

The Main Criteria for Determining Disability in Children with Congenital Cleft Upper Lip and Palate according to the International Classification of Functioning (ICF)

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Article History:

Submitted: 15.01.2020

Revised: 16.03.2020

Accepted: 20.04.2020

ABSTRACT

The role of social rehabilitation of children with congenital facial pathology is very large. Children with this pathology undoubtedly belong to the disabling group and are in urgent need of medical and social protection based on a multidisciplinary approach, involving a neonatologist, a pediatrician, an orthodontist, a maxillofacial surgeon, an ENT doctor, a speech therapist, a teacher, a psychologist, and a social worker. When determining disability for children with congenital cleft upper lip and palate by ICF, it is very important to assess the

clinical condition of the child according to the criteria for the severity of cleft upper lip and palate, followed by their calculation.

Keywords: face pathology, cleft, disability in children, lip, palate

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DOI: [10.31838/srp.2020.4.62](https://doi.org/10.31838/srp.2020.4.62)

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INTRODUCTION

Medical and social rehabilitation of children with congenital maxillofacial pathology is extremely relevant. It consists in the fact that congenital malformations of the maxillofacial and craniofacial regions in children are widespread and differ in the severity of anatomical and functional disorders, the difficulty of social adaptation of patients, economic aspects, and therefore are one of the most important problems of medicine [1, 2, 3].

Congenital and hereditary pathology of the maxillofacial region in children occupies a leading place in maxillofacial surgery. According to WHO, about 3.2 million children are born with various developmental disabilities every year in the world. Malformations of the maxillofacial region occupy the 3rd place among other congenital malformations in children. Of these, 70% are congenital clefts of the upper lip and palate; 30% are other forms of craniofacial dysostosis [2].

In the Republic of Kazakhstan, according to the National Genetic Register, from 2.5 to 3.0 thousand children with congenital malformations are born every year, of which at least 350 are children with a congenital cleft upper lip and palate. According to statistics from the Department of health of Astana and the region, the birth rate of children with congenital cleft lip and palate in Astana (Kazakhstan) for the period from 2005 to 2009 is 1 sick child per 964 newborns. According to statistical data for 2010, the prevalence of congenital and hereditary pathology of the maxillofacial region on the territory of the Republic of Kazakhstan on average was 1 child with a congenital malformation of the maxillofacial region in 889 newborns, and in environmentally unfavorable regions of Kazakhstan - 1 in 422 [1, 3].

In the etiological aspect, congenital malformations of the face belong to a multifactorial pathology: endogenous and exogenous, as well as genetically determined factors. Complex anatomical and functional disorders of the child's organs and body systems lead to maladaptation and disability [10].

The purpose of this study is to evaluate the effectiveness of the criteria of the score system for determining disability in

children with congenital cleft upper lip and palate according to the International classification of functioning (ICF).

MATERIAL AND METHODS

In the clinic of the Department of orthopedic and pediatric dentistry of "Astana Medical University" JSC, we conduct examinations, comprehensive treatment and follow-up rehabilitation with a multidisciplinary approach for children with congenital facial pathology from different regions of the Republic of Kazakhstan (Fig.1.). Among them children with various pathologies have been monitored in recent years:

- Congenital cleft of the upper lip and palate - 829;
- Ectodermal dysplasia -7;
- Pierre Robin syndrome-5;
- Goldenhar syndrome -3;
- Franceschetti syndrome -1;
- Other unclassifiable defects-5.

The pre-assessment and examination of all children is performed in the clinic by a dental surgeon, an orthodontist, and a dental therapist.

In the Department of neonatal surgery of JSC "National Scientific Center of Motherhood and Childhood" and the Perinatal center No. 1 in Astana, on the first day of the operation – cheiloplasty – is performed by a dental surgeon. Further, the surgeon performs preoperative preparation for the subsequent stages of the operation and sets the timing of surgical intervention according to the indications.

Congenital malformations of the maxillofacial region are characterized by the severity of anatomical and functional disorders, and the difficulty of social adaptation of patients. In 96.4% of cases of children with congenital cleft lip and palate there are various dental anomalies, postoperative secondary deformities.

Children with severe craniofacial pathology were sent for consultations in Almaty, Moscow, St. Petersburg, South Korea, and Turkey. A child with Goldenhar syndrome was operated on in Astana by a team of surgeons invited from the UK.

