



JOURNAL OF ORAL MEDICINE AND CRANIOFACIAL RESEARCH

журнал стоматологии и краниофациальных исследований

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СОПУТСТВУЮЩИЕ ПОРОКИ РАЗВИТИЯ У ДЕТЕЙ С ВРОЖДЕННОЙ РАСЩЕЛИНОЙ ГУБЫ И НЕБА



http://dx.doi.org/10.26739/2181-0966-2020-1-11

АННОТАЦИЯ

На базе 2-клиники Самаркандского государственного медицинского института проведен анализ лечения 122 пациентов с различными видами врожденных расщелин верхней губы и неба. В результате проведенных исследований выявлено, что врожденные расщелины верхней губы и нёба встречаются в сочетании с аномалиями развития других органов. Основными из них были- короткая уздечка губ и языка у 76 пациентов, низкий индекс массы тела у 97, деформация позвонка у 6, косолапость у 19, врожденное плоскостопие у 5, врожденная дисплазия тазобедренного сустава у 2, дефекты развития мочевыделительной системы у 19, пренатальная энцефалопатия у 46, врожденная мышечная недостаточность у 15 детей. Анализ проведенного исследования показал, что для достижения высокого уровня медицинской помощи детям с данной патологией требуется четкая и единая, взаимосвязанная тактика всех специалистов, участвующих в лечении ребенка с врожденной расщелиной губы и нёба.

Ключевые слова: врожденные расщелины верхней губы и нёба, пороки развития, лечение, дети.

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ACCOMPANYING DEFECTS OF DEVELOPMENT IN CHILDREN WITH CONGENITAL CLEFT OF LIP AND PALATE

ABSTRACT

An analysis of the treatment of 122 patients with various types of congenital clefts of the upper lip and palate was performed on the basis of the 2nd clinic of the Samarkand State Medical Institute. As a result of our studies, it was found that congenital clefts of the upper lip and palate are found in combination with developmental abnormalities of other organs. The main ones were short frenulum of lips and tongue in 76 patients, low body mass index in 97, vertebral deformity in 6, clubfoot in 19, congenital flat feet in 5, congenital dysplasia of the hip joint in 2, defects in the development of the urinary system in 19, prenatal encephalopathy in 46, congenital muscle failure in 15 children. Analysis of the study showed that to achieve a high level of medical care for children with this pathology, a clear and uniform, interconnected tactics of all specialists involved in the treatment of a child with congenital cleft lip and palate is required.

Keywords: congenital clefts of the upper lip and palate, malformations, treatment, children.

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БОЛАЛАРДА ЛАБ ВА ТАНГЛАЙ ТУҒМА ЁРИҚЛАРИ БИЛАН РИВОЖЛАНАДИГАН БИРГАЛИКДАГИ НУҚСОНЛАР

АННОТАЦИЯ

Самарқанд давлат тиббиёт институтининг 2-клиникасида юқори лаб ва танглай туғма кемтикли 122 нафар беморни даволаш таҳлили ўтказилди. Олиб борилган тадқиқотлар натижасида юқори лаб ва танглайнинг туғма битмаслиги бошқа аъзолар ривожланишининг аномалиялари билан биргаликда кечиши аниқланган. 76 беморда тил ва лаб тутқичининг туғма калталиги, 97 беморда тана вазнининг пастлиги, 6 холатда умуртқа поғонаси деформацияси, 19 холатда туғма маймоқлик, ясси оёқлик -5, чанок-сон буғимининг дисплазияси-2, сийдик чиқариш тизимининг ривожланиш нуқсонлари-19, перенатал энцефалопатия-46 ва мушакларнинг туғма етишмовчилиги-15 холатда кузатилди. Тадқиқот таҳлили шуни курсатдики, ушбу патологияга эга булган болаларга юқори даражадаги тиббий ёрдам курсатиш учун касалликни даволашда иштирок этадиган барча мутахассисларнинг аниқ ва ягона, узаро боғланган тактикаси талаб этилади.

Калит сўзлар: клиникасида юкори лаб ва танглай туғма кемтиклари, туғма нуксонлар, даволаш, боолалар.

