DENTOFACIAL CHARACTERISTICS OF MALAY PATIENTS WITH REPAIRED CLEFT LIP AND PALATE IN KELANTAN

by

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DEDICATION

I would like to dedicate my research work to my late parents, my beloved family and most of all to those with cleft lip and palate from the bottom of my heart.

Date: 26th April, 2006.

Md. Badrul Alam

provide me a grant (Grant No.304/PPSG/6131222), Statistician and other helping hands that have really contributed much in assisting me to compile things together and have supplied all necessary datum in connection with my research work.

While staying in Malaysia in the pursuit of knowledge of wisdom. my mother, brothers and sisters, nephews and nieces and above all my friends and admirers have always been supportive and cooperative alongside my other family members.

Finally I should not forget to thank my wife Dr. Zinat Ara Ferdousi and my lovely children who have always inspired me to obtain this degree. They have missed my company and as well as I have missed their affectionate care in the course of staying apart for a period of one and a half years.

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LIST OF ABBREVIATIONS

Abbreviations	Name
A	Point A
AA	African American
ACPA	American Cleft Palate Craniofacial association
Adj.	Adjusted
AL	Arch length
al	Alare
ANB	Angle between point A-nasion and nasion- point B
A.N.S	Anterior nasal spine
B	Point B
CCCDC	Combined cleft and craniofacial deformities clinics
CLP	Cleft lip and Palate
CL/P	Cleft lip with or without cleft palate
CL	Cleft lip
CP	Cleft palate
ch	Chelion
cph	Crista philter
en	Endo canthion
ex	Exocanthion
fig.	Figure
gn	Gnathion
Go	Gonion
Icc	Intra-class correlation co-efficient
ICAW	Inter canine arch width
ISPPDMW	Inter permanent premolar or deciduous molar width
IFPMW	Inter permanent molar width
LFH	Lower facial height
LI/MP	Angle between the long axis of the lower central incisors to the mandibular plane.
ls	Labiale superiois

li	Labiale inferioris
me	Menton
MMA (MxP/MP)	Maxillo-mandibular angle
MP	Mandibular plane
MxP	Maxillary plane
n	Nasion
NSCLP	Non-syndromic cleft lip and palate
NAC	North American Caucasian
no.	Number
prn	Pronasale
Ref.	Reference
r/n	Registration number
S	Sella
SCLP	Syndromic cleft lip and palate
sn	Subnasale
sto	Stomion
Sl	Sublabialae
SNA	Angle between sella -nasion and nasion- pointa.
SNB	Angle between sella -nasion and nasion- pointb.
SE	Standard error
SD	Standard deviation
UI/MxP	Angle between the long axis of the upper central incisors to the maxillary plane.
zy	Zygoin

SIFAT- SIFAT PERGIGIAN DAN MUKA PESAKIT-PESAKIT BERBANGSA MELAYU YANG MENGALAMI REKAHAN BIBIR DAN LELANGIT YANG TELAH DIRAWAT DI KELANTAN

Abstrak

Rekahan bibir dan lelangit (CLP) adalah suatu kecacatan kongenital muka yang sering ditemui. Pesakit CLP mengalami masalah makan, bernafas, infeksi telinga tengah, masalah psikososial dan lain-lain. Rawatannya memerlukan penglibatan pelbagai disiplin yang bermula dari hari pertama dilahirkan dan berterusan hingga peringkat umur 20 ke 21 tahun. Pemanjangan rawatan dari ketika lahir ke alam dewasa amatlah mahal dan membebankan pesakit, keluarga dan masyarakat. Perkembangan muka pesakit CLP selalunya tidak mengikut tumbesaran yang normal walaupun selepas pembedahan pembetulan. Tidak mengira mekanismanya , kecacatan tumbesaran menjadi lebih jelas apabila pesakit mencapai tahap umur matang. Oleh itu, pemahaman tentang anomali pergigian dan muka adalah penting kerana ia menentukan keseimbangan dan keharmonian muka. Ini amat penting untuk mencapai matlamat ideal untuk merawat pesakit rekahan ini.

Satu kajian perbandingan hirisan lintang (cross-sectional) dijalankan dengan pengukuran terus muka, dengan mengukur kifalometri lateral dan acuan gigi tujuh puluh lima orang pesakit rekahan bibir dan muka dan tujuh puluh lima orang pesakit Melayu yang normal. Julat umur adalah antara 8 ke 40 tahun. Subjek kajian dibahagikan kepada tiga kumpulan umur : kanak-

kanak (8-12 tahun), remaja (13-18 tahun), dewasa (19-40 tahun) bagi kedua-dua jantina.