The plan of rehabilitation stages is made by the dentist with the involvement of a pediatrician, ENT doctor, speech

therapist and according to the indicators of a psychologist. Rehabilitation activities are carried out for children after an explanatory conversation with their parents by each of the specialists separately.

Orthodontic treatment is key, since the subsequent feeding of the child and the outcome of surgery depend on the professional provision of orthodontic care. In this case, it is necessary to take into account the complexity of manufacturing orthopedic structures in the conditions of the clinic (the possibility of taking impressions on the little patients). Before the first feeding, the orthodontist makes a preformed plate for the child, which is replaced every month, and an endonasal obturator is prepared for postoperative patients. In the absence of the possibility of manufacturing an orthopaedic apparatus, the presence of comorbidity, to implement full-fledged feeding children artificially we have implemented and widely used special NUK nipples (made in Germany) at the National Scientific Center of Motherhood and Childhood and Neonatal Pathology Department of the 1st City Clinical Hospital of Astana city.

The program of speech rehabilitation of children with maxillofacial pathology is based on early diagnosis of speech disorders, providing timely speech therapy and defectological assistance, taking into account the mental characteristics of children of each age group. Speech therapy assistance is provided by a *speech pathologist*. A feature of this specialist's work is mandatory training in logopedic skills and speech, conducted before surgery and in the early age group. Pre-training greatly facilitates and accelerates the recovery of normal speech after surgery.

The *dentist-therapist* carries out activities to teach parents and children the rules of hygienic care of the oral cavity and work on the rehabilitation of the oral cavity by age. For local remineralizing therapy, modern remineralizing agents are used.

After a course of treatment on the basis of a multidisciplinary principle, children are given advisory opinions with recommendations for all stages of further rehabilitation, on the basis of which the child is issued a disability at the place of residence. Also, during the rehabilitation stages, advisory opinions are prepared for the registration of social benefits for disability in accordance with the main criteria of the ICF.

There is a certain procedure for establishing a disability group in various nosologies according to the order of the Minister of Health and Social Development of the Republic of Kazakhstan dated January 30, 2015 No. 44 "Rules for conducting medical and social examination". Examination of children with defects is carried out by specialists of medical and social examination (MSE) in accordance with 3P. of the Decree of the Government of the Republic of Kazakhstan dated 02.08.2013 No. 771 "Rules of medical and social examination".

Children with congenital pathology of maxillofacial area at medical-social expertise (MSE) for the establishment of groups of disability are sent to experts of Medical–Control Commission (MCC) of outpatient organizations, they bear the fullness and volume of the examination.

But sometimes there are a number of problems, because there are no standards of examination for medical rehabilitation. Often, patients are sent to a meeting of the medical and social expert Commission (MSEC) without research results that reflect the functional state of the patient and are relevant for expert evaluation by the Commission members. The order does not regulate the algorithm of the patient's examination for referral for medical and social examination, and the clinical protocols are of a recommendatory nature [5, 6].

Basic (mandatory) diagnostic examination performed on an outpatient basis is necessary to register a disability: face anthropometry; abdominal ultrasound; chest x-ray; echocardiography. In addition, additional diagnostic tests are carried out: X-ray of the facial bones; CT of the maxillofacial region; CBC; common urine analysis; blood group determination; determination of the Rh factor.

The advisory opinion for determining disability the nosology code is indicated in accordance with the International Classification of Diseases (ICD) of the 10th revision. Taking into account that the point system of criteria for assessing the degree of disability is focused on the syndrome model, it is necessary that while directing to the MSEC, a specific syndrome, stage, phase, and complication should be reflected in the clinical diagnosis.

To determine the degree of disability for children, methodological recommendations are used to determine the degree of general disability, where the recommended estimates range from 10 to 30%, which is extremely insufficient for an objective reflection of reality in order to determine the degree of disability. Specialists of medical and social expertise departments in the regions can interpret these percentages in different ways, since they are of a recommendatory nature.

When determining disability, the following criteria are provided:

- Features of child care and feeding;
- Preoperative orthopedic and orthodontic care;
- Surgical and speech therapy services;
- Assessment of the child's somatic and neurological status;
- Diagnosis of concomitant defects;
- Assessment of the severity of maxillofacial malformation;
- Assessment of postoperative status;
- Evaluation of anatomical and aesthetic data, speech formation data in the postoperative period.



