Criteria for diagnosing the so-called persistent cloaca

Two of Peña's statements, made without any evidence and contrary to the available scientific research, fundamentally changed the entire pediatric colorectal surgery. First, to justify posterior sagittal anorectoplasty (PSARP), he announced that in ARM the anal canal is absent. Second, he announced that the type of ARM previously thought to be a vaginal fistula was persistent cloaca. After that, these patients began to operate as patients with a true cloaca and the results of these operations were as if they had no urethra, vagina, and anal canal [1]. Some pediatric surgeons have noticed that urological problems occur after correction of the urogenital sinus and are absent if the surgeons solved only the rectal problem. It has been suggested that severe complications of the urinary system are due to surgery, and not congenital pathology [2,3].

Diagnosis and treatment of the so-called persistent cloaca represent the whole complex of modern ideas about anorectal malformations. Therefore, we will analyze them using a typical example (**Figure 1**).

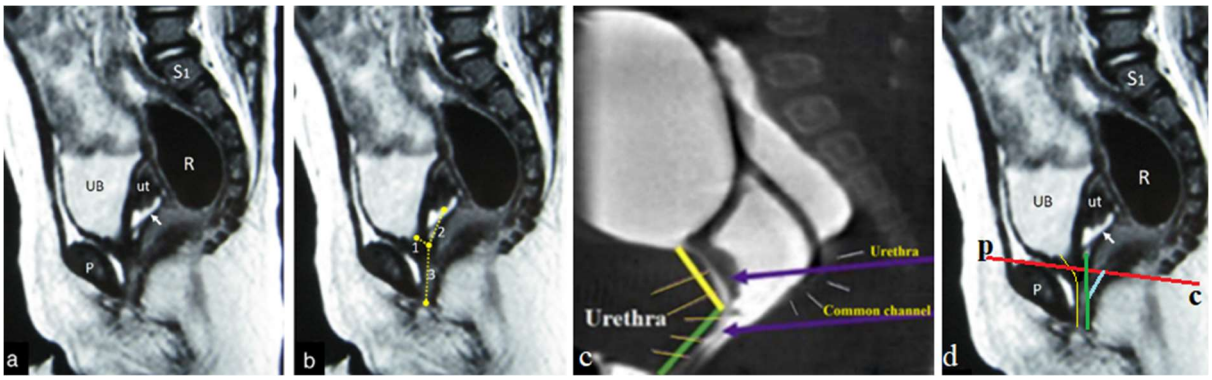

Figure 1. **(a,b).** From AbouZeid and Mohammad [4]. Signature of the authors of the article: "Demonstration of the abnormal pelvic MRI anatomy in a case of cloaca (22-month-old). **(a)** Note: the vagina is distended with fluid that appears hyperintense (white arrow) like the urinary bladder. The lower end of the distended vagina marks for the level of urogenital confluence. **(b)** The urethral length (1) is measured as the distance between the bladder neck and the urogenital confluence; the vaginal length (2) is measured as the distance between the uterine cervix and the urogenital confluence; and the common channel (3) is measured as the distance between the urogenital confluence and the vestibule (just below and behind the lower end of pubic symphysis". **(c).** Study from Wood et al [5]. The purple arrows are drawn by the authors of this article. Where "common channel" is indicated, two channels are visible, separated by a gray strip, which represents the walls of the urethra and of the narrow part of the vagina. As in normal, the urethra approaches the vagina and continues its course to the vulva next to the vagina. I marked it with yellow lines. Behind the vagina, a well-contrasted rectum is visible, ending at the level of the PRM (pubococcygeal line). Below it, a poorly contrasted closed anal canal (white lines) is visible. **(d).** My interpretation of Figure 1.a. The urethra, as in normal, approaches the vagina and continues to be located next to it, as in figure (c). Below the pubococcygeal line is the anal canal (blue), which enters the vagina.

Firstly, this study, repeating the mistakes of other authors, made without the introduction of catheters and without contrasting the anal dimple. Interpretation

of the location of the urethra indicates ignorance of normal anatomy. None of the channels are visible in the pictures. And there is no evidence of any urogenital confluence.

Secondly, if the separation of the cloaca has already occurred with the formation of the urethra, vagina and anal canal, then their confluence in the common channel is scientifically impossible.

