except to dictate the discharge summary when the child goes home because they don't see the complications. And they always ask, "How are we going to learn to manage the complications?"

In science there are certain researchers who are in a class by themselves. Everyone knows who they are. In this century, Jim Watson is in a class by himself. In pediatric surgery, Hardy Hendren is in a class by himself. And my only concern is for those children who will be born with cloaca after he retires. We did not know yet if his disciples have been able to reach the rarefied level he is at. So we all hope he keeps working until they do.

DR. THOMAS C. MOORE (Torrance, California): It will be a long time before another pediatric surgeon comes along with the remarkable determination, consummate ability as a technical operating surgeon and stamina of a Hardy Hendren.

My personal interest in imperforate anus is of long standing and goes back to 1952 when I reported an experience with 120 cases of imperforate anus from the Indiana University Medical Center at the Baltimore meeting of the Society of University Surgeons (*Surgery* 1952;32:352–265). This was a time of great interest in the then recent 1948 report of Jonathan Rhoads Operation for the repair of imperforate anus at birth and without a prior colostomy and by a simultaneous combined abdominal and perineal approach (*Annals of Surgery* 1948;127:552–556). I still consider this report of Jonathan Rhoads to be the most important contribution to the management of imperforate anus in the 20th century. The most important component of this approach, not known at the time, was the carrying out of this repair at birth and within the critical first hours after birth in order to achieve maximum "use it or loose it" activity-driven establishment of hard-wired ano-brain sensory neocortical neurocircuitry. This important and quite narrow window of opportunity for the establishment of this neurocircuitry from clinical and laboratory studies in cats, rats, monkeys, and man has been shown to be some 7 days from birth.

The awesome price of colostomy and wait in the management of imperforate anus in the newborn was clearly demonstrated in a 1991 article by the eminent Professor Jan Molenaar and associates of Rotterdam (*J Pediatr Surg* 1991;26:587–590) in which they reported that 40 of 40 imperforate anus infants managed by colostomy and delayed posterior saggital anoplasty "will never acquire normal continence." In the past 11 years, I have reported and urged the use of repair of imperforate anus at birth and without a prior colostomy (*J Pediatr Surg* 1987;22:1167, *J Pediatr Surg* 1990;25:276–277, and *Challenges in Pediatric Surgery*, R. G. Landes, Austin, TX 1994) and in the last 3 years have initiated with Professor Erik Heineman of Auckland, NZ an International Imperforate Anus Initiative toward this end and approach to achieving maximum continence in newborns with imperforate anus. No discussion of imperforate anus would be complete without recognizing anew the long forgotten and most important contribution of the 19th century, the June 4, 1892 report in the *British*

Medical Journal (1892;1:1187–1188) by Harrison Cripps of St. Bartholomew's Hospital in London of the successful use of the posterior saggital report of five cases of imperforate anus in newborn infants without a colostomy and without morbidity or mortality.

I wish to ask Dr. Hendren if he has done any of these complicated repairs in the immediate newborn period without a prior colostomy and if these operations might be less demanding in time and operator stress/fatigue in this immediate newborn period considering that some day even he may start to age.

DR. W. HARDY HENDREN (Boston, Massachusetts): Dr. Moore has made an important point, that we should repair major malformations as soon as feasible. This has been the trend in many types of pediatric surgery in recent years. It is common place to do immediate repair of many conditions which were formerly delayed or staged. Examples are Hirschsprung's disease, bladder exstrophy, and congenital cardiac anomalies. These cloaca babies, however, are often distended and in poor general condition at birth, precluding complex reconstruction. I would be reluctant to undertake, in a neonate, a long procedure which often needs three to four blood volumes of intraoperative fluid replacement to maintain metabolic homeostasis.

Let me add that none of the complex reconstructive work we have shown could have been done without the wonderful pediatric anesthesia which allows us to do these mega operations and finish with a little patient who is in good shape and who then spends the next day or two on ventilatory support in the intensive care unit. This has given us the ability to do surgery today which was not possible 3 to 4 decades ago.