ORIGINAL ARTICLE ÖZGÜN ARAŞTIRMA

Recommendations and Clinical Experiences in Neonatal Intensive Care Monitoring of Cleft Palate Patients

Yarık Damak Hastalarının Yenidoğan Yoğun Bakım Takibinde Öneriler ve Klinik Tecrübelerimiz

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Abstract

Introduction: Cleft palate is a congenital craniofacial malformation, whose pathophysiology has not been fully elucidated. Most newborns with cleft palate require hospitalization in the neonatal intensive care unit. In the clinical management of these patients, specialized clinical approaches tailored to this specific patient group and interdisciplinary collaboration are crucial. Our study aimed to assess the hospitalization procedures of newborns with cleft palate in the neonatal intensive care unit and share valuable clinical insights from this experience. Through this research, we aim to contribute to the enhanced understanding and management of cleft palate cases in newborns, emphasizing the importance of specialized and interdisciplinary care.

Materials and Methods: Between the years 2013-2021, newborns diagnosed with cleft palate and admitted to the neonatal intensive care unit were retrospectively analyzed in terms of gestational age, birth weight, duration of hospital stay, and additional anomalies.

Results: One hundred thirty-five infants were diagnosed with cleft palate, among whom 46 (34%) had additional anomalies. Statistically significant differences were observed in parameters such as gestational age, birth weight, Small for Gestational Age (SGA), and duration of hospitalization when comparing these groups. Notably, in SGA newborns, the incidence of additional anomalies was higher.Following multinomial logistic regression analysis, it was found that hospitalization for more than one week was independently associated with the presence of an additional congenital anomaly.

Conclusion: Cleft palate is a common congenital malformation often accompanied by additional deformities. Since issues related to nutrition and respiration can be present in most of these patients, close postnatal monitoring is crucial. Hence, managing patients admitted to the neonatal care unit involves early multidisciplinary assessment followed by a long-term clinical follow-up process, ensuring the healthy growth and development of these patients.

*Tetyana Waldheim and Gamze Gürsoy are corresponding authors and equally contributed to this study.

Keywords
NICU, cleft palate, craniofacial abnormalities, neonatal feeding issues, multidisciplinary care, congenital malformation

Anahtar kelimeler
Yeni doğan yoğun bakım ünitesi, yarık damak, kraniyofasiyal anomali, yenidoğanın beslenmesi, multidisipliner yaklaşım, konjenital malformasyon

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Öz

Giriş: Yarık damak, patofizyolojisi tam olarak aydınlatılamamış bir konjenital kraniyofasiyal malformasyondur. Beslenme problemleri ve solunum sıkıntısı gibi nedenlerden ötürü, yarık damaklı bebeklerin yenidoğan yoğun bakım ünitesine yatışı gerekebilmektedir. Yarık damaklı yenidoğanların klinik yönetimi ve multidisipliner yaklaşım, bu hastaların tedavisinde kritik bir rol oynamaktadır. Çalışmamızda, yarık damaklı yenidoğanların yenidoğan yoğun bakım ünitesine kabul süreçlerini değerlendirmeyi ve klinik yaklaşımımızı paylaşmayı amaçladık.

Gereç ve Yöntem: 2013-2021 yılları arasında yarık damak tanısı konulan ve yenidoğan yoğun bakım ünitesine kabul edilen bebekler gebelik haftası, doğum ağırlıkları, yatış günleri ve ek anomalileri açısından retrospektif olarak analiz edilmiştir.

