Congenital anomalies and malformations of the urinary system as a background of chronic renal disease in children in the region of Mirny in Yakutia

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An inspection, examination and treatment of 2273 children of both sexes aged from 1 month to 17 years of Mirny in Yakutia region. Have been diagnosed the high level of pathology of the urinary system in children and adolescents. Found that children with congenital anomalies and malformations of the urinary system are at risk for the development of chronic pyelonephritis in the future, and so require special medical check-up and preventive measures.

Keywords: children, adolescents, organs of the urinary system, congenital abnormalities and malformations of the urinary tract, pyelonephritis.

Introduction: The pathology of the urinary system is a significant place among the diseases of childhood, maybe not so much the frequency as to severity and seriously of the forecast. One of the distinctive aspects of contemporary Pediatric Nephrology and Urology is the desire for early detection of various anomalies of the urinary system, in order to implement early prevention and early treatment. There are critical periods of fetal development, during which the action of adverse factors can form a variety of abnormalities of the kidneys and urinary tract infections that occur more frequently among the diseases of other organs and systems. The clinical significance of defects the urinary system is very variable: from minor anatomical changes that do not cause functional impairment, to severe malformations, leading to progressive renal failure and subsequent disability. In 30 – 40% of patients with abnormalities of the kidneys and urinary tract develop complications such as pyelonephritis, hypertension, secondary stone formation and shrinkage of the kidney [1,2,3].

According to many researchers, most patients with congenital disorders of the urinary system are no specific symptoms, which leads to late diagnosis in some cases it is delayed from 2 to 8 years [2,3,4,10].

This is evidenced by the prevalence of operated patients with malformations of the kidneys older children, which indicates the possibility of a long latency of the disease is not accompanied by severe clinical symptoms. For most children the disease is detected in the survey randomly [4,5]. At the present time attracts the attention of specialists connective tissue dysplasia. Given polyiorgans and polysystem lesions in undifferentiated connective tissue dysplasia is difficult to find a medical

specialty in which the pathology would be of great practical importance. According to the literature in 13% of patients with the connective tissue dysplasia revealed abnormalities of the kidneys in the form of changes in the number, structure and location. The most common nephroptosis, atony pyelocaliceal system, a doubling of kidney and /or urinary tract. Connective tissue dysplasia is the basis for immune disorders, adherence to infection, including urinary tract [5,7].

Congenital malformations are the actual health problem because of their high prevalence [1,3]. Each year, according to WHO 7.9 million babies in the world accounted for 6% of children with congenital malformations. In Russia, the results of monitoring conducted in accordance with the Health Ministry Order № 268 of 10.09.1998 "On the monitoring of congenital malformations in children", the frequency of congenital malformations ranging from 0.27% in Dagestan to 2.47% in St. Petersburg. However, the prevalence of birth defects according to official statistics is low, because the epidemiological studies conducted in some regions of Russia, reveal higher levels from 2.75% in Yekaterinburg to 45.7% in North Ossetia [3,4]. According to official statistics, the prevalence of different congenital abnormalities in the Republic of Sakha (Yakutia) of 4.2/1,000 in the Mirny area – 1.9/1,000.

Malformations of the urinary tract are in fourth place in the structure of congenital malformations, and over the past decade, their prevalence has increased significantly. They occupy a leading position in the structure of causes of disability of children, leading to hardening of the renal parenchyma with the development of terminal chronic renal failure [5,7,8].

It should be noted a continuous increase in the prevalence of diseases of the urinary system as an adult and child population not only of the Mirny region, but also for the Republic of Sakha (Yakutia) in general. The frequency of chronic diseases of the urinary system in the region reaches 27.2%.

Activities aimed at improving the provision of medical and preventive care for children with congenital malformations of urinary system, should focus on early detection, prevention of risk of complications of this disease, to study the optimal form of follow-up [2,3,9,10].

Objective: to study particular pathology of the urinary system, the frequency of congenital anomalies and malformations, and their role in the formation of a chronic disease of the urinary system in children and adolescents in the region of Mirny in Yakutia.

Materials and methods: As part of a contract with "Alrosa" for the period from 1993 to 2011 years in nephrology and urology Children's wards of hospitals in Novosibirsk was examined and treated 2273 children Mirny in Yakutia region in age from 1 month to 17 years. Among the children surveyed the Mirny were – 72.2%, of the cities Lensk, s.s. Good luck, Ayhan, and other –

27.8%. The distribution of children's social status had no significant differences. All subjects lived in the Far North since birth. The examination program included the study of history, diagnosis, comorbidity, and anomalies of other organs and systems. Complex examination of children, along with the standard methods included a renal function tests and instrumental methods: ultrasound of the urinary system, excretory urography, cystoscopy, cystography, isotopic scintigraphy, and radiography, computed tomography, ECG, UCG, ultrasonography of the abdomen, on the testimony of a pelvic ultrasound. Based on the results of a survey of patients carried out verification of clinical diagnosis. According to the standard classification of ICD – X review of each child was exposed to primary diagnosis and comorbid conditions. For statistical processing of the material used the statistical software package Statistica 6. The survey was conducted with informed consent from children, their parents, with the permission of the Ethics Committee.