Figure 1: Children with congenital face pathology

To establish disability group for children by severity and disability certain criteria are used, but there are no quantitative assessment of the degree of violation of functions of organism and accordingly, the degree of restriction of functions that does not exclude the subjective factor in the evaluation during the examination.

In order to provide objective clinical characteristics of congenital pathology of the maxillofacial region of the child,

violations of physiological functions of the maxillofacial area in the framework of the scientific project of the Ministry of Health of the Republic of Kazakhstan, we have developed criteria that characterize the severity of the pathology, estimated in points. These criteria are currently used by specialists in the work of the Commission of Medical and Social Expertise in determining the disability group for children.

Criteria for assessing the severity of congenital cleft lip and palate according to the score system

1.1. Evaluation of the function of food intake (feeding)	
Evaluation criteria	Score
1. No problem when eating: the formation of a bolus of food is impaired, swallowing is physiological.	0
2. Mild problems: chewing, food bolus formation disorders, and swallowing are associated with functional scarring of the upper lip and soft palate. The presence of a residual defect acquired after surgery in the vestibule of the oral cavity, in the anterior part of the hard palate, along the median line of the soft palate.	1
3. Moderate nature of problems: violation of sucking, chewing, swallowing, associated with insufficient activity of the walls of the pharyngeal ring, with an insufficiently formed bottom of the nasal passage after surgery. The presence of paresis of the palate, residual defect acquired after surgery.	2
4. The expressed nature of the problems: violation of sucking, chewing, ingestion of food in the nasal cavity, aspiration of liquid food into the respiratory tract, feeding in a forced semi-vertical position using special nipples, the presence of a preformed plate for separating the oral cavity from the nasal cavity.	3
1.2. Assessment of breathing:	
Assessment criteria	Score
1. No problems: nasal breathing, position of the tongue in the anterior part of the upper jaw.	0
2. Mild problems: respiratory disorders or mouth breathing associated with abnormalities and deformities of the dental system after surgery.	1
3. Moderate nature of problems: respiratory disorders associated with the deviation of the nasal septum after surgery; hypertrophy of the nasal mucosa.	2

4. The pronounced nature of the problems: violation of nasal breathing, lack of pulmonary ventilation, leading to hypoxia and anemia; reduction of the protective function of the nasal cavity - providing filtration, heating and humidification of the air.	3
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1.3. Assessment of speech formation, palatopharyngeal insufficiency	
Evaluation criterion	Score
1. No problems: long movable palate, the high dome of the palate, the anatomy of the palate is restored, the function and articulation of speech is good.	0
2. Mild problems: speech disorders of a phonetic nature, delayed psycho-speech development, a residual defect in the vestibule of the oral cavity, on the border of the hard and soft palate, a short inactive palate, a residual defect in the anterior part of the hard palate.	1
3. Moderate problems: closed rhinolalia; scarring of the upper lip, the area of the hard and soft palate; Gothic palate, violation of articulation of sounds, delay in speech development and the emotional-volitional sphere, nasal speech and voice timbre. Diminished hearing.	2
4. Pronounced nature of problems: open rhinolalia, communication of the oral cavity with the nasal cavity. Absence of anatomical conditions for full-fledged palatine-pharyngeal closure, violation of interaction between articulation and respiratory muscles.	3
1.4. Assessment of the child's weight-a weight defect associated with a malformation of the oral cavity.	
Assessment criteria	Score
1. No problems: there is no deficit of weight of the child	0
2. A child's weight deficit less than 30%	1
3. Child weight deficit in the range of 30-40 %	2
4. Child weight deficit in the range of 40-50%	3
1.5. Evaluation of orthopedic and orthodontic treatment	
Assessment criteria	Баллы
1. Does not require orthodontic treatment	0
2. Fixed constructions (braces), myobrases.	1
3. Removable orthodontic plate that expands the upper jaw, myobrases, trainers.	2
4. Orthopedic preformed plate before the first feeding with replacement 1 time a month up to 3 years, vibrating obturator with a massage brush, surgical intervention.	3