Keputusan kajian ini menunjukkan bahawa ukuran dimensi lingkungan gigi dan kaitan insisor bahagian atas dan bawah ke planar yang berkenaan pesakit CLP mempunyai perbezaan signifikan dengan semua kumpulan yang normal berlainan umur. Panjang lingkungan anterior-posterior. dimensi transvers dibahagian interkanin, inter kedua premolar kekal atau bahagian molar kedua desiduos, bahagian inter pertama molar kekal dan ukuran di bahagian sudut untuk sentral insisor atas dan bawah di planar maksila dan mandibel yang berkenaan didapati lebih tinggi bagi kumpulan yang normal pada semua peringkat umur ($p<0.05$). Lebar filtrum dan sudut pengukuran skeletal anterior-posterior didapati lebih tinggi dalam subjek rekahan sementara ketinggian bibir atas dikalangan subjek yang normal pada semua peringkat umur adalah lebih tinggi secara signifikannya. Kajian juga menunjukkan terdapat bahagian muka tertentu lebar hidung, nasal, filtrum dan ketinggian bahagian atas anterior muka) dan ukuran skeletal (anterior-posterior: Sella-Nasion-Point B, SNB dan Point A-Nasion- Point B, ANB; menegak, sudut maksila-mandibel. MMA dan ketinggian anterior muka bawah, (ALFH) adalah unik bagi subjek kajian kanak-kanak, remaja dan dewasa.

Sifat-sifat pergigian dan muka pesakit Melayu yang telah dirawat rekahan bibir dan lelangitnya di Kelantan seperti yang dijelaskan oleh kajian yang dijalankan adalah berguna untuk mengembangkan protokol rawatan yang lebih baik. Dengan itu, ia kan menyumbang kepada kualiti kehidupan yang lebih baik kepada pesakit CLP dan keluarganya.

Dentofacial Characteristics of Malay Patients with Repaired Cleft Lip and Palate in Kelantan

ABSTRACT

Cleft lip and palate (CLP) is the most common congenital facial defect. Patients with CLP suffer from feeding, breathing, middle ear infections as well as psychosocial and other problems. The treatment requires interventions from multiple disciplines which start from the first day of life and continue up to the age of 20 to 21 years. Prolong treatment from birth to adulthood is expensive and is a burden for the patient, family and for the society. The facial development of CLP patients often does not follow usual development of growth even after corrective surgeries. Growth impairment becomes progressively apparent as patients reach maturity regardless of the mechanisms. Therefore, an understanding about the anomalies of dentofacial region is important as it determine the facial harmony and balance. This is essential to achieve the ideal goal of treatment of cleft patients.

A cross-sectional comparative study was done by direct facial measurements, by measuring lateral cephalometry and dental cast of 75 treated cleft and 75 noncleft Malay subjects. The age range was from 8 to 40 years. The study subjects were divided into three age groups: children (8 to 12yrs), adolescent (13 to 18 yrs) and adult (19 to 40 yrs) of both genders.

Results of this study revealed that the measurements of dental arch dimensions and the upper and lower incisors' relation to the respective plane of CLP patients were significantly different from that of the noncleft subjects of all age groups. The anterior-posterior arch length, transverse dimensions in inter canine region, inter second permanent premolar or deciduous second molar region, inter first permanent molar region and in angular measurements for upper and lower central incisors to the respective maxillary and mandibular plane were found significantly higher in noncleft groups in all ages ($p<0.05$). The philtrum width and skeletal anterior-posterior angular measurements were found significantly higher for the cleft subjects while the upper lip height was significantly higher for noncleft subjects among all age groups. The study further revealed that certain facial (nasal width, nasal, philtrum and upper anterior facial heights) and skeletal (anterior-posterior: Sella-Nasion-Point B, SNB and Point A-Nasion-Point B, ANB; vertical: Maxillo-mandibular angle, MMA and anterior lower facial height, ALFH) measurements were unique to children, adolescent and adult study subjects.

These dentofacial characteristics of Malay patients with repaired cleft lip and palate in Kelantan, as revealed by the present study, would be useful to develop a more ideal treatment protocol. It will, therefore, contribute a better quality of life to the CLP patients, their families and the society.

CHAPTER ONE

INTRODUCTION

CHAPTER ONE

INTRODUCTION

1.1 Background:

Cleft lip, with or without clefting of the primary and secondary palate, is the commonest congenital deformity of the oro-facial region. It is a deformity in which there is a primary mesoectodermal deficiency (Harris et al., 1991).

Clinically, when cleft lip and palate (CLP) appears with other (usually two or more) malformations in recognizable pattern, it is classified as syndromic CLP (SCLP). If it appears as an isolated defect or if syndromes cannot be identified, the term non-syndromic CLP (NSCLP) is used (Wong et al., 2004). More than 400 syndromes have been already associated with CLP (Papadopulos, et al., 2003).

