Tethered cord, sacral ration, and anorectal malformation.

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I. Tethered cord

This problem has been of concern to all pediatric surgeons for several decades. Numerous articles are devoted to her, the purpose of which was to determine the significance of spinal cord anomalies in patients with anorectal malformations (ARM) in postsurgical outcomes of bowel function, lower urinary tract symptoms, and lower-limb neurological abnormalities. Regarding neurological symptoms, the diagnosis and treatment of which is under the jurisdiction of neuropathologists, there is a consensus since untethering surgery is effective in neuro-motor symptoms in selected patients with ARM [1,2]. Most researchers did not find any improvement in bowel and urinary tract function after surgery [1,2,3,4]. Only Destro et al have shown improvement in urodynamic symptoms after neurosurgery [5]. However, as the study by Frainey et al showed, "Assessment of urodynamic data revealed that neither pre- nor post-TCR (tethered cord release) urodynamics predicted continence status" [6].

The fashion for statistical analysis of operated patients on this topic is surprising because there is not a single article on the study of the pathological physiology of ARM. However, playing with numbers without understanding pathological physiology leads to conflicting results.

1. It is known that more than 75% of children with ARM have other associated malformations. The most frequent malformations seen were genitourinary (28%) and spinal anomalies (26%) [7]. In another study, occult spinal dysraphism (tethered spinal cord, spinal lipoma, syringomyelia) was found at MRI in 57%, including anal stenosis, recto-vestibular, and recto-perineal fistulas [8].

When analyzing these and similar works, the following questions arise:

A) Is this spinal anomaly the cause of poor ARM reconstruction results (urinary incontinence, fecal incontinence, and chronic constipation)?

B) What should be done if spinal anomalies are found?

C) For what purpose, by what method, and when to carry out studies of the sacral spine?

To answer the first question (A) we compare the long-term results of different operations by the same group of surgeons. After cutback anoplasty in boys with

perineal fistula, the results were "Amongst 46 respondents (67%; median age 12.3 (5-29) years), overall fecal control was comparable to controls. All patients had voluntary bowel movements; 98% of patients and 97% of controls were socially continent; 67% of patients and 64% of controls were totally continent. Constipation amongst patients (33 vs 3% in controls; $p < 0.0001$) declined significantly with age. Outcomes by bowel function score were good in 85%, satisfactory in 15%, and poor in 0%. Prevalence of LUTS and age at completion of toilet training were comparable to controls" [9].

After anterior sagittal anorectoplasty in girls with perineal and vestibular fistulas, the bowel functional outcome was good in 68% of patients, satisfactory in 26% of patients, and poor in 6% of patients [10].

Of the patients with rectourethral fistula treated with PSARP, by BFS score, 39% had a good functional outcome, 27% had a moderate outcome, 9% had a clearly poor score and 24% were living with an ACE [11]. In Table 1, the results of various operations are shown.

	good	satisfactory	poor	ACE
Cutback (perineal males)	85%	15%	0	0
ASARP (perineal, vestibular females)	68%	26%	6%	0
PSARP (urethral males)	39%	27%	9%	24%

The types of ARM listed in the table are mistakenly associated with a good outcome. From a scientific point of view, they are united by the presence of a functioning anal canal. And the difference in outcomes is due to different surgical methods. After cutback anopasty, some of the patients suffered from chronic constipation to a slightly greater extent than control individuals. This can be explained by the late diagnosis of ARM, when by the time of the operation the megarectum had developed. Pull-through surgery through the anterior or posterior approaches destroys the anal canal, which affects the results. However, in the anterior approach, in contrast to the posterior approach, the muscle complex, including the puborectalis muscle, is not damaged, which explains the difference in the outcomes of operations.

Another study by these authors is important for our analysis. "Of 89 patients (median age 15 years, range 5-29 years), MRI was available in 90%. Spinal cord anomalies were found in 34%, comprising a filum terminale lipoma in 30%, low conus medullaris in 10%, and thoracolumbar syrinx in 6%. The long-term functional outcomes for patients with SCAs (spinal cord anomalies) who had VF/PF and RUF may not differ significantly from patients with the same type of ARMs and a normal spinal cord. (p = not significant for all). The results favor a

conservative approach to their management in the absence of abnormal neurological findings in the lower limbs" [12].

