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THE FREQUENCY AND MANAGEMENTS OF PIERRE ROBIN SEQUENCES AMONG PATIENTS ATTENDING WITH ISOLATED CLEFT PALATE.

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ABSTRACT

Among the cleft lip and palate population, the most common diagnosis is cleft lip and palate at 46%, followed by isolated cleft palate at 33%. Cleft lip and palate may be isolated or associated with another anomalies. Robin sequence (RS) is one of these abnormalities, characterized by: isolated cleft palate, micrognathia and glossoptosis. RS may be isolated or part of several syndromes. In this study only isolated RS are selected for determination the frequency and the managements. From 200 patients with clefts malformation,60 patients presents with isolated cleft palate . Out of 60 patients with isolated cleft palate 2 cases were diagnosis as isolated Pierre robin sequences (0.03), male: female ratio(1:1). One of them treated by anterior tongue repositioning, while the other by conservative management.

KEYWORDS: Freguency, pierre robin, cleft palate.

AIM OF STUDY

Evaluation the incidence of isolated pierre robin sequences among patients presented with isolated cleft palate and the effective managements.

INTRODUCTION

"Among cleft lip and palate patients, the most common diagnosis is cleft lip and palate at 46%, followed by isolated cleft palate at 33%, then isolated cleft lip at 21%. Males are predominant in the cleft lip and palate population, whereas isolated cleft palate occurs more commonly in In females".[1] cleft lip and palate may be isolated or associated with another anomalies. Robin sequence (RS) is defined as a triad of symptoms: isolated cleft palate, micrognathia, and glossoptosis.[2] RS may be part of several syndromes, e.g., Treacher-Collins syndrome[3,4] One of the most common genetic syndrome associated with Robin sequence is *Stickler syndrome.*[5] Robin sequence, was reported after Pierre Robin whose first report appeared in 1923.[6] It is "Generally agreed that the pathophysiologic events in this syndrome are as follows: the receding chin fails to support the tongue. The retroposed tongue impinges against the posterior wall of the pharynx, obstructing inspiration and impeding feeding. Birth prevalence estimates have varied from 1/2000 to 1/30,000.[7] Bush andWilliams Suggested 1/8500"[8].

MATERIAL AND METHODS

-This a prospective study is include 200 patients with clefts malformation. Only sixty cases with isolated cleft palate, attending to the department of maxillofacial surgery between may 2013 to December 2016.

-Only nonsyndromic Pierre robin selected for this study (isolated Pierre robin).

-All the patients was examined by pediatrician to exclude the syndromatic one.

RESULT

From 200 patients with clefts malformation,60 patients presents with isolated cleft palate (30%) table 1. Consisting 41 female and 19 male (1-0.4) table 2. Out of 60 patients with isolated cleft palate 2 cases were diagnosis as isolated Pierre robin sequences (0.03) table 3,one of them was female and the other was male, male:female(1:1).One of them only with family history of cleft malformation 50% table 4. Both them suffering from upper airway obstruction, specialy during supine position and feeding through nasogastric tube. One of them treated by anterior tongue repositioning, while the other by conservative management.

Table: 1 Distribution of cleft lip.

Type of cleft	No.	%
Cleft lip with or without palate	140	70%
Cleft palate only	60	30%
Total	200	100%

Table: 2 Sex distribution of cleft lip and palate.

Cleft type	Sex		Ratio(Male:Female)
	Male	Female	
Cleft lip with or with out palate	96	44	2.1 -1
Isolated Cleft palate	19	41	0.4-1
Total	115	85	1.3-1

Table: 3 The sex distribution of pierre robin in isolated cleft palate.

Sex	No. of isolated cleft palate	No.of pierre robin cases	Incidence
female	41	1	0.02
Male	19	1	0.05
Total	60	2	0.03

Table: 4 The incidence of family history in pierre robin sequences.

No. of pierre cases	Positive histort	Negative history	Incidence
2	1	1	**50%**

DISCUSION

Out of 200 patients with clefts malformation,60 patients presents with isolated cleft palate (30%) and predominantly in female and this agree with (RICHARD A et al - 2007)[1] that the isolated cleft palate represent about (33%) of total cleft deformities and higher incidence in female. In this study only 2 cases out of 60 cases with isolated cleft palate are presents with pierre robin sequences, in review of literatures there is no study reported the incidence of Pierre robin in isolated cleft palate, but generally is rare malformation, (Ted L Tewfik 2015) reported that the prevalence of pierre robin approximately 1 per 8500 live birth, and also like in this study he was reported that male to female ratio (1-1). In this study one case with family history of cleft malformation while the other without, that the exact causes of pierre robin sequences are unknown, it May be genetic or due to mechanical obstruction. The growing will be faster during the first few years of life, that explain the cause may be due to mechanical obstruction during intrauterine life. Regarding the airway, one case treated by anterior tongue repositioning, while the other by conservative management .Smith and Senders, found that 63% of infant responded to conservative management without surgical intervention by keeping the infant in the side lying or prone position.[9]

SUGGESTION

1. Precise preoperative examination by the pediatrician to exclude other abnormalities that may be effect the management.
2. The conservative managements of the airway should be considered.

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