

Improvement of Quality of Medical Services for Patients with Primary Immuno Deficiencies

Zaitseva E.V.¹, Voronina L.I.¹

¹Ural Federal University, Yekaterinburg, Russia

E-mail: l.i.voronina@urfu.ru

Correspondence Author: Zaitseva Ekaterina Vasilevna

Candidate of Sociology, Associate Professor, Associate Professor of the Department of Theory, Methodology and Legal Support of the Ural Federal University named after First President of Russia B.N. Yeltsin, Ekaterinburg, Russia

E-mail: katia_zai@mail.ru , e.v.zaitceva@urfu.ru

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ABSTRACT

The purpose of the study, the results of which are presented in the article is the analysis of the organization and provision of medical services to patients (children) with primary immunodeficiencies. Object of research – the quality of medical services to patients (children) with primary immunodeficiencies. Subject of research – the conditions necessary for quality medical services for patients with primary immunodeficiencies (children). The authors, using the results of the analysis of theoretical literature and legal acts, reveal some of the characteristics of medical services for patients – children with primary immunodeficiencies: legal, organizational aspects, as well as such additional provision of quality medical services as drug coverage. Identify issues that impede quality medical services for patients with primary immunodeficiencies. The theoretical conclusions made by authors supported by analysis of the situation on provision of medical services to patients (children) with primary immunodeficiencies in the Sverdlovsk region, as well as the results of the survey of satisfaction with medical services for patients with inborn errors of immunity. The

authors also underline the link between quality of care and social aspects of life (visit to the municipal nurseries, kindergartens and schools). The practical significance of the study is to develop recommendations for improving the quality of organization and provision of medical services to patients with diseases associated with primary immune deficiencies.

Keywords: medical services, quality, patients with primary immunodeficiencies, legal, social aspects

Correspondence:

Zaitseva Ekaterina Vasilevna

Candidate of Sociology, Associate Professor of the Department of Theory, Methodology and Legal Support of the Ural Federal University named after First President of Russia B. N. Yeltsin, Ekaterinburg, Russia

E-mail: e.v.zaitceva@urfu.ru

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INTRODUCTION

Inborn errors of immunity (IEI or primary immunodeficiencies – hereinafter referred to as PID) are a large and ever-increasing group, including more than 450 different diseases caused by defects in some components of the immune system (mainly cells and proteins). Although PID is a rare disease, its individual species are more common than other known inherited diseases. Together, they represent an important group of diseases that, in the absence of treatment or poor-quality treatment, become chronic, last a lifetime and are characterized by a severe course with fatal consequences. To reduce the number of patients with diseases based on primary immunodeficiency, timely diagnosis at an early age and subsequent correct treatment are important. If these conditions are not performed, then the diseases become chronic, and patients become disabled, which subsequently creates many problems both for the grown-up child and his relatives, and healthcare institutions, and for the health system as a whole. Therefore, the identification of problems that impede the quality of medical services for children patients with primary immunodeficiencies seems relevant.

MATERIAL AND METHODS

The concept of “medical service” (unlike other branches of government) is not often used in research on medical topics, even in topics on healthcare management. At the same time, the application of this concept is relevant because it allows researchers to evaluate various characteristics of medical services for patients – children with primary immunodeficiencies (legal, organizational), as well as drug provision as an additional condition for the quality of medical services. Also, the use of the concept of “medical

service for patients with primary immunodeficiencies” allows us to identify problems that impede quality. The Federal Law dated 21.11.2011 No. 323-FZ “On the Basics of Protecting Citizens’ Health in the Russian Federation” defines the following: “Medical service – a medical intervention or a set of medical interventions aimed at preventing, diagnosing and treating diseases, medical rehabilitation and having independent finished value”. Accordingly, medical care is considered as a set of measures aimed at maintaining and (or) restoring health and including the provision of medical services [1].

The key in this definition is the concept of “medical intervention”, using which it is possible to evaluate services as different types of medical activities carried out in relation to the patient: these are preventive, research, diagnostic, medical, rehabilitation activities, medical examinations and (or) medical manipulations, and also artificial termination of pregnancy [1]. Directly the citizen, as the recipient of the service either agrees to medical intervention or refuses medical intervention.

Accordingly, a set of medical interventions or a set of medical services is a treatment. At the same time, the purpose of such interventions (or services) is to eliminate or alleviate the manifestations of the disease, or diseases, or conditions of the patient, restore or improve his health, ability to work, and quality of life.

But in general, in normative legal acts regulating the provision of medical services, the conceptual apparatus is not built and poorly perceived by patients, which causes difficulties for researchers due to legal uncertainty and ambiguity. Basically, in the conceptual apparatus used in regulatory legal acts, the concept of “medical care” [2] is used, which is a combination of different medical services.