Bulgular: Toplamda 135 bebeğe yarık damak tanısı konuldu, ve bu bebeklerin 46'sında (%34) ek anomaliler tespit edildi. Hastaların gebelik haftası, doğum ağırlığı ve hastanede kalış süreleri açısından yapılan karşılaştırmalarda, yaşa göre doğum ağırlığı düşük bebeklerde ek anomali sıklığının yüksek olduğu, ve ek bir konjenital anomalisi bebeklerin hastanede kalış süresinin arttığı gözlendi

Sonuç: Yarık damak, sık görülen bir doğumsal malformasyon olup genellikle ek deformitelerle birlikte ortaya çıkar. Bu hastaların çoğunda, beslenme ve solunum ile ilgili sorunlar görülebileceğinden, hastaların doğum sonrası yakın takibi önemlidir. Yenidoğan bakım ünitesine bu nedenle yatışı yapılan hastaların izlemi, erken dönemde multidisipliner bir değerlendirme yapılması ve ardından uzun vadeli klinik takip sürecinin başlatılması, hastaların sağlıklı bir şekilde büyümeleri ve gelişmeleri için hayati önem taşır.

Introduction

Orofacial clefts (OFC) represent congenital malformations that affect the lips and oral cavity (1). According to data from the USA, the prevalence of cleft lip with or without cleft palate is 1 in 1000, and isolated cleft palate incidence is 1 in 2500, making it the second most common congenital anomaly, also with variations based on race and gender (2-4). A newborn with a cleft palate can lead to psychosocial and economic challenges for both the patient and the family (5). Cleft lip (CL), cleft lip and palate (CLP), and cleft palate (CP) are the three primary types of OFCs. OFCs can be a part of a syndrome, and accompanying symptoms can indicate an underlying genetic disorder (5). The etiology of OFCs is intricate, involving numerous potential genetic and environmental factors, and the mechanism still needs to be understood fully (4,6,7). Orofacial development is a multifaceted process influenced by many factors, such as cell growth, proliferation, migration, apoptosis, differentiation, and cell and tissue fusion. These coordinated events necessitate the involvement of multiple signaling pathways and transcription factors. It has been identified in previous studies that several impactful signaling pathways and molecules, including the Msh homeobox and T-Box (TBX) gene families, the sonic hedgehog signaling pathway, transforming growth factors, and bone morphogenetic proteins (7).

Based on the systematic review by Maarse et al. (8), the accuracy of ultrasonography in diagnosing orofacial clefts in low-risk patients ranges from 0% to 70%. Fitzsimons et al. (9) diagnosed prenatal clefts in 39% of 406 children, with a higher ratio for cleft lip and palate (78%), 56% for cleft lip, and 1% for cleft palate. Prenatal diagnosis assists in planning delivery and future care; however, it can also negatively impact parents, necessitating proper support (10). Notably, a significant percentage (72%) in Fitzsimons's study (9) were not diagnosed antenatally. Among diagnosed cases, the majority (82.5%) presented with unilateral/bilateral cleft lip and palate, while a portion (6.9%) of cleft palates were identified over a month after birth. Isolated cleft palates can be challenging to detect during newborn assessments. Surgical requirements and the number of primary operations vary based on the cleft type, making multidisciplinary follow-up in specialized centers crucial. Cleft-associated challenges such as feeding difficulties, behavioral issues, speech and language impairments, facial growth retardation, dental anomalies, and hearing loss are common. To effectively address these complications, a multidisciplinary team approach is essential for pediatricians to address medical concerns, cleft surgeons handle surgical interventions, clinical nurse specialists provide perinatal care and nutritional support, pediatric dentists and orthodontists oversee dental health, speech and language therapists manage speech and language disorders, audiovestibular physicians and audiologists monitor hearing, and geneticists advise on genetic aspects (10).

Infants born with a cleft lip or palate should undergo an assessment for potential airway obstruction or breathing difficulties. If no signs of airway compromise are present, an evaluation for feeding and growth can be conducted. Newborns with cleft lip/palate should be screened for possible congenital abnormalities. Our

neonatal intensive care unit's clinical management for cleft palate can be summarized in Figure 1.

The airway and breathing assessment begins with a thorough examination of the child, involving a visual inspection of the infant's head, neck, and chest. Indicators of airway concerns encompass symptoms such as stridor, heightened breathing effort, utilization of accessory muscles, retractions, and changes in skin color such as cyanosis or a dusky appearance. Infants displaying these symptoms should be admitted to the hospital.