Thirdly, it is obvious that the length of the urethra, measured in this way, has nothing to do with the truth. At the same time, the length of the urethra became the justification for difficult urological operations. Halleran et al's in an article performed with serious methodological errors reported supposedly normal urethral length limits for girls aged 6 to 36 months. The length of the minimum boundary differs from the maximum one by a factor of 3 [6]. The maximum limit of normal (4.7 cm) exceeds the maximum limit in adult women (3.5 cm) [7]. Peña and Levitt believe that a urethra less than 1.5 cm long is the cause of urological problems. However, if nearly 30% of healthy adult women have a 2 cm urethra, why is a urethra less than 1.5 cm a problem in girls under 3 years of age? The suggestion that isolating the urethra and lengthening it by creating an additional canal can improve its function is unscientific. The urethra, which is a sphincter, stops responding to high bladder pressure after denervation, necessitating intermittent bladder catheterization. Violation of the blood supply to the urethra turns it into a fibrous tissue with possible narrowing, up to stenosis.

Fourth, the presence of a bowel below the pubococcygeal line indicates the presence of a functioning anal canal. The fact that pediatric surgeons have no idea about the anal canal is the result of the activities of Peña and Levitt and their collaborators. There is no scientific information in their articles. All their claims are false. However, every pediatric surgeon had the opportunity to analyze the articles of Peña and Levitt. And if they did not do this or did not understand that a statement not supported by research or other scientific evidence has no value, then this does not relieve them of responsibility for the fate of patients whose urinary system and anal canal were destroyed instead of preserving of all elements created by nature.

Fifth, all ideas about persistent cloaca have no scientific basis. They are announced by Peña without scientific research or references to the research of other scientists. Operations that are intended for the true cloaca destroy the urological system. So, for example, it is known that any striated muscle contracts in response to an impulse emanating from a nerve ending. If you cross the branch that innervates the flexor of the finger, then the finger will stop bending. Smooth

muscle sphincters connected with other organs by a reflex arc cease to participate in reflexes if they are denervated [8,9].

In figure 1b, there is supposedly a long common channel, for which there is no evidence. Here is the operation proposed by Bischoff in such cases [10] (**Figure 2).**

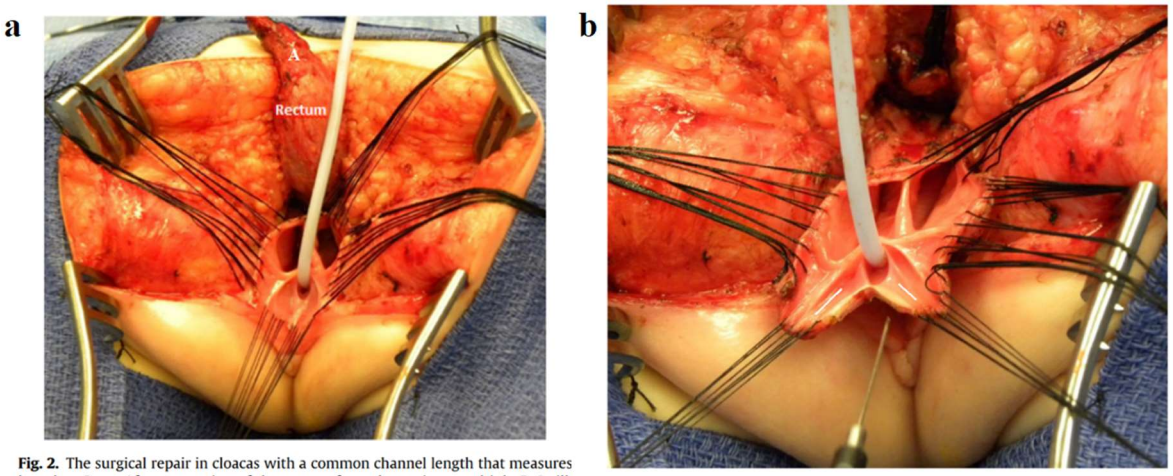

Fig. 2. The surgical repair in cloacas with a common channel length that measures less than 3 cm. After separation of the rectum from the vaginas, multiple 5–0 silk stitches are placed around the urogenital sinus; 5 mm from the clitoris and in the lateral and posterior wall of the vaginas.

Fig. 3. After the total urogenital mobilization is completed, what used to be the common channel is divided in the midline to become part of the neo-labia.

Figure 2. From Bischoff's article. Total urogenital mobilization [10].

(a) The rectum is covered with peritoneum and has a wide lumen. Above it, the section of the intestine, isolated from the tissues of the perineum, has a narrow lumen, since it is a closed, denervated and devascularized internal anal sphincter (A). To easily approach it, the surgeon had to cross the puborectalis muscle. And in order to pull out the rectum, it had to be separated from the levator plates. The complete destruction of the anal canal does not give any chance for the normal processes of fecal retention and defecation.

