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Original Article

Demographic characteristics of cases diagnosed with cleft palate

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Abstract

Aim: The aim of this study is to retrospectively evaluate the cleft palate cases in our neonatal clinic and to examine the associated clinical features, predisposing factors, accompanying anomalies, additional findings, treatment approaches and follow-up results.

Material and Methods: The study included babies that were diagnosed with cleft palate in our Neonatal Intensive Care Unit between January 2014 and December 2018. The etiological risk factors, demographic characteristics, clinical features and concomitant malformations of the patients included in the study were retrospectively recorded from the patient files and the database system of our hospital.

Results: A total of 70 cleft palate patients were observed over a five year period. Of these, 30 (42.9%) were female and 40 (57.1%) were male. The mean gestational age was 38 ± 2.9 weeks and the mean birth weight was 2845 ± 700 grams. 77.1% of the patients were found to additionally have cleft lips. 27 patients (38.5%) had cardiac defects, 18 (25.7%) had central nervous system anomalies and 4 (5.7%) had hypothyroidism. 24 (34.3%) of the parents were consanguineous. The median age at the time of the initial operation was 13 months for cleft palate cases and 5 months for cleft lip cases.

Conclusion: As a result, treatment and follow-up of patients with cleft palate defect requires a multidisciplinary approach. These patients should undergo a thorough examination and evaluation. Since many syndromes or malformations may accompany the palate defects, these cases should be investigated with respect to genetic disease and other system anomalies.

Keywords: Cleft lip; cleft palate; congenital malformation; newborn

INTRODUCTION

Cleft palate has a multifactorial etiology and originates from the insufficient closure of bone and soft tissues forming the upper bone of the jaw and the roof of mouth in the embryonic period (1,2). Cleft palate-lip and isolated cleft palate are two separate clinical conditions with differences in embryology, etiology and epidemiology (3). While the incidence of cleft lip (with or without cleft palate) is between 1/300 and 1/2500, the incidence of solely cleft palate varies between 1/1000 and 1/1500 (4,5). In Turkey, the incidence of cleft palate-lip has been reported as 0.95/1000, while the incidence of only cleft palate has been reported to be 0.77/1000 (6,7). In a study conducted in our country with 1229 patients, it was reported that 64,6% of the cases included cleft palate-lip, while 35,6% consisted of isolated cleft palate (6). While isolated cleft lip or cleft lip with cleft palate is more common in men, isolated cleft palate is more common in women (8).

Cases can exclusively consist of cleft palate, or they can exhibit accompanying anomalies. The incidence of accompanying anomalies in patients born with cleft palate is reported to be 10-25% (9). Our study aimed to conduct a retrospective analysis of cleft palate cases monitored in our newborn clinic and to assess the associated clinical features, additional findings, predisposing factors, accompanying anomalies, treatment approaches and follow-up results.

MATERIAL and METHODS

Patients

70 infants diagnosed with cleft palate and followed-up at the İnonu University Turgut Ozal Medical Centre Newborn Intensive Care Unit between January 2014 and December 2018 were included in the study. Approval for this study was granted from the Scientific Researches and Publications Ethical Board of our university. The etiological risk factors, demographic characteristics, clinical features and laboratory findings of the participants were retrospectively recorded from patient files and the database system of our hospital. Potential accompanying system anomalies were evaluated using various imaging

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techniques (ultrasonography, echocardiography, and magnetic resonance imaging). The cleft palate patients were divided into two groups based on additionally having cleft lip or not. The group in which cleft palate was not accompanied with cleft lip was classified as isolated cleft palate, while the group that demonstrated both cleft lip and palate was classified as the cleft palate-lip group. The accompanying malformations were classified according to the organ system they affected (cardiovascular system, central nervous system, extracranial skeletal system, craniofacial disorders, urogenital system, endocrine system, eye and ear). The day that the temporary palate apparatus was placed and the day that patients began to be fed through breastfeeding or bottle were recorded. The age at which the cases received their correction operations after discharge was recorded from the database system of our hospital.

Statistical analyses

Average, standard deviation, median, minimum-maximum, number and percentage values were used in the descriptive statistics of the data. Logistic regression analysis was used to find out the necessary risk factors. The results were assessed within a 95% confidence interval and the significance level was accepted as p <0,05. SPSS program (V.22,0; SPSS, Chicago, Illinois, USA) was used in the analyses (10).