Results and discussion: on the basis of the data revealed that pathology of the urinary system, as the underlying disease was diagnosed in 500 (22.0%) of children treated in 2273. In addition, 236 children and adolescents (10.4%) was observed as a concomitant disease. Thus, the overall (primary and concomitant disease) pathology of the urinary tract occurred in 736 (32.4%) patients.

When analyzing the frequency of occurrence of diseases of the urinary system in children Mirny region revealed that during the period from 1993 to 1998 years it amounted to – 26.2%, from 1999 to 2004 – 24%, and from 2005 to 2011 years – 23.9%. The findings suggest that the frequency stability of hospital pathology urinary system. By gender differences were found. In the age group from 1 month to 3 years old was diagnosed in 19%, from 4 to 7 years – 33%, from 8 to 12 years – 36% from 13 to 17 years – 12% of cases. Analysis of the structure of urinary system disease in children and adolescents in 1994 – 2011 years showed that of 500 children with chronic kidney disease and urinary tract infections in 425 (85.0%) admitted there was a secondary chronic pyelonephritis, with 79.6% of patients developed against the backdrop of obstructive anomalies of the kidneys and vesicoureteral reflux (VUR) and 20 4% – on the background dysmetabolic nephropathy (DMNP). In 58 (11.6%) of children marked tubulointerstitial nephritis and various forms of chronic glomerulonephritis: glomerulonephritis with nephrotic component, hematuric and hereditary (in 24, 17, 15 and 2 children, respectively). Tubulointerstitial nephritis in all children proceeded against DMNP. Chronic renal failure was diagnosed in 6 (1.2%) boys – teens 13 – 15 years with bilateral ureterohydronephrosis.

The study of genealogical and obstetrical history has shown that parents and relatives have suffered from various diseases of the urinary system, and in 52% of cases – the mother. More than

half of those observed in combined pathology of the urinary system, and infectious-inflammatory diseases of the genitals. Pathological course of pregnancy as a threat of miscarriage, preeclampsia, low water levels, anemia occurred in 89%.

Comprehensive survey of children with diseases of the urinary system showed a high index of congenital malformations of various organs and systems, among which first place is occupied by urinary tract malformations (hydronephrosis, ureterohydronephrosis, a doubling of the urinary tract, VUR 2 – 4 degrees, hypoplasia, agenesis, double renal cystic disease). Hydronephrosis and ureterohydronephrosis often seen in boys. A doubling of the urinary tract and renal anomalies are more common in girls. In 49.6% of children showed a combination of malformations of the kidneys with abnormalities of the heart, 45% – with the anomalies of the gallbladder.

An examination of 425 children and adolescents suffering from chronic pyelonephritis, it was found that 209 patients (109 girls and 100 boys), chronic pyelonephritis proceeded against a background of undifferentiated connective tissue dysplasia – the main group. Group consisted of 216 patients (100 girls and 116 boys).

In the clinical status of children and adolescents of the main group there were: asthenic constitution (90%), underweight (79%), various types of violations of posture (100%), flat (90%), chest deformity (29%), joint hypermobility (89%), functional instability of the cervical spine (45%), hypotension (57%), muscular dystonia (63%), various types of arrhythmias (86%), mitral (12%) and tricuspid valve (6%), added the chord of the left ventricle (39%), their combination (9%), vegetative-vascular dystonia (76%), and various anomalies of the gallbladder (77%). Installed a high percentage of foci of chronic infection of nasopharynx (79%), pathology of the vision (myopia, hyperopia, astigmatism) in 19% of children under 9 years and 37% for teenagers, varicose veins of the lower limbs (5%), hypotonic constipation (39%), hemorrhoids (1%), periodontal (3%), multiple disembriogenesis stigma (eye, dental, ear, skin).

For the children of the main group was characterized by: the diversity, intensity, combination, two-sidedness of various developmental abnormalities (99% and 14% respectively). In the main group of symptoms of pyelonephritis in the foreground of urinary symptoms. Every second child with connective tissue dysplasia pyelonephritis proceeded microsymptoms, latent, and detected by chance during outpatient examination. A decrease in renal function by tubular type. There was a high incidence of membranodestruction process, compared to the comparison group, as evidenced oksalaturia high (100% and 29% respectively), and crystalluria (79% and 15% respectively). Pyelonephritis in children with connective tissue dysplasia had a higher degree of inflammatory activity, which in combination with a longer period of acute (21 days in the

intervention group and 12 days in the comparison group) and frequently relapsing (89% and 27% respectively), can lead to the development of chronic renal failure in a shorter period of time.