This opinion is shared by other authors. For example, Stathopoulos et al concluded that although lower urinary tract dysfunction is common in patients with ARM, a normal spine or spinal cord does not exclude neurovesical dysfunction. Myelodysplasia or vertebral anomaly does not determine lower urinary tract dysfunction [13]. Di Cesare et al concluded that for ARM patients the prognosis in terms of continence depends mainly on the type of malformation and is not complicated by the association with neurospinal dysraphism. In their series neurosurgical treatments did not have any effect in improving the continence of ARM patients and conservative management of neurospinal dysraphism did not expose the patients to the sequelae of progressive deterioration, reported elsewhere, requiring rescue neurosurgery [14]. Long-term functional outcome in patients with ARM and TSC undergoing untethering surgery is equivalent to that in those without TSC [15]. In some articles, phrases are allowed in the conclusions that are unacceptable in scientific papers. For example, Tsuda et al stated that "Patients with TC were more likely to have poor bowel function, but this did not reach statistical significance" [16]. From a scientific point of view, if they did not find statistical differences then they had no worse bowel function. Levitt et al studied 934 patients with anorectal malformations, 111 of whom had magnetic resonance imaging of the spine. Based on a huge amount of data, they concluded that no solid evidence supports the concept that tethered cord by itself affects the functional prognosis of patients with anorectal malformations. In addition, there is no good evidence demonstrating that surgical untethering improves the prognosis [17].

Thus, numerous studies have established that from 36% to 53% of patients with ARA are diagnosed with the tethered cord. However, the incidence of postoperative complications does not depend on the presence or absence of neurospinal dysraphism, as well as on the combination with esophageal atresia, heart defects, etc.

Analysis of the literature shows that poor bowel function was often observed together with lower urinary tract dysfunction, which can be explained by one cause that disrupts the nerve supply to both systems [18]. If numerous studies do not associate poor results with the tethered cord, and most types of ARM differ from each other only in the distance of the fistula from the anal fossa and with pull-through surgery this difference is leveled, then it becomes obvious that poor results are due to destructive methods of surgery, and depend on the volume of denervation of the anorectal zone and lower urinary tract **(Figure 1).**

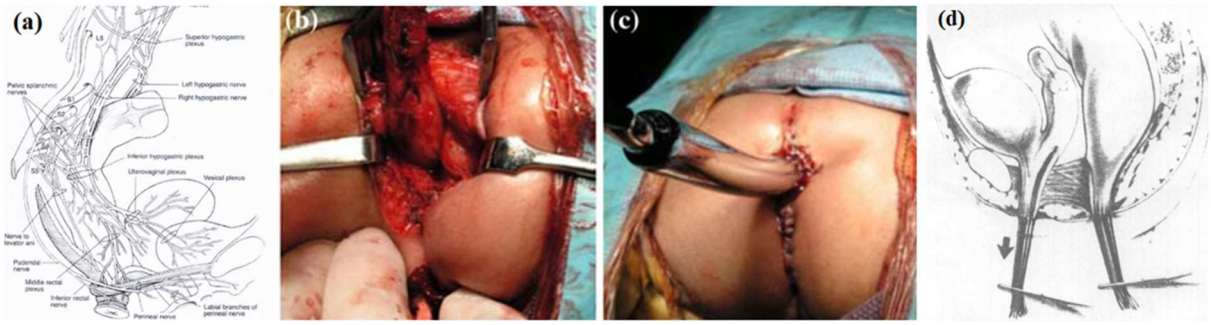

Figure 1. (a) Scheme of the nervous supply anorectum from the article Bharucha [19]. **(b-c)** Stages of PSARP. **(d)** Scheme of total urogenital mobilization [20].

Images (b) and (c) show that with PSARP, there is extensive denervation of the pelvic organs. During the PSARP operation, Peña was unable to discern the puborectalis muscle in the muscle complex and began to argue that it did not matter in the retention of feces. He also did not see the nerve plexuses that he crossed during the operation. This is described as follows: -"Putting traction on the rectum makes it possible to identify bands and vessels of its wall. Since these impede its being pulled through, they must be cauterized and cut" [21].

The total urogenital mobilization, proposed by Peña for the correction of the so-called cloaca, is accompanied by complete denervation of the pelvic organs, which inevitably leads to a violation of their function. These are the basics of medicine. Nevertheless, Muller et al, based on a statistical analysis of as many as 25 observations, published «...the first study, which highlights the impact of different types of spinal dysraphism on functional outcome in patients with cloaca" [22]. They found that "The sacral ratio was abnormal (**below 0.74)** in 18 cases out of 25 (72%)" [21]. This study cannot be taken seriously, firstly, because "Normal values for anteroposterior view ranged from 0.52 to 1.12, with an **average of 0.74** "[23], or 0.514-0.936 [24]. Secondly, treatment outcomes with different SR values were not compared.