The feeding assessment involves critical components such as observing the infant's behaviors, length, frequency of feeding, and overall volume consumed. Additionally, evaluations for signs of noisy breathing, effort during feeding, episodes of emesis or nasal regurgitation, coughing, choking, or gulping are paramount. Breastfeeding is encouraged if the infant can effectively latch and sustain proper feeding. Special cleft palate nipple bottles or feeders can facilitate feeding in cases where additional support is required.

Materials and Methods

Between 2013 and 2021, infants diagnosed with cleft palate and admitted to the neonatal intensive care unit (NICU) were retrospectively analyzed regarding gestational weeks, birth weights, duration of hospitalization, and associated additional anomalies.

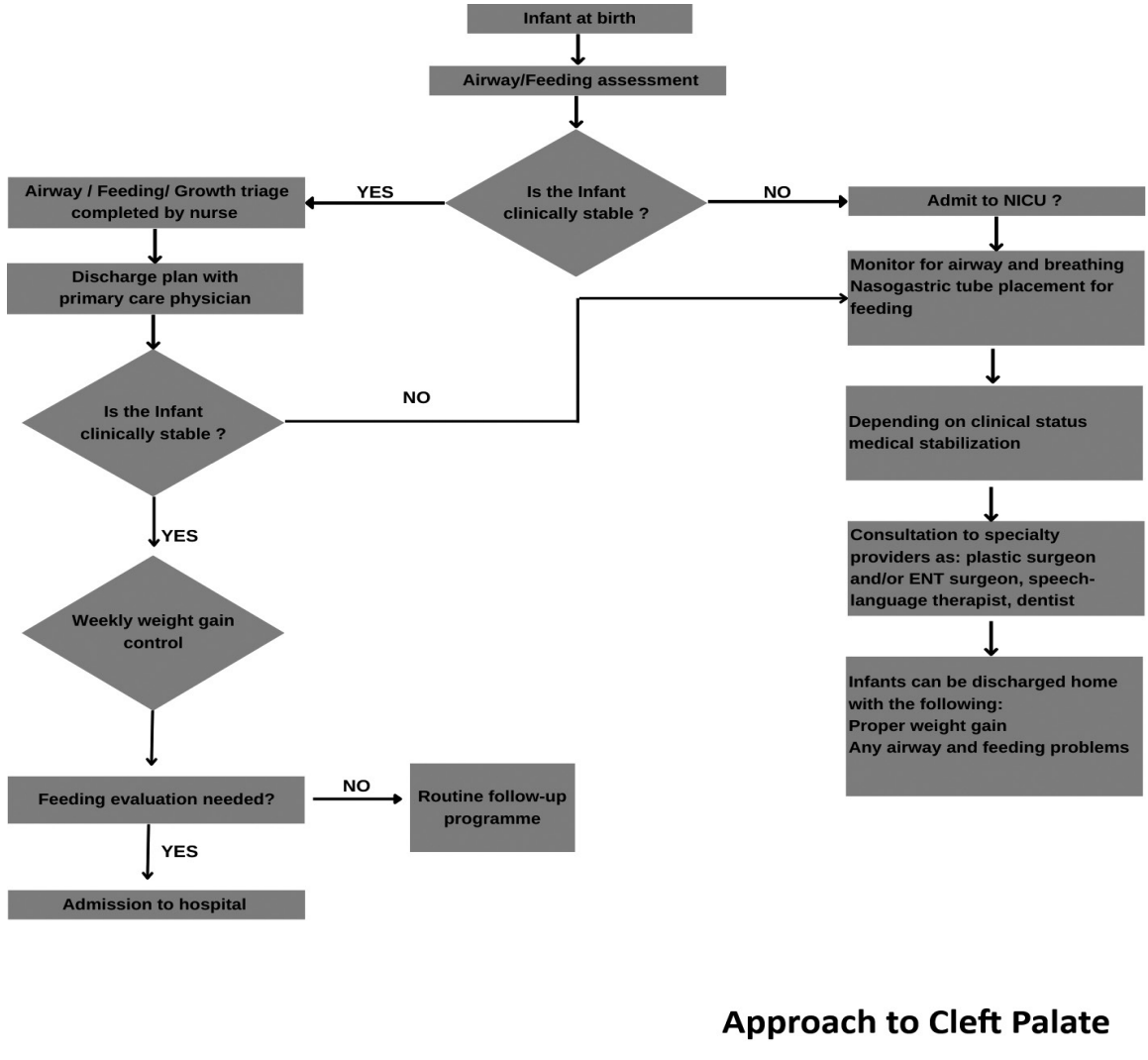

Figure 1. Algorhythm for evaluation and follow-up of cleft palate.

Ethics committee approval was obtained from the Ankara City Hospital Clinical Research Ethics Committee with number E2-21-1026.

Results

Between 2013 and 2021, there were 22,348 NICU admissions and 135 cases diagnosed with cleft palate. Among the diagnosed cleft palate patients, 51.9% were male. The mean birth weight was 2969.70 grams, ranging from a minimum of 1000 grams to a maximum of 4900 grams. Nineteen patients (14.1%) were classified as small for gestational age (SGA), irrespective of gender. The mean gestation week was 37±2.1, with a minimum of 28 weeks. Out of the total, forty-six patients (34.8%) exhibited extra congenital abnormalities (Table 1). Notably, SGA patients showed a higher prevalence of additional congenital

Table 1. Congenital abnormalities with cleft palate anomalies

		Number of patients
Syndromes	Pierre Robin Sequence	6
Total Patients: 11	Fraser Syndrome	1
	Fetal Alcohol Syndrome	1
	Velocardiofacial Syndrome	1
	Klippel-Feil Syndrome	1
	Septo-optic Dysplasia (Morsier Syndrome)	1
