

Dear editor,

The Journal of Pediatric Surgery published the statistical results of an examination of children with anorectal malformations who were under the supervision of the surgical departments of 10 hospitals. These data were published in two articles [1,2]. The authors have done a great study, which, unfortunately, is presented with serious methodological errors.

1). The articles contain assertions as reliable scientific data, with reference to the articles. However, there is no evidence in these articles. At the same time, there are no references to articles in which there is evidence contradicting the opinion of the authors. For example, one of the articles [2] begins with the sentence "Children born with an anorectal malformation (ARM) often have associated anomalies of the sacrum and spinal cord which impair the normal functioning of sensory innervation, sphincter control, and colonic motility needed for fecal continence. [1,2]". In these articles, without evidence, it is stated that the poor results of posterior sagittal anorectoplasty (PSARP) are due, firstly, to the absence of the anal canal, and, secondly, to a violation of innervation due to vertebral abnormalities. However, these assumptions are wrong.

2) Embryological studies have shown the cranial-caudal migration of the internal anal sphincter (IAS) into the pelvic tissue in ARM's rats [3]. In all patients with visible fistulas, a manometric study found normal anal pressure and a positive anorectal reflex [4,5]. X-ray studies found normal function of puborectalis muscle (PRM) and levators plates [5]. In a study by Kraus et al. it has been shown that in 90% of boys with urethral fistulas the distal intestine is in constant contraction under the influence of muscles [6], which is an accurate description of the normal function of the anal canal. Thus, in all cases with visible fistulas, and at least 90% of boys with urethral fistulas have a functioning anal canal and no information has been received that its sensitivity differs from the normal anal canal.

The authors refer to Rome III diagnostic criteria for fecal continence, where functional disorders in adults are considered and there is no definition of incontinence as ≤ 1 stool accident per week, as indicated in the article. The fecal incontinence less than once a week as a standard criterion, despite the use of laxatives and enemas is too big a deviation from the physiological norm. These comments apply to all other references. They do not contain evidence of those assumptions that are indicated in the peer-reviewed articles.

3). The analysis of a statistical study is limited to a statement of numbers, and the conclusions are contradictory. For example, "Patent foramen ovale and patent ductus arteriosus were the most commonly identified cardiac anomalies, seen in

193 (58.0%) and 146 (43.8%) patients, respectively" [1]. It turns out that these defects at the same age are observed with the same frequency in the general population (62% and 45%) [7].

The frequency of various anomalies of spine and sacrum does not exceed 20.6 (Tethered spinal cord syndrome) [1]. Meanwhile, the rate of occult spinal dysraphism in asymptomatic infants with cutaneous stigmata ranged from 12% for patients with asymmetrically deviated gluteal crease to 55% for those with other isolated cutaneous stigmata. "The clinical significance of such lesions remains unclear" [8]. Many researchers confirm that in most children with abnormalities in the spine and spinal cord, the fecal and urine continence is not disturbed [9, 10, 11]. Surgical treatment is recommended in rare cases when symptoms appear. In patients with ARM, surgical treatment does not improve sphincter function [12, 13].

Based on a statistical analysis, the authors conclude that "The type of ARM was the only factor identified early in life that predicted fecal continence in children born with an ARM". But they negated this scientific fact in the discussion. "However, the presence of associated sacral and spinal anomalies may also be contributing to impaired continence...", again referring to articles in which there is no evidence.

4). Is it possible to predict the outcome of the operation? And why is it necessary?

The present study confirms the well-known fact that the more severely subtypes of ARM, the worse the functional result. However, in each case it is impossible to predict the outcome of the operation, since in the least severe of ARM, which is the perineal fistula, the incidence of fecal incontinence is 40%.

Since the spinal examination does not affect the treatment and does not allow for the doctor to predict the outcome of the treatment, then it has no meaning. Although the US and MRI of the spine in newborns with ARM does not make sense, and overly aggressive, but they are relatively harmless.

X-ray examination of the pelvis in newborns should be of substantial value in order to justify the use of ionizing radiation. First, statistical correlation does not mean a causal relationship. Secondly, such a correlation does not guarantee diagnostic value. Thus, for example, patients with rectobladder neck fistula and tethered cord have dismal prognosis for bowel control, unrelated to their sacral ratio status [14]. Neurospinal cord dysraphism may be present despite normal sacral ratio [15]. The long-term functional outcomes for patients with 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 [9]. In patients with cloaca no significant difference was found regarding fecal prognosis based on sacral ratio [16].

Some authors are of the opinion that the sacral ratio is useful for diagnostics. However, there is not a single article where this opinion proved. For example, the article by Bischoff and co-authors showed that there is a 75% chance that the tethered cord patient will have a lower ratio than the non-tethered cord patient [17]. The article shows correlation ($P < 0001$), not diagnostic accuracy of the method.

Other authors believe that «... a sacral ratio as a part of the VACTERL screening can help the surgeon identify which patients need closer urologic follow up" [14]. Does X-ray examination make sense in order to determine the need of urological examination if the high association between ARM and renal anomaly is known (22%) [1]? Isn't it better to carry out ultrasonography of the urinary system and pelvis for all children with ARM?