Conclusion. Based on the analysis of the literature on the combination of an ARM with spinal dysraphism, the following conclusions can draw:

1. Spinal dysraphism with ARM occurs much more often than in healthy children.

2. Spinal dysraphism has no effect on the function of the urinary and fecal systems.

3. Neurosurgery is recommended only in the presence of neurological symptoms because it does not affect the function of the urinary and anorectal systems.

4. Poor results are due to pull-through surgical (posterior approach anterior approach, laparoscopically assisted), which, to varying degrees, destroy the nature-created anal canal and innervation of the pelvic organs.

II. Sacral ratio

In 1997, Peña and Levitt et al argued that "no solid evidence supports the concept that tethered cord by itself affects the functional prognosis of patients with anorectal malformations" [17]. At the same period (1995), Peña suggests measuring the sacral ratio, since "An accurate diagnosis and evaluation of the sacrum allows us to establish, with reasonable accuracy, functional prognosis in most children" [25]. Thus, he revives the scientifically rejected idea of the absence of the effect of sacral pathology on the function of the pelvic organs in patients with ARM.

While the sacral ratio (SR) is formally suggested for use for counseling families of children with ARM, the most important reason Peña and Levitt and their staff defy common sense is to protect PSARP. This is a palming of another reason, allegedly responsible for the poor results after PSARP. In this way, they want to convince the audience that PSARP is the ideal method, and the poor results after the operation are due to the pathology of the spine. Instead of investigating the real causes of disastrous results to improve the quality of treatment, they constantly spur the discussion on this topic with their repeated "research". We are faced with the task of answering the following questions: 1) What is the relationship between SR and sacral dysraphism? 2) Is there a dependence of treatment results on SR? 3) What should a scientist do: look for the best ways to diagnose and treat ARM, or look for an excuse for his failures?

1) **What is the relationship between SR and sacral dysraphism?**

First, as shown above, even though sacral dysraphism occurs in patients with ARA in approximately 36%, it does not affect the severity of postoperative complications. Second, Chang et al, using MRI of the lumbar spine in 76 patients, showed that "The prevalence of spinal cord anomaly was not correlated to SR ($p = 0.39$) [26]". “Neurospinal cord dysraphism may be present despite a normal sacral ratio” [27].

2) **Is there a dependence of treatment results on SR?**

Minneci et al showed that spinal anomalies and the lateral sacral ratio were not associated with continence. Type of ARM was the only factor that predicted fecal continence in children with ARM [28]. The incidence of tethered cord (TC) among patients with ARM is 36%. The incidence of TC among patients with RBNF was 30% (11 of 37). It correlated with SR value and was higher with lower

SR. Patients with rectobladder neck fistula and TC have a dismal prognosis for bowel control, unrelated to their SR status [29]. "Although the SR was different in patients with sacral agenesis it was no different in continent, partially continent or incontinent patients, and thus it is of no practical value in identifying patients likely to have fecal incontinence" [30].

Groups of physicians from the Peña and Levitt departments persistently publish articles that argue, “That a lower SR correlated with the presence of urologic and renal malformations” [31]. The Levitt group has published 10 articles on this topic, and the Peña group has published -13. They state the well-known fact that in patients with ARM, in almost 36% of cases, SR is less than 0.4 and almost always they have poor results. However, these authors, in contrast to previous researchers, did not compare the two groups of patients with ARM, i.e., with SR <0.4 and SR> 0.4. In other studies, a comparison of these two groups showed that the results of treatment are independent of the value of SR [28, 29, and 30]. Thus, the fact that patients with ARM often have an SR <0.4 does not affect treatment outcomes. The stronger the dissection of the perineum, the worse the function of the pelvic organs.

3. The goal declared by the proponents of the SR calculation is astonishing. Even if predicting the poor pelvic floor function because of calculating SR was scientific, it would be worthwhile if we could prevent a poor result or at least improve treatment outcomes. But it makes no sense and is not necessary to devote numerous articles to warn parents about the inevitability of bad results, since the results are bad in all patients and cannot be good after the destruction of the anal canal. After PSARP, during which the internal anal sphincter is excised, the muscle complex is transected, including the puborectalis muscle, as well as the levator plates are detached from the rectum and the pelvic organs are denervated, there are two variants of anorectal function. If stenosis of the created perineal fistula prevails, then the patient suffers from severe constipation. If there is no severe narrowing of the perineal fistula, fecal incontinence of varying degrees prevails. Even theoretically, there can be no other outcomes. In addition, the more severe the perineal dissection, the more likely urinary problems are.

