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CONCOMITANT ABNORMALITIES AND DEFECTS IN CHILDREN WITH URINARY MALFORMATIONS

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Backgraund: Urinary malformations are a congenital pathology that occurs in most boys with anorectal defects. An analysis of the literature showed very contradictory and insufficient information regarding the tactics of treating urinary fistulas in children with concomitant anomalies and malformations of other organs and systems. Among them, urethral (bulbar and prostatic) and vesical anastomoses are most often diagnosed. Patients with urinary malformations, unfortunately, have an unfavorable "functional" prognosis. Unsatisfactory results are aggravated in children with concomitant anomalies and malformations of other organs and systems.

Aims: Improving the results of treatment of urinary malformations in children depending on concomitant anomalies and malformations

Materials and methods: The work is based on the results of treatment of urinary malformations in 37 children. All children, along with routine and general clinical examination methods, underwent: excretory urography, cystography, ureterography, voiding cystography, ultrasound of the urinary tract (pelvis), fistuloirrigography, screening tests.

Results: In 37 examined children, 69 variants of concomitant anomalies and malformations were found. The largest group was concomitant pathology of the urinary system, which was detected in 17 children (24.6%). Staged corrective operations were performed depending on the "clinical dominant" of one or another pathology. In a number of cases, in relation to the identified lesions of the urinary tract, it was necessary to adhere to expectant tactics. This was done if information about a specific nosological unit allowed in general terms to predict an unfavorable outcome of the operation, and there were also aggravating moments.

Conclusions: With urinary malformations, most of the associated anomalies and defects remain unrecognized or are diagnosed with a great delay. Erroneous interpretation of the form of the urinary anastomosis, and as a result, unreasonable tactics of surgical correction without taking into account associated concomitant

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defects of the spine, anorectal system, congenital heart defects, brain, leads to the development of complications with severe progressive structural and functional changes in various organs and systems with the development of disability and a decrease in the quality of life of children.

Key words: Urinary malformations in children, concomitant anomalies, treatment results.

Relevance: Urinary malformations are congenital pathologies found in most boys with anorectal defects, most commonly including two types: urethrorectal (bulbar and prostatic fistulas) and vesicorectal fistulas. Differentiating these types is essential from both a prognostic perspective and in selecting the appropriate treatment method. Children with a fistula opening into the prostatic urethra exhibit a higher rate of associated anomalies (60%) compared to patients with a bulbar urethral fistula (30%).

Vesicorectal fistula is considered the "highest" type of malformation among boys; fortunately, its frequency is only 10%. However, associated anomalies are observed in 90% of these children. The ureters and vas deferens are in close proximity to Lieto's bladder triangle and the rectum, requiring caution to avoid damage during surgery. Patients with urinary malformations, unfortunately, have an unfavorable "functional" prognosis. Only 15% of them achieve voluntary bladder emptying by the age of three. These children suffer from urinary incontinence and progressive pathological changes in the urinary tract and other organs and systems.

Objective: To improve treatment outcomes for urinary malformations in children by selecting differentiated corrective strategies based on concomitant anomalies and developmental defects.

Materials and Methods: This study is based on the results of treating urinary malformations in 37 children with associated anomalies of the urinary tract and other organ systems in the Pediatric Surgery Department of the Tashkent Pediatric Medical Institute between 2016 and 2021.

All children underwent routine and general clinical examination methods, including excretory urography, cystography, ureterography, voiding cystography, ultrasound of the urinary tract (pelvic area), fistulography, and screening tests.

Results and Discussion: A total of 37 (100%) children with urinary malformations were analyzed. Among them, 23 (62.2%) had a urethrorectal fistula, and 14 (37.8%) had a vesicorectal fistula. Schematic variants of urinary fistulas are presented in Figures 1 and 2.

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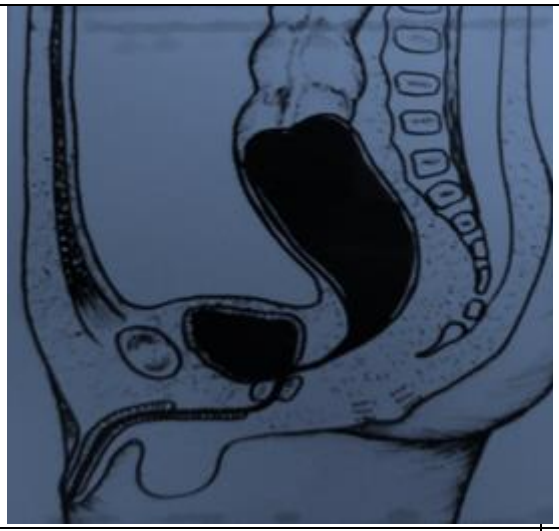
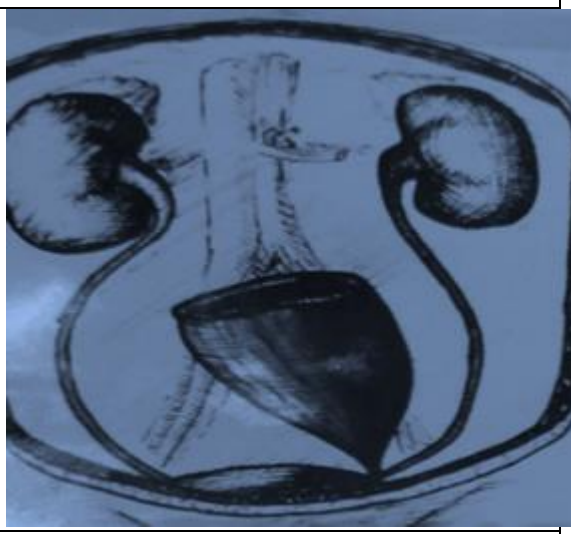
Figure 1. Vesical fistulas