There is no specific date or time of the existence of cleft lips but it is assumed or rather considered that cleft individuals have existed since mankind. In a study on neonatal cleft lip and cleft palate repair by Sandberg et al., 2002, it was found that surgical cleft lip repairs have been reported as early as 390 AD in China.

History also reveals that these unfortunate victims of nature were scorned, ridiculed or even ostracized from society regardless of their social, intellectual, or economic status. Some were worshipped as deities whereas others were feared and even condemned to death (Philip, C, 1990).They were considered

by some early civilizations as partly divine. On the other hand others disgusted them and thought they were demons (Hermann. H. V, 2000).

The Romans preferred to threw away their malformed children from the Tarpeian Rock (Nordhoff, 1997). Also, numerous sayings suggest what a pregnant woman should do or more often what they should not do, in order to avoid giving birth to a cleft child (Hermann, N. V. 2000).

It was not until the sixteenth century, immensely through the efforts of the eminent barber-surgeon Ambroise Par'e and his student Franco, a more descriptive and human approach was followed to surgically close 'hare lips'. Attempts by others to close palatal clefts had a high risk of failure and infant mortality (Philip, C, 1990).

Over a period of time, many surveys have been conducted to estimate the incidence of CLP in a total population. There is some variation in incidence between racial groups (Malcolm et al., 2000). The incidence varies according to the type of cleft, racial group and sex. Cleft lip and palate ranges from 3.6 /1000 live births for Indians to 0.5 per 1000 for negroes.(Mars,M 1998).In Malaysia the incidence of cleft lip and palate is 1 / 941 live births(MOH, 1998) and in Kelantan it is 1 / 700 live births (Halim and Singh, 2000).

The etiological factors include heredity, intermarriage, maternal environment, demographic factors and other factors like intra-uterine posture, drugs, vitamins, alcohol consumption, smoking, infections, diet etc. (Haq, M. E, 1995).

There are multiple morpho-functional problems associated with CLP patients and as well as a tremendous psychological stress to their parents.

The analysis of facial symmetry is complex (Riden, K, 1998). Craniofacial under growth of subjects with cleft lip and/or palate is caused by the lack of intrinsic growth potential of the nasomaxillary complex (Suzuki et al., 1993). The degree of collapse in the maxillary dental arch varies according to cleft type (Suzuki et al., 1992).

The study by Singh et al (2004) suggests that rehabilitation of the patient with CLP must be based on a through knowledge of their growing potential, because even after surgery the patients' tissue development often does not follow usual growth trajectories. Anomalous growth of the mid-facial area after corrective surgery may be associated with several complications such as unclear pattern of speech, velopharyngeal deficiencies, skeletal deformities, and dental malformation, which contribute to facial disfigurement and low self-esteem. To overcome problems that often occur after post surgical procedures, the abnormal dentofacial features is one of the most important factor that needs to be addressed to prevent less successful outcome. Thus, post surgical results are not predictable because the response of growing tissues to surgical repair is variable, leading to phenomena such as midfacial hypoplasia (Dahl, E, 1970).

1.2 Problem statements:

The facial features are one of the most important structures of an individual as a social being. Besides functional problem any abnormality in the facial area brings great psychological effect not only to the CLP patient but also to the effected family and the society.

The individuality of the human face is an important phenomenon in one's life. No part of our anatomy provides more information like face. Infact, no part of our anatomy is more vital to our success and existence. Minor alterations in the size, shape, position, and proportions of our face results in major perceptible differences, and subtle differences between two people are instantly recognizable (Jacobson, R. L, 1995). Whatever is the type of cleft, it causes severe distortion of the face both before and after surgery. The treatment of CLP patients starts from the very first day of life and continues up to the age 20 -21 years and it is a heavy burden to the patients, family and also for the society. Even than, it is found to have many problems remain with the CL/P patients, especially with the facial form and the speech, which have tremendous psychosocial impact to the effected patients, especially in the growing age. Figure 1.1 shows severe asymmetry in the face of a young adult Malay woman even after treatment which develops low self esteem to the victimized.

Figure1.1: A) Frontal and B) profile view of an adult repaired CLP Patient

A study by Capelozza et al (1993) found that the CLP patient's facial morphology has been studied extensively with varied results. He added that the reasons for differences are due to different treatment protocol for the same type of cleft patients, functional changes resulting from the cleft, and inherited growth pattern (Chierici et al., 1973; Bishara et al., 1976; Ross,1987;Capelozza et al.,1993).