To the most widespread of congenital defects of development refers congenital cleft of upper lip and palate. The birth rate of children with the fact of defect is composed of about 86% anomalies of jaw-facial area and 20-30% defects of development of man [9]. Basically, clefts of lip and palate are considered polygenic multifactoral diseases. They may occur as isolated defect of development and was been one of the symptoms of congenital syndromes (syndrome Van-der-Vud, Pier Robin and others). Besides, in majority of sick children with congenital clefts of lip and palate are occurred accompanying congenital defects of development of other organs and system (heart, lungs, kidneys and others), and it is also important to take into consideration in forming plan of treatment.

Immediately, after birth of a child with these anomalies of development in maternity hospital is established exact clinicanatomical diagnosis and determined the plan of surgical and conservative treatment. In case of need child is moved to the surgical department of newborns where is carried out examination, revealed congenital defects of development of other organs and system, diagnosed accompanying somatic and nervous diseases [1,7,8]. In the presence of accompanying defects of development, severe somatic diseases, diseases of central nervous system the time of surgical treatment are separated. Surgical rehabilitation must be finished not later 5-6 years with that estimation that the child can go to ordinary school [1,7]. Children with Daun, Little, children cerebral paralysis and other rough pathology of organism on contraindications may be operated in later estimations [1,2,5]. Therefore, in the period new birth is composed of individual plan of preparation of patient to the surgical operation with taking into consideration of gravity of defect of development in the maxillary-facial area accompanying defects and somatic status condition of the child [4,6,7,10].

Aim: To reveal accompanying defects of development (ADD) in children with congenital cleft of lip and palate.

Materials and methods: For the examination and correction 122 patients from them with congenital cleft of lip -68(55.74%), palate- 13(10.66%), one-sided- 96(88.07%), two-sided- 13(11.93%) patients, girls- 47(43.12%) and boys accordingly-62 (56.88%) were in the clinic № 2 of SamMI during 2004-2014. 54 patients from them complete cleft of palate -52 (96.30%), with incomplete- 2 (3.70%), girls-19 (35.19%), boys -35(64.81%) were with non closure palate. In primary referencing these children was carried out complete clinic-laboratory examination including clinical, laboratory and instrumental examinations of patients with examination of

qualified specialists such as jaw-facial surgeon, INT doctor, geneticist, orthopedist, pediatrician in necessarily neurologist, hematologist and orthopedist-traumatologist.

Carried out research was showed that the children with congenital cleft of upper lip and palate accompanying defects of development were registered in the area of head, neck, extremities and trunk and were often combined [3,5].

The main of them were short frenum of lip and tongue in 76 patients, low index of body weight in 97, deformation of vertebra in 6, club-foot in 19, congenital flat-foot in 5, congenital dysplasia of hip joint in 2, defects of development of urinary system in 19, prenatal encephalopathy in 46, congenital muscular wryneck in 15 children.

In the process of research the functions of kidneys were established pyuria in 19 children with congenital cleft of upper lip and palate. These patients were carried out special urological examination in which was revealed congenital stricture of prerenal (8) and pelvic area of urethra (6) was carried out ultrasound examination of urinary system. On evidence of excretion urography in 14 patients were revealed pyelcctasis, in 8 hydronephrolic transformation from the direction of urethra obstruction. Cystic-urethral reflux II degree as manifestation of congenital dysplasia stoma of urethra was established in 5 children.

In combination of pathology the child was examined by particular specialists (neurologist, endocrinologist, cardiologist and others), were carried out suitable recommendations, in the case of need was carried out treatment. The children with accompanying defects of development of extremities and congenital muscular wryneck after training features of nursing and feeding, examination of orthodontist and making of obturator for closing of palate defects during feeding were directed for treatment to the orthopedist-traumatologist before cheilorhinoplasty that is till three-four months of age.

Psychoneurologist carried out medico-pedagogical rehabilitating measures with parents and then with patients about medicamental corrective therapy, psychotherapy. All children without exception were examined by neurologist; in the case of need they were taken electroencephalographically examination. Children with encephalopathy got adequate treatment till I stage of operation and further was carried out prophylaxy, observation and the treatment of the specialist [6,7.10]. After made diagnosis and type of hereditary transmission(in the presence of the same) by geneticist were prognosticated the degree of birth risk in proband or members of his family of other children with CCL and P.