Skeletal System	Polydactyly	2
Total Patients: 8	Pes equinovarus 2	2
	Skeletal Dysplasia 2	2
	Achondroplasia 1	1
	Diastematomyelia 1	1
Cardiac Anomalies	Tetralogy of Fallot	2
Total Patients: 7	Coarctation of Aorta 2	2
	Hypoplasia of Arcus Aorta 1	1
	Atrial Septal Defect (ASD) 1	1
	Atrioventicular Septal Defect (AVSD) 1	1
Chromosomal Anomalies	Trisomy 21 2	2
Total Patients: 7	Trisomy 13 2	2
	Trisomy 18 1	1
	Monosomy 5 1	1
	Chromosome 8q Duplication 1	1
Central Nervous System	Encephalocele 2	2
Total Patients: 4	Holoprosencephaly 1	1
	Agenesis of the corpus callosum (ACC) 1	1
Gastrointestinal System	Anal Atresia 2	2
Total Patients: 4	Hernia of Diaphragm 1	1
	Omphalocele 1	1
Renal System	Hydronephrosis 2	2
Total Patients: 4	Polycystic Kidney 1	1
	Duplex Collecting System 1	1
Genital System	Cloacal Malformation 1	1
Total Patients: 1		

anomalies, with 15 out of 19 SGA patients (78.95%) having such anomalies. Longer hospitalization durations were linked to the presence of additional congenital anomalies in comparison to isolated cleft palate cases. Patients with multiple abnormalities experienced hospital stays lasting over one week. Odds Ratio (OR) was 15.6 (4.2-57.1) with a significance level of p: 0.001.

The most prevalent coexisting anomalies were cardiac and skeletal abnormalities. Among the 46 patients, seven had cardiac anomalies, including Fallot tetralogy, coarctation of the aorta, atrioventricular septal defect, and hypoplasia of the arcus aorta. Eight patients exhibited skeletal system abnormalities such as polydactyly, pes equinovarus, achondroplasia, and diastematomyelia. Additionally, seven patients presented with chromosomal anomalies; trisomy13 was the most frequent, affecting three patients and others were identified with trisomy 21, trisomy 18 and monosomy 5. Six patients were diagnosed with Pierre Robin Sequence, and four had renal anomalies, including polycystic kidney, duplicated collecting system, and hydronephrosis.