The clinical picture of the disease control group of children were the predominant complaints of abdominal pain (77%), recurrent fevers (59%) and asthenovegetative manifestations (75%). Hypertension occurred in 18% of the comparison group of children, mainly with cystic dysplasia and congenital hydronephrosis. In 56% of children in both groups were observed functional disturbances of urodynamics (neurogenic bladder dysfunction).

The structure of secondary pathogens of chronic pyelonephritis significant differences have been received mostly recorded in both groups: E.coli (58,6%), Proteus S. and other Enetrobacter (13,1%), Pseudomonas (13,2%), Candida (3.7%) etc. In 32% of children registered with the mixed infection – E.coli in combination with Staphylococcus spp. and Enterococcus spp.

To achieve the full clinical and laboratory remission in children and adolescents of the main group, as compared with controls were necessary for longer courses of antibiotics, uroseptic therapy, which may be explained by passage of urine during hypotension, urinary tract, the presence of membranodestruction process and lack of vitamin B 6.

Particular attention was paid to the study of reproductive function in adolescents of both groups. In 29% of girls – teen core group were diagnosed with various disorders of the reproductive system (sexual development lag, delayed emergence and prolonged menstruation, irregular menstruation as the dysmenorrhea and the hypomenstrual syndrome), 19% identified inflammatory diseases of upper and lower genital ways. According to a pelvic ultrasound revealed a variety of changes (hypoplasia of the uterus, multifollicular transformation of ovarian, retrodeviation uterus, saddle uterus, etc.). In 59% of adolescent boys with connective tissue dysplasia as diagnosed by a variety of disorders of the reproductive system (delayed sexual development, sexual development disharmonious, varicocele, a condition after surgery for varicocele, phimosis, inguinal hernia, hypoplasia of the right testicle, hypoplasia of the left testicle, hypoplasia of both testicles, gynecomastia).

All children carried out a comprehensive treatment with antibiotics, uroseptik, probiotics, vitamin therapy and symptomatic agents. During the period 1993 – 2011 years for congenital anomalies and malformations of the urinary tract, surgical treatment was 125 children (28.1%), and 6% for repeated surgery. All patients were discharged with improved and detailed advice on diet, a regime of drug therapy, herbal medicine, surveillance in the community pediatrician, nephrologist, urologist, and other specialists.

Conclusion:

Analysis of hospital morbidity in children and adolescents Mirny in Yakutia region for the period 1993 – 2011 years showed that the pathology of the urinary system was observed in 21.9% of patients. Among the examined children were treated and dominated the age group from 4 to 12 years. The increase of birth defects and developmental abnormalities during the observation in 2.5 times. Chronic pyelonephritis in 41.8% of children and young people proceeded against a background of undifferentiated connective tissue dysplasia. Congenital anomalies of the urinary system is one of the underlying causes of chronic disease.

REFERENCES:

1. Arkhipov V.V. Pediatric nephrology at the turn of the century: results and prospects / V. V. Arkhipov, A.V. Papayan // Reference pediatrician. – 2005. – № 1. – P. 55 – 59.

2. Bashkirova E.G. Clinical supervision for children with congenital malformations of urinary system: Textbooks / E.G. Bashkirova, T.I. Sadykova, A. I. Safina. – Kazan, 2008. – 34 p.

3. Pediatric nephrology / ed. M.S. Ignatova. – M. : Medical News Agency, 2011. – 696 p. – ISBN 978-5-8948-1867-2.

4. Ignatova M.S. Actual problems of childhood nephrology at the beginning of the XXI century / M.S. Ignatova // Pediatrics. – 2007. – Vol. 86, № 6. – P. 6 – 13.

5. Kadurina T.I. Connective tissue dysplasia in children (clinical picture, diagnosis, treatment): Thesis dis ... dr. med. sciences: 14.00.09 / Kadurina T.I. – St.Petersburg, 2003. – 43 p.

6. Makovetskaya G.A. The problem of chronic kidney disease in children / G.A. Makovetskaya // Pediatrics. – 2008. – Vol. 87, № 3. – P. 134 – 136.

7. Timofeeva E.P. Connective tissue dysplasia in children with congenital abnormalities of the urinary system: Thesis dis ... candidate med. sciences: 14.00.09 / Timofeeva E.P. – Tomsk, 1997. – 23 p.

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