The medical service, despite its specifics, nevertheless, has common aspects with other types of services. So, the following definition of a service is presented in the text of the National Standard of the Russian Federation: it is the result of direct interaction between the contractor and the consumer, as well as the contractor's own activities to satisfy the needs of the service consumer [3]. A medical service is classified as a socio-cultural (intangible) service, and the patient is considered as a consumer, customer, and user of services.

The application of all-Union State Standard 150 9000 gives reason to consider the provision of services as an action (or a set of actions), as a process or a set of separate stages for the implementation of which resources are required, compliance with the technological process of execution, control, evaluation of results (result and service process).

The most well-established concept of service quality is as follows: it is a set of characteristics of a service that determine its ability to satisfy the set or expected needs of a consumer. At the heart of any service is the principle of priority of the needs and interests of the customer. A similar principle defines the basic requirement for the provision of medical services: this is the priority of the interests of patients.

Methods used in the study:

1) system analysis, which includes the study of the elements of the health care system: this is a service and its quality characteristics; conditions necessary for the provision of services (legal, organizational, personnel, financial; additional conditions – drug provision);

2) the sociological method, namely, a survey using a written questionnaire. Answers from respondents-parents with children with diseases caused by primary immunodeficiencies – reveal problems that reduce the quality of service for this group of patients. Theoretical conclusions about systemic problems (organizational and regulatory) are confirmed by the results of a sociological study conducted by the authors of the article in the Sverdlovsk region, where the number of patients with orphan diseases is 483 people, including 332 children. 56 parents of children and adult patients with primary immunodeficiency living in the Sverdlovsk region (Russia) were interviewed. Respondents were asked questions, the answers to which allowed clarifying the organizational and legal problems that exist in the health system and reduce the quality of the services for the diagnosis and support of patients with primary immunodeficiencies.

Results and discussion

The discourse of quality of medical services is broad and raises serious scientific discussion. The concept of “quality of care” is a very complex construct. In addition to monitoring compliance with standards, is important the quality of health services to the needs and expectations of patients [4]. Analyze the extent to which medical services have the necessary characteristics, and thus how the quality of services meets the needs of patients with illnesses on the basis of identified primary immunodeficiency. First of all, according to the Federal law No. 323-FZ and articles, in particular, the recognition, observance and protection of the rights of citizens to medical assistance in accordance with the generally recognized principles and norms of international

law. This right is specified by Decree of the Government of the Russian Federation No. 403 of 26.04.2012, in accordance with which information about a patient with an orphan disease, is included in regional registers. Then formed the Federal register of persons suffering from life-threatening and chronic progressive rare (orphan) diseases, leading to reduced life expectancy of citizens or their disability [5]. The quality management of the register indirectly affects the quality of subsequent medical services for this category of patients. It means that the contents of the register reflect all the information required by regulation [6], on the basis of which made medical decisions about treatment, prescription drugs and so on. In regions of highly qualified specialists (allergists, immunologists, etc.) keep such registers.

Thanks to the maintenance of the register, conditions are created to ensure such characteristics of medical services as targeting and personification. So, when maintaining the register, personified data on the needs of a particular patient in drugs are necessarily generated.

In the constituent entities of the Russian Federation, executive bodies of state authority in the field of health care must approve the relevant Procedures, the application of which allows providing citizens living in a particular region with medicines and specialized medical nutrition products at the expense of regional budgets. In the event of a conflict caused by difficulties in the provision of drugs, the information in the register can always clarify the legitimacy of the claims of the parties (patients or their representatives and health facilities).

The next characteristic of medical services for patients with PID-based diseases is accessibility. For such patients, a complex of medical manipulations (or medical services), including diagnosis, genetic counseling and treatment, including immunoglobulin replacement therapy, bone marrow transplantation, hematopoietic stem cells and gene therapy, can be provided by medical institutions of levels II and III [7]. That is why in each region they develop the Procedure for the provision of medical care to patients with diseases based on PID, including high-tech, in specialized medical institutions. Undoubtedly, the existence of Article 34, which gives the concept of “specialized”, “high-tech medical care”, is undoubtedly a positive point, since it allows a differentiated approach to assessing the quality of both different types of care and the services that are their integral component. "

1. Specialized medical care is provided by specialists-doctors and includes the prevention, diagnosis and treatment of diseases and conditions (including during pregnancy, childbirth and the postpartum period) that requiring the use of special methods and sophisticated medical technologies, as well as medical rehabilitation.

2. Specialized medical care is provided in stationary conditions and in a day hospital.

3. High-tech medical care, which is part of specialized medical care, includes the use of new complex and (or) unique treatment methods, as well as resource-intensive treatment methods with scientifically proven effectiveness, including cell technology, robotic technology, information technology and genetic methods engineering developed on

the basis of the achievements of medical science and related **branches of science and technology**”[1].