It was carried out by the orthodontist ensuring full feeding of child (obturators, disjunctive plastics and etc.), preand post-orthodontic treatment, notice and treatment of secondary deformation of tooth-jaw system, restoration of tooth set. Periods of active observations by doctor-orthodontist are: new born period, before and after operation, changeable bite period, and further systematically observation till complete formation of permanent bite [6,7,10,13]. Logopedist carried out logopedic treatment before and after uranoplastics, training of muscular structure of soft palate, pharynx wall, the development of speech respiration, pre- and post-operative logopedic treatment, development of phonemic hearing, formation of connected speech. Periods of active observation of logopedist are periods in 1-2 years, 4-5 years and further 7-13 years. It has been carried out estimation of somatic status by the pediatrician individually chosen type of feeding and volume of nutrition, evaluated micro flora of oral cavity, immune status, carried out prophylaxy and in the case of need anemic treatment, rickets and hypotrophy, realized preparation for surgical treatment during the first year. These children were actively observed prophylaxy by ENT doctor taking into consideration that the possibility of hit of food from oral cavity to nasal and pharyngonasal cavity may result in to the development of chronic rhinitis, eustachitis, otitis and as consequence to the reduce of hearing.

Even in the absence of evident complaints to the function of cardiovascular system, once-only electrocardiographically research in 74 (60.66%) children with congenital cleft of lip and palate was revealed disturbance of automatism function, myocardial, because of which the patients were examined by cardiologist and got preoperative corrections of disturbances.

Success of surgical treatment in majority of patients depended on timely conduction of complex preparation in conditions of dispensary observation [7,12,14]. Surgical treatment began from 3 months in satisfactory somatic and neurological status.

Heading and coordinating the work of doctorsconsultants the surgeon-dentist determined data and volume of surgical treatment, controlled timeliness referral children to the consultation of particular specialists and carrying out prophylactic treatment, together with them determined absolute and relative contraindication to the operation. It shouldn't forget in decision of question operation dates and higher responsibility of pediatrician giving permission to the surgical treatment. His conclusion about contraindication or indication to the operation of a child having accompanying CDD or somatic diseases, the pediatrician confirmed conclusions of particular specialists (cardiologist, nephrologist, urologist, neurologist, pulmonologist, endocrinologist and others).

Absolute contraindication to the operation is insufficiency of blood circulation IA and IIB degrees.

Relative contraindications are: sharp evident dystrophic changes in myocardium, acute somatic and infectious diseases and not earlier 2 months after recovery hypotrophy I-III degrees, rickets I-III degrees in fastigium period, atopic dermatitis in the stage of recrudescence, anemia with containing of erythrocytes less 3.5x10¹²/l, hemoglobin less 120g/l, colored finding less 0.75, chronic diseases during 6 months after recrudescence, chronic nephritis with manifestations of renal insufficiency, dermatitis and eczema in the facial area, affection of mucousa. Children with relative contraindications were carried out specific treatment accompanying pathology (in the out-patient or profile departments) and on favorable dynamic was allowed operative treatment [7,10]. But in consideration of earlier address to the specialized medical aid from the first days of the life and timely diagnostics of accompanying defects of development, primary cheilorhinoplasty were carried out at the age of 2-3 months, but qualified surgical treatment of children with congenital cleft of upper lip and palate was finished during 12-16 months of life.

Conclusions: Detected in children with cleft of upper lip and palate external stigma of disembryogenesis- congenital defects of development of extremities, kidneys and other changes of organs have systemic character, progress which is conditioned with dysplasia of connective tissue. For extension of complex aid and provision complete rehabilitation of patients with congenital cleft of lip and palate is necessary synchronic work of specialists: geneticist, logopedist, hematologist, otorhinolaryngologist, orthodontist, pediatrician, psychiatrist, orthopedist, surgeon, for timely detection of accompanying defects of development and diseases with the purpose of conduction of full, active preoperative preparation and qualified staged correction of all defects of development.

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