(b) Into the bladder through the urethra a white catheter was inserted. The distal urethra is dissected (white lines). The entire genitourinary complex (truncated urethra and two vaginas) from the surrounding tissues was isolated so that the opening of the urethra was visible for continuous or intermittent catheterization of the bladder. However, the bladder and urethra are denervated and the blood supply to the urethra is impaired.

(c) The Bischoff article makes statements that defy common sense:

1. "Patients with a common channel length shorter than 3 cm can be repaired through a posterior sagittal approach. After the separation of the rectum from the vagina, the surgeon should perform a maneuver called total urogenital mobilization. To accomplish this, the posterior wall of the vagina and the entire common channel is divided to expose the urethral and vaginal openings. " [10].

This phrase suggests that the transection is made over the common canal and confirms what is shown in Figure 2, i.e., the distal urethra is cut off.

2. "In most cases the total urogenital mobilization is sufficient for the urethra and vagina to reach the perineum together without tension. In some cases the posterior vaginal walls still require some extra mobilization. Once we are done with the total urogenital mobilization, what used to be the common channel is divided in the midline, creating two flaps that will become part of the neo-labia (Figure 3)". It follows from this segment that, first, the surgeon cuts off a part of the urogenital zone, which will go to decorate the perineum, and then what is left pull with tension to attach to the skin.

3. Cloaca with a common channel length between 3 and 5 cm « is probably the most challenging group of malformations due to the extent of the common wall between the three structures (urethra/bladder, vagina, and rectum) » [10]. This misconception about a common wall is often found in the work of Peña and his followers. After the end of the cloacal stage, the urethra, vagina, and anal canal have their own walls, which in infants are difficult to separate from each other. But firstly, there is no need for this. Secondly, it becomes apparent when fatty tissue appears between the walls (**Figure 3.a**) [11]. With these cloaca's "transabdominal urogenital mobilization is performed. This maneuver consists in bringing the urogenital sinus inside the abdomen and continuing to divide all the avascular pelvic attachments of the urethra, bladder, and vagina. With this maneuver we can sometimes gain between 1–2 cm of extra length" [10]. Since the beginning of TUM, the urethra has been shortened. Then, during the abdominal stage, it is released from the vessels and nerve fibers. This tube may become longer, but it is no longer the urethra, which before this operation was a smooth muscle internal sphincter surrounded by an external striated sphincter **(Figure 3. b)** [7].

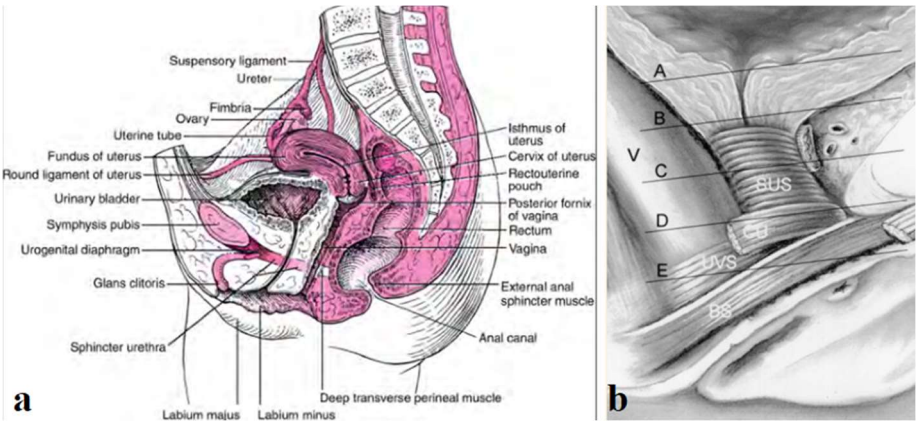

Figure 3. (a) From Gross Anatomy [11]. Из статьи Umek et al [7].

4. Bischoff further states that "when the common channel has a smooth trajectory to the bladder, it should be left as urethra to be used as a conduit for bladder catheterization" [10]. I think that the channel that leads the catheter to the bladder is nothing more than a remnant of the urethra.

Early results. Reoperations were in 104 out of 570 patients with cloaca. "The most common types of reoperations needed in patients with cloaca include persistent urogenital sinus, acquired vaginal atresia or stricture, acquired urethral atresia or stricture, rectal prolapse, neo-vagina prolapse, and complex catastrophic complications (dehiscence, abscess, and fistula) that we consider as failed attempted repairs "[10].

Long-term urological outcome. When the common channel is shorter than 3 cm, about 20% of the patients will require intermittent catheterization to be able to empty their bladders. On the other hand, when the common channel is more than 3 cm, 80% of the patients require intermittent catheterization". According to Warne et al up to 50% of patients may have urinary incontinence or may be dependent on clean intermittent catheterization after cloaca repair [12].