The regional strategy for the provision of medical services allows to systematically satisfy the diverse needs of people with orphan and chronic diseases and disabilities. Regional models of care have important implications for people with these diseases. However, ethical issues related to orphan diseases, disability and regionalization are just begin to study [8; 9]. To establish the sequence of actions of doctors in each region, a procedure is being developed for providing medical care to patients with any disease, including diseases associated with PID. As a rule, the contents of this document include the following items: conditions and form of care, average duration of treatment, list of diagnostic and treatment services, content of drug therapy, criteria for the effectiveness of treatment. Also in such a document indicate the name of medical institutions located on the territory of a particular subject of the Russian Federation, where they can provide the necessary type of care for this category of patients. An example is the document developed in the Sverdlovsk region [7]. By purpose, this type of document is similar to the administrative regulation of public services.

According to experts, for the treatment of children-patients with diseases caused by inborn errors of the immune system, or primary immunodeficiency, are needed immunology and permanent locations for therapy, which, unfortunately, do not exist in many municipal hospitals, is the only regional children's hospitals. Note that ensuring this condition is one of the challenges in the modernization of primary health care, namely, providing access to primary health care as close to its place of residence or training. However, there remain the questions of organization of medical services, implementation of funding hospitals receiving patients with orphan diseases that require expensive treatment, this problem is relevant for other countries [10].

Further, any medical service should be provided through the application of specific standards of medical care. The

standard establishes General provisions for the development of clinical recommendations (treatment protocols) that determine the volume and quality of rendering medical assistance to the patient with specific disease, with a with a specific syndrome or in specific clinical situation. To some extent, the purpose of this kind of standard can be compared with the administrative standard for the organization and provision of public services, namely, the determination of volume and quality indicators. But for the analyzed categories of patients there is no standard of primary health care for disease caused by human immunodeficiency virus (HIV), although these standards are for adults [11].

In general, it can be recognized that the basic conditions are created to provide medical care and, accordingly, the provision of medical services for the analyzed category of patients. But, according to some experts, despite the large organizational, legislative work to provide medical care to patients with orphan diseases, in particular patients with PID, there is currently no concept for the further development of this branch of medicine [12].

There are certain reasons for this judgment. Most nosologies are not included in the list of orphan diseases, which allows, unfortunately, to state that at present there is no proper state support for patients with PID. This problem is also considered by other researchers, for example, in China, a list of 121 orphan diseases has been compiled, but the system of providing social security and the provision of medical services is not yet ideal due to problems of diagnosis and the correct classification of diseases, also [13].

Assessment of patients health care provided allows you to draw a picture of the real situation in the healthcare system. Of course, there is the relationship between quality of care and patient satisfaction [14]. Many patients believe that their participation in the polls will lead to changes in the quality of health services and improving the quality of medical services [15].