RESULTS

A total of 70 cleft palate patients were followed-up in our newborn intensive care unit over a five year period. It was found that 24 (34.3%) of the patients had isolated cleft palate and 46 (65.7%) additionally had cleft lip with cleft palate. 21 patients were found to have a prenatal diagnosis. As stated in Table 1, 30 (42.9%) of the patients were female and 40 (57.1%) male. The average birth week of the patients was 38 ± 2.9 and their average birth weight was 2845 ± 700 gr, while the mean age of the patients' mothers was found to be 30 (18-45).

Odds ratio values were not found to be statistically significant due to the logistic regression conducted with the variables of consanguineous marriage history, mother's age, history of cleft palate in the family, diabetes mellitus, the state of smoking during pregnancy and chronic hypertension (Table 2).

Table 1. Demographic, clinical features and predisposing factors of the patient's group

Gestational age, mean ± SD, (week)		38 ± 2.9
Birth weight, mean ± SD, (g)		2845 ± 700
Male gender, n (%)		40 (57.1)
Cesarean section, n (%)		38 (54.2)
Age of mothers at birth, median (min-max)		30 (18-45)
Consanguineous marriage, n (%)		23 (32.8)
The presence of risk factors in pregnancy	Preeclampsia, n (%)	4 (5.7)
	Oligohydramniosis, n (%)	2 (2.8)
	Polyhydroamniosis, n (%)	1 (1.4)
	Diabetes mellitus, n (%)	6 (8.5)
	Premature rupture of membranes, n (%)	4 (5.7)
	Chronic hypertension, n (%)	2 (2.8)
	A history of febrile illness, n (%)	1 (1.4)
	History of cleft palate in previous pregnancy, n (%)	5 (7.1)
	Drug/smoking during pregnancy (%)	10 (14.2)
	The total number of mothers with risk factors in pregnancy, n (%)	21 (30.0)

Table 2. Isolated cleft palate and cleft lip-palate made by comparing the variables of patients with risk factors logistic regression odds ratio values in results

Risk factors	p	Odds ratio
Consanguineous marriage	0.314	0.573
Age of mothers	0.648	1.023
History of cleft palate in previous pregnancy	0.906	1.131
Diabetes mellitus	0.374	0.448
Smoking during pregnancy	0.408	2.572
Chronic hypertension	0.294	0.248

Table 3. Classification of malformations accompanying cleft palate according to affected organ system

Organ system	n, (%)	Organ system	n, (%)
Cardiovascular system	**27 (38.5)**	**Central nervous system**	**18 (25.7)**
PDA	7 (10.0)	Hydrocephalus	3 (4.2)
ASD	5 (7.1)	Agenesis of corpus callosum	3 (4.2)
Pulmonary stenosis	3 (4.2)	Periventricular leukomalacia	4 (5.7)
VSD	2 (2.8)	Cerebellar vermis hypoplasia	2 (2.8)
Tetralogy of Fallout	2 (2.8)	Others	6 (8.5)
Others	8 (11.4)		
Extracranial skeletal system	**8 (11.4)**	**Craniofacial disorders**	**11 (15.7)**
Syndactyly	4 (5.7)	Micrognathia	7(10.0)
Polydactyl	3 (4.2)	Microcephaly	2 (2.8)
Hypoplasic thumb	1 (1.4)	Hypertelorism	2 (2.8)
The Eyes	**5 (7.1)**	**Urogenital system**	**5 (7.1)**
Microphtalmia	2 (2.8)	Hypospadias	3 (4.2)
Anophtalmia	1 (1.4)	Hydronephrosis	2 (2.8)
Eyelid deformity	2 (2.8)		
Endocrine system	**4 (5.7)**	**The Ears**	**4 (5.7)**
Hypothyroidism	4 (5.7)	Ear deformity	4 (5.7)
Additional syndrome	**6(8.5)**		
Pierre Robin sequence	4 (4.2)		
Down syndrome	1 (1.4)		
Trisomy 13	1 (1.4)		

PDA: Patent Ductus Arteriosus; ASD: Atrial Septal Defect; VSD:Ventricular Septal Defect

It was found that the first correction surgery of cleft palate patients was performed at a median age of 13 months, and the first correction surgery of the cleft lip patients was performed at a median age of 5 months. In patients with cleft palate, the most commonly observed accompanying defects after cleft lip were, in sequence, cardiac defects, central nervous system defects, craniofacial defects, musculoskeletal system defects, urogenital system defects, eye-ear defects and hypothyroidism (Table 3).