5). What is the reason for poor results of ARM treatment?

The peer-reviewed articles do not specify the method of surgery for ARM, but this only proves that it is a question of posterior sagittal anorectoplasty (PSARP). There are two ways to determine the effect of the operative method on long-term results: (A) comparing the results of different surgical methods, and (B) analyzing anatomical changes when performing different methods.

(A). **After PSARP** "... the rate of continence varied by ARM subtype ($p=0.002$), with the highest rate seen in patients with perineal fistula (60%) and lowest in cloacal exstrophy (0%)" [2]. In males treated for low ARMs with **cutback anoplasty** the "...overall fecal control **was comparable to controls** ($p = NS$) » [18].

(B). During **PSARP**, the puborectalis muscle intersects, the internal anal sphincter excised, the levator plates is cut off from the rectum, and the S2-S4 spinal nerve roots which provide innervation to the rectum, intersects (**Figure 1**).

After **cutback anoplasty**, only the subcutaneous portion of the external anal sphincter intersects. In the healthy people the contraction of this part enhances of the short-term fecal retention during a rise in rectal pressure.

As a result of a large statistical study [2], the authors conclude that "...the type of ARM was the only factor identified early in life that predicted fecal continence in children born with an ARM". Obviously, the wider was the operative field and trauma to the tissues of the perineum, including the muscles and nerves

responsible for fecal retention and defecation, the worse was the postoperative result.

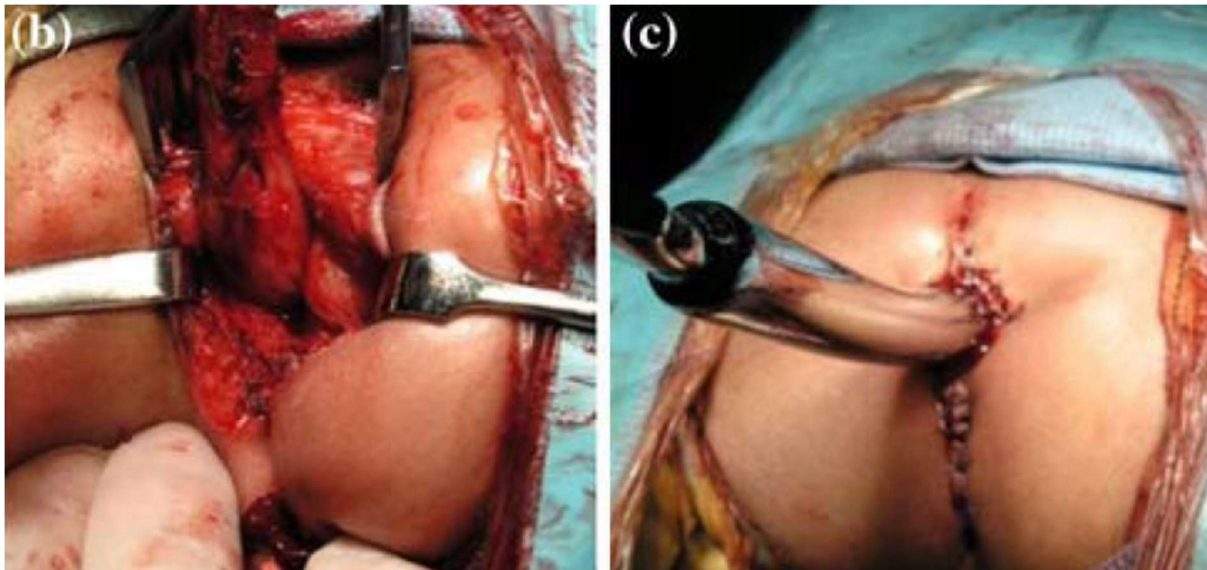


Figure 1.

The steps of the posterior sagittal approach in the girl with vestibular fistula. From the article Shehata [19]. All sections of the descended intestine, including the located in the tissues of the pelvis, were isolated from the surrounding tissues with the inevitable intersection of the neural pathways.

Analysis of articles published in the Journal of Pediatric Surgery, allows to draw the following **conclusions**:

1. There is no possibility and there is no need to predict the outcome of surgical treatment of newborns with ARM.
2. Most children with ARM (with perineal and vestibular fistulas, at least 90% of boys with urethral fistula and patients without fistula) have an ectopy of the functioning anal canal.
3. Poor results of treatment after PSARP are caused by damage of the muscles and nerves that ensure the function of the ectopic anal canal.
4. It is not proven that the presence of spinal defects worsens the prognosis of treatment of ARM. The detection of these defects does not affect the tactics of treatment and these studies do not make sense in the absence of clinical symptoms. X-ray examination of the pelvis (sacral ratio) is not justified.

Respectfully

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Dr Levin--I am not able to accept your Letter to the Editor as it is really a review paper and not an LTE.

Thanks-Whit

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Editor-in-Chief

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