The facial morphology of CLP patients deviates from the norm (Dahl,E, 1970; Hermann et al., 1999).Patients having only incomplete cleft lip show normal development (Dahl, E,1970), while those, having complete cleft lip shows growth anomaly in the facial morphology (Dahl, E, 1970).

Several investigations have suggested that the deviation of facial morphology of children, adolescents, and adults are due to surgical intervention (Hermann et al., 1999, Dahl, E., 1970, 1971)

Hermann et al (1999), confirmed in his study that UCCLP is a localized deviation because the other part of the craniofacial region shows no abnormality, except where the growth is directly influenced by the surgical intervention.

The effect of palatal surgery on maxillofacial growth is observed most clearly in those aged 10-20 years. Although patient may have a normal appearance early in life, he or she may begin to develop a definite flattening of the mid-face and dento-occlussal malrelation, particularly during adolescent (Will, P and Rapley, 2003).

The CLP patient suffers from a lot of problems throughout their life. The anomalies associated with these children vary from mild to severe and can cause complex distortion in the facial features (figure1.2).

Figure 1.2 A. before repair B. after repair of CLP patient

General problems of CLP patients (immediate problems):

Feeding:

Following the birth of a child with a cleft, one of the foremost concerns is to ensure that the child has adequate nutritional intake for growth. Because of the lack of closure between the oral and nasal cavities, new born often have difficulty maintaining enough negative pressure (suction) to suck from the breast or bottle.

Dental problem: Children with CL/P require the same routine dental care as all other children, and a local dentist can usually provide this preventive care. However, approximately 75%of all children with cleft present with defects involving the

part of the upper jaw that houses the teeth and the many have missing or extra teeth, teeth that are malformed or incorrectly positioned, or have problems with eruption.

Hearing and speech language problem:

Hearing:

Malformations of the head and neck including cleft palate, was identified as one of seven high risk factors for hearing loss in infants (Friedman et al., 1992). Almost 94% of children with cleft palate have continuous fluid in the middle ear, frequently causing conductive hearing loss (Charkins H., 1996)

Speech-language:

In addition to sound distortion that can result from hearing impairment, structural differences and the lack of closure between the oral and nasal cavities may also cause speech sound to become distorted due to hyper nasality.

Psychosocial problem:

The birth of an infant is an exciting and stressful event, even when the pregnancy, labor, and delivery go well. For the parents of an infant born with facial anomaly, however, it can be a devastating experience. Feeling of fear, guilt, resentment; inadequacy, shame, and grief are common among parents and family members of babies born with cleft deformities. Such feeling can interfere with parent infant bonding and may have psychosocial effects on both the infants and family members. Infants with the above problems in combination with abnormal facial appearance can heighten parental stress and decrease acceptance from society (Sandberg et al., 2002).

1.3 Justification of the Study:

There are many studies regarding CLP patients, including facial characteristics. The dentofacial characteristics of CLP patients were previously reported either on operated or unoperated cases by many researchers. There are few available studies on the quantitative evaluation of the facial features. Till now no one evaluated the repaired CLP patients based on direct anthropometry, lateral cephalometry and on study cast to describe their soft and hard tissues and dental arch dimensions as a whole.

The diagnosis and evaluation of CLP patients was based on clinical examination, mainly on cephalometry and study cast or from photographic analysis. Recently few morphological studies are found on CLP patients by 3D (three dimensional) system. But there are no available quantitative data for Malay patients with repaired CLP of different age groups in Kelantan, Malaysia.

The management of cleft lip and /or palate starts from the first day of life. The treatment needs of children with CLP are numerous and complex, requiring specialized and coordinated care of a multidisciplinary team. A substantial number of children undergo surgery after birth and treatment continues up to the age of 18 - 20 yrs, which is undoubtedly lengthy and costly.

The study is basically focused on dento-facial and skeletal characteristics of Malay patients with repaired CLP in Kelantan. To achieve the best morpho-functional reconstruction (with facial balance) in patients with CLP, it is important to have a complete understanding of the fundamental anatomy and function of this equilibrium in normal subjects as well as in patients with CLP (Markus et al., 1992).

The present study was consisted in a cross-sectional facial morphologic study. The purpose of this study was to locate and describe abnormalities of dentofacial area of Malay patients with repaired cleft lip and palate as compared with same noncleft ethnic group in Kelantan with an aim to contribute to the knowledge of differences concerning the dentofacial characteristics in individuals with Malay patients with repaired CLP in Kelantan.

The incidence of cleft lip and palate is not much higher as compared to many other diseases. Undoubtedly, its psychosocial impact is very high, especially due to the facial features and speech for the sufferer's as well as his /her close associates. Usually the CLP patient needs a long-term treatment by a group of specialists which is very expensive. The lengthy series of treatment of CLP patients from birth to adulthood is heavy burden for the patients, family, and also for the society.