DISCUSSION

During our study, cleft palate was found in 70 cases in our hospital; while 24 cases had isolated cleft palate, 46 were found to have both cleft palate and lip. The most common anomaly accompanying cleft palate were determined to be cardiac defects.

Palate and lip deformities occur as a result of more than one factor (1,11,12). Advanced maternal age, oligohydramnios, vitamin deficiency (especially folic acid), parental consanguineous marriage, drug use in pregnancy (such as steroids and anticonvulsive drugs), drugs, alcohol, smoking, exposure to radiation during pregnancy, disease during pregnancy (gestational diabetes, rubella and toxoplasma infections) are considered as possible causes (1,11-13). In our study, the median maternal age was found to be 30 (18-45). During pregnancy, gestational diabetes mellitus was present in 6 of the mothers, chronic hypertension and oligohydramnios were seen in 2, and drug (salicylate, anticoagulant and insulin) use was seen in 10. 23 patients (32,8%) were found to have consanguineous parents. 70% of cleft palate-lip patients were non-syndromic cases with sporadic defects and 50% of isolated cleft palate patients were also non-syndromic (5). Conditions such as Pierre Robin sequence, trisomy, and various syndromes have been reported to be associated with cleft palate-lip (1,11,12). This risk was found to be increased for parents whose previous children had cleft palate-lip (1,11,12). The siblings of 5 (7,1%) of our cases were found to have a history of cleft palate, while 6 (8,5%) had a coexisting syndrome. Skeletal system defects, facial appearance anomalies, cardiac defects, central nervous system and cerebral anomalies have been reported as frequently seen accompanying anomalies in cleft palate patients (1,6). 27 (38,5%) of our patients exhibited cardiac anomalies, while central nervous system anomalies were found in 18 (25,7%), craniofacial anomalies in 11 (15,7%) and skeletal system defects in 8 (11,4%). For this reason, it will be appropriate to conduct genetic research, perform echocardiography in the fetal and postnatal periods to search for cardiac defects and to conduct a detailed physical examination and medical imaging after birth to

check for accompanying anomalies in patients with cleft palate defects.

With the increase in the use of three and four dimensional ultrasonography during pregnancy, there has been a reported increase in the number of patients who are diagnosed early for cleft palate anomalies in the prenatal period (14). A detailed ultrasonography and early diagnosis during the prenatal period is important for providing information to families and allowing the delivery of the fetus in a suitable environment. While 21 (30%) of the patients in our study were found to have a prenatal diagnosis, the remaining patients were not evaluated with detailed ultrasonography during the prenatal period.

In patients born with cleft palate, the primary problems of significance are feeding difficulties and frequent lower respiratory tract infections resulting from aspiration (15). Surgical correction operations in cleft palate-lip patients are performed in later periods so that jaw development is not hindered (16). Patients with substantial cleft palate cannot produce sufficient sucking power and face difficulty in feeding through breastfeeding or bottle (17). For this reason, a temporary palate apparatus (feeding plaque), which helps to improve swallowing functions and tongue movements, can be used until correction operations are performed. In cleft palate-lip cases, the anatomic defects are corrected using a two-stage closure method consisting of lip correction between 3-5 months of age and palate correction between 12-18 months (16-18). In our study, the median age at the time of the first correction operation was five months for cleft lip and thirteen months for cleft palate These patients should be followed-up with a multidisciplinary approach by neonatologists, plastic and reconstructive surgeons, otolaryngologists, audiologists, speech therapists and orthodontists. These patients and their families should receive consultancy regarding genetic and psychosocial factors when necessary. Through such a team approach, it will be possible to provide care to patients in which their familial, social, emotional, physiological and educational needs are met.

CONCLUSION

In conclusion, the treatment and follow-up of patients with cleft palate defect requires complete teamwork. These patients should receive a detailed examination and evaluation. Since a great number of syndromes or malformations can accompany palate defects, these cases should be examined for potential genetic disease and other system anomalies. The patient's family should be informed about cleft palate being a common anomaly that can be treated successfully and emphasis should be given that follow-up and treatment by healthcare centres with expertise on the issue will lead to a greater chance of success.

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