This is evidenced by the figures shown by Peña in his article, where he first proposes to study SR: - "Patients with voluntary bowel movements and no soiling were classified as totally continent; 40.8% of the series belong to this group. Distributed by diagnosis, it varied from 100% in cases with rectal atresia or perineal fistula, 65.9% in those with vestibular fistula, 34% in those with bulbar fistula, 31.6% in those with cloacas, 26.3% in those with prostatic fistula; none of the patients with vaginal fistula or bladder-neck fistula was totally continent.

Constipation was detected in 43.1% of all patients and was more frequent in those with simple defects. Urinary incontinence was found in 19% of patients with cloacas who had a common channel shorter than 3 cm and in 68.8% of the patients who had longer common channels" [25]. These figures convincingly show that the higher the surgeon's scalpel rises, the worse the functional results of the treatment.

Conclusion. In control subjects, the sacral ratio of 0.4 is the lower limit of the norm. In children with ARM, an SR of less than 0.4 is observed from 36 to 57% of cases. However, the SR value, firstly, does not always indicate spinal dysraphism. Second, treatment outcomes and severity of complications are similar in patients with both low and normal SR. Treatment results depend on the type of ARM. The wider the dissection of the perineum, the worse the results. Calculation of SR does not make any sense, because it does not provide any useful information neither for the doctor nor for the patient and his parents. Articles claiming the value of SR have no scientific basis.

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References

1. Totonelli G, Messina R, Morini F, et al. Impact of the associated anorectal malformation on the outcome of spinal dysraphism after untethering surgery. Pediatr Surg Int. 2019 Feb;35(2):227-231. doi: 10.1007/s00383-018-4400-8.
2. van den Hondel D, Sloots C, de Jong TH, et al. Screening and Treatment of Tethered Spinal Cord in Anorectal Malformation Patients. Eur J Pediatr Surg. 2016 Feb;26(1):22-8. doi: 10.1055/s-0035-1563673.
3. Uchida K, Inoue M, Matsubara T, et al. Evaluation and treatment for spinal cord tethering in patients with anorectal malformations. Eur J Pediatr Surg. 2007 Dec;17(6):408-11. doi: 10.1055/s-2007-989277.
4. Valentini LG, Selvaggio G, Erbetta A, et al. Occult spinal dysraphism: lessons learned by retrospective analysis of 149 surgical cases about natural history, surgical indications, urodynamic testing, and intraoperative neurophysiological monitoring. Childs Nerv Syst. 2013 Sep;29(9):1657-69. doi: 10.1007/s00381-013-2186-5.
5. Destro F, Canazza L, Meroni M, et al. Tethered Cord and Anorectal Malformations: A Case Series. Eur J Pediatr Surg. 2018 Dec;28(6):484-490. doi: 10.1055/s-0037-1606844.
6. Frainey BT, Yerkes EB, Vani S Menon VS, et al. Predictors of urinary continence following tethered cord release in children with occult spinal

dysraphism. J Pediatr Urol. 2014 Aug;10(4):627-33. doi: 10.1016/j.jpurol.2014.06.008.
7. Nah SA, Ong CCP, Lakshmi NK, et al. Anomalies associated with anorectal malformations according to the Krickenbeck anatomic classification. J Pediatr Surg. 2012 Dec;47(12):2273-8. doi: 10.1016/j.jpedsurg.2012.09.017.
8. Scottoni F, Iacobelli BD, Zaccara AM, et al. Spinal ultrasound in patients with anorectal malformations: is this the end of an era? Pediatr Surg Int. 2014 Aug;30(8):829-31. doi: 10.1007/s00383-014-3546-2.
9. Kyrklund K, Pakarinen MP, Taskinen S, Rintala RJ. Bowel function and lower urinary tract symptoms in males with low anorectal malformations: an update of controlled, long-term outcomes. Int J Colorectal Dis. 2015 Feb;30(2):221-8. doi: 10.1007/s00384-014-2074-9.
10. Kyrklund K, Pakarinen MP, Koivusalo A, Rintala RJ. Bowel functional outcomes in females with perineal or vestibular fistula treated with anterior sagittal anorectoplasty: controlled results into adulthood. Dis Colon Rectum. 2015 Jan;58(1):97-103. doi: 10.1097/DCR.0000000000000239.
11. Kyrklund K, Pakarinen MP, Koivusalo A, Rintala RJ. Long-term bowel functional outcomes in rectourethral fistula treated with PSARP: controlled results after 4-29 years of follow-up: a single-institution, cross-sectional study. J Pediatr Surg. 2014 Nov;49(11):1635-42. doi: 10.1016/j.jpedsurg.2014.04.017.
12. Kyrklund K, Pakarinen MP, Taskinen S, et al. Spinal cord anomalies in patients with anorectal malformations without severe sacral abnormalities or meningomyelocele: outcomes after expectant, conservative management. J Neurosurg Spine. 2016 Dec;25(6):782-789. doi: 10.3171/2016.4.SPINE1641.
13. Stathopoulos E, Muehlethaler V, Rais M et al. Preoperative assessment of neurovesical function in children with anorectal malformation: association with vertebral and spinal malformations. J Urol. 2012 Sep;188(3):943-7. doi: 10.1016/j.juro.2012.04.117.
14. Di Cesare A, Leva E, Macchini F, et al. Anorectal malformations and neurospinal dysraphism: is this association a major risk for continence? Pediatr Surg Int. 2010 Nov;26(11):1077-81. doi: 10.1007/s00383-010-2686-2.
15. Inoue M, Uchida K, Otake K, et al. Long-term functional outcome after untethering surgery for a tethered spinal cord in patients with anorectal malformations. Pediatr Surg Int. 2017 Sep;33(9):995-999. doi: 10.1007/s00383-017-4127-y.