The child born with a congenital CLP has not only physical defects but also many other problems facing him as he matures and seeks his way in life. How successful this individual will be in overcoming his handicap will depend largely upon the combined efforts of the group to assess, plan and treat the multiplicity of problems associated with his anomaly. One of the important tools for successful treatment is the quantitative measurement of age and ethnicity matched analytical anthropometrics data. Anthropometrics studies are integrated part of craniofacial surgery and syndromology (Farkas et al., 1992).

Malaysia is a multiracial country comprises the Malays, Chinese, Indians and others of mixed origin with various ethnic differences (Ong , M., 2004). This study will benefit, especially during orthognathic and orthodontic treatment from the aesthetic and functional point of view.

Measurements are now typically undertaken with the aim of contributing to the planning of surgical reconstruction involving the skull or jaw, necessitated by injury or by abnormality, if it affects eating effectiveness or appearance. Facial measurements can also contribute to monitoring of progress after surgical repair and even detecting possible regression, etc. (Atkinson, K. B., 1996).

In Kelantan (Malaysia), the treatment of operated cleft lip and palate patients has become a multidisciplinary management of a group of specialists since 1993 and most of the CLP patients are operated during infancy or early child hood. As a result there is little opportunity to examine persons with untreated clefts at later stages of development. Nevertheless the treatment demands more and more attention considering the magnitude of the prevailing problems. This investigation focuses primarily on the postoperative CLP patient's dentofacial characteristics.

Many studies on CLP patient's treatment, etiology and embryology have been reported. There are, however, some studies, which describe either the facial soft tissue or hard tissue but no similar studies that describe the dentofacial characteristics of Malay patients with repaired CLP in Kelantan.

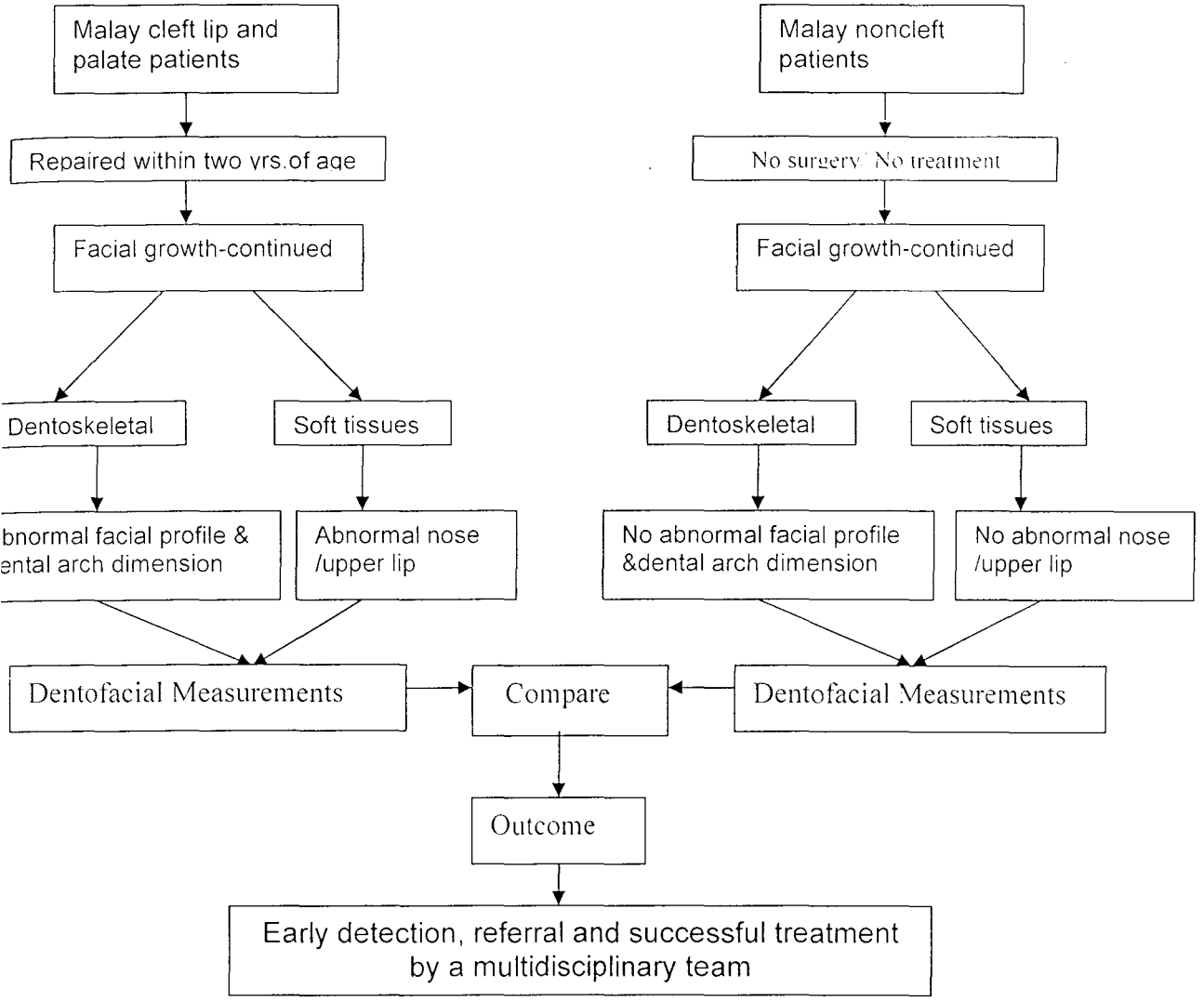
'A primary aim in treatment is to normalize facial appearance. To achieve this goal, it is important to be able to assess the normal facial morphology and any divergence from the norm' (Ayoub, A, 1999).