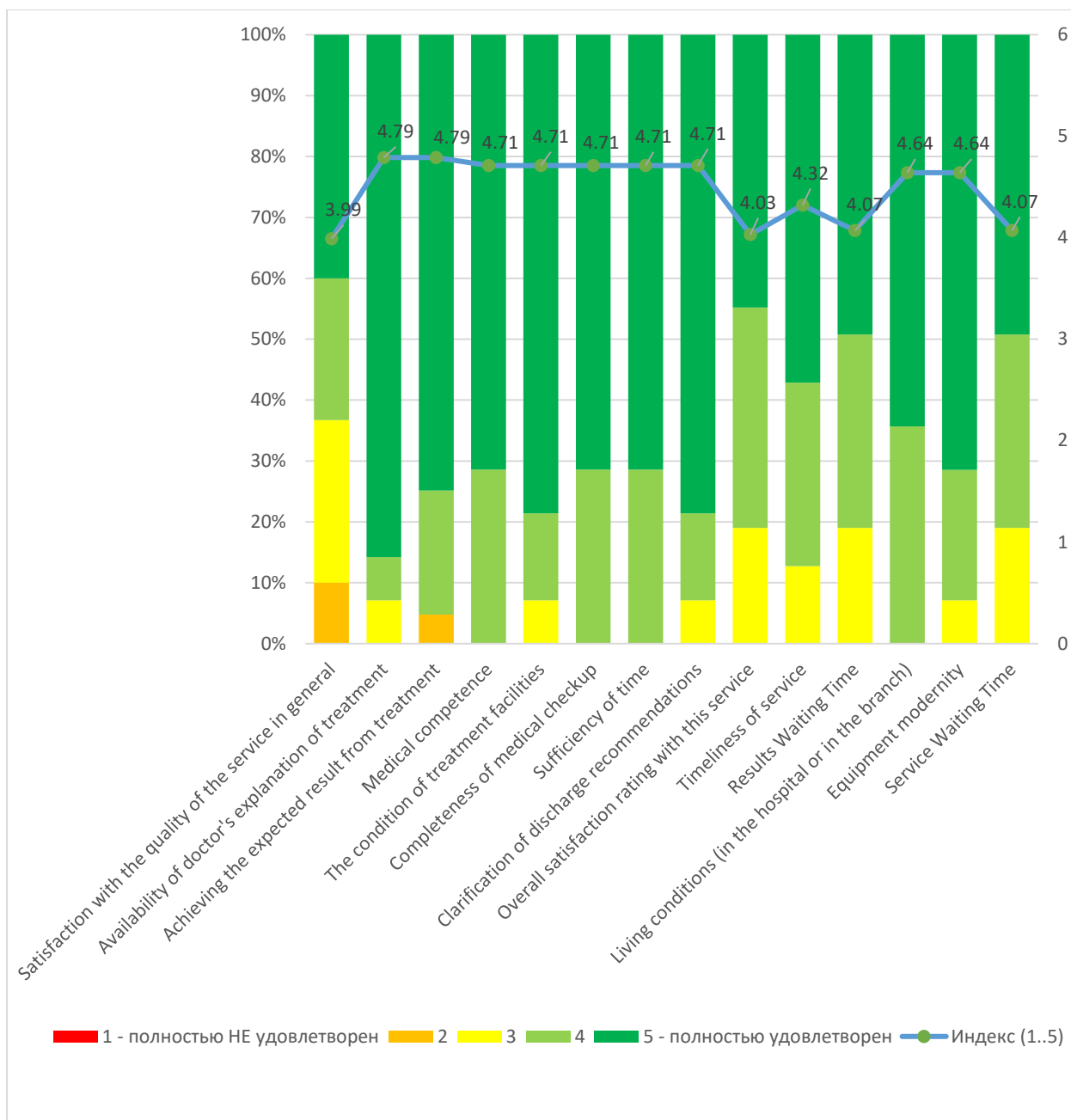


Fig. 1: Satisfaction level of substitution therapy patients (Represented by the parents of minor patients)

In the application study, we attempted to identify the level of satisfaction of patients with PID (or inborn errors of immunity, then – IEI) various medical services in the process of obtaining replacement therapy. Integral evaluation made up 4.49 points. Informants dissatisfied with many parameters, but especially in the quality of service (3.99 points). This figure is slightly asymmetrically reflects the other indicators listed in figure 1. Respondents put a higher estimate for the other components of medical services. However, the lack of coordination between physicians and untimely provision of medical services, the lack of a unified centre of rendering of services in medical support, duration of waiting in a queue or necessary medications reduce patients' satisfaction with these services. Key causes of dissatisfaction in a variety of medical services is determined by the following important parameters. It is the timeliness of

the provision of services: if the service is provided very late, that time has already been lost, timely treatment is not started, it creates a threat to health. Further, this long waiting lines or long waiting period for results, which further exacerbates the situation. Here's the overall evaluation of satisfaction with the service: if the service is not effective, then the patient or his representative is wasted time, money and sometimes disappointing, overall results, similar to ours we see in other studies [16]. In most medical institutions, for example, in the Sverdlovsk region, there are no permits for the use of high-tech care in the treatment of patients with PID (taking tablets or an intravenous drug), which they can receive only in level 4-5 institutions, immunology departments that have permission to use specific preparations. It should be note, that specialized units are not exist in every subject of the Russian Federation.

Also in this study, the opinion of the respondents on the basis of which medical institution assist should be provided, respectively, open immunology departments, create permanent places for substitution therapy, infectious boxes for planned therapy. At the same time, the respondents' opinions were clarified according to three parameters: type of institution, specialization of the department, and geographical location.

According to informants, it is necessary to open immunology departments and create permanent places for substitution therapy at the city or district level, a department, perhaps in the form of a hospital or an outpatient institution, specializing in general therapy.

We found that the opinion of respondents depends on their place of residence. So, respondents from Yekaterinburg equally prefer both outpatient clinics and hospitals, while residents of the region often prefer to receive substitution therapy in hospitals (72.7%). This is due to the fact that in a situation with a day hospital, patients living in the region will have to solve problems with living in the regional center or transport in order to get to the house and back, given that the area of the Sverdlovsk region is almost equal to the area of some European states. If necessary, other prohibitively heavy expenses for treatment, food necessary for patients with PID (IEI), receiving services in a hospital is unacceptable.

The answers of residents of Yekaterinburg and the region to the question on the level of specialization are equally symmetrical, 70% each: residents of the capital of the Urals – for diagnostic specialization and general therapy, representatives of provincial cities – for general therapy. Regarding the opening of infectious boxes for planned therapy, the respondents do not have a consolidated opinion: the largest percentage of respondents (39.1%) think that it is better to open such boxes at the city hospital.