1.8 Assessment of the severity of maxillofacial malformation

1.6. Anatomic-aesthetic and functional disorders after cheiloplasty		
Condition of the lip, nose, and vestibule of the oral cavity after cheiloplasty surgery.	Degree of deformation	Score
1. The shape of the lip is correct, mobile, the vestibule is deep, there is no scarring.	no	0
2. Scarring of the filtrum of the red border of the lips, the wing of the nose, a short frenulum of the upper lip, flattening of the nose, and the absence of a vestibule.	first	1
3. Short flattened and excessively high upper lip, red border screwed into the mouth, rough scarring deformation of the lip and nose	second	2
4. Disfigurement of the skin of the upper lip and nose, impaired mobility of the upper lip. Deformity of the red border, the vestibule of the mouth and nose.	third	3
1.7. Anatomical and functional disorders after uranoplasty		
Morphological and functional data of the operated palate	score	
1. Long, movable palate, the anatomy of the palate is restored, the function and articulation of speech is good.	0	
2. Short, sedentary palate, residual defect in the anterior part, narrowing of the alveolar arch in the anterior-lateral part, impaired articulation of speech, nasal pronunciation.	1	
3. Residual defect on the border of the hard and soft palate, shortening of the palate, unformed uvula, nasal speech.	2	
4. Scarring, shortening of the palate, residual defect (one or two), slurred speech, Gothic palate .	3	
1. Congenital unilateral cleft of the upper lip without deformity of the skin-cartilage section of the nose.	1	
2. Congenital unilateral cleft of the upper lip with deformity of the skin-cartilage section of the nose.	2	
3. Congenital unilateral cleft of the upper lip, alveolar process, upper lip and palate with deformity of the skin-cartilage section of the nose.	3	

4. Congenital bilateral cleft of the upper lip, alveolar process, upper lip and palate with deformity of the skin and cartilage of the nose.	4
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Quantitative assessment of disability in congenital cleft upper lip and palate		
Risk	Score	International Classification of Functioning
No or low risk	0-10	No problem, or low -5-24%
Average risk	10-20	Moderate problems 25-49%
High risk	20-25	Severe problems (high, intense) 50-95%

Calculation of the severity criteria with congenital cleft lip and palate in a clinical example

Patient	Birth year	Diagnosis	Criteria	Score
Patient 1 (boy)	2014	Congenital full cleft upper lip, alveolar ridge, hard and soft palate on the right. Condition after heylo-uranium plastics Q 37.5	1.1	1
			1.2	2
			1.3	2
			1.4	1
			1.5.	2
			1.6.	3
			1.7.	3
			1.8.	3
Patient 1 (girl)	2013	Congenital full cleft upper lip, alveolar ridge, hard and soft palate on the right. Condition after heylo-uranium plastics Q37.5	1.1	2
			1.2	2
			1.3	2
			1.4	1
			1.5	2
			1.6	3
			1.7	3
			1.8	3

During the calculation, the following results are obtained:

- 5-24% - mild problems: low risk of disability.

The highest score of the criteria was 1.6; 1.7, the correction for these criteria indicators, where a child has a secondary deformity, is carried out at the age of 12 to 14 years.

As part of this study, we have assessed the degree of disability and the effectiveness of the current system for establishing disability in the Republic of Kazakhstan. A very important positive situation in determining the degree of disability of patients is that since 2005 there has been a unified information system, the Central database of disabled children in the Republic of Kazakhstan, which is integrated with the Central database, employment agencies and social programs.

The new international classification of functioning, unlike the previous version of 1980, has certain advantages and includes a number of new concepts. It focuses on the overall range of functioning of the human body – in the case of a child with disabilities, it indicates the need to assess all of his or her capabilities, not just physiological limitations. The new ICF includes different degrees of disability, placing them in a "broad continuum of the functioning of the human body. Disability is not viewed as something that affects a minority of people, but as a universal experience of humanity. At some point in their lives, most people become disabled to some extent" [8, 9].

It presents the degrees of functioning, ranging from "no violations" to "violations of the functions of the entire body".

Classification is not limited to making a medical diagnosis. WHO argues that by shifting the focus from cause to effect, the ICF gives equal status to all health conditions [7].

SUMMARY

The Role of social rehabilitation of children with congenital facial pathology is very large. Children with this pathology undoubtedly belong to the disabling group and are in urgent need of medical and social protection based on a multidisciplinary approach, involving a neonatologist, a pediatrician, an orthodontist, a maxillofacial surgeon, an ENT doctor, a speech therapist, a teacher, a psychologist, and a social worker.

When determining disability for children with congenital cleft upper lip and palate by ICF, it is very important to assess the clinical condition of the child according to the criteria for the severity of congenital cleft lip and palate, followed by their calculation.

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