An objective and easily reproducible description of the form and characteristics of the facial and skeletal is prerequisite in longitudinal and comparative studies, as documentation, follow-up and communication among cleft lip and palate team members (Boric,V et al., 1993)

Thus a complete morphological feature of repaired cleft lip and palate Malay children, adolescents and adult of Kelantan have been evaluated along with the population norm of the peer age group.

The aim of this study was to measure the dentofacial characteristics that expected to be influenced by cleft defects. This study would be very helpful in diagnosis, treatment planning, evaluation, follow-up and also for research purpose which was also a demand for the clinicians.

Conceptual Framework (figure1.3)



CHAPTER TWO

OBJECTIVES

CHAPTER TWO

OBJECTIVES

2.1 General Objective:

To study the dentofacial and skeletal characteristics of Malay patients with repaired cleft lip and palate in Kelantan

2.2 Specific Objectives:

2.2.1 To determine the dentofacial and skeletal characteristics of Malay repaired cleft lip and palate children, adolescent and adult patients and noncleft Malay children, adolescent and adult in Kelantan

2.2.2 To compare the dentofacial and skeletal characteristics between Malay repaired cleft lip and palate children, adolescent and adult patients and Malay noncleft children, adolescent and adult in Kelantan

2.3 Research Hypothesis:

The dentofacial and skeletal characteristics of Malay repaired cleft lip and palate children, adolescent and adult patients are different from Malay noncleft children, adolescent and adult in Kelantan

2.4 Operational Definition:

1. Malay: The subjects included in this study, whose parent were Malay (Malay language, Muslim culture and Malay ancestor) in origin.

2. CLP groups (subjects): who born with a congenital defect either in lip or palate or in both regions in Kelantan.

3. Noncleft groups (subjects): The subjects who have no CLP and fulfilled the selection criteria.

4. Repaired CLP patients: Cleft lip and palate patients (same homogenous group) who underwent repair of complete cleft lip and palate either unilateral or bilateral.

5. Dentofacial: Included the structures below the upper eyelids (McWilliams, B. J, 1992) which includes facial surface soft tissue measurements and dental and skeletal relation to the respective jaws.

6. Children: Cleft lip and palate patients and normal subjects included in this study were from age 8 to 12 years (Gander, M. J, et al., 1981).

7. Adolescent: Cleft lip and palate patients and normal subjects included in this study were from age 13 to 18 years (Gander,M. J et al., 1981).

8. Adult: Cleft lip and palate patients and normal subjects included in this study were from age 19 to 40 years. (Ferrario et al., 1997).

CHAPTER THREE

LITERATURE REVIEW

CHAPTER THREE

LITERATURE REVIEW

3.1 Over view of cleft lip and palate:

3.1.1 Definition of CLP:

Cleft lip with or without cleft palate is a congenital malformation characterized by partial or complete clefting of the upper lip, with or without clefting of the alveolar ridge or the hard or soft palate. This definition exclude midline cleft of upper or lower lip and oblique facial fissure going towards the eye (www.icbd.org).

3.1.2 Epidemiology:

Cleft lip and cleft palate (CL/P) is one of the commonest congenital facial defects worldwide. The reported incidence varies from 0.5 to 3.6 per 1000 live birth (Mars, M, 1998). Studies in Denmark by Jensen et al., (1988) have demonstrated an increased incidence of cleft births between 1942 and 1981. Between 1938 and 1942 there were 1.5 per 1000 births, which increased to 1.89 per 1000 births between 1978 and 1981. Similar increases have been reported by Rintala (1986) and Srivastava and Bang (1990) in Finland and Kuwait respectively (Bellis, T.H et al., 1999).