The survey included questions on provision of medicines. The developers of the questionnaire took into account the following circumstances. In the health care system of the Russian Federation there is no single standard treatment of patients whose diseases are due to PID. Accordingly, the territorial funds of obligatory medical insurance (further-TFOMI) in some regions pay for the purchase of specific health care institutions essential drugs or compensate the expenses of parents who purchase these necessary drugs. And in other regions TFOMI refuse such actions. Such actions of TFOMI are only a consequence of the fact that, in accordance with Federal law No. 323-FZ (article 16, paragraph 10, 44, 83 n. 9) providing citizens with drugs intended for the treatment of diseases included in the List of life-threatening and chronic progressive rare diseases, leading to reduced life expectancy of a citizen or disability should be funded at the expense of means of budgets of subjects of the Russian Federation [1]. These guarantees and commitments specified by Government Decree of 18 October 2013 № 932 “On the programme of state guarantees of free rendering to citizens of medical care for 2014 and the planning period of 2015 and 2016”. So, in the text of the resolution stated that at the expense of budgetary appropriations of budgets of constituent entities of the Russian Federation providing citizens duly registered on the territory of the Russian Federation of pharmaceuticals for the treatment of diseases

included in the life-threatening and chronic progressive rare (orphan) diseases, leading to reduced life expectancy of the citizen or his disability [17]. But the conditions for the implementation of such rights at the level of constituent entities of the Russian Federation is not provided: the cost of drugs for the treatment of orphan diseases is not provided in the total amount of funds for financing of health organizations. This leads to a serious problem.

We also need specific conditions for the administration of drugs, which is also established by the standard: under the supervision of a doctor or direct administration by a qualified, specially trained employee (in a medical institution), as well as compliance with special storage conditions of the drug. To ensure these conditions, patients are sent to inpatient departments of medical organizations or day care units [18]. In a situation where there are a minimum number of such inpatient departments or outpatient departments, their geographical remoteness from the place of residence of patients, organizational problems arise that also reduce the quality of medical services.

The researchers also tested the following hypothesis: despite the fact that in some constituent entities of the Russian Federation legislative and executive authorities are making efforts to provide the necessary medical care for patients with rare diseases, in Russia as a whole, to ensure the existing need for treatment in the required volumes only at the expense of regional budgets does not seem possible. Therefore, the proportion of patients provided with medications and medical nutrition products necessary for medical reasons (from the number officially registered in the Register) for individual nosologies ranges from an average of 33% to 90% [19].

Accordingly, the researchers found out from the survey the extent to which patients were provided with the necessary medicines; what medicines are missing. The results of the study allow the following conclusion. According to respondents, the degree of provision of patients with PID (IEI) with the necessary medications and drugs must be much more better. Thus, less than a third of families (28.1%) are fully provided with medicines and drugs. Almost a third of respondents (31.3%) indicated that their families were provided with only some drugs.

Of the 24 possible nosologies in individuals with PID, 22 nosologies exist in the regional segment of the Federal Register. Moreover, there is an increase in the number of patients with these diseases. So, over the seven months of 2013, another category was added to the list of nosologies, respectively, the number of patients who require drug therapy has increased (108 patients, of which children – 64 people, adults – 44 people). The average cost of a prescription for an orphan drug is 204,167.1 rubles. [20].

It should be noted that the residents of Yekaterinburg are better provided with the necessary drugs than the residents of the region. If the informants of Yekaterinburg noted that they are fully provided in 38.9% of cases, then the residents of the region – only in 10%. Also, the availability of drugs is affected by income – the higher the income, the better (according to informants) they are provided with the necessary medicines and drugs (Figure 2). Thus, the hypothesis of the study of a

relatively low degree of provision of drugs and preparations was confirmed.

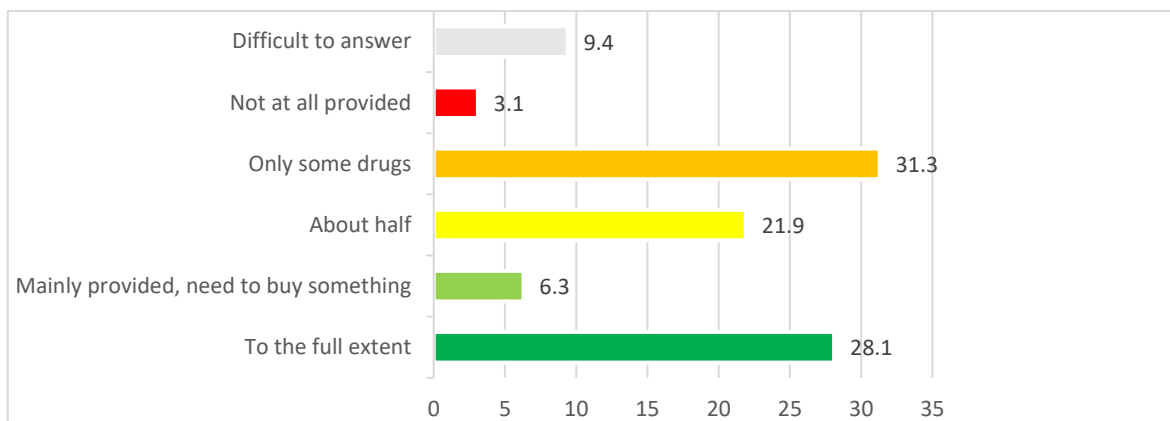


Fig. 2: The degree of provision of patients with IEI with the necessary medicines and drugs (% of the number of respondents)