I have questions that should worry all pediatric surgeons:

1. Why do the authors of the articles do not examine the state of the urinary system in these patients before surgery, even though there is evidence of its normal function [2,3]?

2. Can there be at least one proof of the confluence of the already formed urethra, vagina and anal canal, if this contradicts embryology?

3. What is the need for surgical expansion of the vagina in infants?

4. Is the TUM operation justified in terms of functional results, if there were no such terrible outcomes before its application?

5. Why is the presence of the anal canal and the possibility of its preservation not being investigated?

6. How can surgeons remodel the perineum in infants with ARM without knowledge of anatomy and physiology [2,3]?

Conclusion

1. Children with the so-called persistent cloaca are ARM with a fistula in the vagina. They have a normally functioning bladder, urethra, and anal canal. Unlike the usual form of ano-vaginal fistula, they have a narrowing of the distal part of the vagina of different lengths. Because of this, some of them have an obstruction between the expanded and narrow parts of the vagina with the formation of

hydrocolpos. Early drainage of hydrocolpos and diverting colostomy are the only procedures that are needed in neonates. Bougienage of the vagina, and operational expansion in the pubertal period should be carried out by gynecologists. It remains only to save the anal canal by perforation of the perineum [13]. Then to suture ano-vaginal fistula. These children may be completely healthy.

2 I accuse Peña, Levitt, Bischoff et al in that they without any scientific research fraudulently introducing false information about anorectal malformations among pediatric surgeons, which leads to the destruction of the genitourinary and anorectal function of children with the so-called persistent cloaca. Because of this, surviving children become disabled for life.

References

1. Levin MD. Discussion about methods of X-ray diagnostics of anorectal malformations in children. Review. https://www.anorectalmalformations.com/_files/ugd/4d1c1d_4a9958669dca432db0b56eb2bd625873.pdf
2. Kittur DH, Vora RM. Persistent Cloaca: A Long-term Follow-up Study. J Indian Assoc Pediatr Surg. Apr-Jun 2017;22(2):126-127. doi: 10.4103/0971-9261.202685.
3. AbouZeid AA. Achieving Full Anatomical Correction in Girls with cloacal anomalies: A Necessity or an Overdoing? The journal of genital surgery. Volume 1, January 2019, Page 1-10 DOI:10.21608/JGS.2019.7646.1015.
4. AbouZeid AA, Mohammad SA. The cloacal anomalies: Anatomical insights through a complex spectrum. J Pediatr Surg. 2019 Oct;54(10):2004-2011. doi: 10.1016/j.jpedsurg.2019.04.005.
5. Wood RJ, Reck-Burneo CA, Dajusta D, et al. Cloaca reconstruction: a new algorithm which considers the role of urethral length in determining surgical planning. J Pediatr Surg. 2017 Oct 12;S0022-3468(17)30644-9. doi: 10.1016/j.jpedsurg.2017.10.022.
6. Halleran DR, Thompson B,Fuchs M, et al. Urethral length in female infants and its relevance in the repair of cloaca. J Pediatr Surg. 2019 Feb;54(2):303-306. doi: 10.1016/j.jpedsurg.2018.10.094.
7. Umek WH, Kearney R, Morgan DM, et al. The axial location of structural regions in the urethra: a magnetic resonance study in

nulliparous women. Obstet Gynecol. 2003 Nov;102(5 Pt 1):1039- 45. doi: 10.1016/j.obstetgynecol.2003.04.001.
8. Bharucha AE. Pelvic floor: anatomy and function. Neurogastroenterol Motil. 2006 Jul;18(7):507-19. doi: 10.1111/j.1365-2982.2006.00803.x.
9. Palit S, Lunniss P, Scott SM. The physiology of human refecation. Dig Dis Sci. 2012 Jun;57(6):1445-64. doi: 10.1007/s10620-012-2071-1
10. Bischoff A. The surgical treatment of cloaca. Semin Pediatr Surg. 2016 Apr;25(2):102-7. doi: 10.1053/j.sempedsurg.2015.11.009.
11. Gross Anatomy. 6th Edition. By K.W. Chung and H. M. Chung/ Wolters Kluer.Lippincort. Williams & Wilkins.
12. Warne SA, Wilcox DT, Ransley PG. Long-term urological outcome of patients presenting with persistent cloaca. J Urol. 2002 15 Oct;168(4 Pt2):1859-62; discussion 1862. doi: 10.1097/01.ju.0000030712.17096.0d.
13. Levin MD. The pathological physiology of the anorectal defects, from the new concept to the new treatment. Eksp Klin Gastroenterol. 2013; (11):38-48. (PubMed).