CL/P is most prevalent among Native Americans (3.6 /1,000 live birth), less common in Asians (1.7 to 2.1 /1,000 live birth) Caucasians (1 /1,000 live birth) and African Americans (0.3 /1.000 live birth) (Edmondson et al., 1998). Overall, Asians, Americans, Indians, Alaskans and Japanese have a higher incidence of CLP in comparison to Caucasians while Negroid races have a lower incidence of oral clefting (Papadopulos, N.A et al., 2003)

There is some variation in incidence between racial groups (Malcolm et al., 2000). The incidence in Malaysia based on the report of CCCDC (2004), Kelantan , approximately 1000 babies born with a cleft lip and / or palate every year, which is about 1:500 live birth. In 1988, a published case control study found that CLP was the 2nd commonest birth defect in Malaysian population after musculoskeletal deformities (Goh,P. P and Yeo,T.C, 1988).The morbidity of CLP is higher in Kelantan than in other states in Malaysia (Kazumichi Fujioka and Yumiko Yamauchi, 2004).

Overall males are more affected than females. Isolated cleft lip and cleft lip and palate are more common in males (2:1). Isolated cleft palate is more common in females (2:1). The frequency of cleft lip and palate is 50%, Cleft palate is only 30%, Cleft lip is only 20% and Cleft lip and alveolus only 5% (Smith L, F.,1991).Numerous studies of congenital anomalies associated with CLP have reported that 98% are isolated CLP and only 2% are syndromic.

3.1.3 Embryology (How does it occur):

Clefting occurs very early in pregnancy, usually before a woman even knows that she is pregnant. The development of an individual is a complicated and delicately balanced process. Knowledge of the evolutionary development of the skull, face, and jaws is important and helpful in understanding the complex events involved in cephalogenesis, which will aid in further development of CLP management as well as research, as it is an established congenital defect (Cate, T et al., 2003). The embryology of facial development is complex and is still not fully elucidated (Sperber., 1989.)

The lip and the palatal regions develop from the embryonic primary and secondary palate. The primary palate contributes the lip, anterior tooth bearing alveolus, and the anterior palate up to incisive foramen. The secondary palate contributes the remaining hard and soft palate. (Wong, F. K et al., 2004).

Normal development of Primary palate:

During early development (In 4^{th} week) the primitive oral cavity (stomodeum) is bounded by five facial swelling, produced by proliferating zones of mesenchyme lying beneath the surface ectoderm. These are the frontonasal, mandibular and maxillary process. The frontonasal process lies above, the two mandibular processes lie below, and the two maxillary processes are located at the sides. The maxillary and mandibular processes are derived from the first

branchial arches. The facial processes are demarcated by grooves that, in the course of normal development, become flattened out by the proliferative and migratory activity of the under lying mesenchyme.

In a 5-week old embryo, localized thickening of ectoderm give rise to the nasal and optic placodes. These placodes will form the olfactory epithelium and the lenses of the eyes and the two- blind ended nasal pits (the primitive nasal cavities) respectively. Proliferation of mesenchyme from the frontonasal process around the opening of the nasal pits produces the medial and lateral nasal process. By the end of the 5^{th} week these nasal processes and maxillary processes fuse to form the primary palate. These processes are essentially the mesodermal tissues covered by ectoderm (Berkovitz et al., 2002).

The maxillary process buds of from its upper border and it continue to grow along with the mandibular process to meet its fellow processes in the midline. During the course of its growth towards the midline, each maxillary process fuses with the lateral process and then with the medial process, before meeting with its fellow of the opposite side to form a primary palate, from which develop the upper lip, and palate anterior to the incisive foramen. These processes are essentially mesodermal tissues covered by ectoderm. During the fusion, the covering epithelium of these processes at the sites of union disintegrates and their mesodermal tissues come in contact with each other and unite. The above union is usually completed within 5 to 6 week in intra-uterine life.

Development of Cleft of Primary palate:

Failure of this union due to lack of disintegration of epithelium or due to any other cause will produce total cleft of primary palate, while partial fusion will produce sub-total clefts.

Normal development of Secondary palate:

The formation of the secondary palate commences between 6 and 9 weeks and completes around the third month of gestation. Three outgrowths appear in the oral cavity; the nasal septum grows downward from the frontonasal process along the midline, and the two palatine shelves or processes, one from each side extend from the maxillary processes towards the midline. At first, the palatal shelves grow downwards and adjacent to the tongue. Gradually, they elevate to a level above the tongue and assume a horizontal position. Contact of these three processes and with the primary palate (and fusion of the tissues then occurs) results in the development of a normal secondary palate (Cate, T et al., 2003).