Another problematic relationship that was identified by the survey results is: the better the patient's health level, the higher the availability of medicines (Figure 3). But the same conclusion can be interpreted in another way: the lower the availability of medicines, the worse the patient with IEI (or his representative) evaluates health.

The logic in interpreting the relationship between health status and the availability of drugs is obvious. According to the respondents, the state of health is deteriorating due to the lack of the necessary drugs; the worse the health, the more drugs are required since there are no drugs due to the inability to buy or receive them, a shortage problem arises, which ultimately affects health.

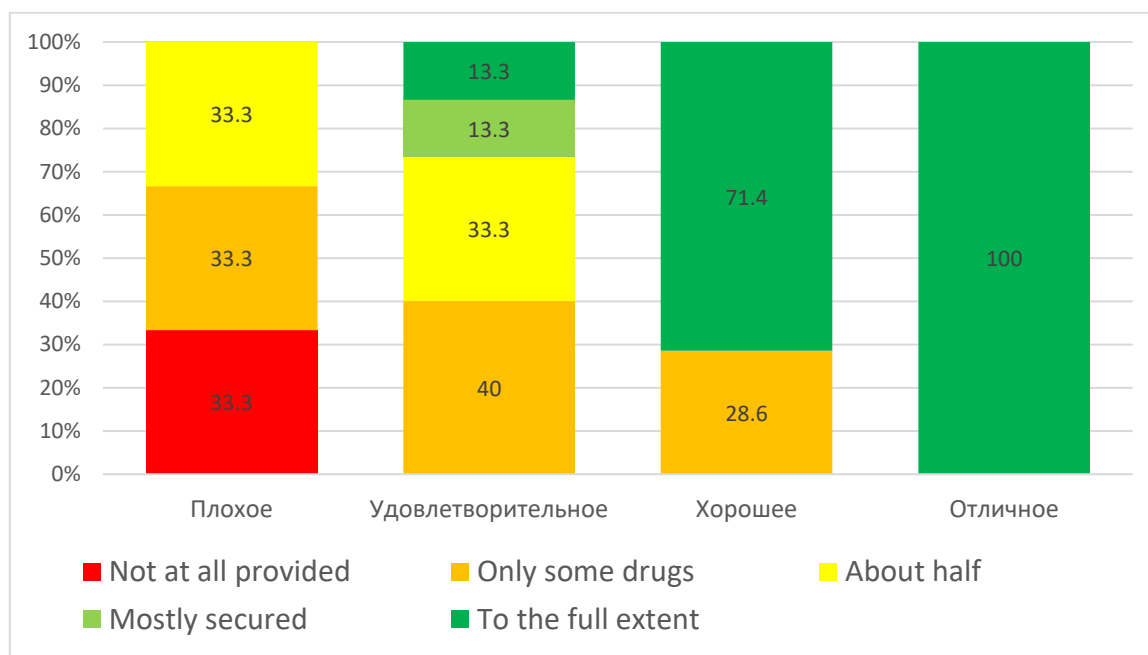


Fig. 3: Provision of necessary medications and drugs, depending on the assessment of the health level of patients with CIE (in% of each health level)

As a result, doctors at polyclinics in which children with a diagnosis of primary immunodeficiency are observed and treated are not able to prescribe drugs which are not included in the standard. In hospitals, these drugs are also absent. And even if the main freelance immunologist in a particular subject of the Russian Federation develops such a standard, because it is not prohibited by law, as a rule, the regional leadership of the executive body of state power in the field of

health does not approve it, fears unforeseen administrative risks. As a result, the patient suffers, and his treatment becomes poor. The absence of a specific vitally important drug in the standard also leads to the fact that in order to avoid potential risk, doctors choose only the only method of treatment (administering the medicine), namely, under control, in a day care center (at the place of residence) or in the regional children's clinical hospital. In this case, a

different method is often ignored, for example, an outpatient or self-made injection if the parent has been trained. Note that this practice has long been established, for example, in the treatment of patients with diabetes mellitus, when the drug is given into the hands and is administered independently by the patient or relative. According to the results of the study, it was found that the lack of drugs for patients with PID (IEI) is due to the

following factors (Figure 4). 36.8% of respondents indicated the economic factor, saying the high cost of medicines. So, 31.1% of informants (from those families who lack drugs) believe that “doctors don’t want to prescribe the necessary drugs”. 21.1% of respondents believe that “doctors do not understand what drugs we need.” According to 21.1%, “there is a long line for the necessary drugs”.