Discussion

Prenatal diagnosis is crucial in connecting parents with the appropriate medical centers before delivery. Our research observed that none of the patients were diagnosed with prenatal ultrasonography. The accuracy of the diagnosis can be influenced by factors such as the sonographer's expertise, gestational age, and the quality of ultrasonography (3). Even though technical constraints might limit prenatal diagnosis during the initial newborn examination, it is imperative to make every possible effort to rule out the presence of a cleft palate.

Caring for infants with orofacial anomalies can be financially burdensome and time-consuming. While admission to the Neonatal Intensive Care Unit (NICU) might not always be necessary for a newborn with an isolated cleft, it is worth noting that it can increase the risk of airway complications and nutritional problems (11,12). Our clinical experience has shown that many newborns are referred to the NICU due to issues related to nutrition and respiration. Although we make efforts to minimize NICU hospitalization, these infants need to be fully enterally fed, and parents need to be educated in this regard. In some cases, a cleft palate is part of a syndrome that necessitates hospitalization. We believe early and accurate prenatal diagnosis is important for appropriate care for infants with orofacial anomalies. Despite the challenges, efforts should be made to avoid unnecessary NICU admissions.

When hospitalization is unavoidable, we prioritize efficiently using healthcare resources by focusing on early discharge whenever feasible. The American Cleft Palate-Craniofacial Association highlights the significance of early multidisciplinary therapy, which is pivotal for achieving optimal outcomes (13).

In our NICU, neonatologists and pediatricians offer primary care and work collaboratively with our craniofacial team, including feeding nurses, plastic and ENT surgeons, and dentists. This collaboration begins as soon as possible to facilitate comprehensive planning for the newborn's treatment. Following a thorough evaluation, we proceed with genetic tests to gain further insights if the baby presents any additional anomalies.

Infants with cleft palate often experience problems with nutrition, such as poor weight gain and dehydration, which is one reason for NICU admission (14). While babies with cleft lip can usually breastfeed, cleft palate typically makes breastfeeding unsuccessful (3). In our clinic, if newborns can breastfeed, we encourage mothers to do so. However, several cleft-specific bottles may be necessary. These bottles can be divided into squeeze bottles (Haberman and Mead Johnson) and rigid bottles (Pigeon and Dr. Brown's). Previous studies indicate no differences in growth outcomes based on bottle type. We assist parents through our nutrition nurse and help them choose the most effective bottle for nutrition (15). A newborn with a cleft lip and palate can be fully bottle-fed unless feeding the baby using a nasogastric tube may be necessary.

Lip taping and nasoalveolar molding might be used during the neonatal period to reduce the severity of cleft abnormalities. However, the effectiveness of these techniques remains questionable, and we lack clinical experience with them in our NICU (16-18).

In our study, we aimed to emphasize the significance of a multidisciplinary approach in addressing the challenges faced by newborns with cleft palate and resolving these issues. Unfortunately, due to the lack of complete data on total respiratory distress, ventilation requirements, and the days of transition to full enteral

feeding for cleft palate infants in our intensive care unit, this information could not be included in the study, thus constituting a limitation of our research.

Conclusion

A cleft palate, often accompanied by additional anomalies, frequently necessitates NICU admission for feeding and breathing issues. Longer hospitalization is linked to additional anomalies. Early multidisciplinary assessment and long-term follow-up are critical for optimal clinical outcomes.

Ethics

Ethics Committee Approval: Ethics committee approval was obtained from the Ankara City Hospital Clinical Research Ethics Committee with number E2-21-1026.

Conflict of Interest: No conflict of interest was declared by the authors.

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