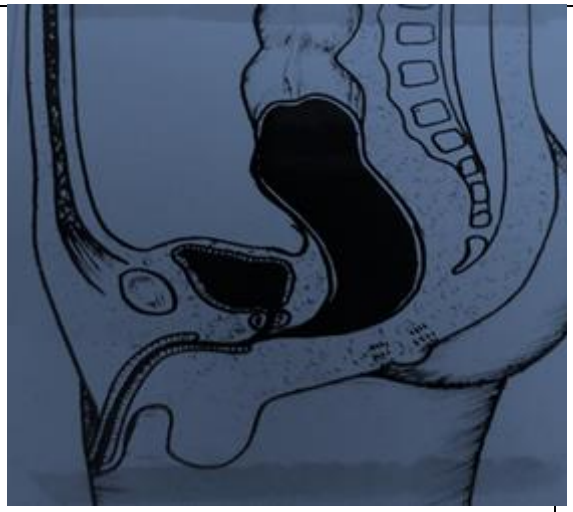


Fistula opening into the bladder neck

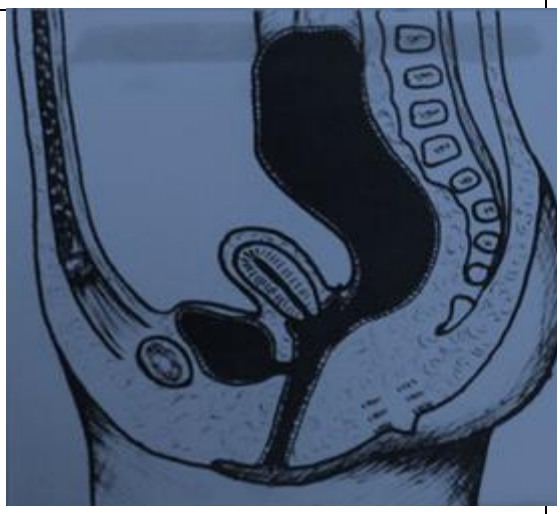


Fistula opening into the ureterovesical segment of the bladder

Figure 2. Urethrorectal fistulas



Fistula opening into the prostatic urethra



Urethra (bladder), intestine, vagina (uterus) opening into a single sinus (Cloaca)

Among the examined patients, most were under one year old - 22 (59.5%). The second most common age group was children under three years - 8 (21.6%). Five (13.5%) patients were aged 4 to 7 years, and two (5.4%) were older than 7 years (Table 1).

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Table 1. Distribution of primary patients by age and nosological forms of urinary malformations (n=37)

Nosological forms	0-1 years	1-3 years	4-7 years	>7 years	Total
Urethrorectal fistula	14 (37.9%)	5 (13.5%)	3 (8.1%)	1 (2.7%)	23 (62.2%)
Vesicorectal fistula	8 (21.6%)	3 (8.1%)	1 (2.7%)	2 (5.4%)	14 (37.8%)
Total:	**22 (59.5%)**	**8 (21.6%)**	**4 (13.5%)**	**3 (8.1%)**	**37 (100%)**

Most children with urinary fistulas had a history of complaints about fecal discharge during urination or urine leakage through the rectal canal.

A thorough examination confirmed that almost every child with urinary malformations had one or more associated congenital conditions. The combination of urinary fistulas with anomalies of other organs and systems is shown in Table 2.

Table 2. Combination of urinary malformations with anomalies of other organs and systems (n=37)

System	Types of anomalies	Total (%)
Uro-nephrological	Renal aplasia/hypoplasia (2), Duplicated kidneys and ureters (2), Congenital hydronephrosis (3), Urethral stenosis (2), Vesicoureteral reflux (2), Megaureter (3), Neurogenic bladder (3)	17 (24.6%)
Cardiovascular	VSD (5), ASD (3), Tetralogy of Fallot (3)	11 (15.9%)
Gastrointestinal	Esophageal atresia (3), Embryonic hernia (2), Ladd's syndrome (3)	8 (11.6%)
Genital	Cryptorchidism (4), Hypospadias (1), Vaginal septum (2), Vaginal atresia (4)	11 (15.9%)
Neurological	Hydrocephalus (3), Spinal hernia (4), Paresis/Plegia (3)	10 (14.5%)
Anorectal	Anal atresia (3), Hirschsprung's disease (1), Intestinal aplasia (1)	5 (7.2%)
Skeletal	Coccyx agenesis (4), Polydactyly (2), Limb aplasia (1)	7 (10.1%)
Total:		**69**

The results show that among 37 examined children, 69 types of concomitant anomalies and defects were identified. A high rate of associated anomalies was

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observed, as every third child had multiple congenital defects either within the same system or in combination with anomalies from different systems.

The largest group consisted of urinary system pathologies, identified in 17 children (24.6%). Notably, complex developmental defects, particularly in the urogenital system, required a strictly individualized and differentiated approach regarding the sequence, timing, and extent of interventions. These conditions often presented with minimal clinical symptoms. Dysuria, pain, and varying degrees of urinary syndrome were rarely observed and were often attributed to the primary disease.

Conclusion: In urinary malformations, a large proportion of concomitant anomalies and defects remain undiagnosed or are identified with considerable delay. An incorrect interpretation of urinary fistula types and, as a result, inappropriate surgical correction tactics without considering associated defects of the spine, anorectal system, congenital heart defects, or brain malformations, leads to severe complications with progressive structural and functional changes in various organs and systems. This results in disability and a decreased quality of life for affected children.

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