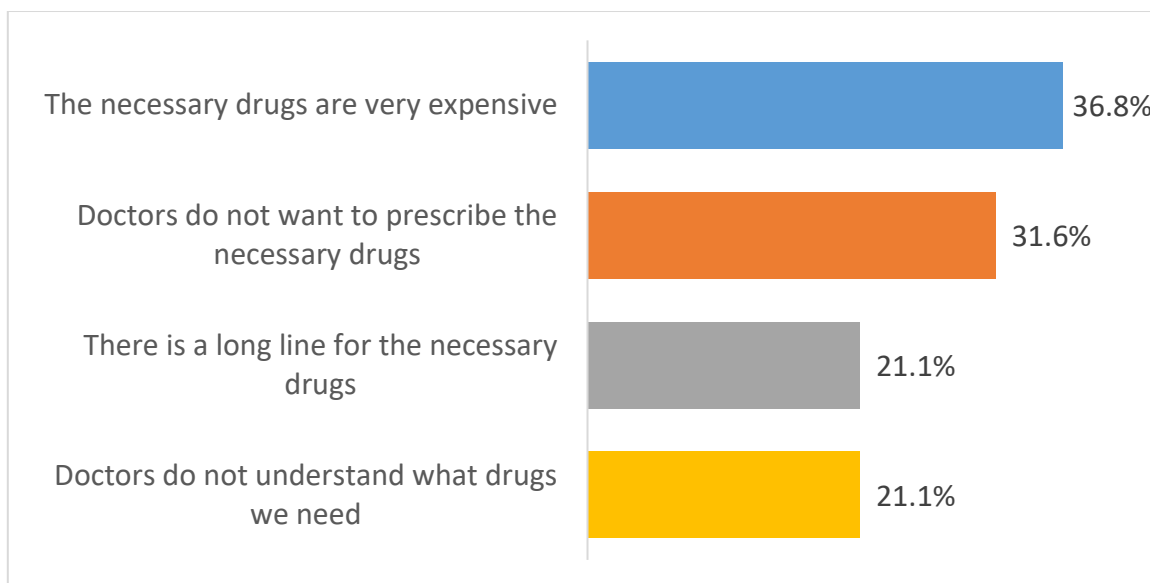


Fig. 4: The main reasons for the lack of drugs (according to informants) in % of the number of respondents. The amount % is more than 100, as several reasons could be mentioned

Thus, informants note that there is a problem in obtaining drugs that occurs due to incompetence or the doctor’s unwillingness to prescribe the necessary drugs. Since we are

talking mainly about drugs that stop the manifestations of IEI, the responsibility lies largely with pediatricians and other specialists.