Development Cleft of Secondary palate:

If the palatal shelves fail to fuse, it results in a defect that varies from a bifid uvula to a complete cleft of the secondary palate. The union between the palatal processes and nasal septum occurs at 8 to 10 weeks intra uterine life. Other possible reasons for palatal clefting involve abnormal growth of the palatal shelves, cell death (post fusion), and the failure of mesenchymal consolidation and differentiations. Cleft of the secondary palate is usually the result of the different morphogenic events when compared to cleft lip with or without cleft palate.

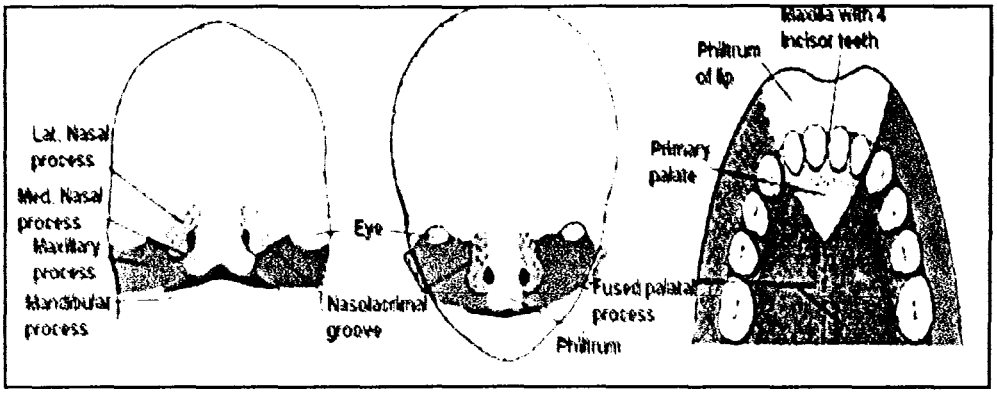


Figure 3.1 shows embryonic processes of lip and palate.

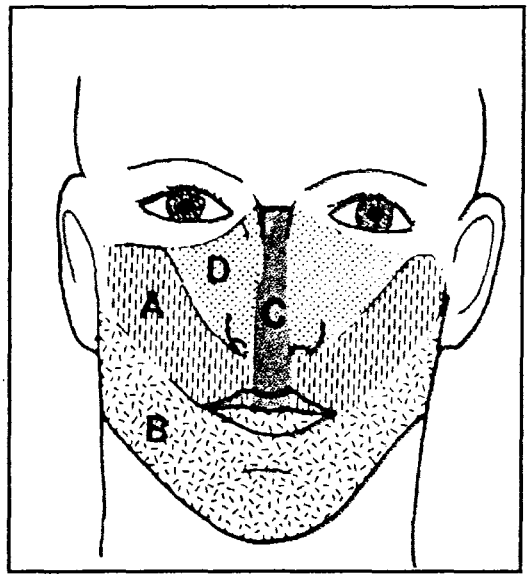


Figure 3.2 shows facial structures developed from the embryonic processes.

A= Maxillary process, B= Mandibular process, C= Medial nasal process , D = Lateral nasal process.

3.1.4 Etiology:

The exact cause of clefting has not been determined, but most examples appear to be of multifactorial inheritance. They are due to a combination of genetic and environmental factors, rather than a single gene inheritance (Edmondson et al., 1998).

The cleft may be part of a syndrome or may occur in isolation (Wong et al., 2004). For some there is a clear familial history of facial clefting, implying a genetic disorder but for the majority of cases the occurrence is sporadic, suggesting that as yet unidentified factors have an important role in the etiology of the condition. Recent literature reveals that wide ethnic and racial variations in the occurrences of cleft lip and/ or palate exist (Wantia et al., 2002).The etiology involves both hereditary and environmental factors. Approximately 40% of CL with or without CP is hereditary. About 20% of isolated CP is hereditary and 5% of cleft cases are associated with syndromes. If there is cleft lip with or without cleft palate, the probability of the deformity in the next sibling is as follows (Smith, 1991):

a) One sibling with cleft and no parent with cleft- 4%

b) One sibling and one parent has cleft -10 to17%

c) No sibling with cleft and one parent with cleft- 2%

If there is cleft palate alone:

d) One sibling has cleft and no parent with cleft is - 2%

e) One sibling has cleft and one parents with cleft -17%

f) No sibling with cleft and one parent with cleft -7%.