16. Tsuda T, Iwai N, Kimura O, et al.Bowel function after surgery for anorectal malformations in patients with tethered spinal cord. Pediatr Surg Int. 2007 Dec;23(12):1171-4. doi: 10.1007/s00383-007-2025-4.
17. Levitt MA, Patel M, Rodriguez G, Gaylin DS, Pena A. The tethered spinal cord in patients with anorectal malformations. J Pediatr Surg. 1997 Mar;32(3):462-8. doi: 10.1016/s0022-3468(97)90607-2.
18. Borg H, Holmdahl G, Doroszkievicz M, Sillen U. Longitudinal study of lower urinary tract function in children with anorectal malformation. Eur J Pediatr Surg. 2014 Dec;24(6):492-9. doi: 10.1055/s-0033-1357299. Epub 2013 Oct 10.
19. Bharucha AB. Pelvic floor: anatomy and function. Neurogastroenterol Motil. 2006 Jul;18(7):507-19. doi: 10.1111/j.1365-2982.2006.00803.x.
20. Peña A. Total urogenital mobilization--an easier way to repair cloacas. J Pediatr Surg. 1997 Feb;32(2):263-7; discussion 267-8. doi: 10.1016/s0022-3468(97)90191-3.
21. deVries PA, Peña A. Posterior sagittal anorectoplasty. J Pediatr Surg. 1982 Oct;17(5):638-43. doi: 10.1016/s0022-3468(82)80126-7.
22. Muller CO, Crétolle C, Blanc T, et al. Impact of spinal dysraphism on urinary and faecal prognosis in 25 cases of cloacal malformation. J Pediatr Urol. 2014 Dec;10(6):1199-205. doi: 10.1016/j.jpurol.2014.05.012.
23. Torre M, Martucciello G, Jasonni V. Sacral development in anorectal malformations and in normal population. Pediatr Radiol. 2001 Dec;31(12):858-62. doi: 10.1007/s002470100006.
24. Bahadir K, Arikan-Ergun B, Elhan AH, et al. Development of Sacral Ratio Percentile Card for Children: A Preliminary Report. Eur J Pediatr Surg. 2021 Nov 30. doi: 10.1055/s-0041-1739424.
25. Peña A. Anorectal malformations. Semin Pediatr Surg. 1995 Feb;4(1):35-47.
26. Chang PCY, Duhc YC , Fu YW, et al. How much do we know about constipationafter surgery for anorectal malformation? Pediatr Neonatol. 2020 Feb;61(1):58-62. doi: 10.1016/j.pedneo.2019.05.010.
27. Arnoldi R, Macchini F, Gentilino V, et al. Anorectal malformations with good prognosis: variables affecting the functional outcome. J Pediatr Surg. 2014 Aug;49(8):1232-6. doi: 10.1016/j.jpedsurg.2014.01.051.
28. Minneci PC, Kabre RS, Mak GZ, et al. Can fecal continence be predicted in patients born with anorectal malformations? J Pediatr Surg. 2019 Jun;54(6):1159-1163. doi: 10.1016/j.jpedsurg.2019.02.035.
29. Samuk I, A Bischoff A, Freud E, Pena A. Tethered cord in children with anorectal malformations with emphasis on rectobladder neck fistula.

Pediatr Surg Int. 2019 Feb;35(2):221-226. doi: 10.1007/s00383-018-4399-x.
30. Macedo M, Martins JL, Freitas Filho LG. Sacral ratio and fecal continence in children with anorectal malformations. BJU Int. 2004 Oct;94(6):893-4. doi: 10.1111/j.1464-410X.2004.05053.x.