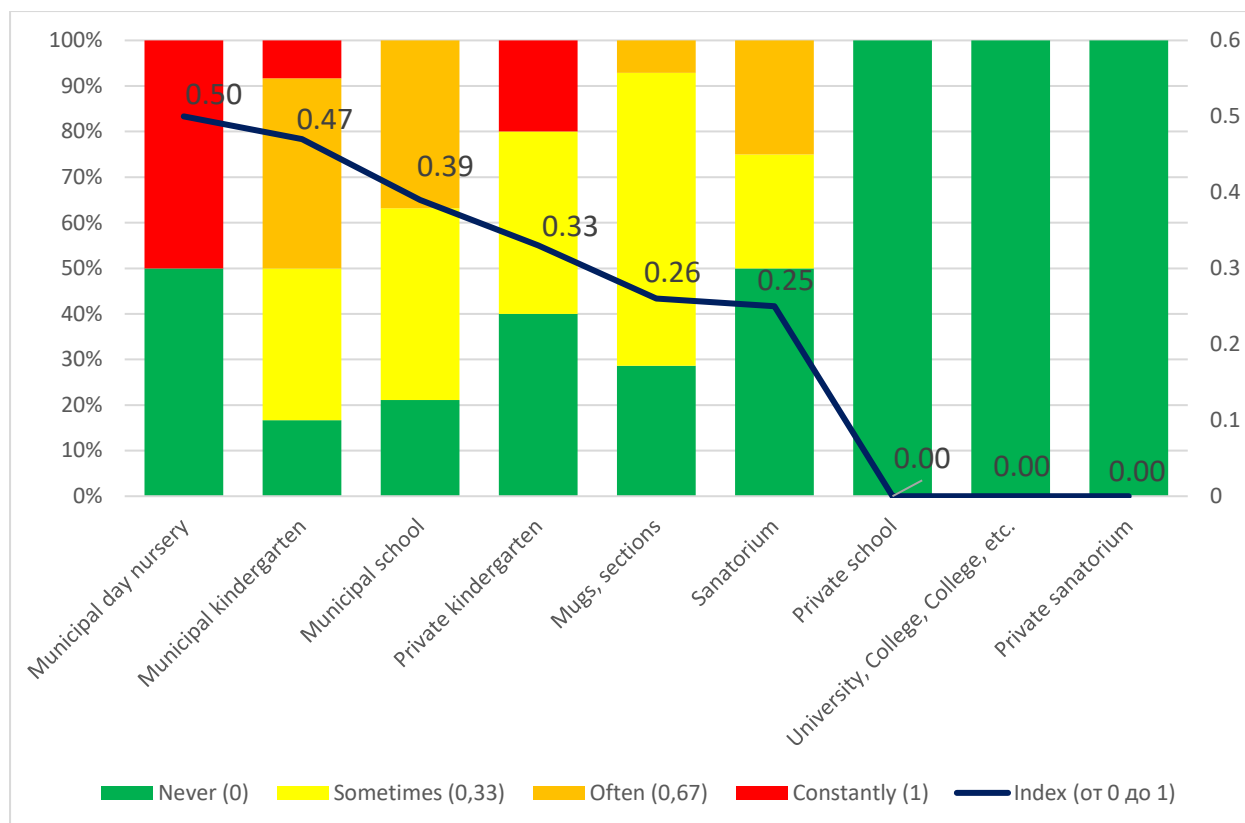


Fig. 5: How often did a family member find it difficult to visit various institutions (% of respondents)

The combination of problems that reduce the quality of medical services for patients with diseases associated with PID poses a risk of lowering the quality of life of the child. If he is a schoolboy and is obliged to attend school, but finds himself dependent on a particular medical institution for a long period of time, he is not able to travel outside the city, because forced to stay in the hospital. The daily routine imposed on the patient by a healthcare institution is not compatible with the educational process schedule. Therefore, this situation prevents full education, entails a child lagging behind the general education program, deliberately puts the future of the child in uncompetitive conditions with respect to peers.

For comparative evaluation of how often family members with children with diseases of the IEI, have various difficulties in educational institutions, researchers Index was calculated by the method of average, which was measured from 0 (all visited never had difficulties) to 1 (all visited constantly experienced difficulties). According to the data presented in figure 5, it is seen that most often the difficulties experienced by family members whose children attend municipal nursery, municipal kindergartens and public schools. Half of the respondents indicates that constantly experienced problems while visiting the municipal nursery. The key cause difficulties for patients with PID (IEI) in the municipal nursery, noted by all informants, is the inability to provide sterile climate in the institution. It is known that often parents bring in a nursery who fall sick or not fully recovered child who may infect others firstly the child with IEI. Certainly, representatives of educational institutions explain the danger of this situation to the parents and to prevent sick

children attended nurseries. However, parents are not always able to determine correctly whether the condition of their child's health risk to other children. Also members of families where there are sick children with PID (IEI), are faced with the lack of special children's institutions, and the unwillingness of the staff of the nursery work with those children. The second place on frequency index is the difficulty associated with attending a municipal kindergarten children. Someone interviewed the patients' parents constantly struggle with, someone occasionally. And only 16.7% of parents surveyed whose children with IEI attended kindergarten, never experienced difficulties. The key cause of difficulties for those who attended kindergarten, the same as for nursery – the inability to provide sterile climate. A third of parents whose children attended kindergarten, noted the lack of specialized children's institutions, 20% of respondents pointed to the unwillingness of personnel to work with these children, the lack of kindergarten health professionals, as well as the presence of aggression to a child with a IEI by other children or their parents.

CONCLUSION

The analysis of theoretical literature, legal acts and practices allows us to conclude the following. Currently, the regulatory framework governing the provision of medical care and, accordingly, specific medical services for patients with PID, is incomplete and difficult to apply, because it includes many acts, which constant changes. At the same time in the list of orphan diseases need to make primary immunodeficiencies, and such a decision should not be delayed. Next, we need to recognize that managing the organization and delivery of

health services is more difficult than in other social services. But it is possible to take into account the accumulated experience in organization of public services, the development and application of administrative regulations for systematization of the normative acts of medical services. A powerful source for making adjustments and improving the quality of medical services are the results of surveys of patients, their representatives. Our study showed dissatisfaction of patients quality of provided services, however, these studies show similar trends [21; 22]. Also the results of the surveys help to identify how the poor quality of health services affect other aspects of children's lives – patients, namely, the quality of socialization. Such complex approach in identifying problems and their subsequent solutions will allow to create conditions for improving the quality of medical services to patients (children) with primary immunodeficiencies.

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